NON ODONTOGENIC TUMORS OF THE ORAL CAVITY

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A TUMOR, by definition , is simply a swelling of the tissue. In the strict sense ,the word does not imply a neoplastic process.

NEOPLASIA , on the other hand can be defined as an abnormal mass of tissue, the growth of which exceeds and is uncoordinated with that of the normal tissues and persists in the same excessive manner after cessation of the stimuli, which evoked that change.

However, for the sake of simplicity, we use both these words interchangeably.

Tumors of the oral cavity may be divided, on the basis of origin into

1. Odontogenic –those which arise from tooth forming tissues.
2. Non-odontogenic- those which arise from any other tissue except the odontogenic tissues.

Here we shall discuss only the latter type, i.e. the non-odontogenic tumors.

Classification of Non – Odontogenic tumors of the oral cavity

Based on the tissue of origin, they are divided into five types :

- Tumors of Epithelial tissue origin
- Tumors of Connective tissue origin
- Tumors of Muscle tissue origin
- Tumors of Nerve tissue origin
- Metastatic tumors of jaws

Epithelial origin

Benign

- Squamous papilloma
- Keratoacanthoma
- Oral nevi

Malignant

- Squamous cell carcinoma
- Basal cell carcinoma
- Verrucous carcinoma
- Spindle cell carcinoma
- Adenoid squamous cell carcinoma
- Basaloid squamous cell carcinoma
- Adenosquamous carcinoma
- Undifferentiated carcinoma
- Lymphoepithelioma and transitional cell carcinoma
- Malignant melanoma

➢ Connective tissue origin

Benign

- Fibroma
- Fibrous histiocytoma
- Lipoma
- Oral hemangiomas
- Lymphangioma
- Myxoma
- Chondroma
- Benign chondroblastoma
- Chondromyxoid fibroma
- Osteoma
- Osteoid osteoma
- Benign osteoblastoma

Malignant

- Fibrosarcoma
- Malignant fibrous histiocytoma
- Synovial sarcoma
- Liposarcoma
- Hemangioendothelioma
- Hemangiopericytoma
- Kaposi's sarcoma
- Chondrosarcoma
- Osteosarcoma
- Non-Hodgekin's lymphoma
- Primary lymphoma of bone
- Burkitt's lymphoma
- Hodgekin's lymphoma
- Multiple myeloma
- Plasmacytoma

Muscle tissue origin

Benign

- Leiomyoma
- Angiomyoma
- Rhabdomyoma
- Granular cell myoblastoma
- Congenital epulis of newborn

Malignant

- Leiomyosarcoma
- Rhabdomyosarcoma
- Alveolar soft-part sarcoma

Nerve tissue origin

Benign

- Neurofibroma
- Neurilemmoma
- Melanotic neuroectodermal tumor of infancy

Malignant

- Malignant peripheral nerve sheath tumor

- Metastatic tumors of jaws

BENIGN TUMORS OF EPITHELIAL TISSUE ORIGIN

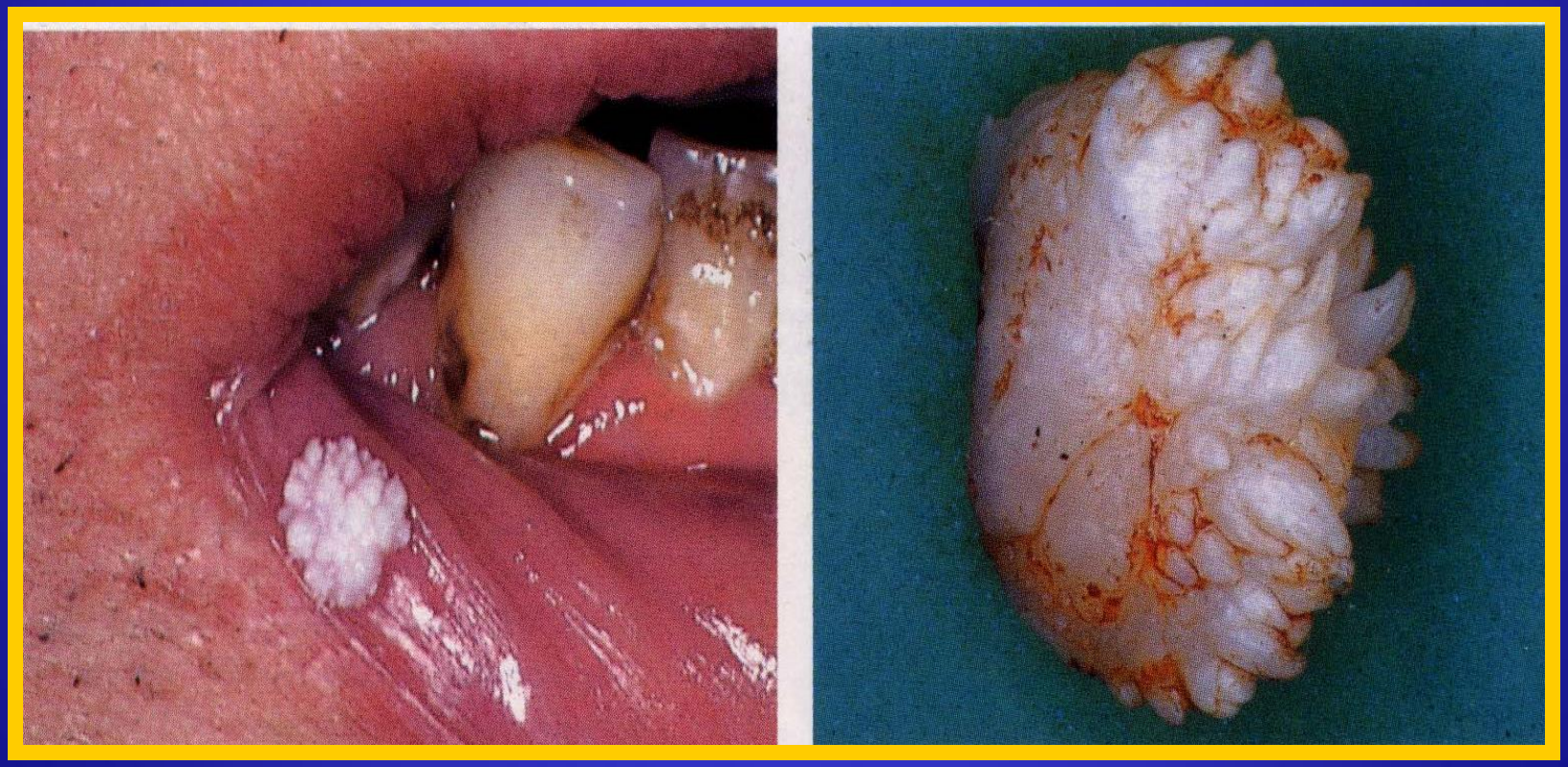
SQUAMOUS PAPILLOMA

- It seems to be associated with papilloma virus (HPV 6 & 11), the one causative in common skin warts (verruca vulgaris).
- All HPV lesions are infective, but oral papilloma do not seem to be contagious.
- It is clinically and microscopically indistinguishable from verruca vulgaris, the virus-induced focal papillary hyperplasia of the epidermis.

Clinical features-

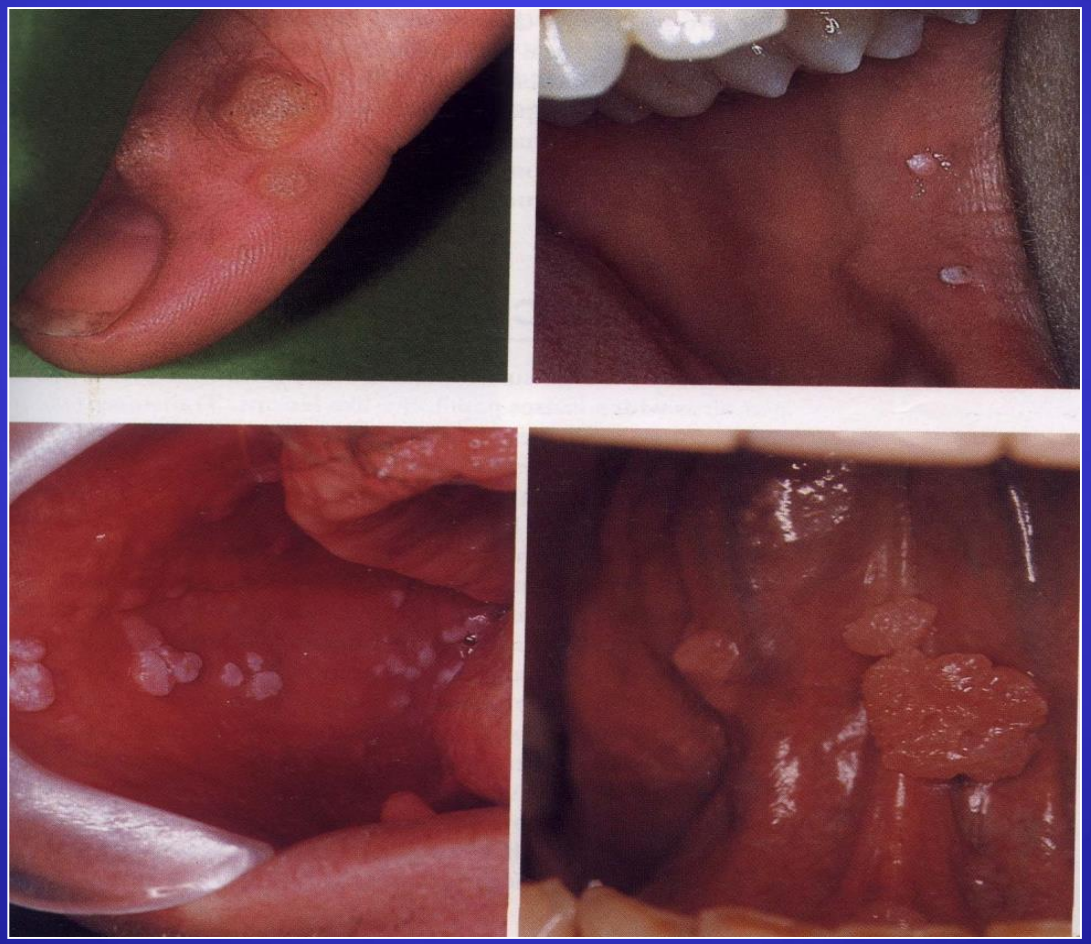
- It is an exophytic, pedunculated, painless growth made up of numerous, small finger-like projections, which result in a lesion with a roughened, verrucous or 'cauliflower' surface.
- Intraorally it is found most commonly on the tongue, lips, buccal mucosa, gingiva & palate, esp. that area adjacent to the uvula.

Squamous papilloma of lip (L) and excised specimen (R)



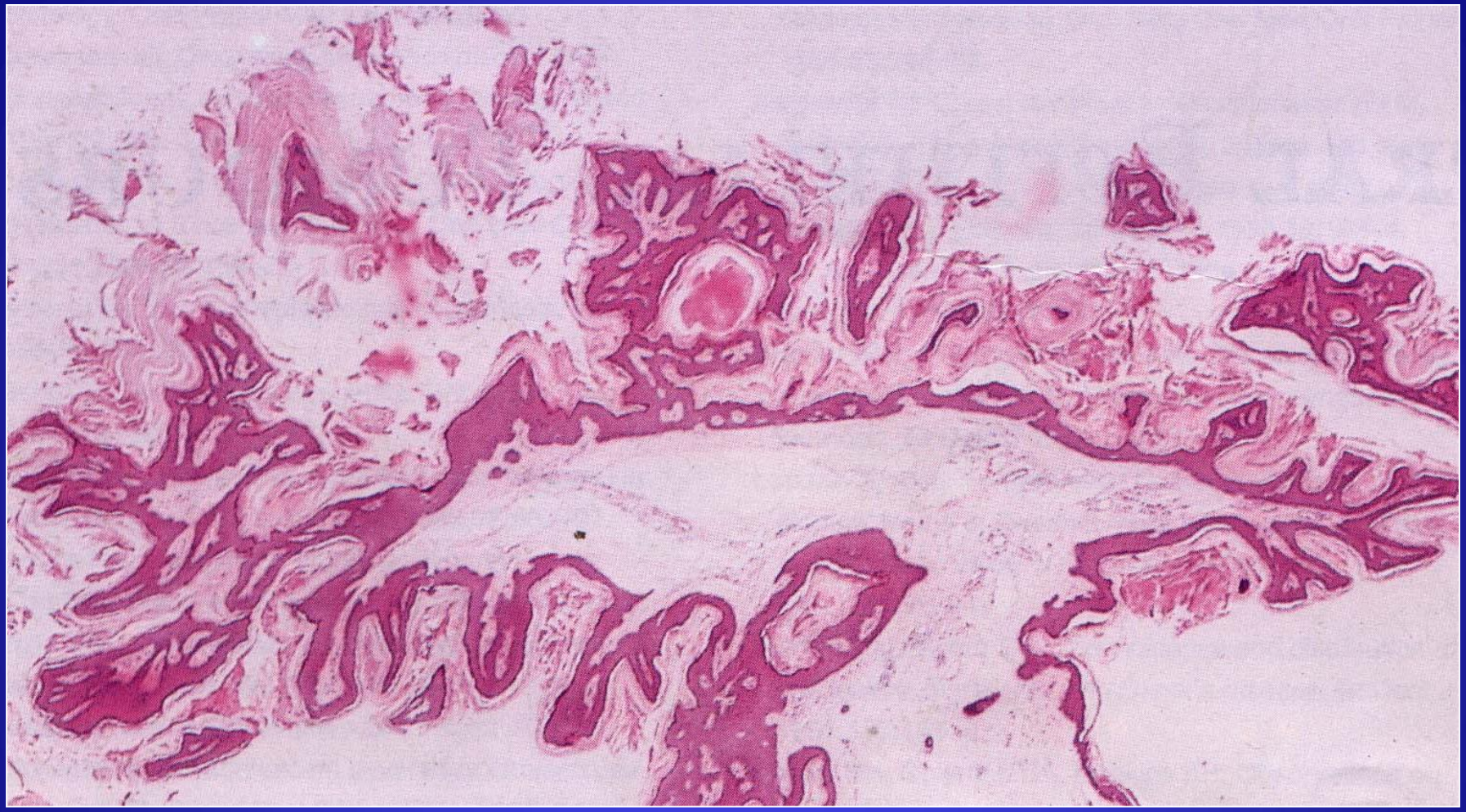
- The common wart is a tumor of skin, analogous to the oral papilloma, but uncommon on the OMM.
- However, these can be seen on the lips and occasionally intraorally, in patients with verrucae on the hands or fingers, because of autoinoculation by finger sucking or finger nail biting.
- Papillomatous or ‘pebbly’ lesions and fibromas of various sites in the oral cavity are recognized as one of the many manifestations of **Cowden's syndrome.**

Verruca vulgaris of skin (L) and oral cavity (R)



Histologic features-

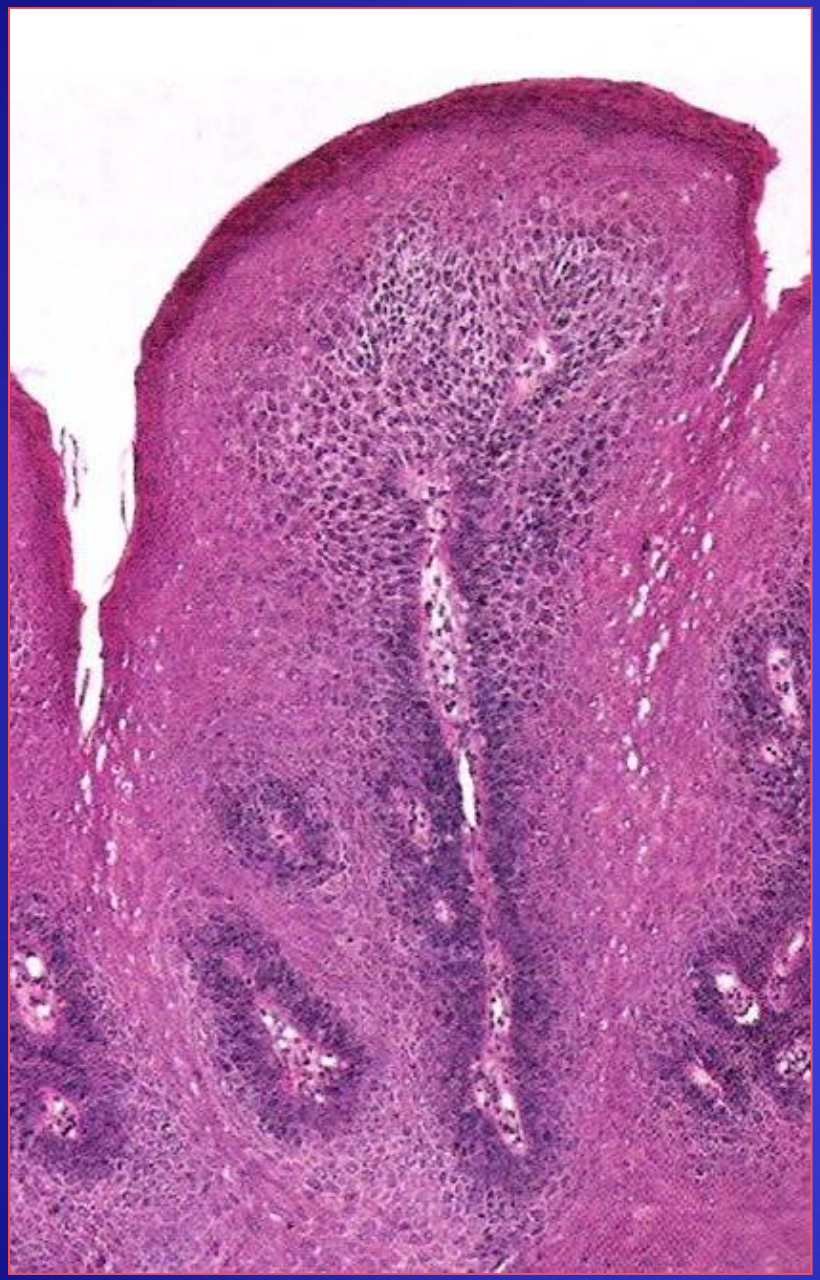
- Many long, thin, finger-like projections extending above the mucosal surface, each made up of a continuous layer of stratified squamous epithelium and containing a thin central connective tissue core, which supports the nutrient blood vessels.
- The essential feature is a proliferation of the spinous cells in a papillary pattern; the connective tissue is supportive stroma only and is not a part of the neoplastic element.
- Occasionally they show marked basilar hyperplasia and mild mitotic activity.
- **Koilocytes**, HPV altered epithelial cells with perinuclear clear spaces and pyknotic nuclei, may or may not be found in the superficial layers of epithelium.



SQUAMOUS PAPILLOMA

Connective tissue processes are covered by hyperkeratinized stratified squamous epithelium

SQUAMOUS PAPILLOMA : Low power view showing a pedunculated squamous epithelial proliferation. There are multiple papillary projections with fibrovascular connective tissue cores.



SQUAMOUS PAPILLOMA : tip of the papillary projection shows mature stratified squamous epithelium with a slightly thickened parakeratin surface layer.

Treatment-

- Excision, including the base of the mucosa into which the pedicle inserts.
- If properly excised, recurrence is very rare.
- Some other methods are -
  conservative surgical excision

  -curettage

  -liquid nitrogen cryotherapy

  -topical keratolytic agents (usually containing salicylic acid and lactic acid)

KERATOACANTHOMA
(self healing carcinoma, molluscum sebaceum, verrucoma)

- It is relatively common low-grade malignancy that originates in the pilosebaceous glands.
- It is considered to be a variant of invasive squamous cell carcinoma.

Etiologic factors :

- Sunlight
- Chemical carcinogens
- Trauma
- HPV
- Genetic factors
- Immunocompromised status

Clinical features :

- Occurs in all age groups but incidence increases with age.
- Men: women =2:1
- More common in fair-skinned people and in sun-exposed areas
- Face, neck and dorsum of upper extremities are the most common sites. Intraoral lesions are quite uncommon. May be seen on lips.
- Lesions are typically solitary elevated, umbilicated or crateriform with a depressed central core or plug. Its often painful and regional lymphadenopathy may be present.

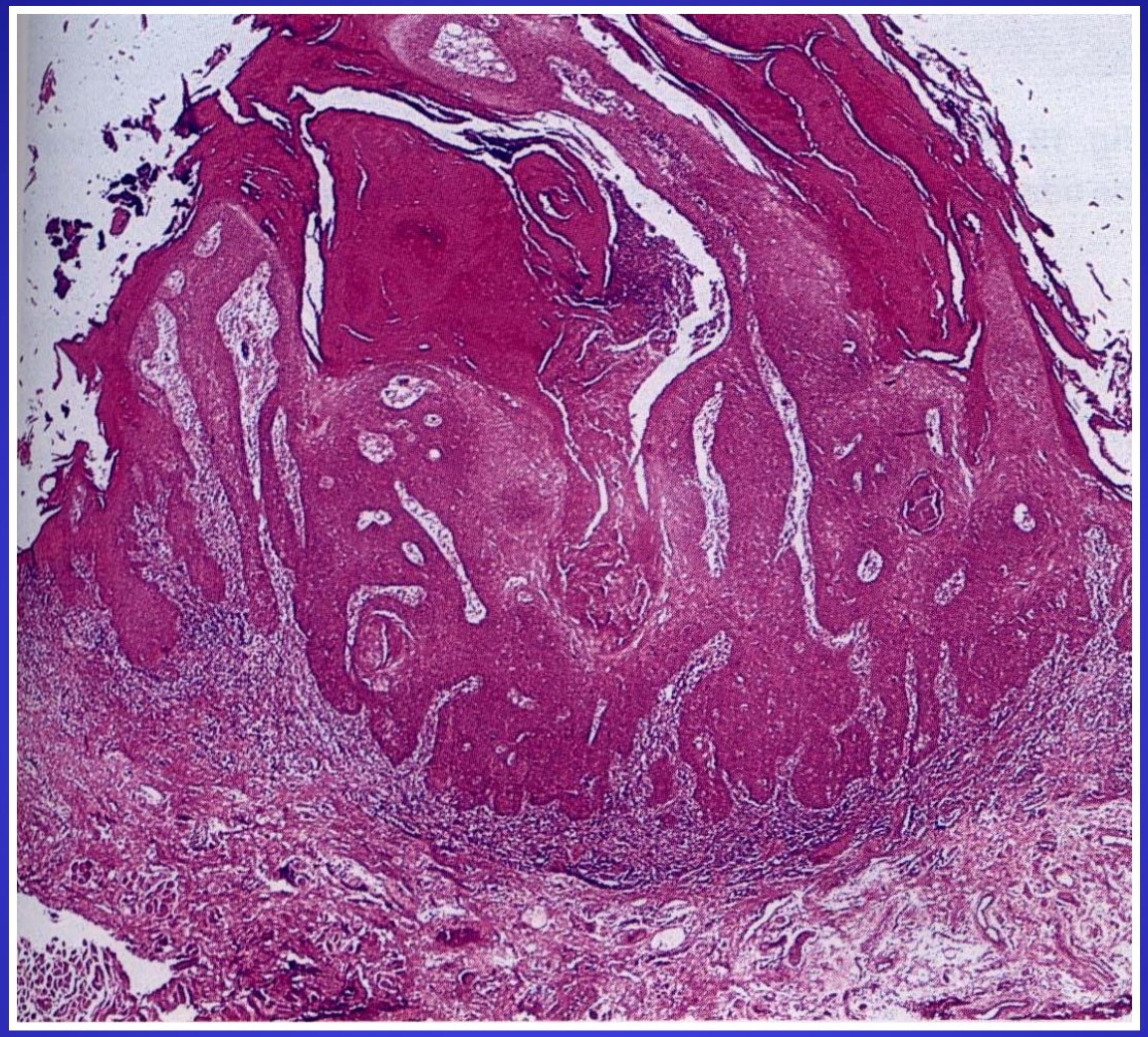
KERATOACANTHOMA

Histologic features :

- Consists of hyperplastic squamous epithelium growing into the underlying connective tissue (C.T.).
- Surface is covered by a layer of para- or orthokeratin with central plugging.
- At the deep leading margin of tumor, islands of epithelium often appear to be invading and usually this area cannot be differentiated from the SCC. Sometimes it may also invade perineural spaces, but this not a distinguishing feature between the two.
- Pseudocarcinomatous infiltration typically presents a smooth, well-demarcated front that does not extend beyond the level of sweat glands. C.T. shows chronic inflammatory cell infiltration.

- Most characteristic feature is found at the margins where the normal adjacent epithelium is elevated towards the central portion of the crater; then an abrupt change in the normal epithelium occurs as the hyperplastic acanthotic epithelium is reached. That's why diagnosis may be impossible if the normal adjacent epithelium is not included in the biopsy.

- The term **squamous cell carcinoma, keratoacanthoma type,** has been introduced for otherwise classic keratoacanthomas, that reveal a peripheral zone formed by squamous cells with atypical mitotic figures, hyper chromatic nuclei, and loss of polarity. These marginal cells may also penetrate into surrounding tissue in a more aggressive pattern.



Keratoacanthoma – deep keratin filled pit is surrounded by hyperplastic epithelium

Treatment-

- Surgical excision
- Patients should be followed for development of new primary skin cancers (SCC).

ORAL NEVI
(oral melanocytic nevus, nevocellular nevus, mole)

- Nevi are hamartomas (a benign tumor-like nodule composed of an overgrowth of mature cells and tissues normally present in the affected part, but with disorganization and often with one element predominating),i.e. benign proliferations of nevus cells in either epithelium or c.t.
- They may be classified as **congenital** or **acquired.** On the basis of histologic location of nevus cells, cutaneous acquired nevi can be classified into 3 categories :

✓ *Junctional nevi*

✓ *Compound nevi*

✓ *Intradermal nevi*

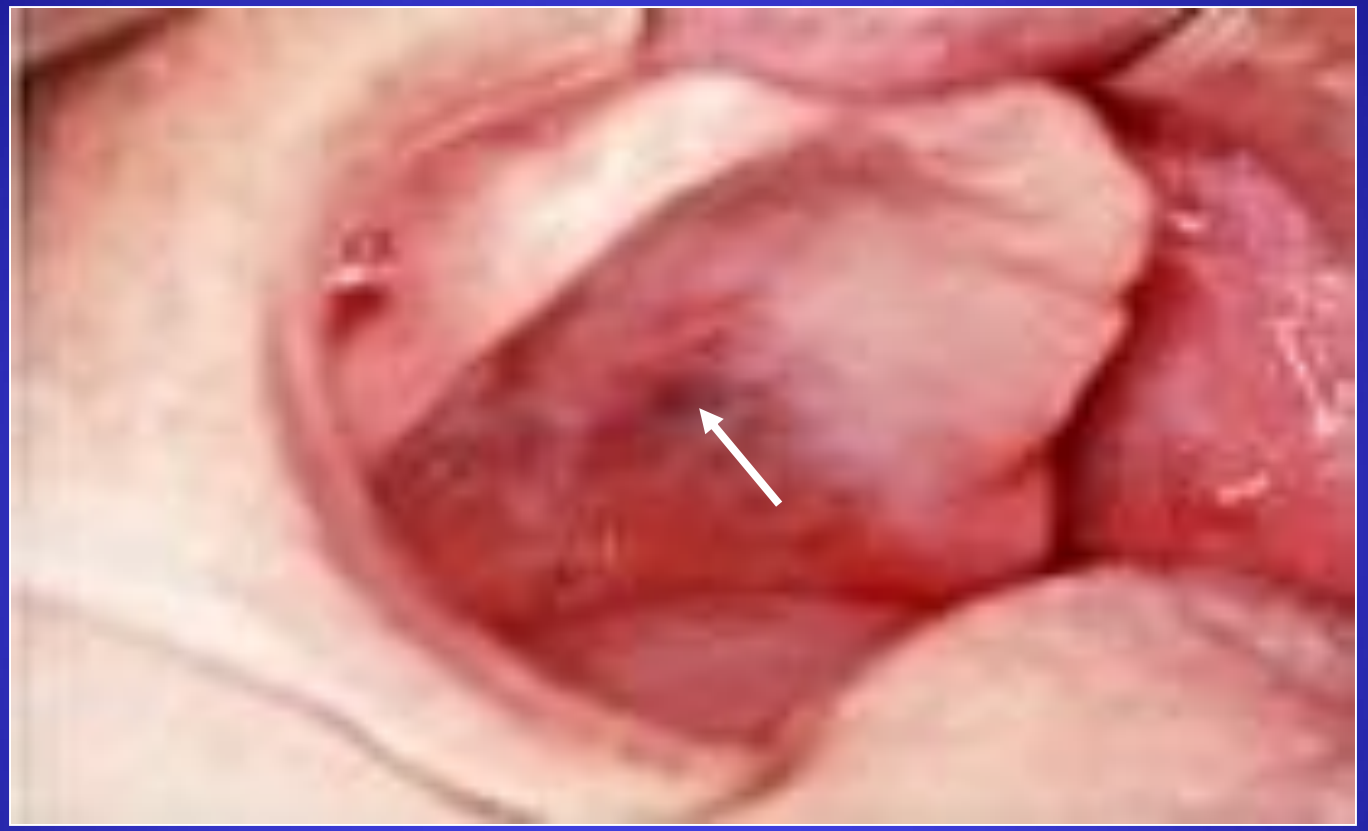
- Junctional nevi that are first noted in infants, children and young adults typically mature into compound nevi. Then during later adulthood, they mature into intramucosal nevi (intraorally, the term intradermal is replaced by intramucosal) which have diminished pigmentation.

- Most common mucosal type is the intramucosal nevus, followed by the common **blue nevus** (commoner in the mouth than in the skin).

Clinical features :

- 85% of oral nevi are found in patients <40 years old.
- Found in all races but more commonly in whites.
- Intramucosal nevi are slightly predominant in women.
- Oral nevi occur most commonly on the hard palate and the buccal mucosa, followed by gingiva, lip, and labial mucosa.
- Mostly asymptomatic.
- Usually pigmented, brown, black or blue.
- They are well circumscribed, raised lesions.
- Sometimes amelanotic, sessile growths which resemble fibromas or papillomas.

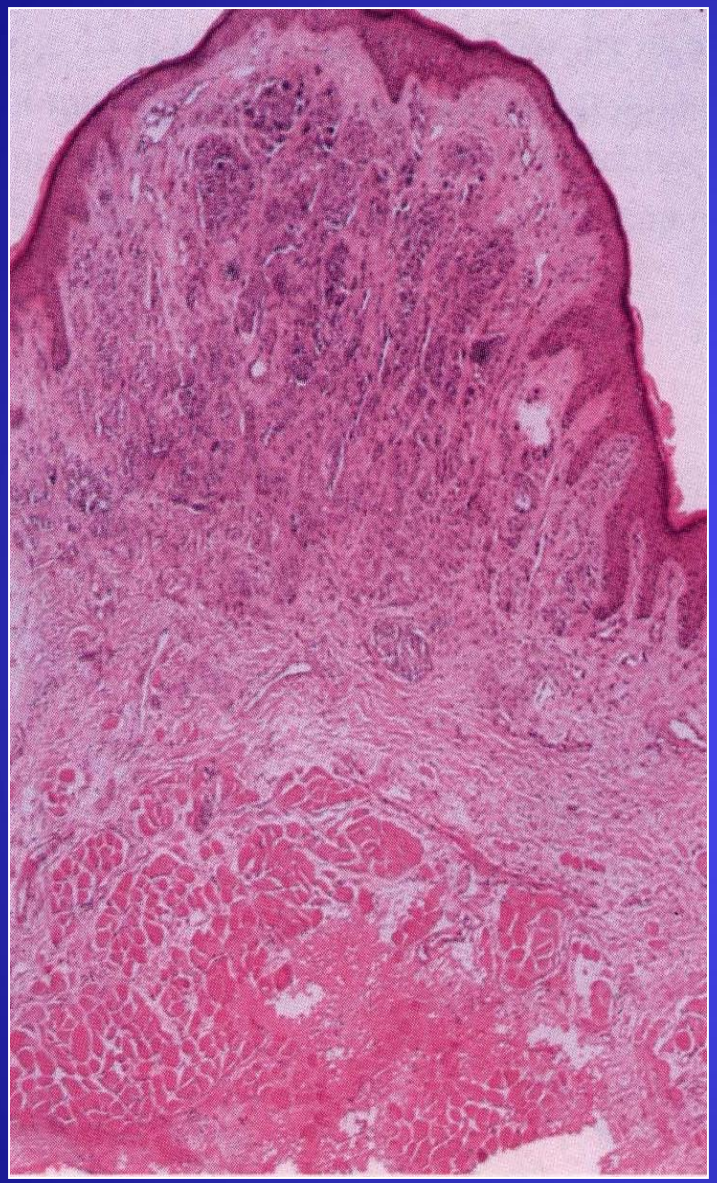
Oral pigmented nevus – macular lesion on the tip of the tongue



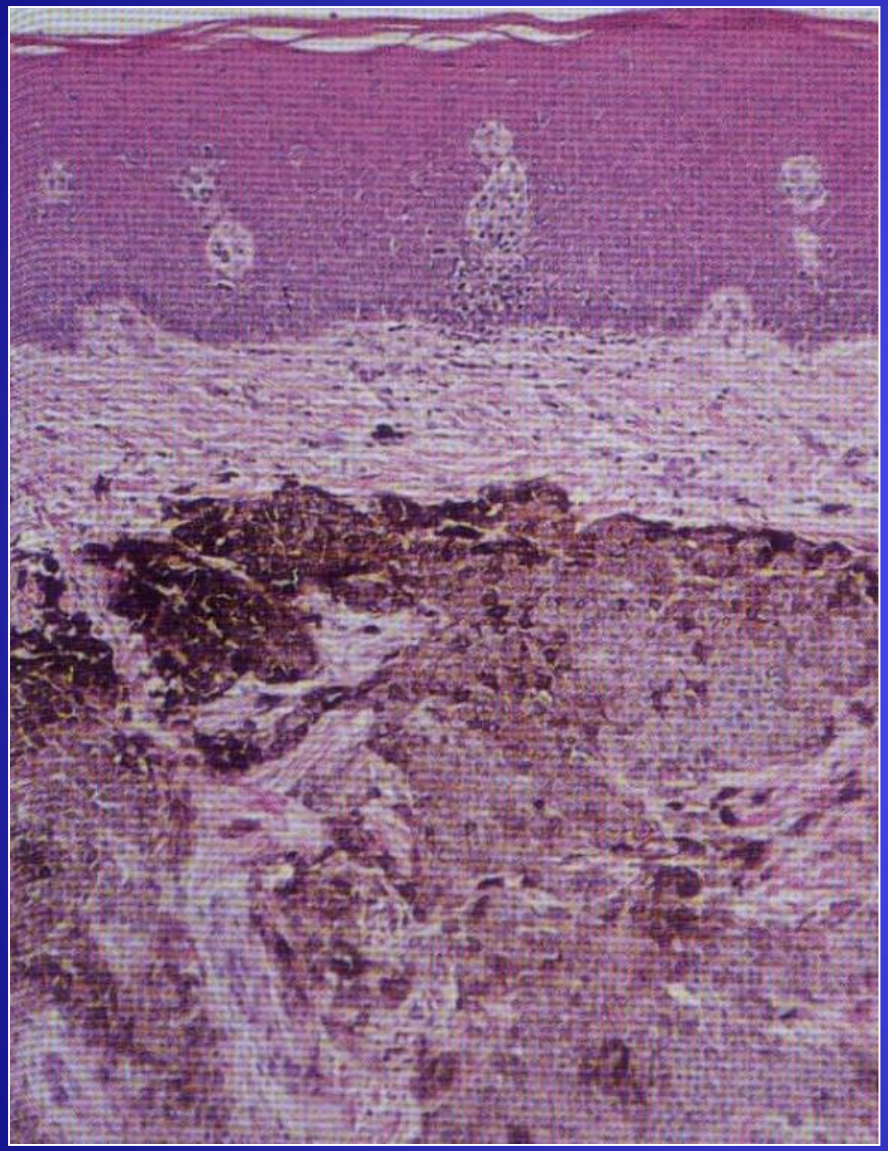
BLUE NEVUS :
oral lesions are found almost always on the palate

Histologic features :

- Nevus cells are assumed to be derived from the neural crest cells.
- They are large, ovoid, round or spindle-shaped cells with pale cytoplasm and may contain melanin pigment granules.
- Nucleus is vesicular and lacks the dendritic processes typical of melanocytes.
- Melanosomes are retained by the nevus cells and are not transferred to the adjacent keratinocytes. They tend to be grouped in sheets or chords called **nests or theque**.
- In the **intradermal nevus**, nevus cells are situated within the dermis and are separated from the overlying epithelium by a well defined band of c.t.

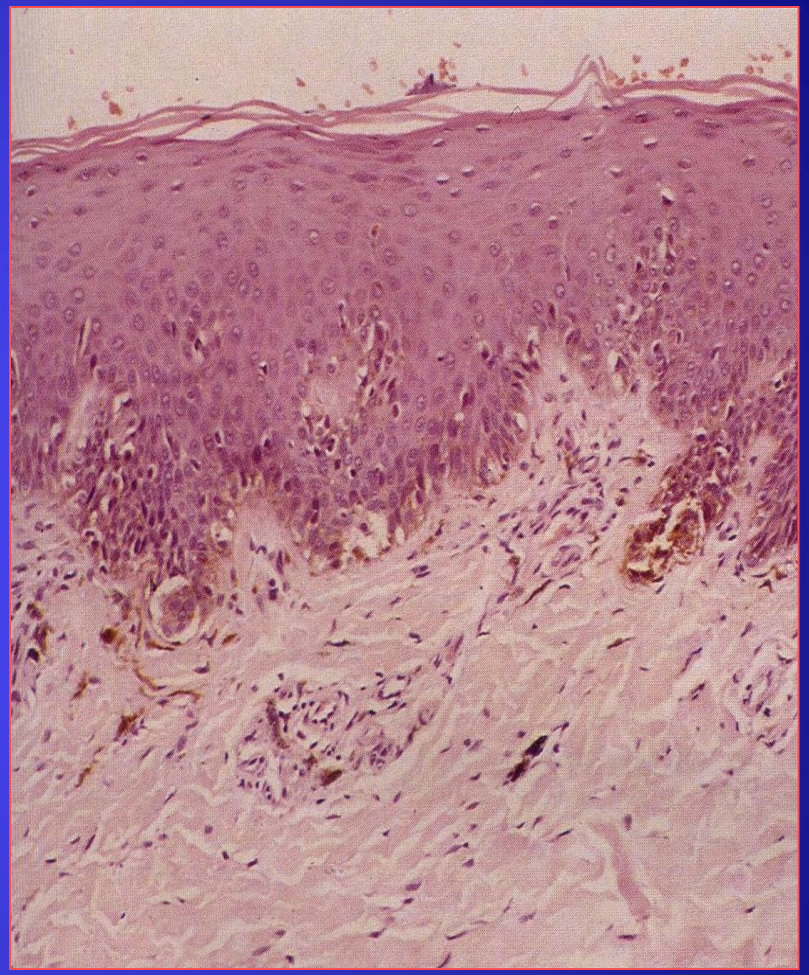


Intradermal nevus – There is absence of junctional activity but nests of lesional cells are present in the dermis.

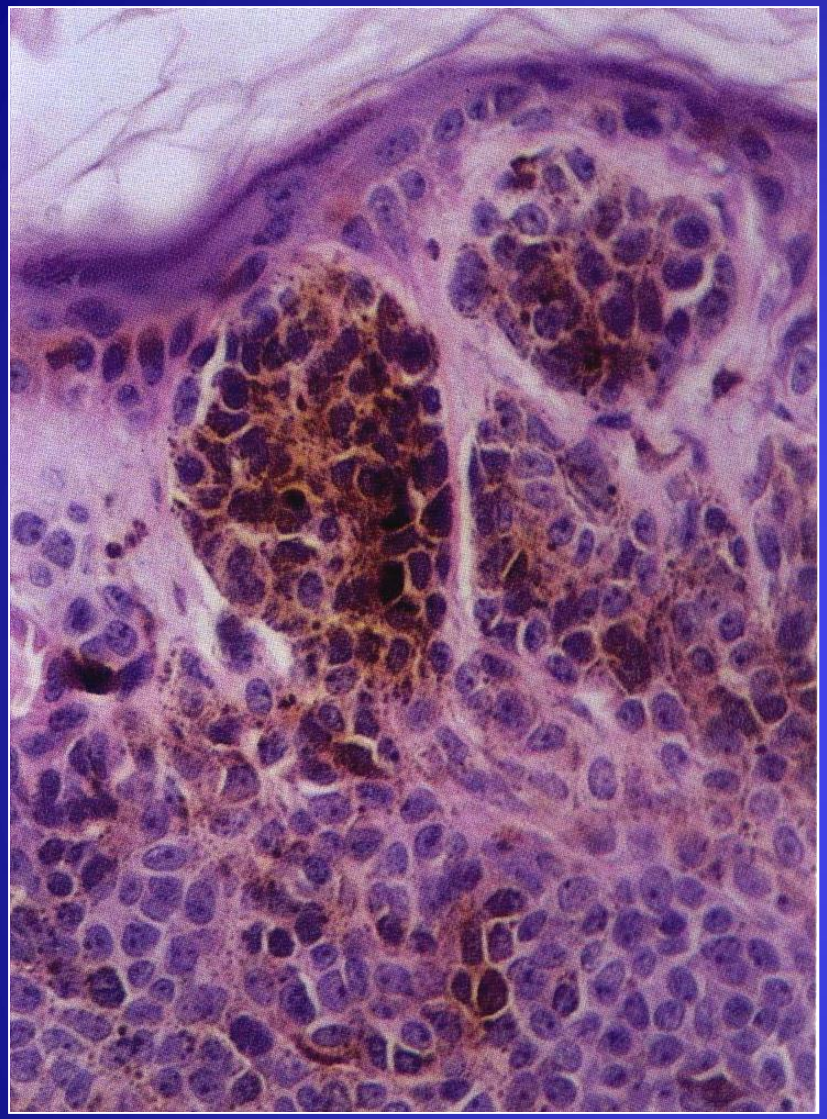


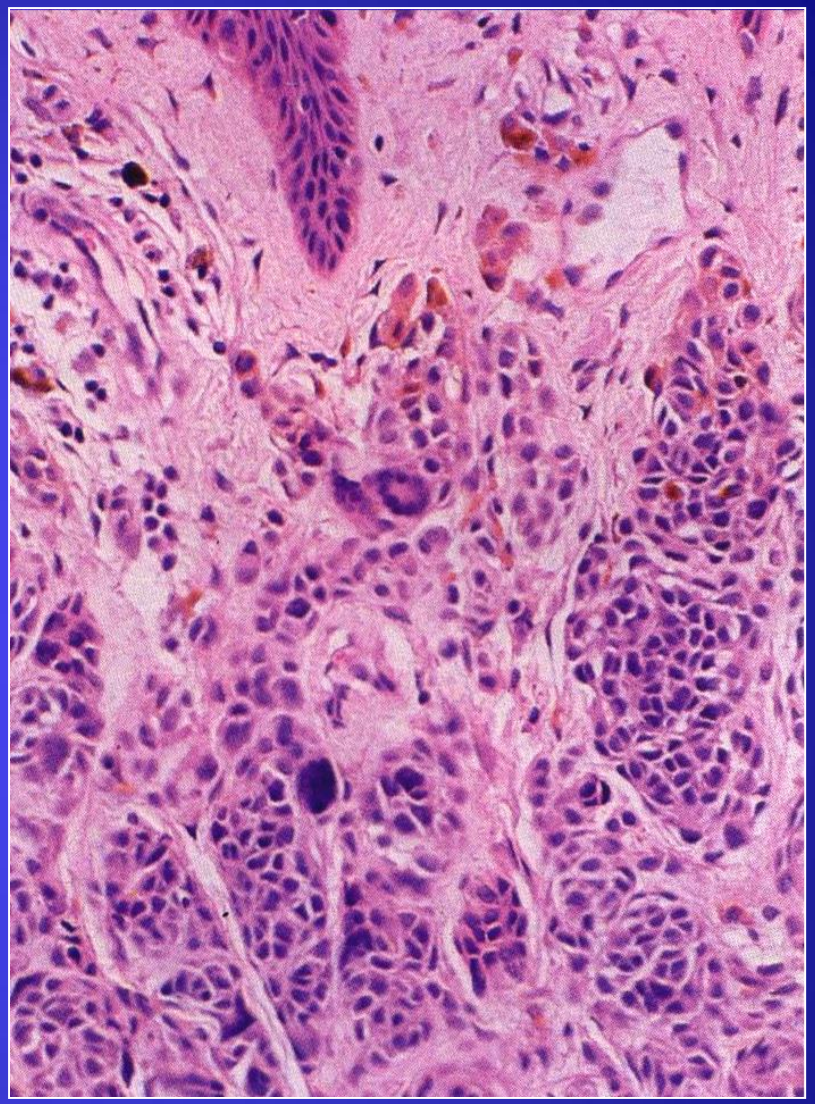
Intramucosal nevus

- In the **junctional nevus**, the nevus cells contact and seem to blend into the surface epithelium, which is usually thin and irregular and shows cells apparently crossing the junction and growing down into the c.t. – *abtropfung* or 'dropping off ' effect. These are commonly known to undergo transformation into malignant melanomas.



- The **compound nevus** shows features of both. Nests of nevus cells are dropping off from the epidermis, while large nests are also present in the dermis.





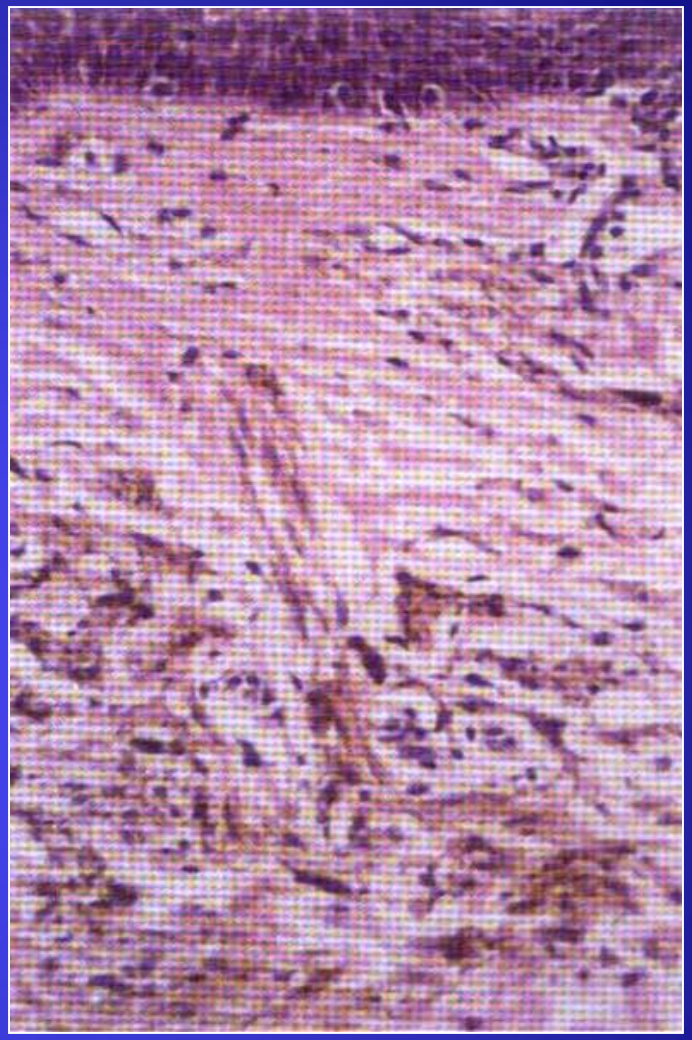
Compound nevus – (L) junctional activity is seen. Nests of melanocytes are seen 'dropping off' from the basal layer of epithelium. **(R)** Nests of nevus cells and multinucleated nevus cells.

- The **blue nevus** is of two types - common
  - cellular

- In the common type, elongated melanocytes with long branching dendritic processes lie in bundles usually oriented parallel to the epidermis, in the middle and lower third of dermis. no junctional activity is seen. Melanocytes are typically packed with melanin granules, sometimes obscuring even nucleus and even extending into the dendrites.

- In the cellular type, an additional cell type is present- a large, round or spindle cell, with a pale vacuolated cytoplasm. These cells are commonly arranged in an alveolar pattern.

Blue nevus – fontana stain shows densely pigmented spindle cells in the corium



Blue nevus – pigmented spindle shaped cells are well separated from the underlying epithelium

Treatment :

- Surgical excision of all intraoral nevi is recommended due to the constant chronic irritation of the intraoral mucosa in nearly all sites.
- Congenital nevi have a greater risk for malignant transformation.

MALIGNANT LESIONS OF EPITHELIAL ORIGIN

BASAL CELL CARCINOMA
(BCC, Rodent ulcer)

- The most common malignancy in humans.
- Most frequently develops on the exposed surfaces of the skin, face and scalp in middle aged or elderly fair-skinned persons.
- It is slow growing and rarely metastasizes but can cause significant local destruction.

Etiology –

- UV light (chronic sun ray exposure)– most important and common cause
- Radiation like X-rays
- Chemicals like arsenic
- Immunosuppression
- Syndromes like xeroderma pigmentosum and nevoid BCC syndrome

BCC is thought to arise from pluripotent stem cells of the basal cell layer of epidermis as well as follicular structures (hair follicle stem cells).

Clinical features :

- Most frequently in the fourth decade of life
- Male :female =2:1
- Most frequently seen on the middle third of face
- Does not arise from the oral mucosa so is not seen intraorally except for invasion from an adjacent skin surface.
- Subtypes of BCC are –

✓ **Nodular BCC –** most common variety. It begins as a slightly elevated papule with a central depression which ulcerates, heals over and then breaks down again. Very mild trauma may cause bleeding. Eventually, the crusting ulcer which appears superficial develops a smooth, rolled border representing tumor cells spreading laterally beneath the skin.

✓ **Pigmented BCC –** in addition to the features seen in the nodular type, this type contains black or brown pigmentation and are seen more commonly in dark skinned people.

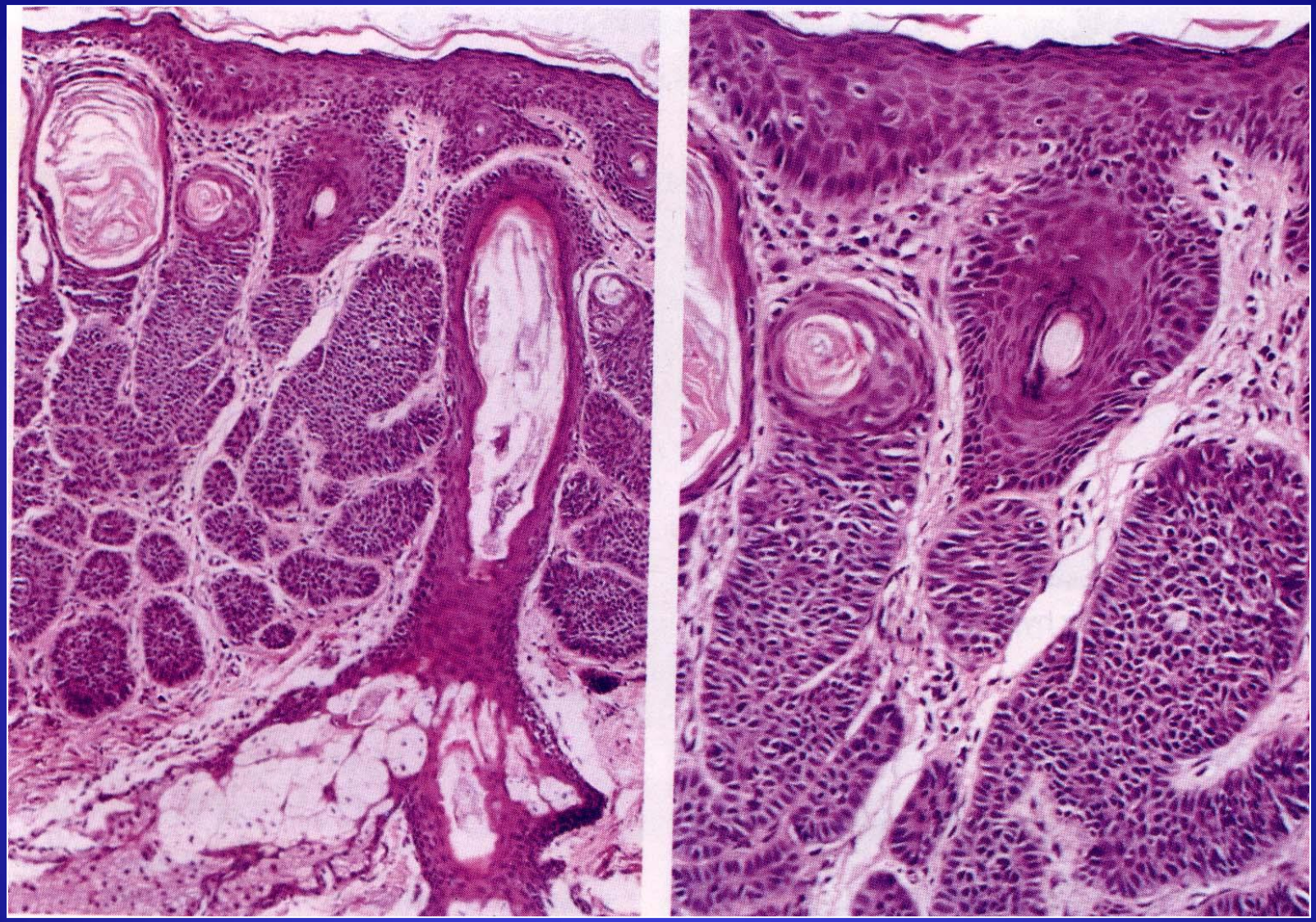
- **Cystic BCC –** may contain translucent blue-gray cystic nodules that may mimic benign cystic lesions.
- **Superficial BCC –** presents as scaly patches or papules, commonly on the trunk, that are pink to red-brown in color, often with central clearing and a thread-like border. Erosion is less common than in the nodular type. Papules may mimic psoriasis or eczema but are slowly progressive & not prone to fluctuation. Numerous superficial BCCs may indicate arsenic exposure.
- **Micronodular BCC –** an aggressive variety. less prone to ulceration, may appear yellow- white when stretched, and is firm to touch. May have a seemingly well-defined border.
- **Morpheaform and infiltrating BCC –** aggressive types with sclerotic (scar-like) plaques or papules, which may be mistaken for scar tissue. Border is usually not well defined and often extends well beyond clinical margins. Ulceration, bleeding and crusting are common.

BASAL CELL CARCINOMA

Lesion seen on the middle third of face, just below the eye.

Histologic features :

- In **nodular and pigmented types** , tumor cells called *basalioma cells* typically have large, oval, hyperchromatic nuclei with little cytoplasm.
- These are arranged in well demarcated islands which appear to arise from the basal layer of the overlying epidermis and invade into the underlying dermis.
- These islands typically show peripheral palisading and cleft formation (retraction artifact) commonly occurs between the nests and stroma because of mucin shrinkage during tissue fixation and staining.
- In **pigmented type**, benign melanocytes in and around the tumor produce large amounts of melanin.
- In the **superficial type**, lobules of tumor cells drop from the epidermis in a multifocal pattern.



BASAL CELL CARCINOMA

Well demarcated islands showing peripheral palisading.

(L) Low power (R) high power view

- **Morpheaform type** exhibit infiltrating thin strands of tumor cells in a dense fibrous stroma. The strands of **infiltrating type** are thicker and have a spiky irregular appearance. Also they lack the scar-like stroma. Peripheral palisading and retraction are much less pronounced in both of them and sub clinical involvement is often extensive.

- Another aggressive variant, **micronodular type**, appears as small nodular aggregates of basaloid cells. Retraction artifacts are less common and sub clinical involvement is often significant.

- BCC admixed with SCC is called **basosquamous carcinoma.**

Treatment –

- Small lesions (<1 cm)– surgical excision, laser ablation or electrodissection and curettage, with 5 mm margins of normal appearing skin.
- Large lesions – radical surgery or radiation therapy
- For sclerosing type or recurrent lesions, **mohs micrographic surgery** should be used, which uses frozen section evaluation of specially mapped and marked surgical specimens to determine whether tumor tissue has been left behind.
- Prognosis is good since recurrence uncommon & metastasis very rare. Death if occurs is usually the result of patient's negligence and local invasion into vital structures.

EPIDERMOID CARCINOMA (Squamous cell carcinoma, SCC)

- It is a malignant neoplasm exhibiting squamous differentiation as characterized by the formation of keratin and/or the presence of intercellular bridges.
- The most common malignant neoplasm of the oral cavity.
- Male: female = 2:1 , except for carcinoma of vermilion border of the lower lip, where there is a strong male predominance.
- Mainly found in the elderly, after the fourth decade of life.
- In India, oral cancer ranks first among all cancer cases in males.
- The mortality rate is the lowest for lip cancer and highest for the tongue.

Etiology :

- Tobacco in its various forms, including smokeless tobacco, is regarded as the main cause, especially when coupled with excess alcohol.
- High exposure to UV radiation is a predisposing agent.
- Leukoplakia
- Poor oral hygeine.
- Diets with low levels of vitamin A and C or inadequate consumption of fruits and vegetables may be a contributing factor.
- Patients who are immunosuppressed are predisposed.
- Rare conditions like xeroderma pigmentosum, Fanconi's anemia, and Bloom's syndrome are some other predisposing agents.
- Risk for oral cancer has been shown to increase in the presence of HPV infection.

Clinical features :

Clinically, all oral cancers have two very characteristic features in the form of ulceration and an indurated margin.

Histologic features :

- Most of the lesions are ***moderately well differentiated*** with some evidence of keratinization.

✓ Consists of sheets and nests of cells with obvious origin from squamous epithelium.

✓ Cells are usually large and show a distinct cell membrane, although intercellular bridges often cannot be demonstrated.

✓ Nuclei are large and may demonstrate a good deal of variability in staining intensity.

✓ Mitotic figures may be found, many of which are atypical.

✓ Most prominent features are individual cell keratinization, and formation of numerous epithelial or keratin pearls.

- In ***Less well differentiated tumors*** the resemblance to squamous epithelium is less pronounced. The characteristic shape of the cells may be altered.

✓ The growth rate is more rapid.

✓ Greater numbers of mitotic figures are present.

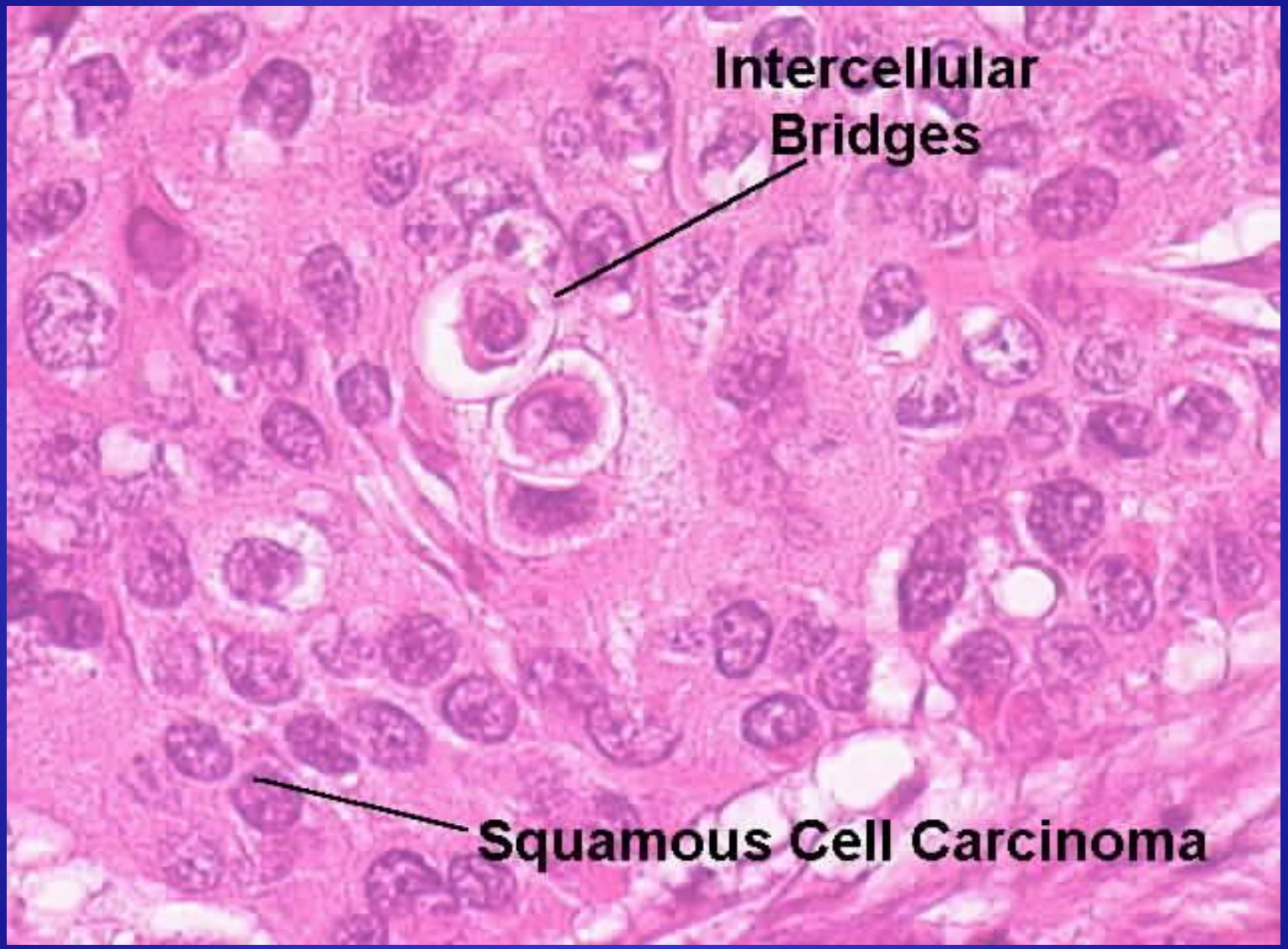
✓ They fail to form keratin.

- The ***poorly differentiated tumors*** bear little resemblance to their cell of origin and will present diagnostic difficulties.

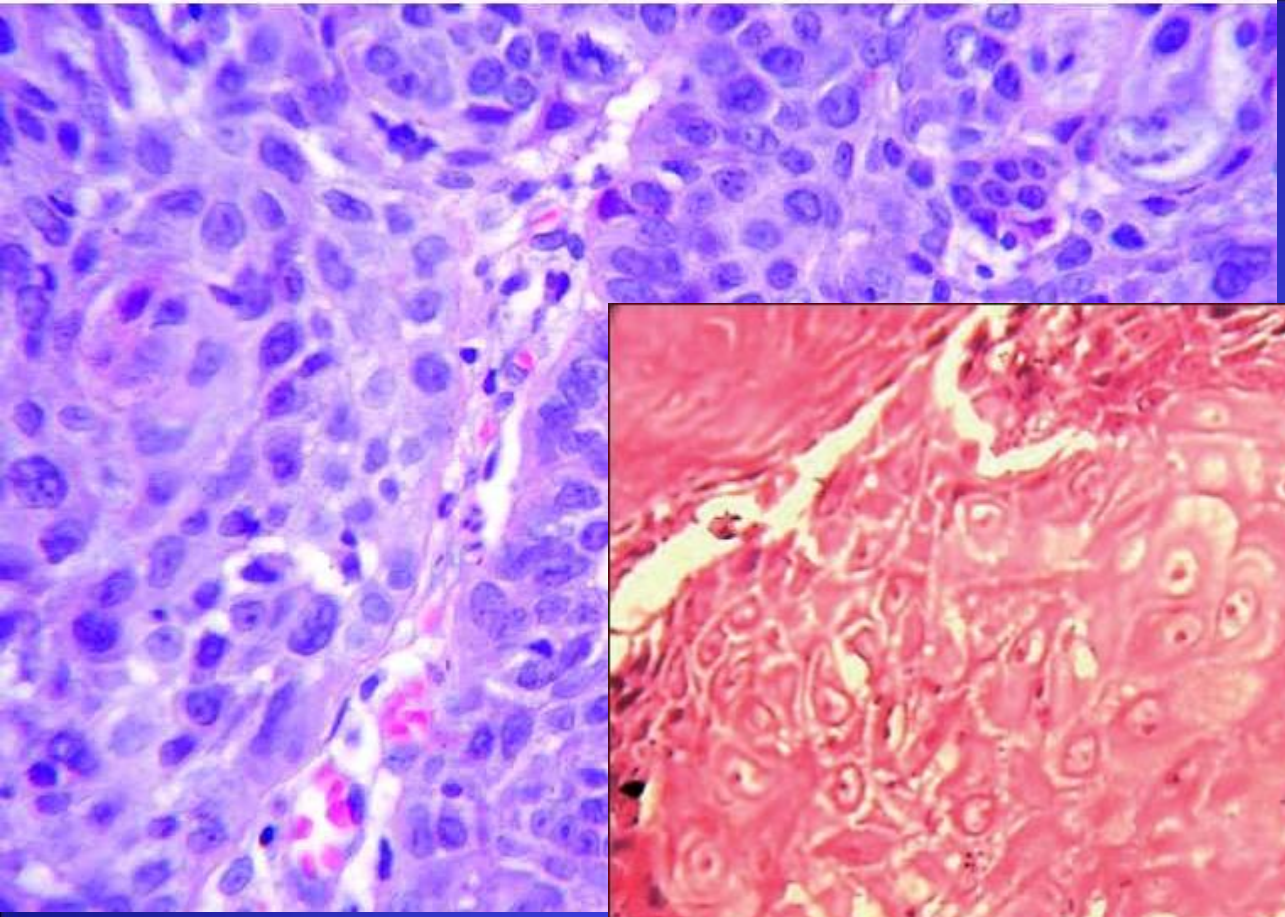
✓ They show an even greater lack of cohesiveness and are extremely vagarious.

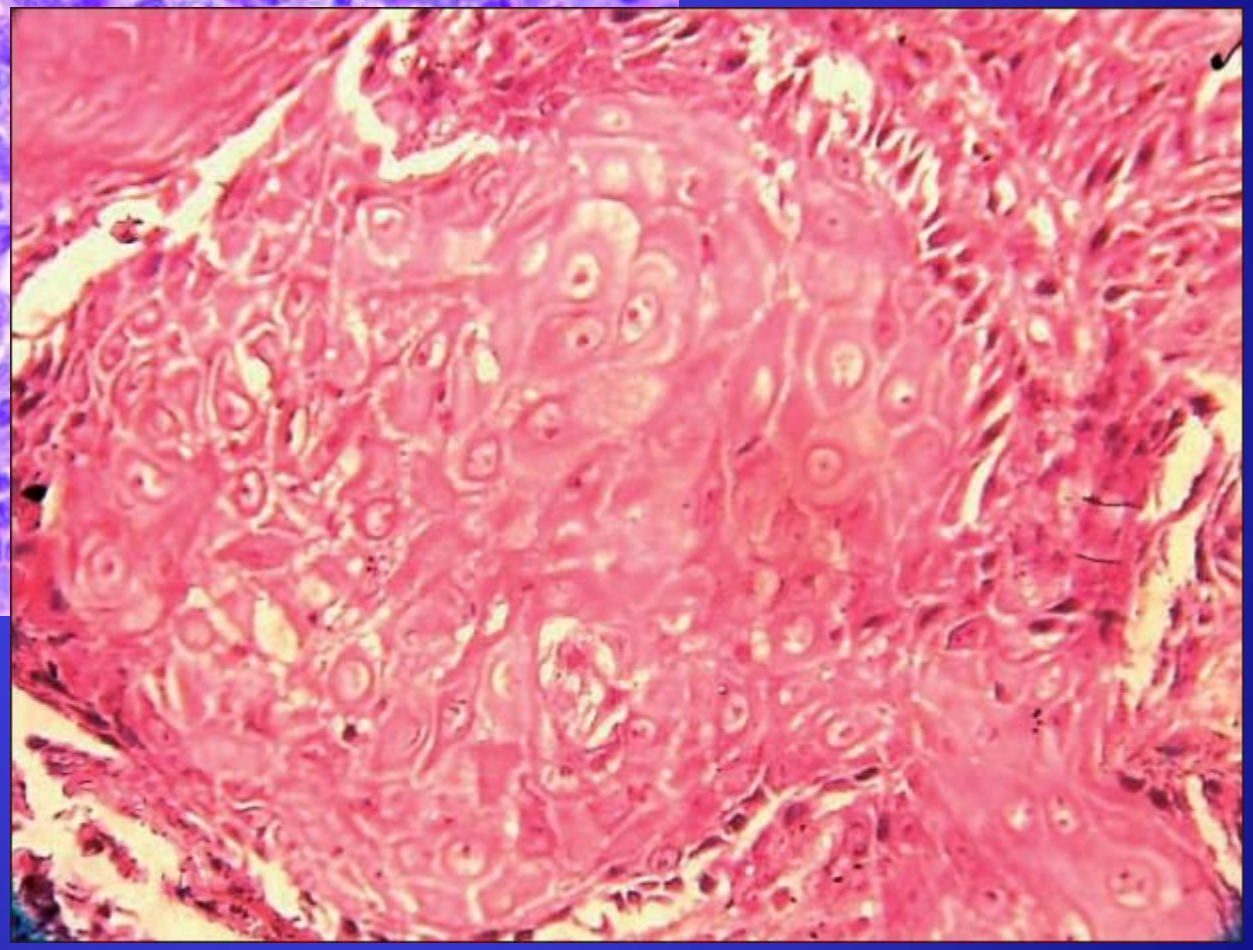
✓ Metastasis involves chiefly the submaxillary and superficial and deep cervical lymph nodes.

✓ Blood stream metastasis is uncommon.

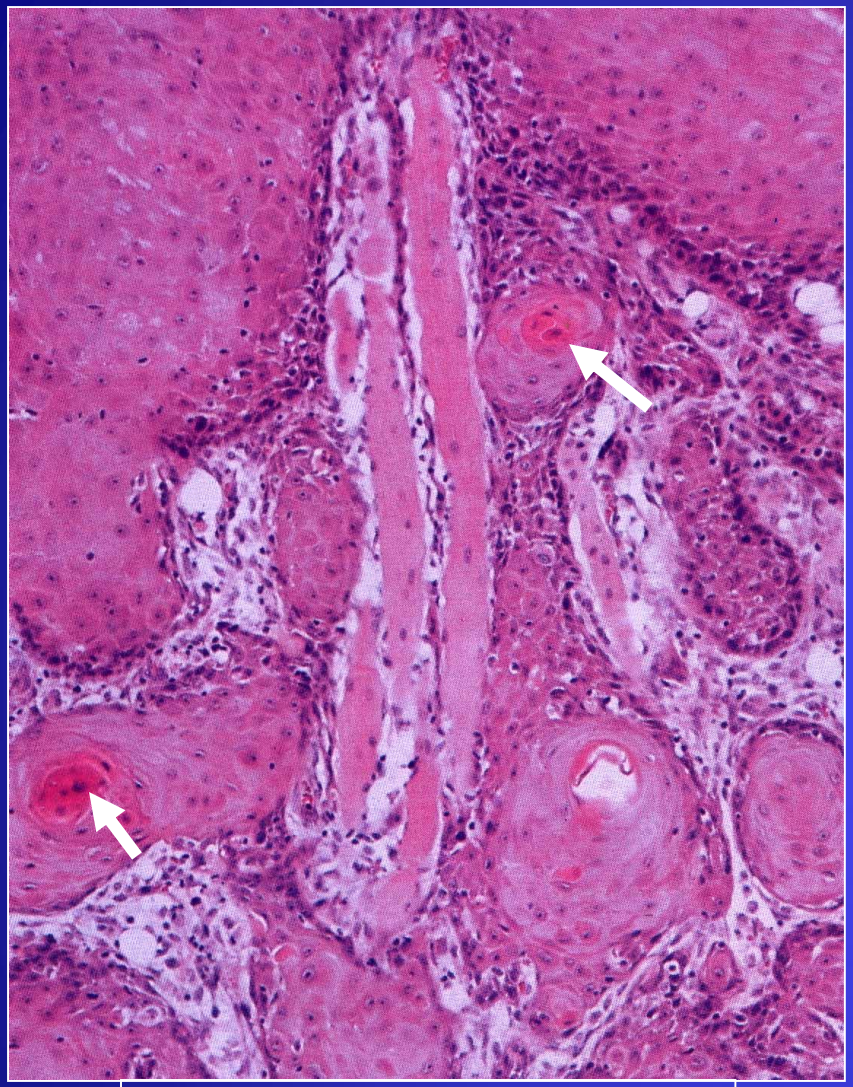


Well Differentiated SCC

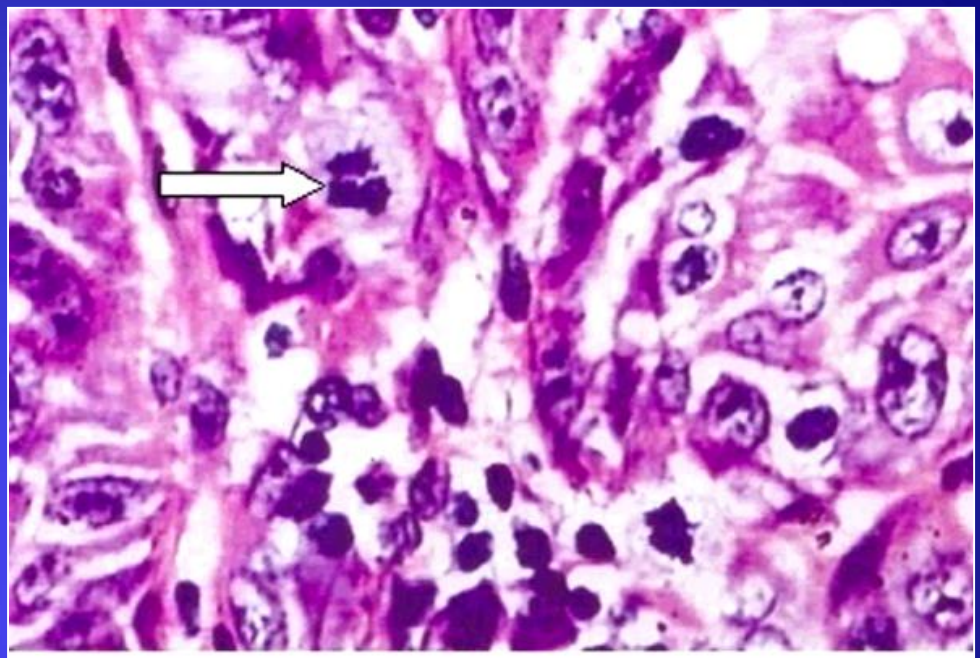




Moderately Differentiated SCC



Arrows show keratin pearls

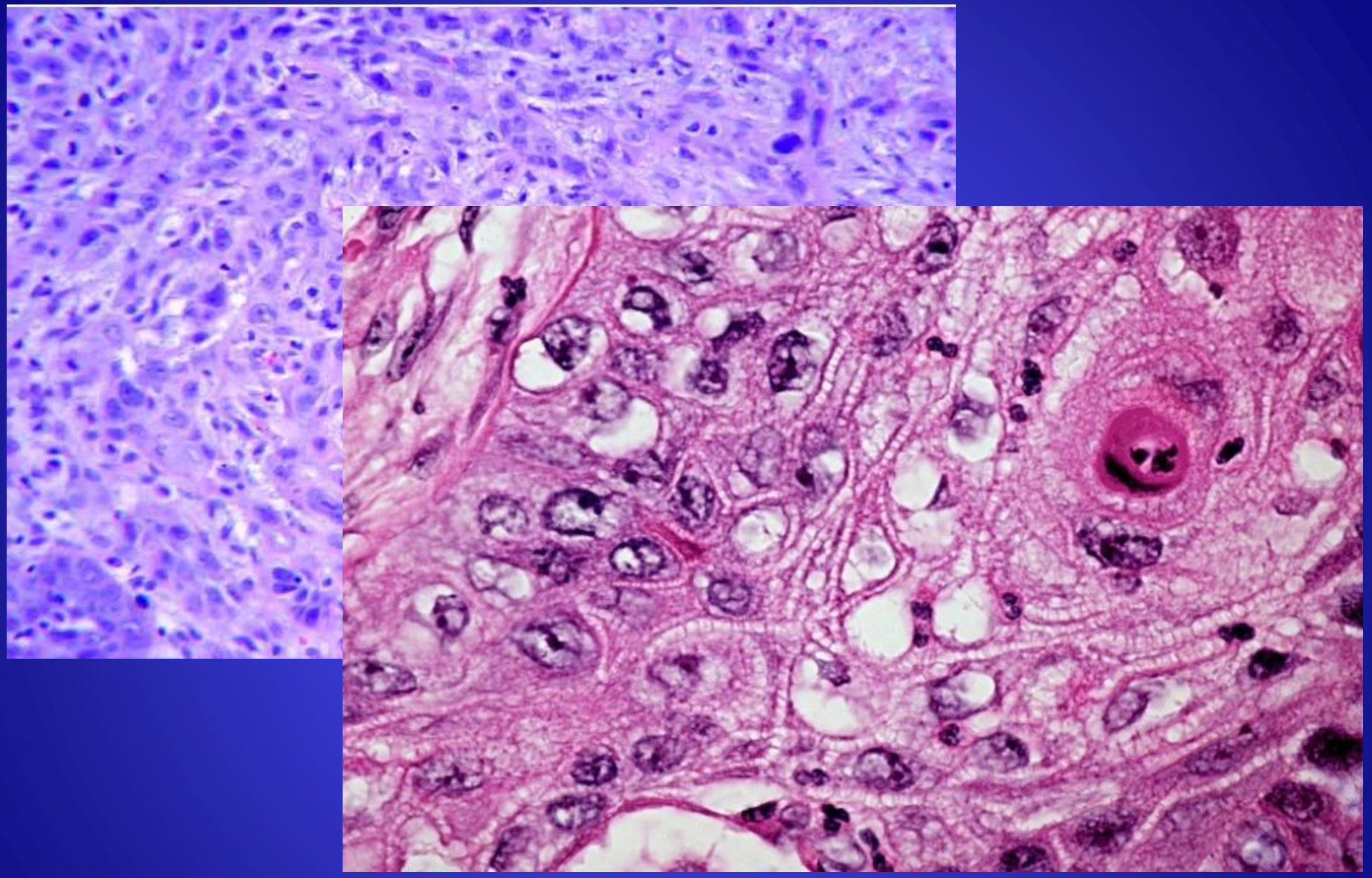


Epidermoid carcinoma

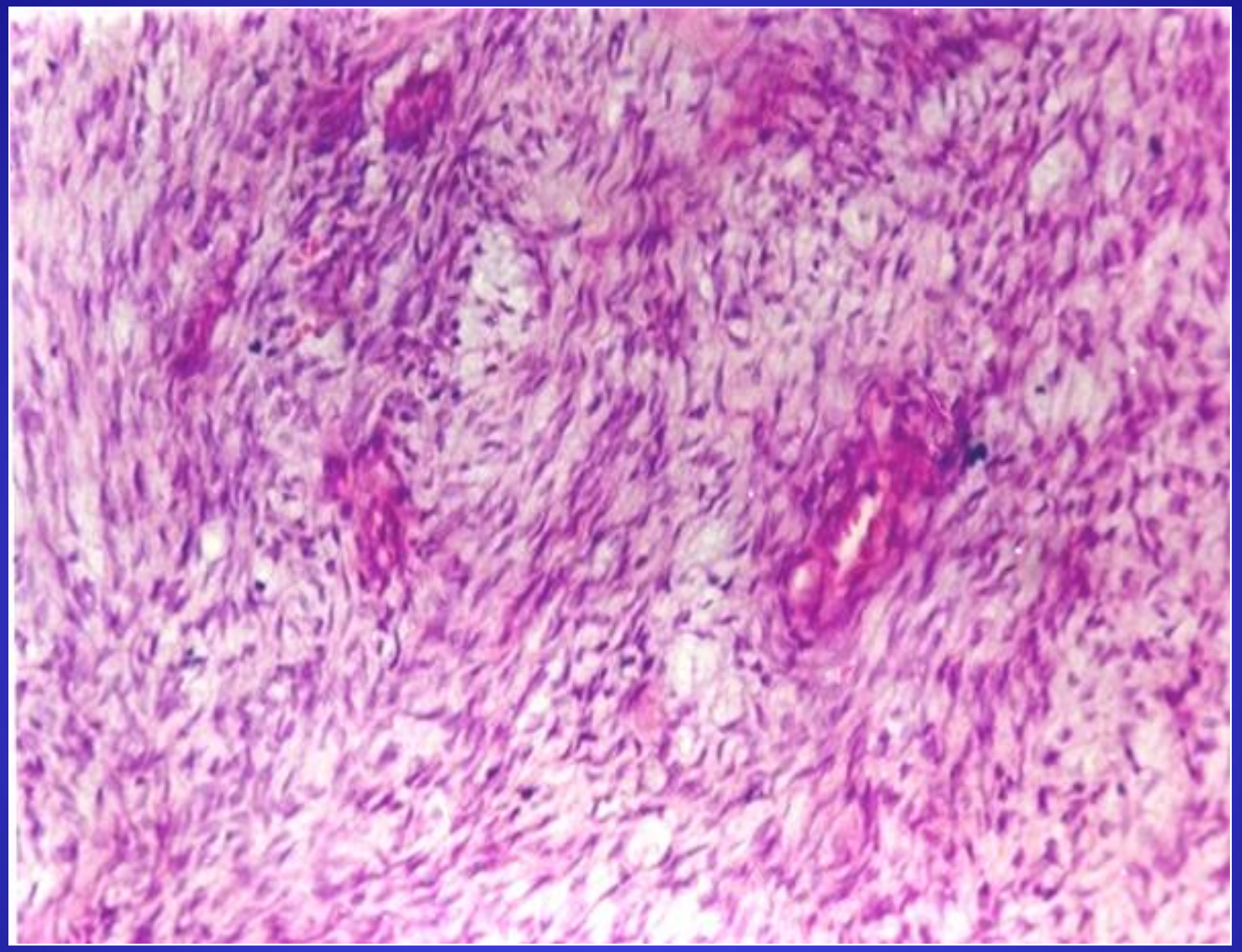
Figure 2-21. Increased number of mitotic figures per field. (H&E stain). p. 149

Arrow shows mitotic figures

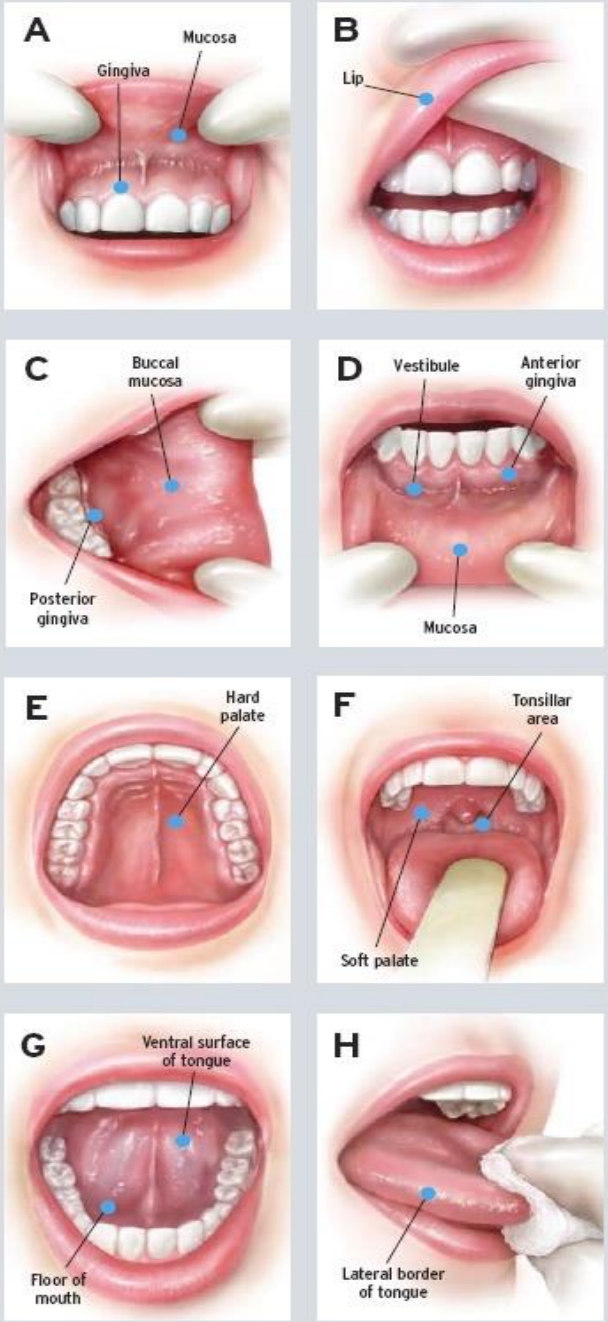
SQUAMOUS CELL CARCINOMA



Poorly Differentiated SCC



Poorly differentiated squamous cell carcinoma showing metaplastic spindle cells.



Here and on the cover: © Krystyna Sroduls ki

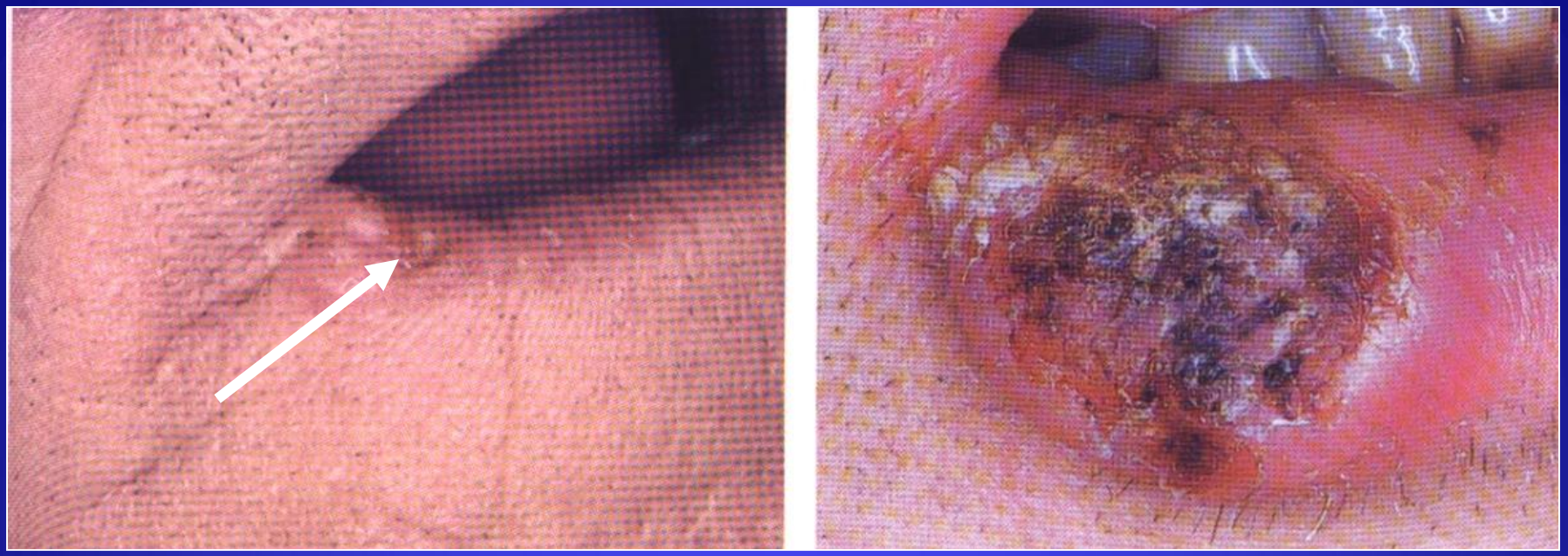
FIGURE 1. A brief screen for oral cancer includes this eight-step examination of the inside of the mouth.

Carcinoma of lip

- Chiefly occurs in the elderly men , in the lower lip.
- One of the most common cause is the use of tobacco, mainly through pipe smoking.

Clinical features :

- Usually begins on the vermilion border of the lip to one side of the midline.
- Often starts as a small area of thickening, induration, and ulceration or irregularity of the surface. As it enlarges, it may create a small crater-like defect or produce an exophytic, proliferative growth.
- Generally slow to metastasize.
- When metastasis does occur, it is usu.ipsilateral and involves the submental or submaxillary nodes. Contra lateral metastasis may occur only if the lesion is near the midline.



Carcinoma lip – (L) early lesion;
(R) Advanced lesion

Histologic features :

- Most lip carcinomas are well differentiated lesions.

Treatment :

- Either surgical excision or x-ray.
- Usually has a good prognosis.

Carcinoma of tongue

- A relationship has been suggested between syphilis and tongue carcinoma but nothing has been proved as yet.

Clinical features :

- It presents as a painless mass or ulcer, which might become painful if secondarily infected.
- May begin as a superficially indurated ulcer with slightly raised borders and may develop into a fungating exophytic mass or infiltrate the deep layers of the tongue, producing fixation and induration.
- Develops on the lateral border or ventral surface of the tongue.
- Lesions near the base of the tongue are esp. insidious, since they may be asymptomatic until far advanced. Even then the presenting feature might be only a sore throat or dysphagia.
- Lesions on the posterior portion are usually of a higher grade of malignancy, metastasize earlier and offer a poorer prognosis, especially because of their inaccessibility for treatment.

SCC of tongue – early to advanced

Treatment and prognosis :

- Treatment is very difficult as no statement can be made about the efficacy of surgery in comparison to that of x-ray radiation.
- Prognosis is not good and basically depends upon the extent of metastasis.

Carcinoma of floor of the mouth

- Smoking, especially pipe or cigar is the most important in its etiology.

Clinical features :

- An indurated ulcer of varying size, situated on one side of the midline.
- More frequently in the anterior portion of the floor. Because of its location, early extension into the lingual mucosa of the mandible, into the mandible proper, into tongue, and even into the sub maxillary and sublingual glands has been recorded.
- Sometimes it may produce limitation of motion of the tongue or slurring of speech.
- Contra lateral metastasis is common as the primary lesion occurs most commonly near the midline, where lymphatic cross-drainage occurs.

SCC of floor of the mouth

Treatment :

- Large lesions are usually not a problem, however, small ones apt to recur
- X-ray radiation and the use of radium give far better results than surgery.
- Prognosis is fair.

Carcinoma of buccal mucosa

- It has a strong predilection of occurrence in men, almost ten times more.

Etiology :

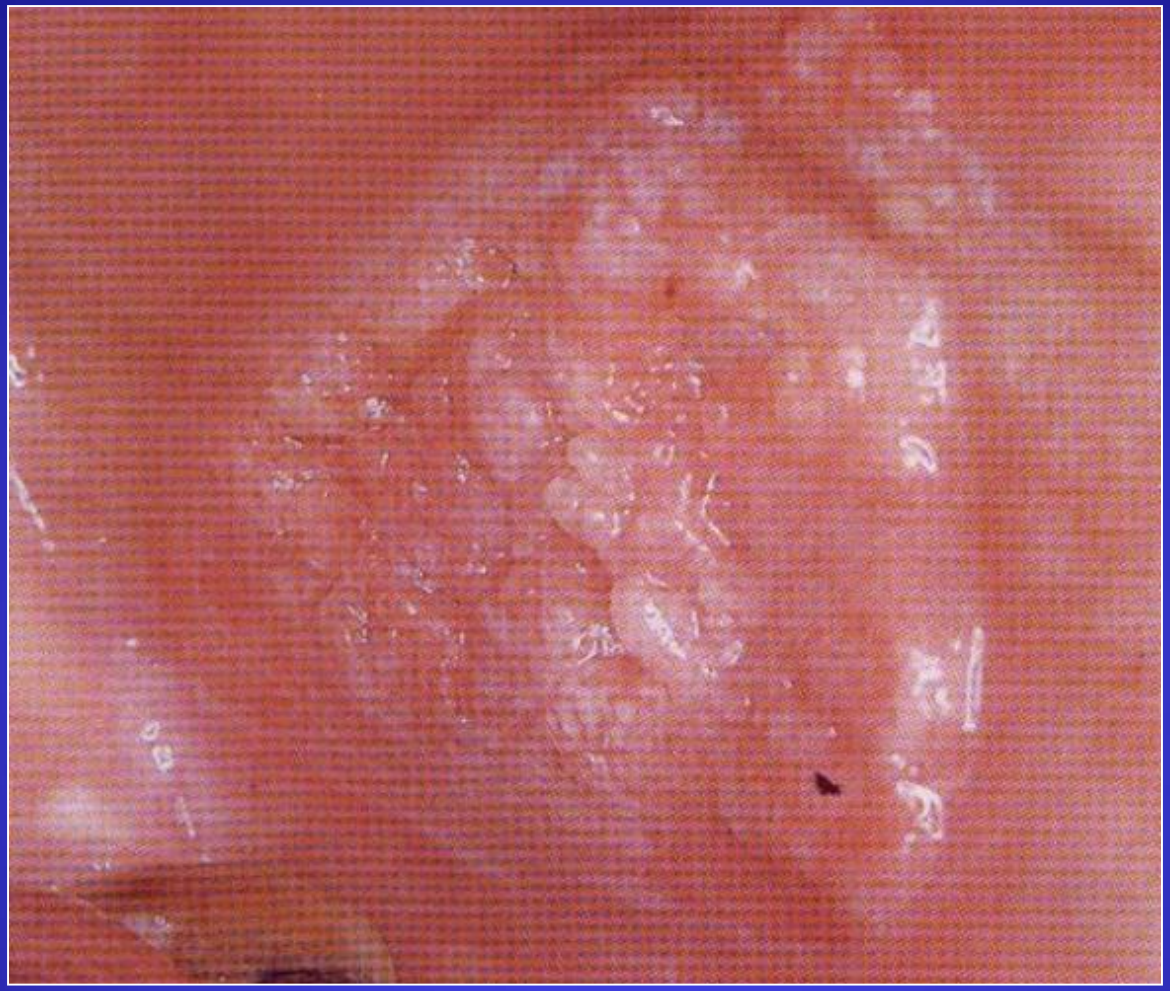
- It has usually been seen to develop in an area against which a person has habitually carried a quid of chewing tobacco for years.

Clinical features :

- Usually develops along or inferior to a line opposite the plane of occlusion.
- Lesion is often a painful ulcerative one where induration and infiltration of deeper tissues is common. Some lesions may even be exophytic.
- Metastasis is very frequent.

Treatment :

- Combined use of surgery or x-ray radiation.



CARCINOMA OF BUCCAL MUCOSA

Carcinoma of gingiva

- Its similarity to common dental infections has frequently led to delay in diagnosis or even to misdiagnosis.

Clinical features :

- More commonly found in the mandibular gingiva.
- Initially presents as an area of ulceration, which may be purely erosive or may exhibit an exophytic growth.
- Arises more commonly in edentulous areas.
- Fixed gingiva is more commonly involved than the free gingiva.
- Erosion of the underlying bone is frequent.
- Metastasis is more common from the mandibular gingiva.

Treatment :

- Since the use of x-rays is very hazardous for bone, its treatment is generally difficult and prognosis bad.

CARCINOMA OF GINGIVA

Carcinoma of the palate

- Not a common lesion of the oral cavity.

Clinical features :

- Poorly defined, ulcerated, painful lesion on one side of the midline. It frequently crosses the midline and may extend laterally to include the lingual gingiva or posteriorly to involve the tonsilar pillar or even the uvula.
- It may invade the bone or occasionally the nasal cavity, while infiltrating lesions of the soft palate may extend into the nasopharynx.
- Metastasis occurs quite commonly.

Treatment :

- Both surgery and x-ray radiation are used.
- Prognosis is not very good.

Epidermoid carcinoma

Figure 2-30. Epidermoid carcinoma of the palate. p. 162

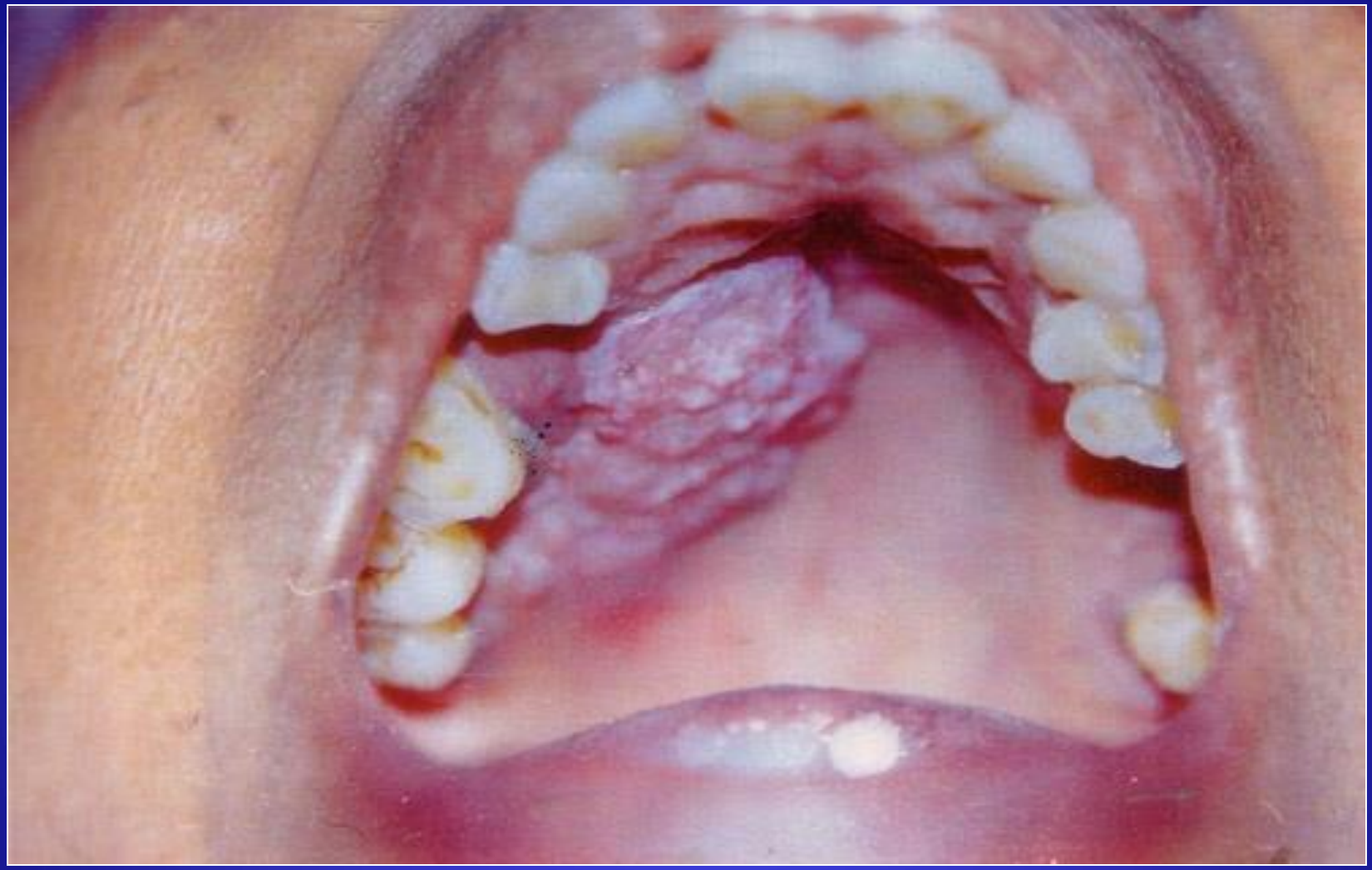
SCC of palate

VERRUCOUS CARCINOMA

- A warty variant of SCC.
- It is a predominantly exophytic overgrowth of well differentiated keratinizing epithelium having minimal atypia, and with locally destructive pushing margins at its interface with c.t.

Clinical features :

- Usually in elderly, on an average between 60-70 years.
- Most commonly on buccal mucosa and gingiva.
- Appears papillary, with a pebbly surface, which is sometimes covered by a white leukoplakic film.



VERRUCOUS CARCINOMA

- Lesions on the gingiva may grow into the soft tissue and invade and destroy the underlying bone.
- Regional lymph nodes are enlarged and tender, simulating metastatic tumor, but this is usually inflammatory.
- Pain and difficulty in mastication are the common complaints.
- The term ORAL FLORID PAPILLOMATOSIS represents the same disease as verrucous carcinoma.
- This disease has a high occurrence rate in tobacco chewers, smokers or snuffers, or in patients having ill-fitting dentures.
- Growth is usually slow and metastasis occurs late, if at all. It may become more aggressive if irradiated.

Histologic features :

- Histology may be extremely deceptive and it may be mistaken for papilloma or benign epithelial hyperplasia
- Generally marked epithelial proliferation with downgrowth of epithelium into the c.t. but usually without true invasion.
- Well differentiated hyperplastic Epithelium is organized into bulbous rete ridges show little mitotic activity, pleomorphism or hyperchromatism.
- Characteristically, cleft like spaces lined by a thick layer of parakeratin extend from the surface deeply into the lesion.Parakeratin plugging also occurs extending into the epithelium. Both these features together constitute the hallmark of verrucous carcinoma.
- Basement membrane is generally intact and usually a heavy inflammatory infiltrate is present in the c.t.

- Areas of verrucous hyperplasia, which describes an exophytic overgrowth of well differentiated keratinizing epithelium that is similar to verrucous carcinoma but without the destructive pushing border at its interface with the c.t., may be seen in association with verrucous carcinoma or SCC.

Treatment :

- Conservative excision
- Risk of anaplastic transformation if irradiated.

VERRUCOUS CARCINOMA

SPINDLE CELL CARCINOMA (carcinosarcoma, pseudosarcoma, lane tumor)

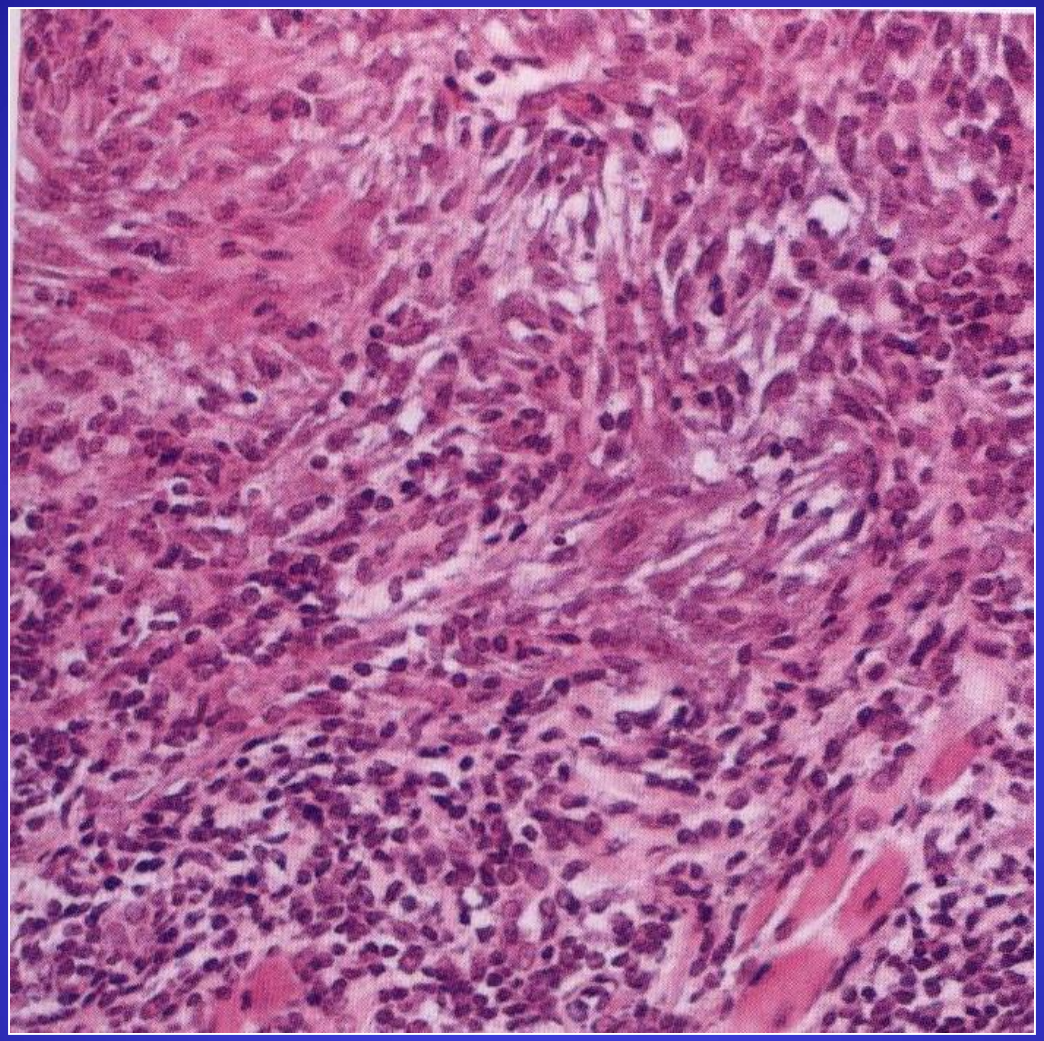
- In a true spindle cell carcinoma, the malignant spindle cells should be of epithelial origin and derived from the squamous cell component of the carcinoma.
- It must be distinguished from SCC that has provoked a reactive fibroblastic proliferation & from a carcinoma in which SCC is accompanied by a sarcoma of fibroblastic type.
- Often the SCC component is inconspicuous & multiple sections or blocks may be needed.

Clinical features :

- Predilection for occurrence in males
- Usually found in lower lip, tongue and gingiva.
- Lesion appears as polypoid, exophytic or endophytic & usually presents with swelling, pain and a non healing ulcer.

Histologic features :

- It is a bimorphic or biphasic tumor, which, although almost ulcerated, will show foci of surface SCC or epithelial dysplasia of surface mucosa, usually just at the periphery.
- Proliferation and 'dropping off' of basal cells to spindle cell elements is a common phenomenon.
- The bulk of the tumor is either fasciculated, myxomatous or streaming in the fasciculated type, elongated cells with elliptical nuclei and pleomorphic cells are found.
- Giant cells and bizarre atypical cells may also be found.
- An inflammatory cell infiltration is often present.



SPINDLE CELL CARCINOMA

Tumor shows a fasciculated pattern with elongated cells.

Treatment :

Surgical removal, with or without radical neck dissection, alone or in combination with radiation.

ADENOID SQUAMOUS CELL CARCINOMA (adenoacanthoma,pseudoglandular SCC)

- A squamous cell carcinoma containing pseudoglandular spaces
- It is produced as a result of acantholysis and degeneration within islands of SCC, but there is no evidence of glandular differentiation or secretory activity.

Clinical features :

- Predilection for occurrence in males
- Usually found in lower lip, tongue and gingiva.
- Lesion appears as polypoid, exophytic or endophytic & usually presents with swelling, pain and a non healing ulcer.

Histologic features :

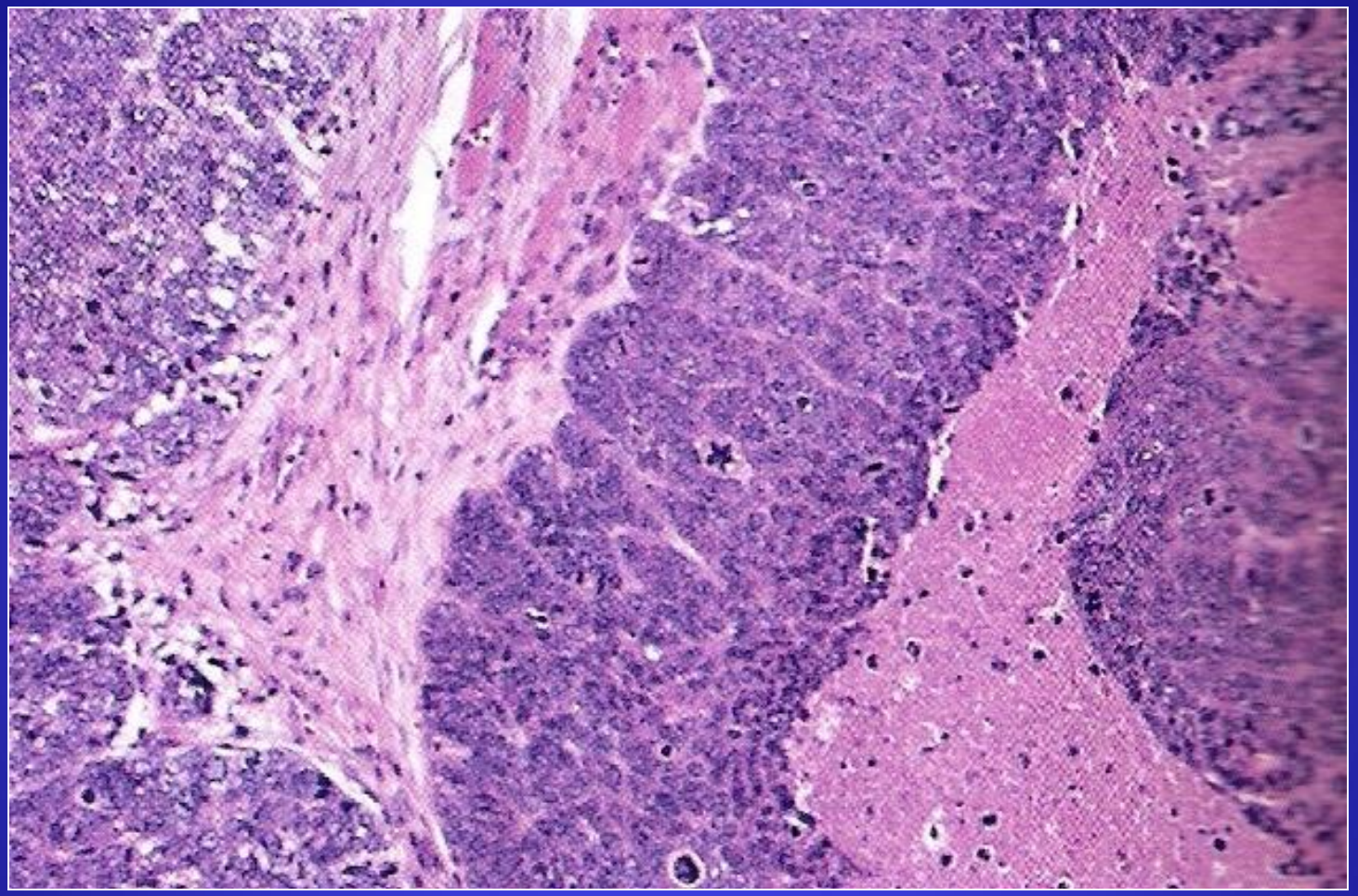
- Surface dysplastic epithelium proliferates into the c.t., as in SCC, but the deep extensions show the characteristic solid and tubular ductal structures.
- These duct-like structures are lined by a layer of cuboidal cells and often contain or enclose acantholytic or dyskeratotic cells.

Treatment :

- Surgical excision.
- Recurrence is common but metastasis is very rare.

BASALOID SQUAMOUS CELL CARCINOMA

- A carcinoma with a mixed composition of basaloid and squamous cells.
- The basaloid component comprises small cells with hyperchromatic nuclei and scant cytoplasm that are crowded together into lobulated sheets or strands, focally connected to the surface epithelium.
- Cells at the periphery of the lobules are often palisaded; more centrally there may be cystic spaces.
- Mitotic figures and areas of necrosis are commonly seen.
- Confusion with ameloblastoma and adenoid cystic carcinoma is to be avoided; a focal squamous cell carcinoma component among the basaloid areas is the most important diagnostic feature.



BASALOID SQUAMOUS CELL CARCINOMA : sheets of basaloid squamous epithelium exhibiting a high mitotic index and tumor necrosis.

ADENOSQUAMOUS CARCINOMA

- A malignant tumor with histological features of both adenocarcinoma and SCC.
- May arise from the ducts of minor salivary glands or from the overlying surface epithelium.

UNDIFFERENTIATED CARCINOMA

A carcinoma that lacks evidence of squamous, glandular or other types of differentiation.

LYMPHOEPITHELIOMA AND TRANSITIONAL CELL CARCINOMA

- An unusual group of malignant neoplasms, which arise from the mucosa of nasopharynx, oropharynx, tongue, tonsil, nasal chamber and paranasal sinuses.
- They exhibit a relatively specific histologic pattern and react in a rather atypical fashion to x-ray radiation.
- This group consists of Lymphoepithelioma, transitional cell carcinoma and undifferentiated SCC.

Clinical features :

- The primary lesion is usually very small, completely hidden, slightly elevated, indurated, frankly ulcerated or presenting a granular, eroded surface.
- Lymphoepithelioma occurs chiefly in the nasopharynx and is such a small lesion that it does not manifest itself clinically before regional lymphadenopathy is apparent. Death is the frequent outcome, though it is radiosensitive.
- Transitional cell carcinoma arises chiefly from the tonsil, base of the tongue and nasopharynx. It is extremely malignant, running a rapid course, metastasizing rapidly and causing very early death.
- Regional lymphadenopathy was the most common presenting symptom followed by sore throat, nasal obstruction, defective hearing, ear pain, headache, dysphagia or epistaxis.
- Average age of patients with lymphoepithelioma was 44 yrs, with transitional cell carcinoma 26 yrs, with undifferentiated carcinoma 56 yrs.

Histologic features :

- Transitional cell carcinoma Consists of cells growing in solid sheets or in cords and nests.
- Cells are moderately large, with lightly basophilic cytoplasm and indistinct cell outlines, with large and round nuclei, showing varying degrees of mitotic activity.
- Stroma exhibits little or no lymphocytic infiltration.
- Lymphoepithelioma consists of cells growing in a syncytial pattern with stroma infiltrated by varying numbers of lymphocytes.
- Cells are large and polyhedral with indistinct outlines, lightly eosinophilic cytoplasm, large oval and vesicular nuclei containing one or two large eosinophilic nucleoli.

Treatment :

- X-ray radiation is the most accepted treatment.
- The complicating factor lies in the relative inability to treat the widespread metastases in the various organs, and this imparts an unfavorable prognosis.

MALIGNANT MELANOMA

- A neoplasm of epidermal melanocytes.
- One of the most biologically unpredictable and deadly of all human neoplasias.
- The **third most common cancer of skin** (after BCC and SCC).
- Earlier it was believed that melanomas develop in nevi, especially junctional nevi. But now it is thought that the lesions which were interpreted as junctional nevi were, in fact, premalignant melanocytic dysplasias of some type.
- Certain lesions considered to be premalignant melanomas are the acquired nevus, dysplastic nevus, congenital nevus and cellular blue nevus.
- Melanomas may also arise de novo.

Etiological factors :

Environmental factors :

- Sun exposure
- Artificial UV sources
- Socioeconomic status
- Fair skin, freckles, red hair
- No. of melanocytic nevi

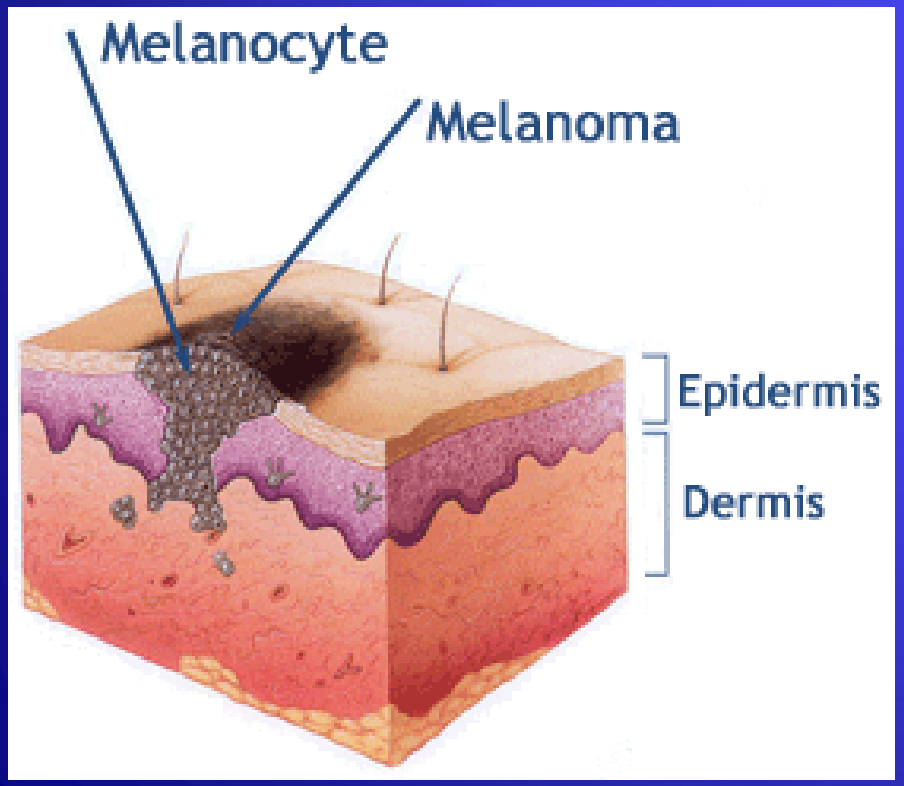
Genetic factors :

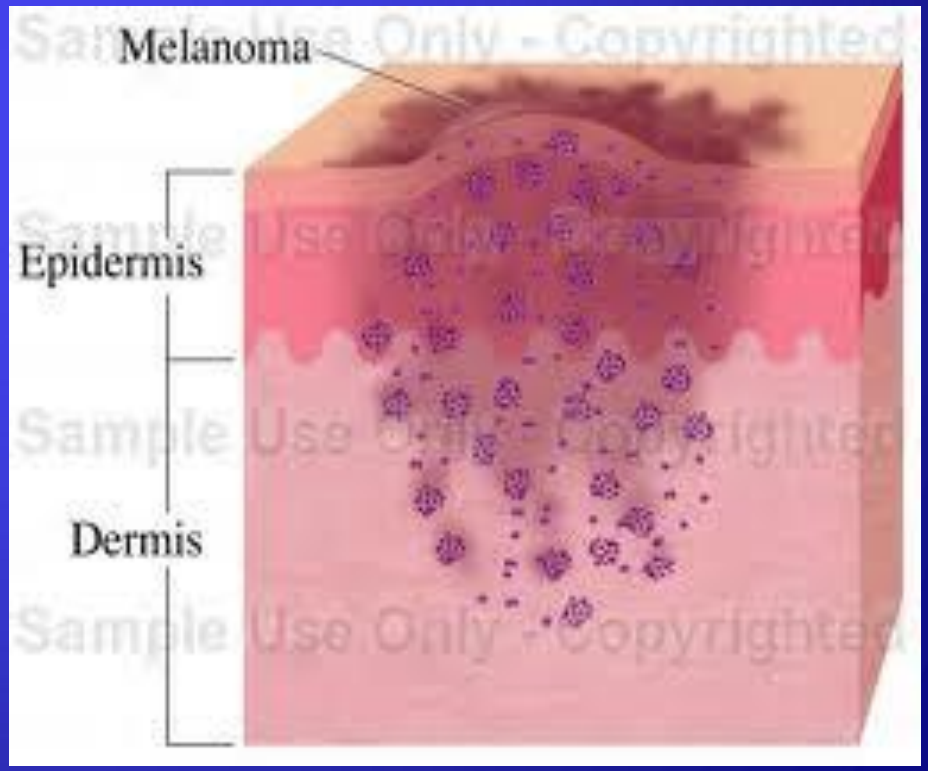
- Familial melanoma
- Xeroderma pigmentosum

Risk factors for oral melanomas are unknown. They have no apparent relationship to chemical, thermal or physical events to which the oral mucosa is continuously exposed. Most of these are thought to arise de novo.

There are two phases in the growth of a melanoma :

- The ***radial growth phase*** is the initial phase which may last for many years and the neoplastic process is confined to the epithelium.
- The ***vertical growth phase*** begins when the neoplastic cells populate the underlying c.t. In this phase, metastasis is possible.





Cutaneous melanomas have been classified into :

Superficial spreading melanoma
Nodular melanoma
Lentigo maligna melanoma
Acral lentiginous melanoma
Mucosal lentiginous melanoma

Clinical features :

SUPERFICIAL SPREADING MELANOMA (SSM)

- Most common cutaneous melanoma in caucasians.
- Exists in a radial growth phase called premalignant melanosis or pagetoid melanoma in situ.
- Presents as a tan, brown, black or admixed lesion on sun-exposed skin, esp. the back.
- Vertical growth phase is characterized by an increase in size, change in color, nodularity and ulceration.

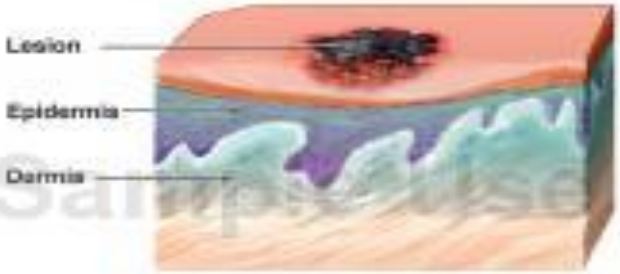
NODULAR MELANOMA (NM)

•No clinically recognizable radial-growth phase, existing solely in a vertical-growth phase.

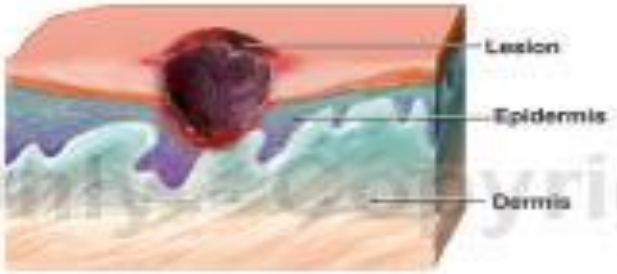
•Presents as a sharply delineated nodule with degrees of pigmentation, may be pink (Amelanotic melanoma – when melanoma cells are poorly differentiated that they no longer can produce melanin) or black.

•Predilection for occurrence on the back and head and neck skin of men.

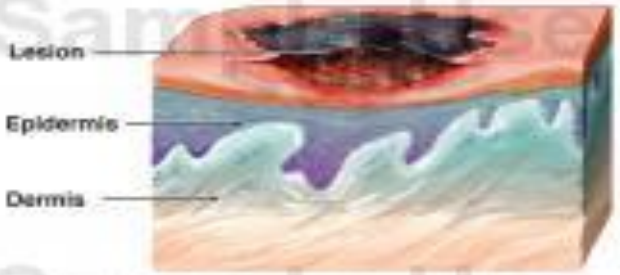
Superficial Spreading Melanoma vs. Nodular Melanoma



Early stage - Small irregular mass which is confined to the epidermis

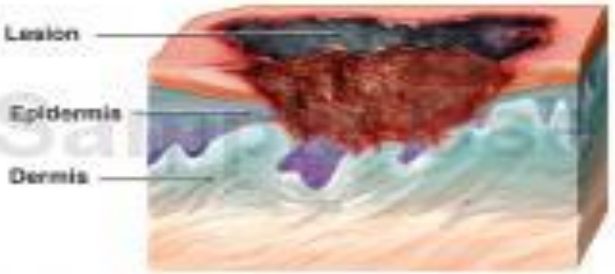


Early stage - Small initial lesion nearly identical in vertical and radical dimensions. It already involves the dermis.

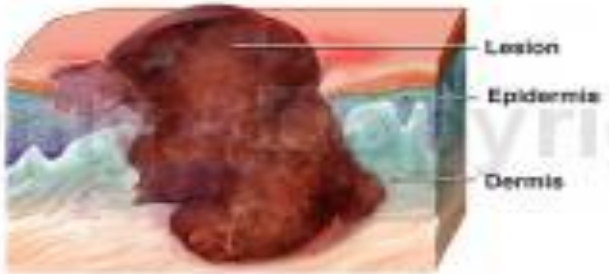


Significant increase in radial diameter of lesion but no significant change to vertical dimension

Aggressive vertical growth into the dermis without significant increase in radial dimension



Significant increase in radial diameter of lesion but only minimal vertical dimension increase



Aggressive vertical growth

LENTIGO MALIGNA MELANOMA (LMM)

- Exists in a radial growth phase which is known as lentigo maligna (melanoma in situ) or melanotic freckle of Hutchinson.
- Occurs characteristically as a macular lesion on the sun exposed skin mainly malar skin of a middle aged and elderly caucasian, more common in women.
- It has pure radial growth phase.
- Nearly 53% of the lesions evolve into Lentigo maligna melanoma , the vertical growth phase of this type.

ACRAL LENTIGINOUS MELANOMA

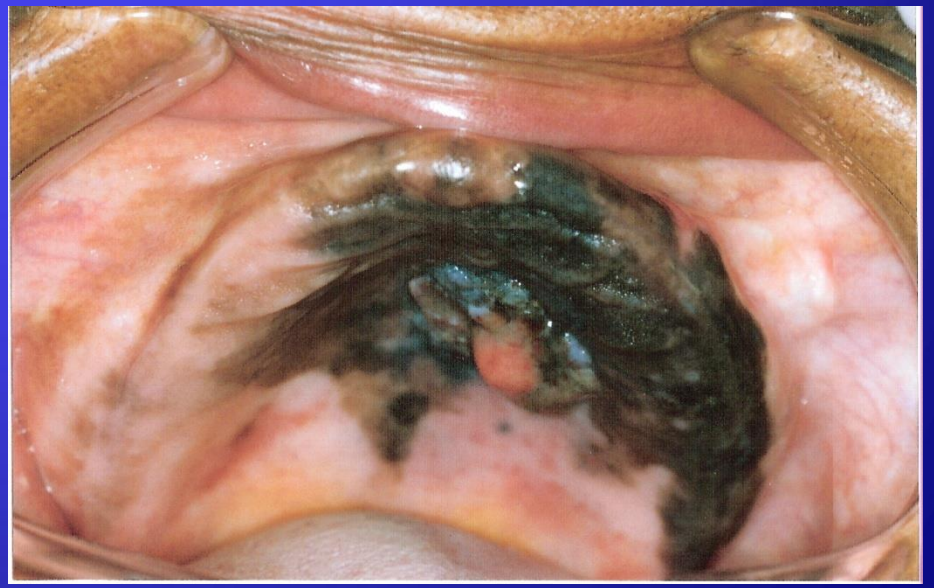
- Develops on the palms and soles, as well as on toes and fingers.
- Characterized by a macular lentiginous pigmented area around a nodule.
- They are extremely aggressive, with rapid progression from the radial to vertical growth phase.

MUCOSAL LENTIGINOUS MELANOMA

- Develop from mucosal epithelium that lines the respiratory, gastrointestinal, and genitourinary systems.

- May occur on any mucosal surface, including conjunctiva ,oral cavity, esophagus, vagina, female urethra, penis and anus.

- Most commonly found in patients of advanced age and follows a very aggressive course.

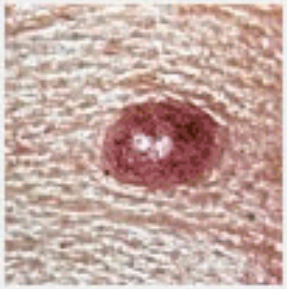
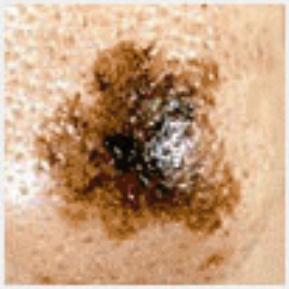


AMELANOTIC MELANOMA

- Presents as an erythematous or pink, Sometimes eroded nodule.
- Only histologic examination can provide the right diagnosis.

The following criteria aid in the clinical diagnosis of melanoma (ABCDE - rule) :

- ASYMMETRY
- BORDER IRREGULARITY – with blurred, notched or ragged edges.
- COLOR IRREGULARITY – pigmentation is not uniform.black, brown, red, tan, white and blue can all appear together.
- DIAMETER – greater than 6 mm. growth in itself is a sign.
- ELEVATION

Normal Mole	Melanoma	Sign	Characteristic
		Asymmetry	when half of the mole does not match the other half
		Border	when the border (edges) of the mole are ragged or irregular
		Color	when the color of the mole varies throughout
		Diameter	if the mole's diameter is larger than a pencil's eraser

Photographs Used By Permission: National Cancer Institute

Asymmetry

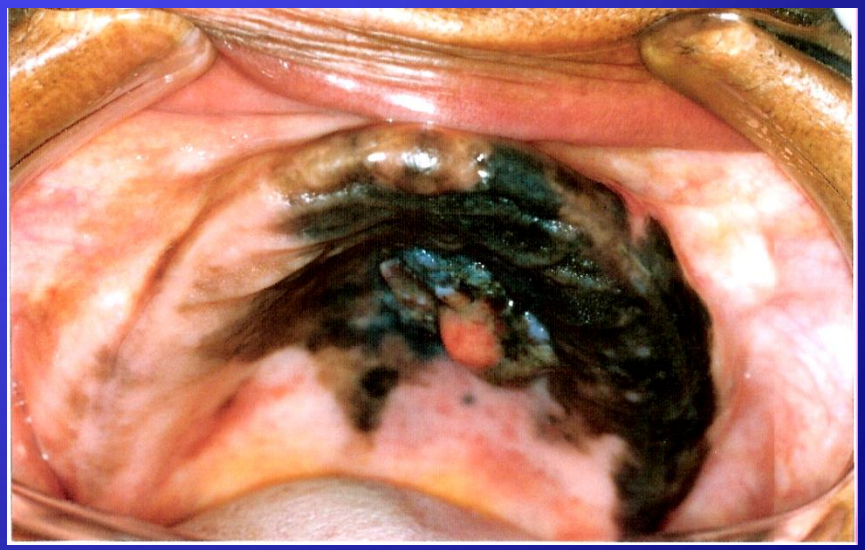
Border irregularity

Color

Diameter: ¼ inch or 6mm

Oral manifestations :

- Twice as common in men than in women.
- Most cases occur between 40 and 70 years.
- Predilection for the palate and maxillary gingiva.
- Usually appears as a deeply pigmented area, at times ulcerated and hemorrhagic, which tends to increase progressively in size.
- Early lesions are macular but with time they may assume nodular growth pattern.
- Oral melanomas exist in superficial spreading, acral lentiginous and nodular types.



Histologic features :

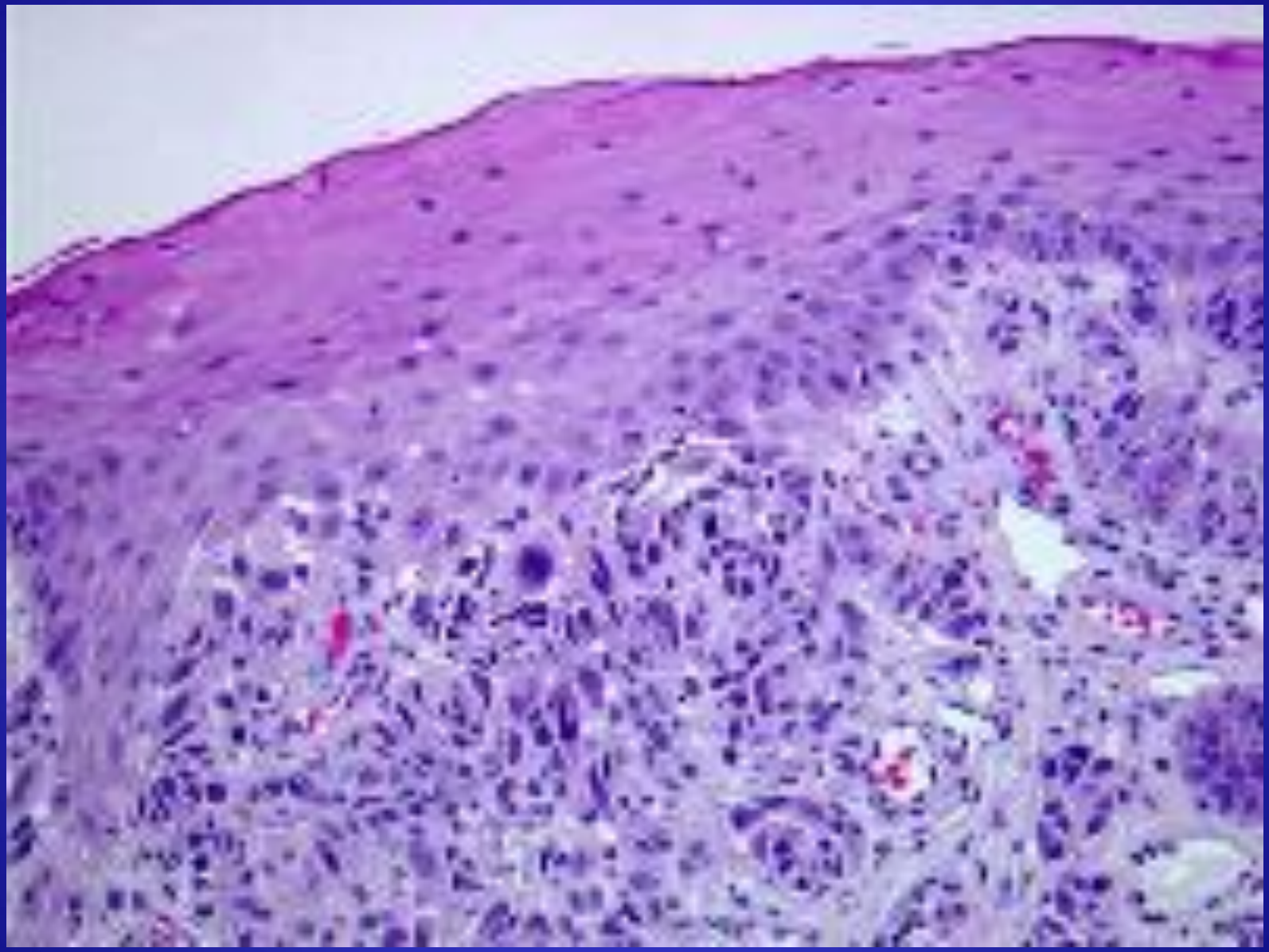
- Malignant cells often nest or cluster in groups in an organoid fashion; however, single cells can predominate.

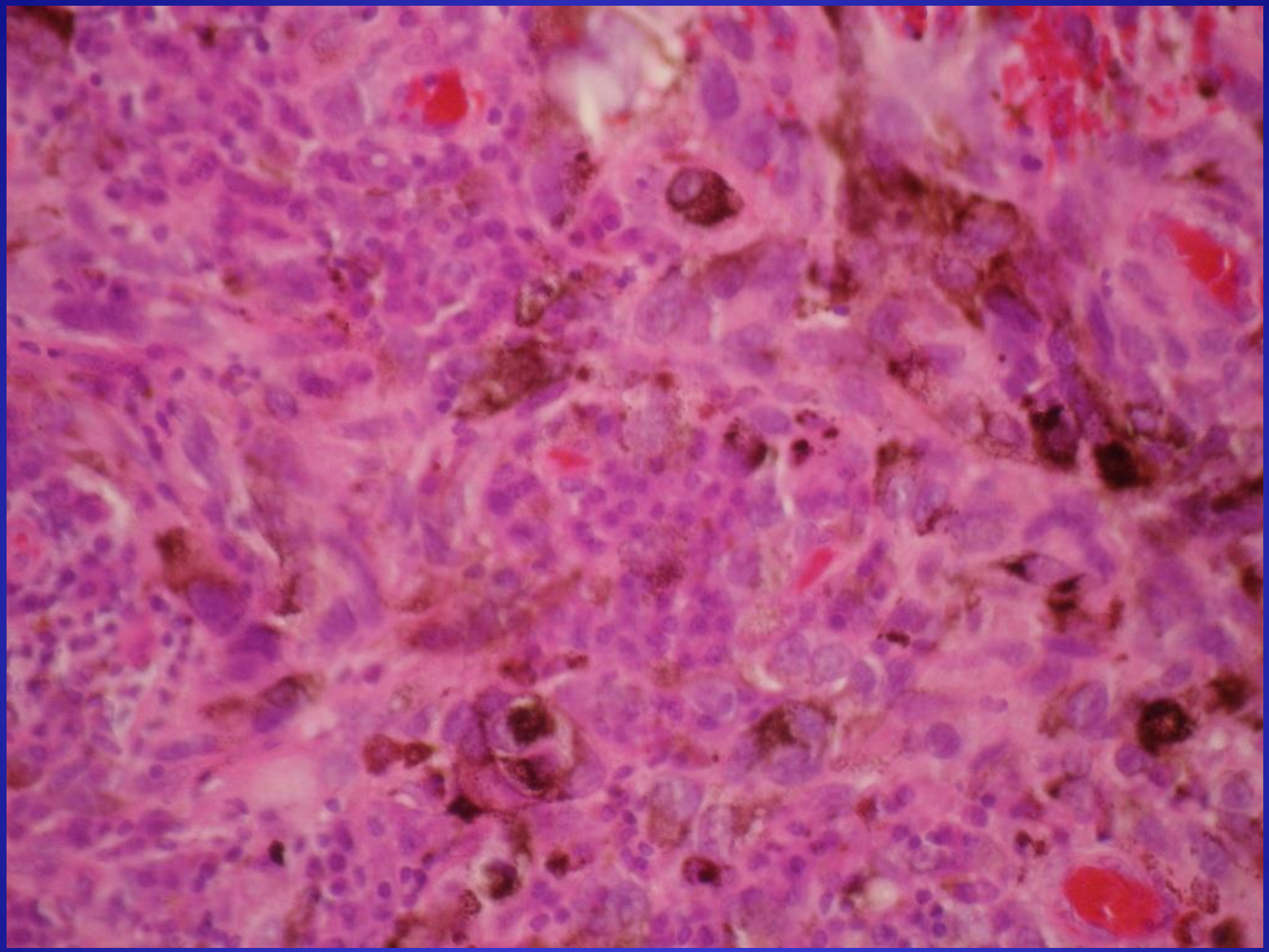
- They have large nuclei, prominent nucleoli, and show nuclear pseudoinclusions due to nuclear membrane irregularity.

- Radial growth phase of **superficial spreading melanoma** is characterized by the presence of large epithelioid melanocytes distributed in a so called pagetoid manner(upward spreading-infiltrating suface epithelium singly or in nests), sometimes known as **'buckshot scatter'(**Proliferation of cells within the epidermis in a random pattern of distribution giving a 'buckshot' scatter pattern, describes random appearance of melanoma cells in the epidermis).

- When melanocytes penetrate the basement membrane, a florid host cell response develops which destroys the tumor cells.

- The vertical growth phase is characterized by the proliferation of malignant cells in the dermis. Melanin pigment is usually scanty.



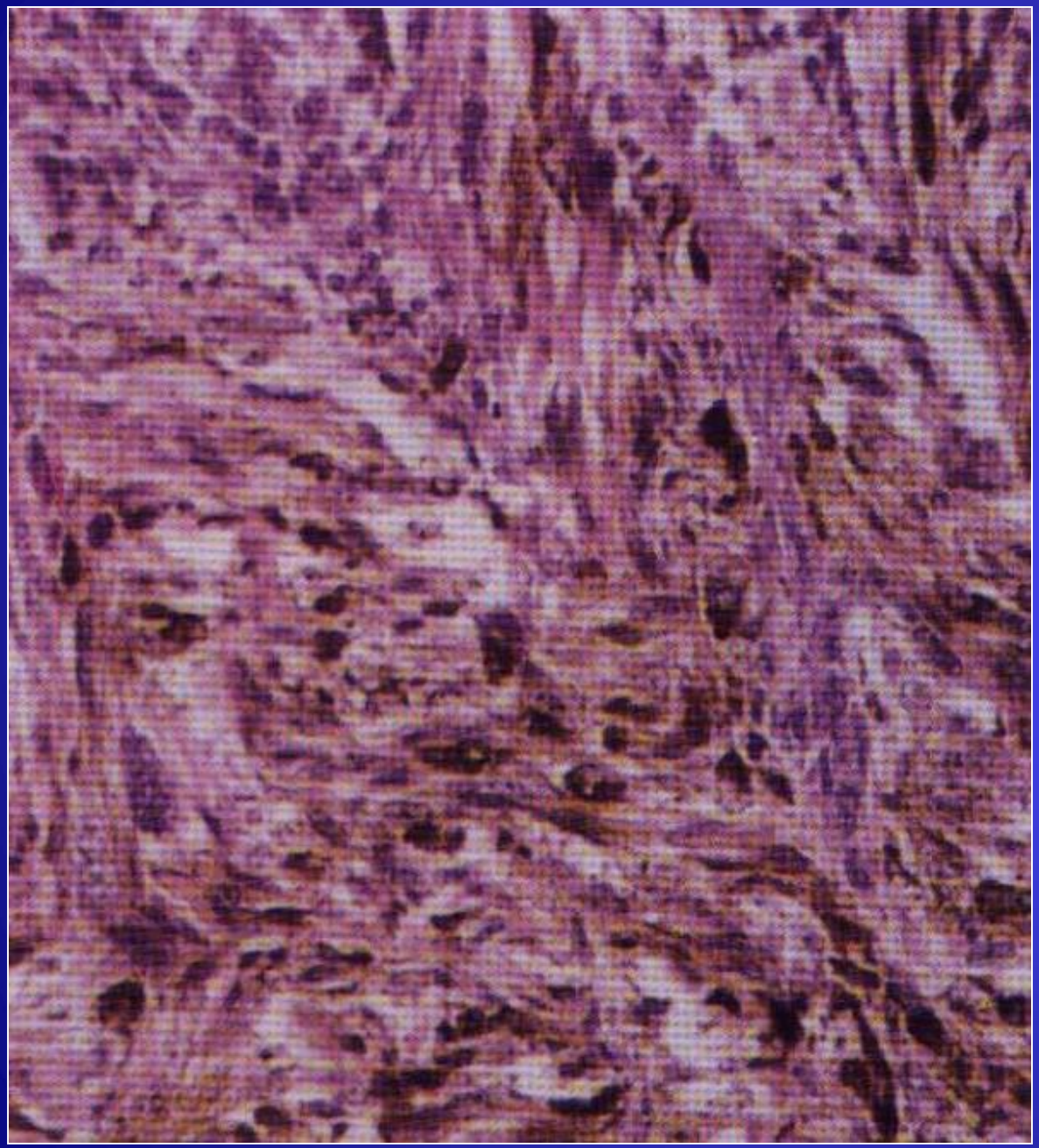


Histopathology of oral melanoma

- **Nodular melanoma** is also characterized by large epithelioid melanocytes within the c.t.

✓ Melanin pigment is usually but not invariably present.

✓ Tumor cells may invade and ulcerate the overlying epithelium and penetrate the deep soft tissues.

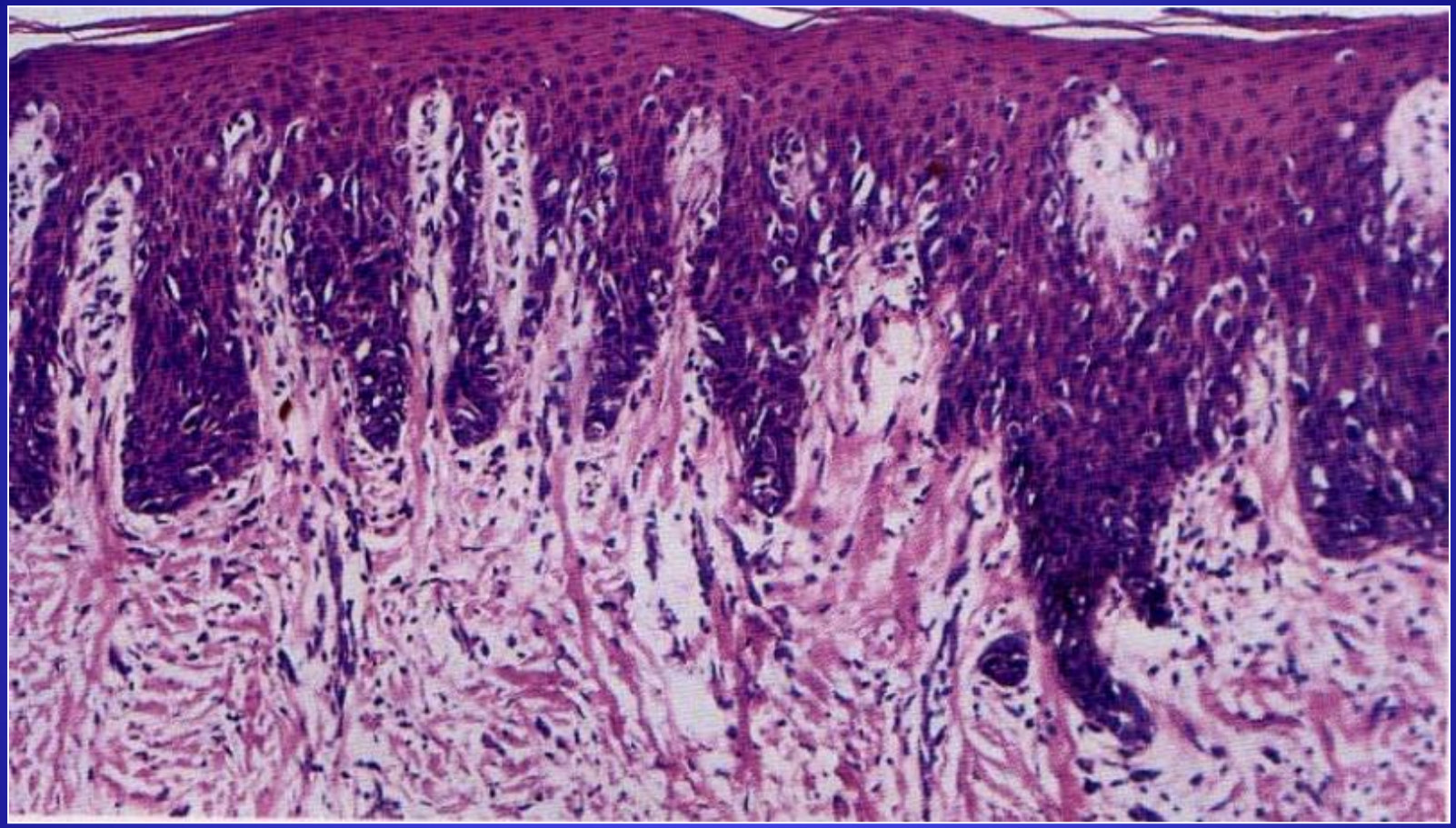


Nodular melanoma

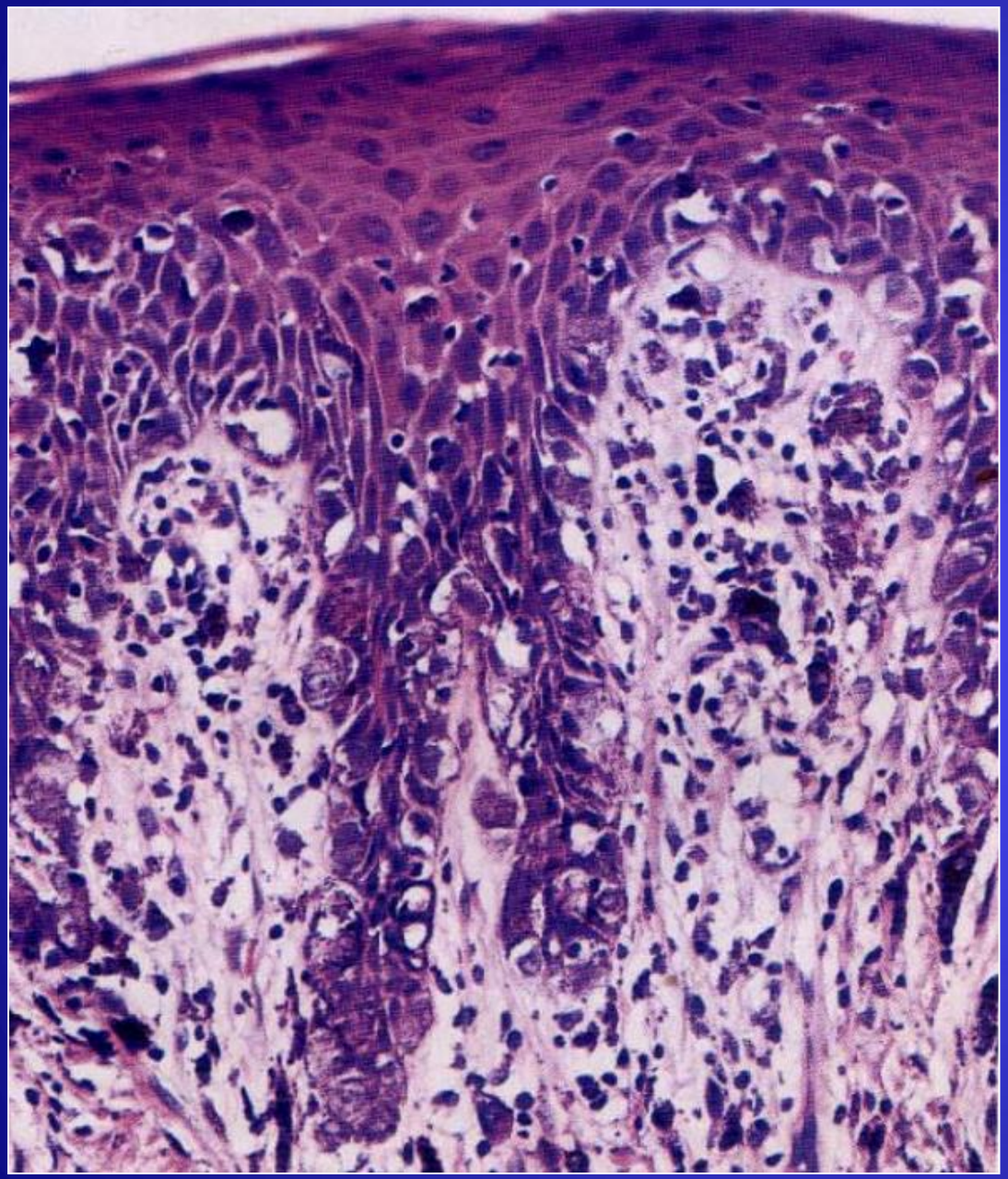
- **Lentigo maligna** is characterized by increased numbers of atypical melanocytes within the basal epithelial layer.

✓ Epithelium is generally atrophic and the dermal collagen shows basophilic degeneration.

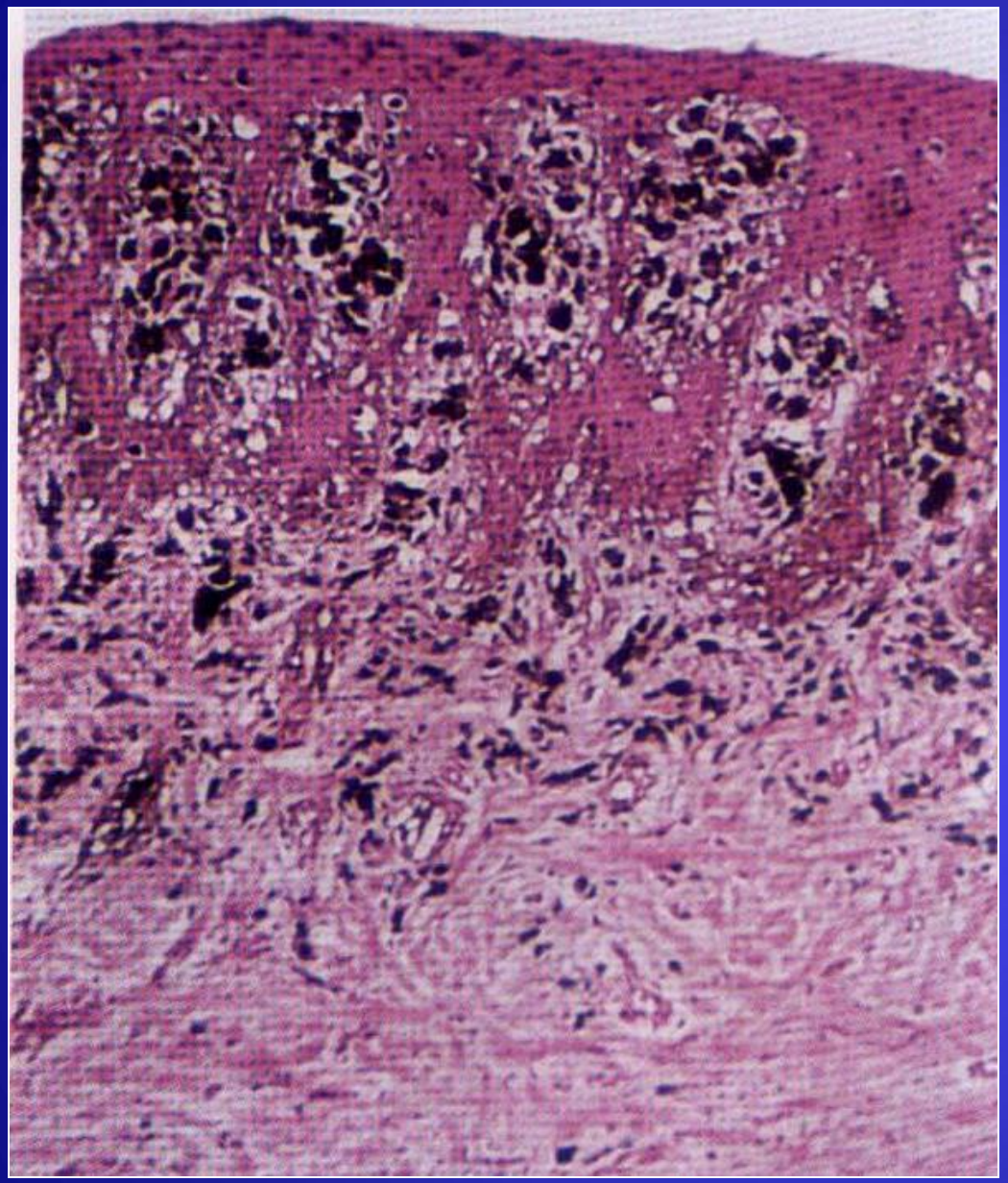
- **Lentigo maligna melanoma** is characterized by invasive spindle cells into the underlying dermis. A lymphohistiocytic infiltrate is usually present.



LENTIGO MALIGNA MELANOMA



LENTIGO MALIGNA MELANOMA



LENTIGO MALIGNA MELANOMA

- **Acral lentiginous melanoma** is histologically similar to LMM. Salient features are –

✓ Lentiginous radial growth phase

✓ Deep vertical growth phase composed predominantly of spindle cells

✓ Psoriasiform epidermal hyperplasia

✓ Intense host cell response

✓ Prominent desmoplasia associated with vertical growth phase.

Treatment and prognosis :

- Surgical excision for cutaneous lesions.
- When lymph nodes are involved, regional lymph node dissection.
- Tumors > 0.75 mm in thickness and located in the so called BANS sites (back, arm, neck and scalp), have a greater tendency to metastasize.
- Surgical excision for oral melanomas, jaw resection and lymph node dissection.
- Women have a much better survival rates upto 50 years and then the rate declines.
- Nodular and superficial spreading melanomas have a much poorer prognosis than the LMM.
- Tumors <0.75 mm rarely metastasize or cause death, regardless of location.
- Oral melanomas have a much poorer prognosis than the cutaneous ones.

BENIGN TUMORS OF CONNECTIVE TISSUE ORIGIN

ORAL FIBROMA (irritation fibroma, focal fibrous hyperplasia)

- Most common benign soft tissue neoplasm of the oral cavity.

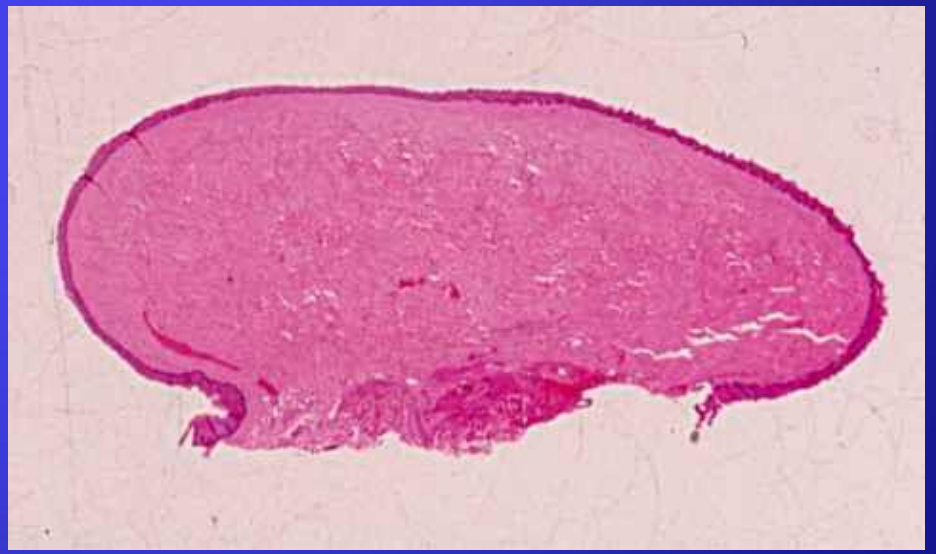
Clinical features :

- May occur at any oral site, most commonly on the buccal mucosa along the plane of occlusion.
- Appears as an elevated nodule of normal color with a smooth surface, and a sessile or pedunculated base.
- A well defined, slow growing lesion, most common in the third, fourth, and fifth decades.
- Females are affected twice more commonly than the males.

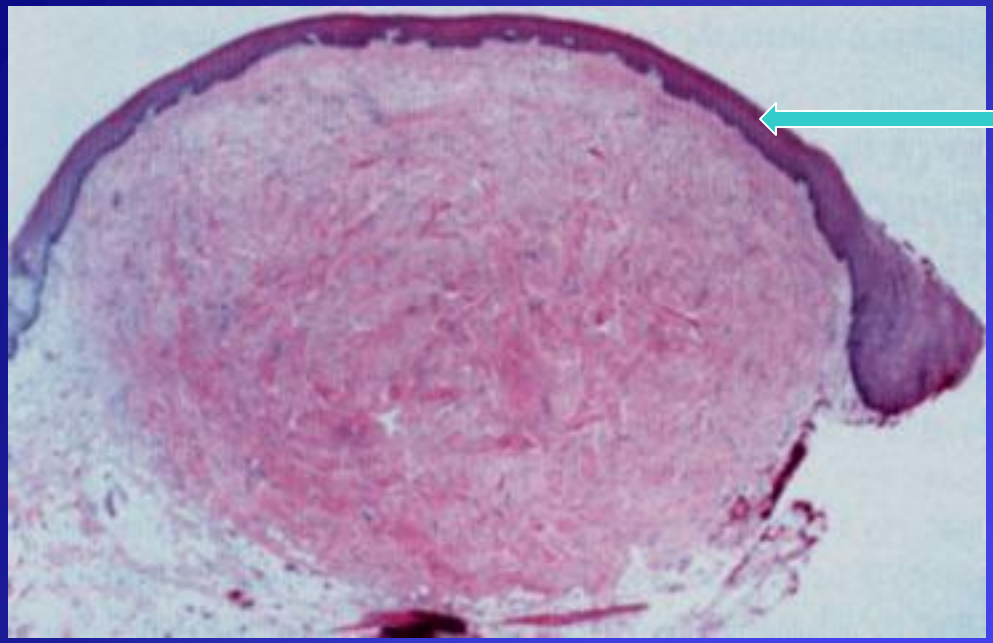
ORAL FIBROMA

Histologic features :

- Consists of bundles of interlacing collagen fibers interspersed with varying numbers of fibroblasts and blood vessels.
- Surface is covered by a layer of stratified squamous epithelium, which frequently appears stretched and shows shortening and flattening of rete pegs.
- Areas of focal or diffuse calcification or even ossification are found sometimes.



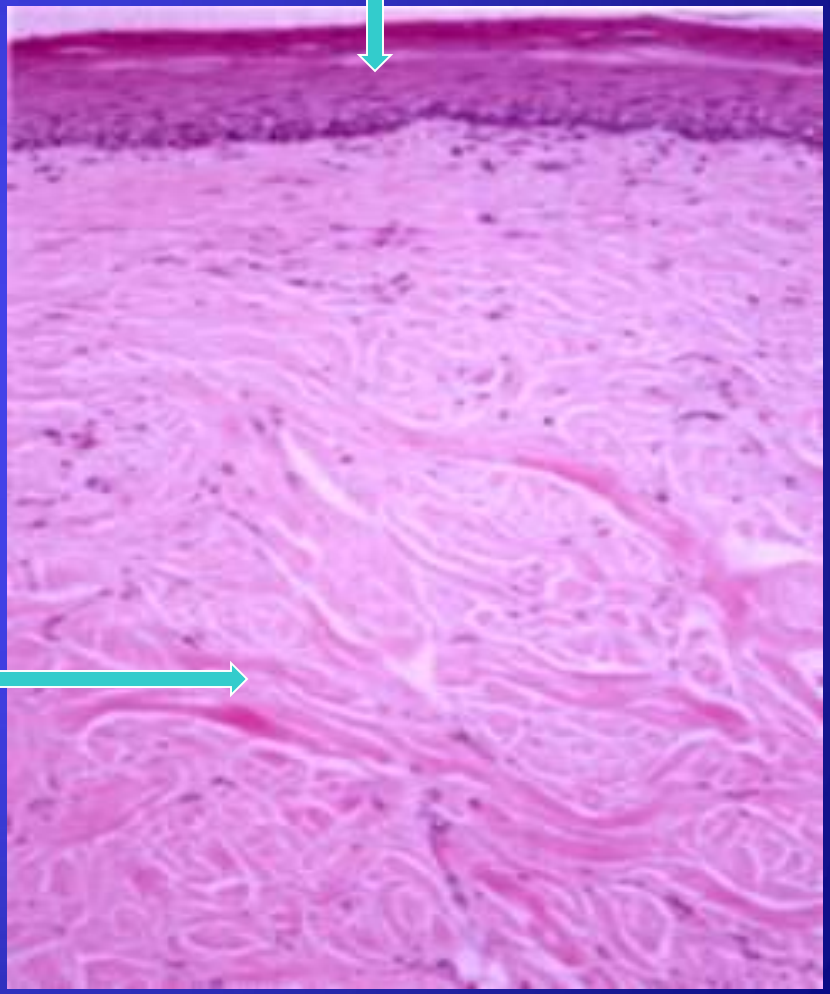
ORAL FIBROMA



Low power view

Atrophic epithelium

Collagen fiber bundles



High power view

Treatment :

Conservative surgery

LIPOMA

- Rare intraoral tumor though it is common in other areas, esp. subcutaneous tissues of the neck.
- Benign slow growing neoplasm composed of mature fat cells.

Clinical features :

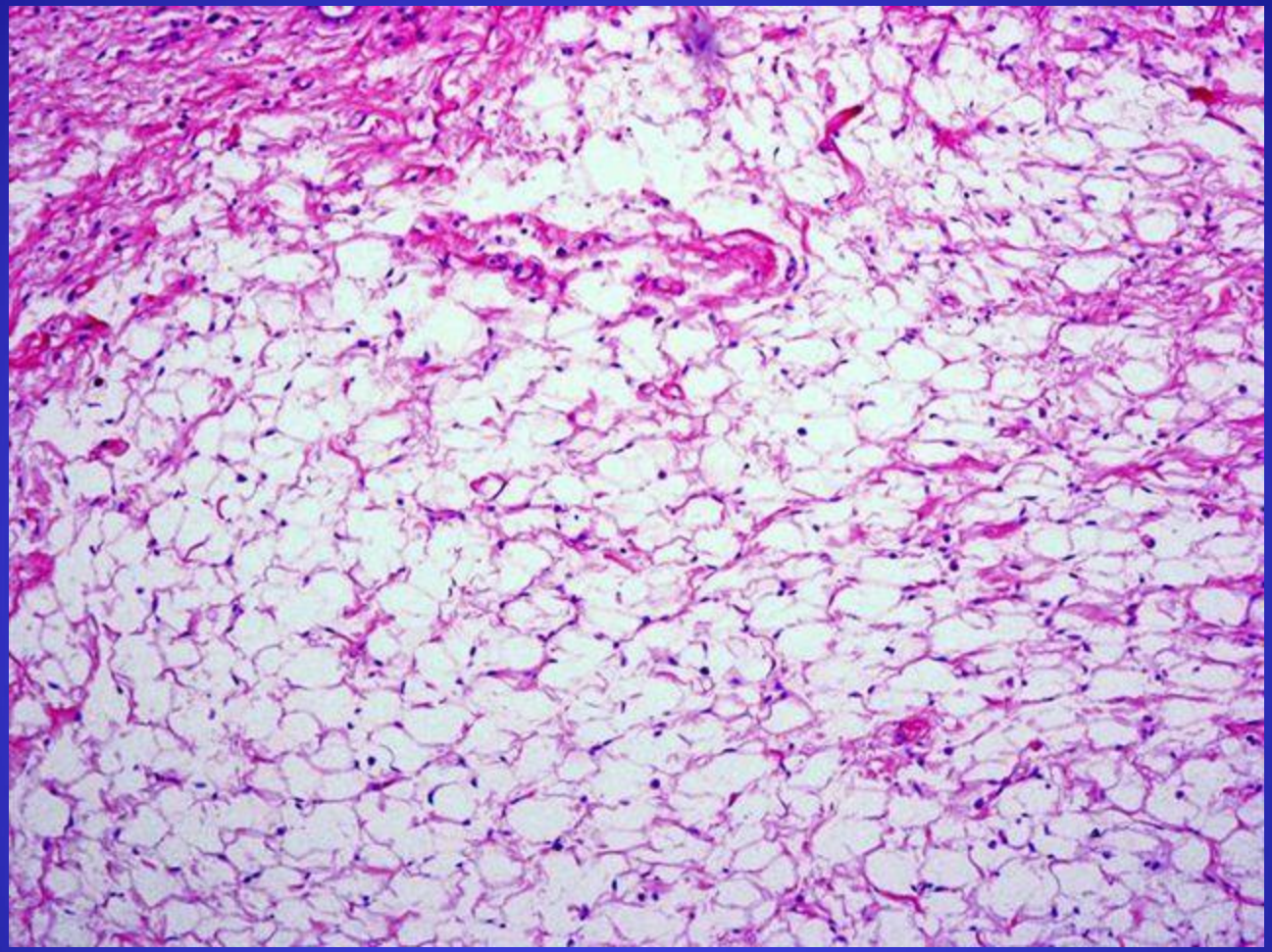
- Usually found in adults.
- Intraorally they occur in the tongue, floor of mouth, buccal mucosa and gingiva.
- Morphologically intraoral lipomas can be classified as DIFFUSE FORM affecting the deeper tissues, and a SUPERFICIAL & ENCAPSULATED FORM.
- Superficial form appears as a single or lobulated, sessile or pedunculated, painless lesion. it presents as a yellowish surface discoloration and well encapsulated. it is freely movable beneath the mucosa.

- Epithelium is usually thin and the superficial blood vessels are readily visible over the surface.
- When palpated, the diffuse form feels like fluid, sometimes leading to a mistaken diagnosis of ‘cyst’.

LIPOMA

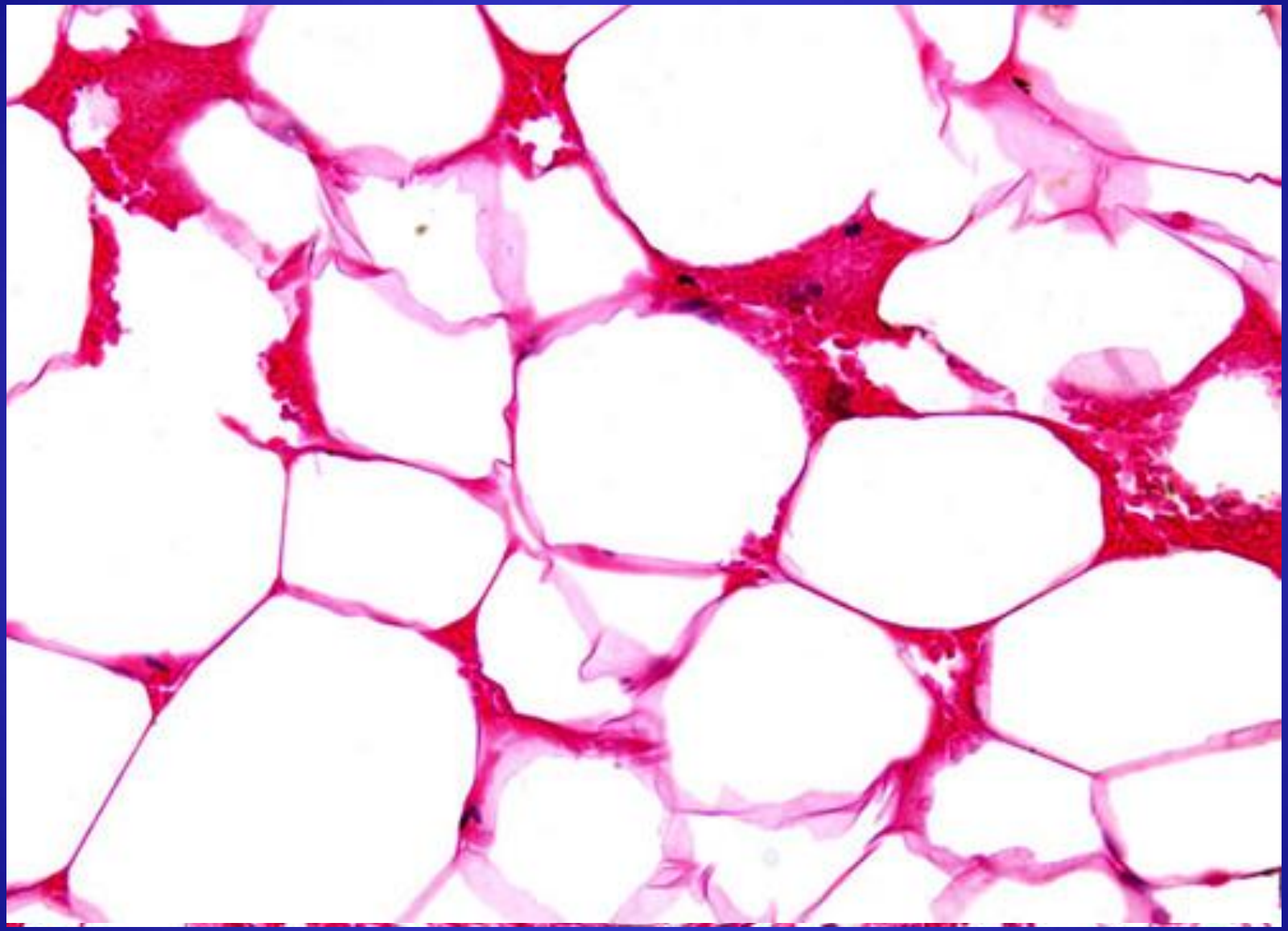
Histologic features :

- Composed predominantly of mature adipocytes, admixed with collagen streaks, and is often well demarcated from the surrounding c.t.
- A thin fibrous capsule may be seen and a distinct lobular pattern may be present.
- Often, lesional fat cells are seen to infiltrate into surrounding tissues, producing long thin extensions of fatty tissue radiating from the central tumor mass.
- When located within striated muscle, this variant is called **intramuscular lipoma**, but extensive involvement of a wide area of fibrovascular or stromal tissues might best be termed **lipomatosis.**

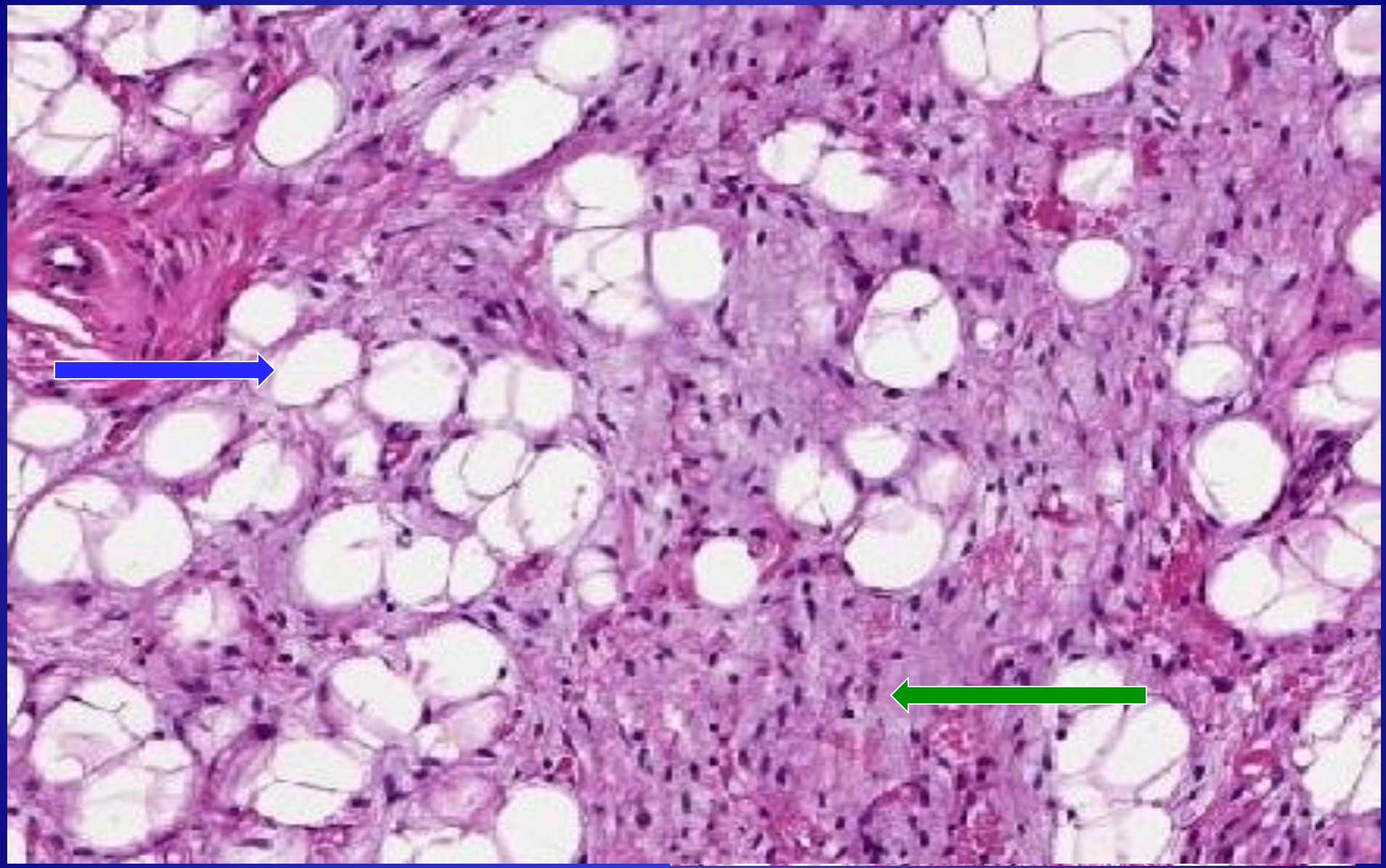


LIPOMA

Mature adipocytes and a connective tissue capsule are distinctly visible.



- Lesions with excessive fibrosis – **fibrolipoma**
- With excess no. of vascular channels – **angiolipoma**
- With a myxoid background stroma –**myxolipoma**
- With spindle cells scattered – **spindle cell lipoma**
- When spindle cells appear dysplastic or mixed with pleomorphic giant cells – **pleomorphic lipoma**
- When spindle cells are of smooth muscle origin – **myolipoma**



Spindle cell lipoma

TREATMENT :

Surgical excision of the tumor.

HAEMANGIOMAS

- They are not present at birth, but manifest within the first month of life, exhibit a rapid proliferative phase, and slowly involute to nonexistent.
- Hemangiomas of the oral cavity are not common but the head and neck are common sites.
- Classification proposed by Watson and McCarthy is as follows:

1. Capillary hemangioma.
2. Venous hemangioma.
3. Angioblastic or hypertrophic hemangioma.
4. Racemose hemangioma.
5. Diffuse systemic hemangioma.
6. Metastasizing hemangioma.
7. Nevus vinous, or port-wine stain
8. Hereditary hemorrhagic telangiectasis.

ETIOLOGY :

Unknown
One hypothesis postulates that placental cells such as the trophoblast, may be the cell of origin for hemangiomas.

Pathophysiology :

The classic sequence falls into three stages :

1) the undifferentiated capillary network stage
2) the retiform developing stage
3) the final developmental stage

Clinical features :

- Occur in infants or children.
- Usually affects whites but rarely occurs in blacks.
- Female: male = 3:1.
- Most commonly affected facial bones are mandible, the maxilla, and the nasal bones.
- **Intramuscular hemangiomas** in the oral region are most commonly seen in the masseter.
- Oral lesions appear as a flat or raised, deep red or bluish red lesion which is seldom well circumscribed.
- They are readily compressible and fill slowly when relieved.

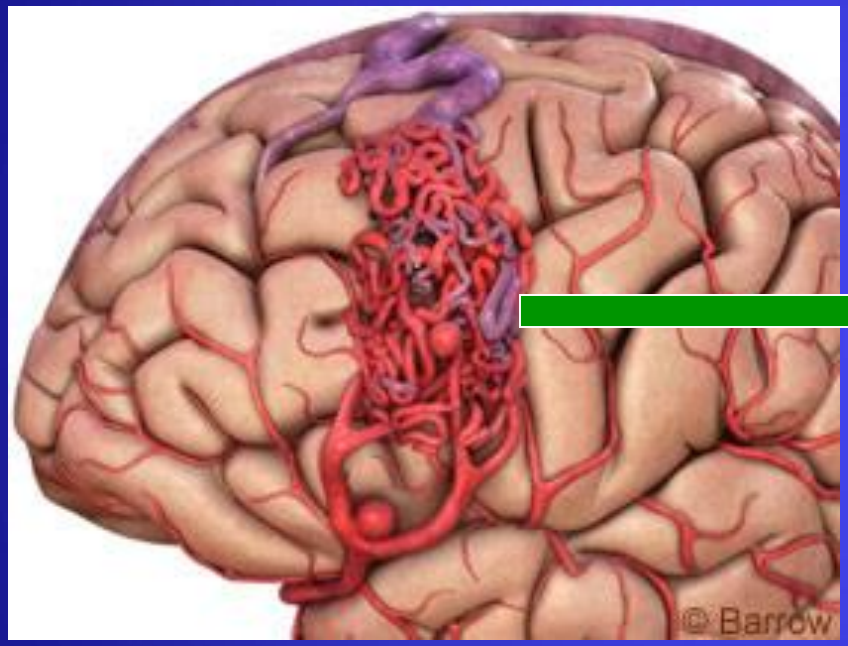
HEMANGIOMA

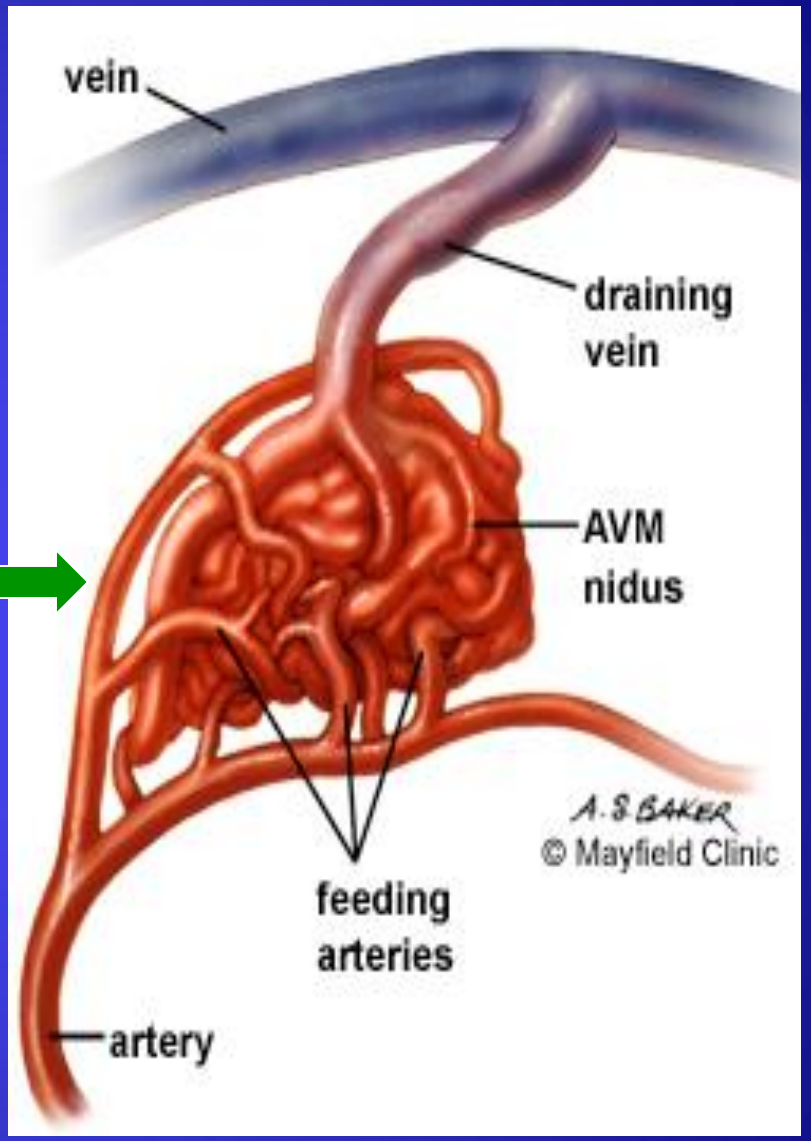
Infant with two red, nodular masses on the posterior scalp and neck

Port wine stain in an infant

Hereditary Hemorrhagic Telangiectasia

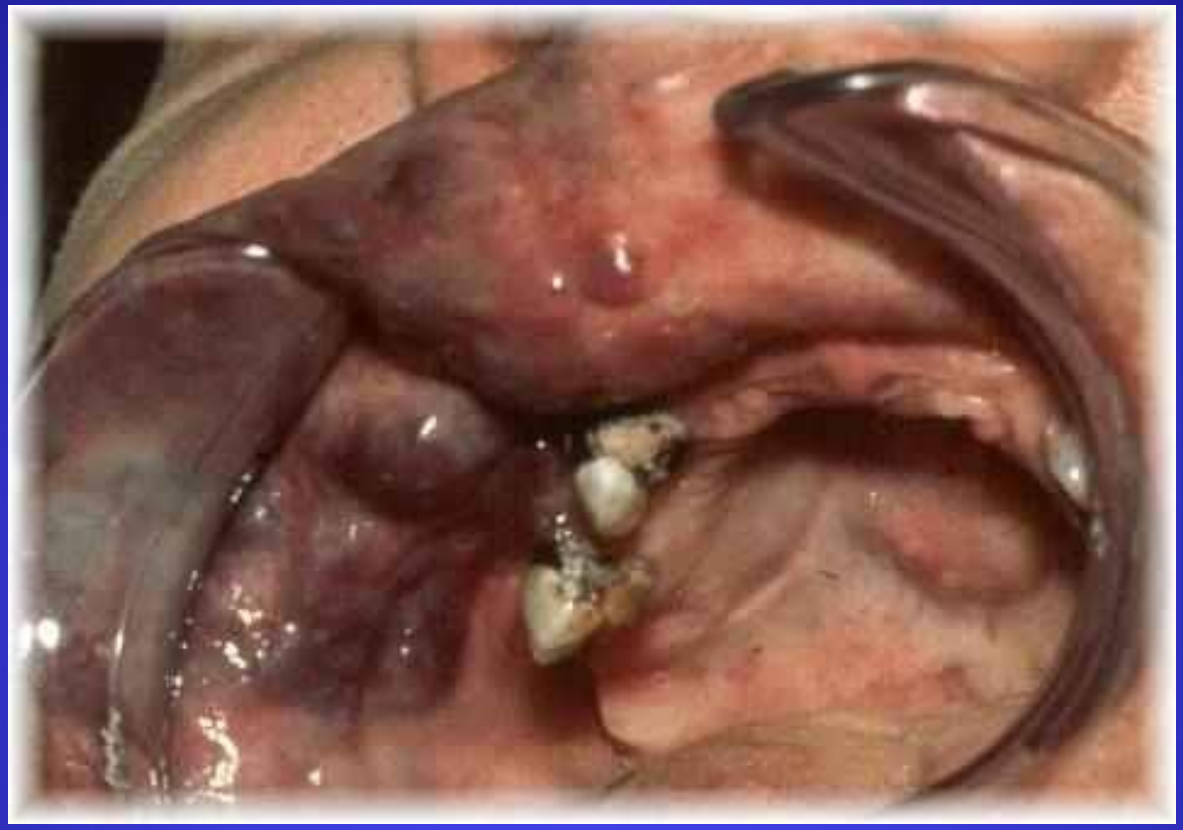
Arterio-venous malformations





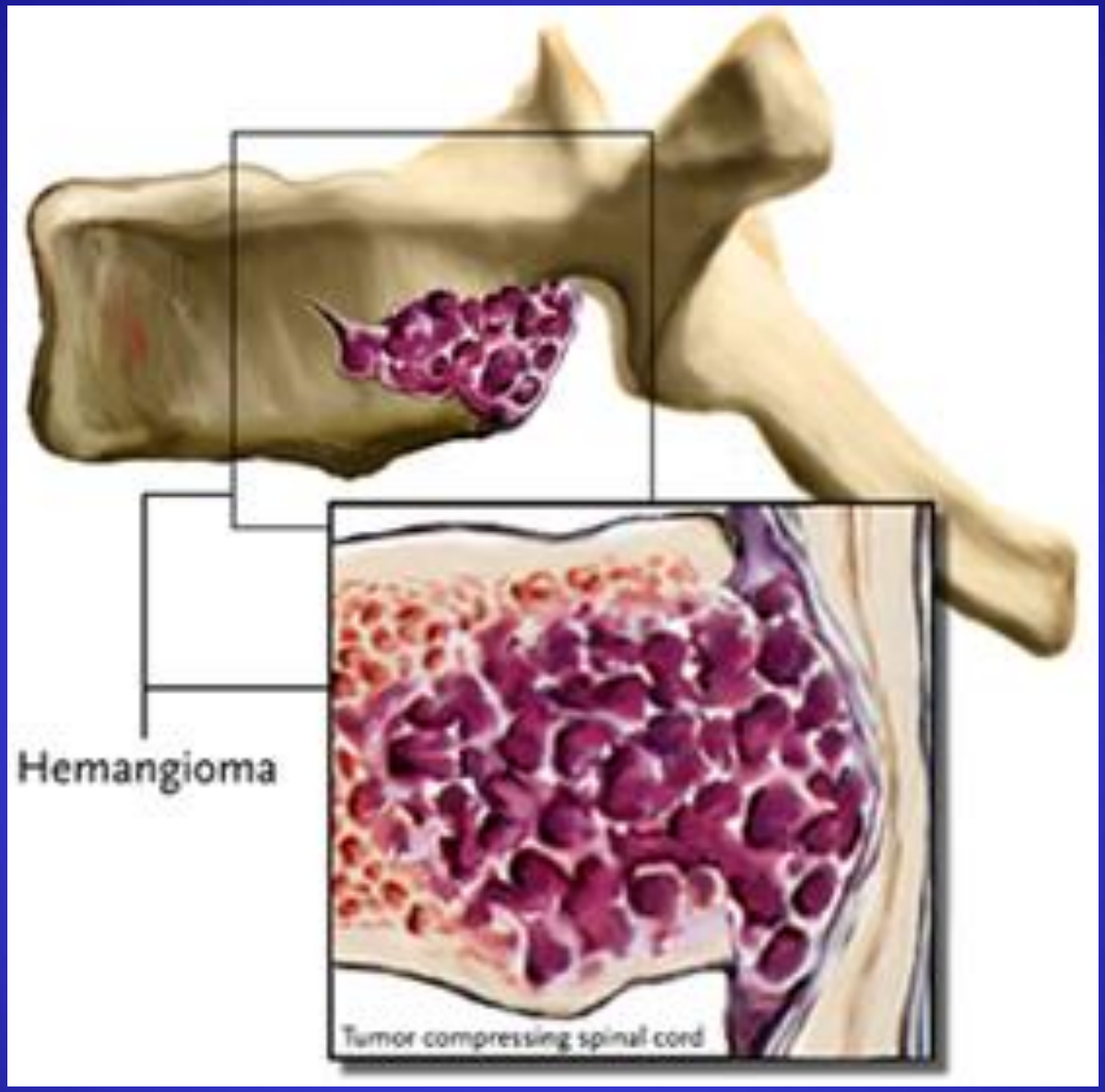
Arterio-venous malformations

- Most common site of occurrence is lip, tongue, buccal mucosa, and palate.
- Two third of the **central hemangiomas** were located in the mandible. They are bone destructive lesions, often resembling cyst and causing root resorption, but vitality of teeth is not affected.
- Some of these present a honey comb appearance on radiographs and sometimes a sun burst appearance.
- In such bony lesions it is always advisable to aspirate fluid contents through a needle before surgically opening the area.
- When it is entirely central within the bone, the prognosis is excellent with adequate surgical intervention, in contrast to those lesions where soft tissue is also involved. Here the lesion become aggressive, invade locally and often recur if not totally eradicated



Diffuse, multinodular cavernous hemangioma in Sturge-Weber syndrome.

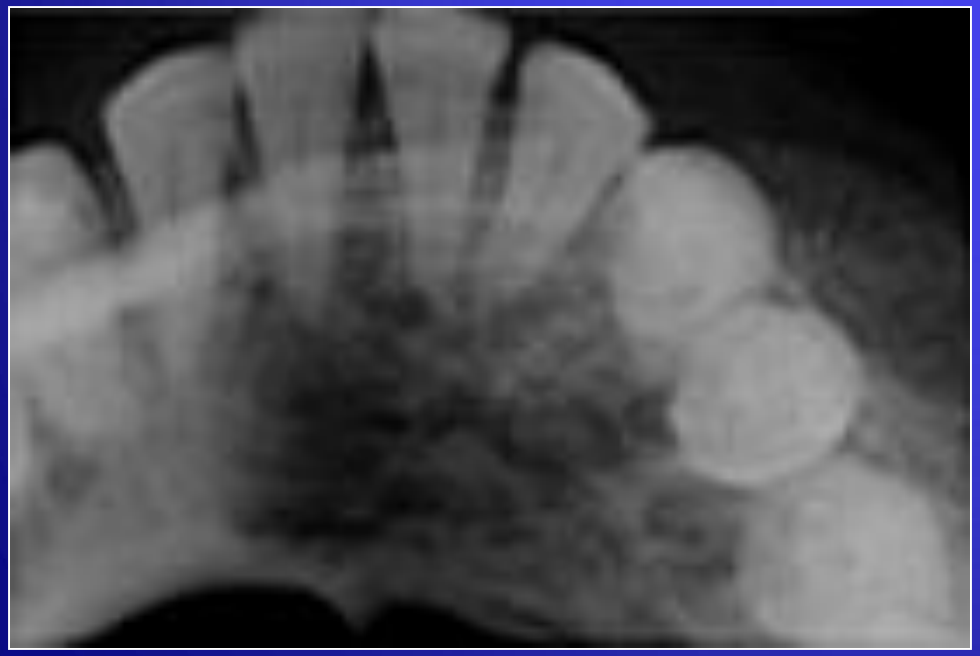
Hemangioma of Tongue



Central Hemangioma showing blood filled spaces

Roentgenographic features :

- Imaging should be considered to determine their extent and flow characteristics.
- Angiography is considered the most definitive of the studies.
- Ultrasonography can be used to determine angiomatous in nature.
- MRI can be used to differentiate a hemangioma from a lymphangioma in the oral cavity.



HEMANGIOMA OF BONE

Histological features :

- Composed of small capillaries lined by single layer of endothelial cells supported by a connective tissue stroma of varying density.
- Bears considerable resemblance to young granulation tissue and is nearly identical with some cases of pyogenic granuloma.
- Cavernous forms consist of large dilated blood sinuses with thin walls each showing an endothelial lining and filled with blood.

Salient histopathologic findings of hemangiomas :

- **Proliferative phase** :

✓ Endothelial cell hyperplasia forming mass.

✓ Thickened (multilaminated) endothelial basement membrane.

✓ Ready incorporation of tritiated thymidine in endothelial cells.

✓ Presence of large numbers of mast cells.

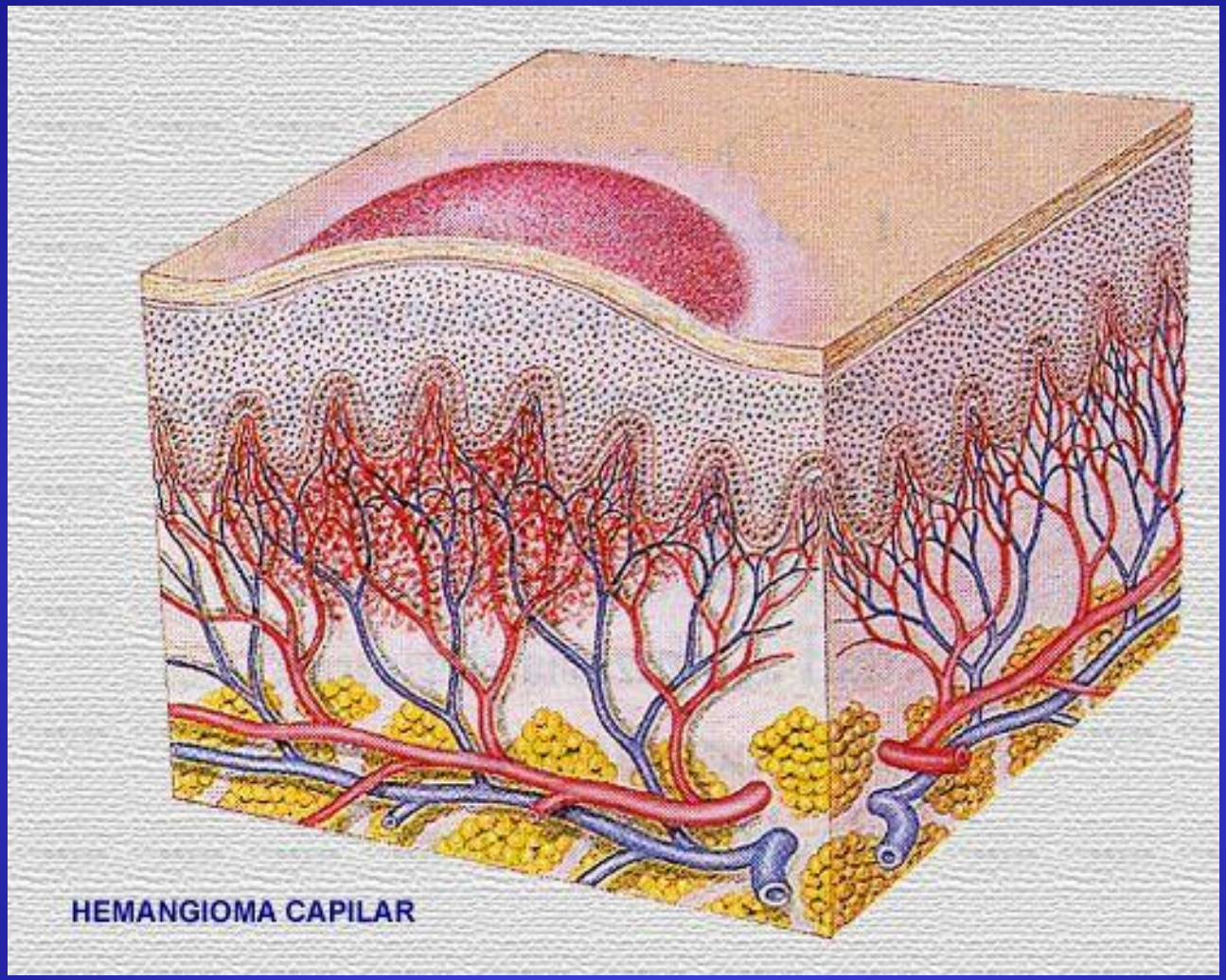
- **Involuting phase** :

✓ Less mitotic activity

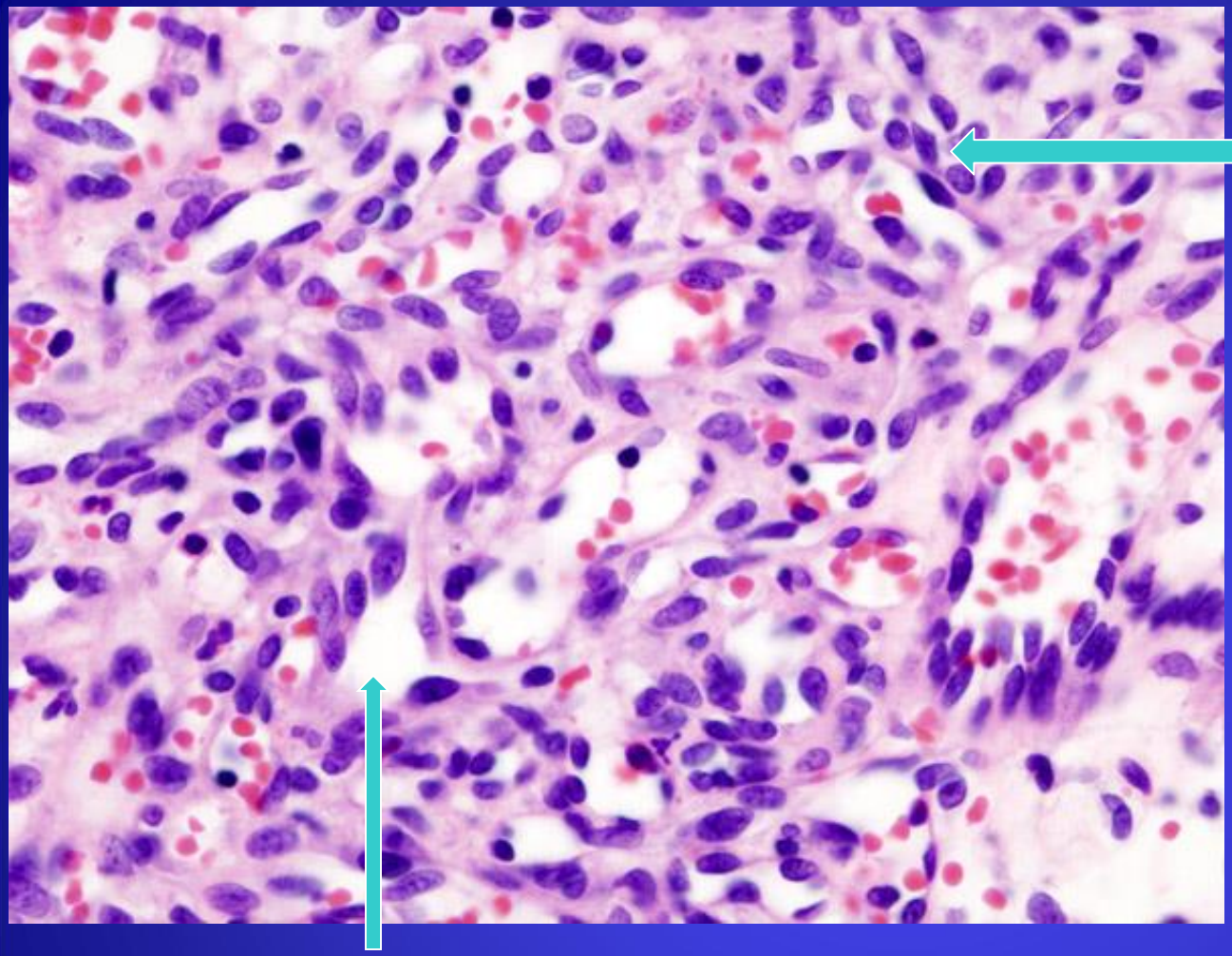
✓ Little or no uptake of tritiated thymidine in endothelial cells.

✓ Foci of fibrofatty infiltration.

✓ Normal mast cell counts.



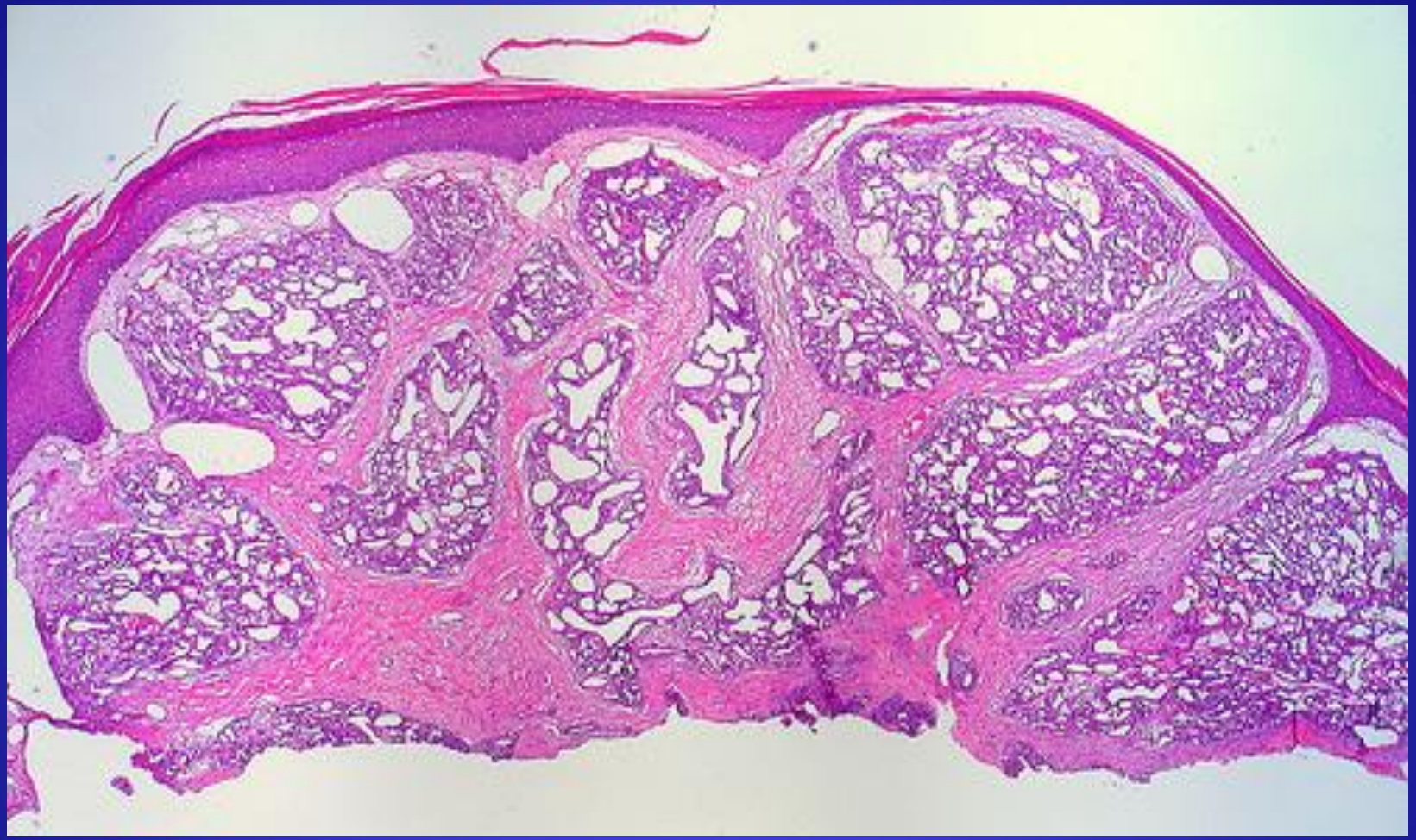
Schematic diagram of Capillary Hemangioma



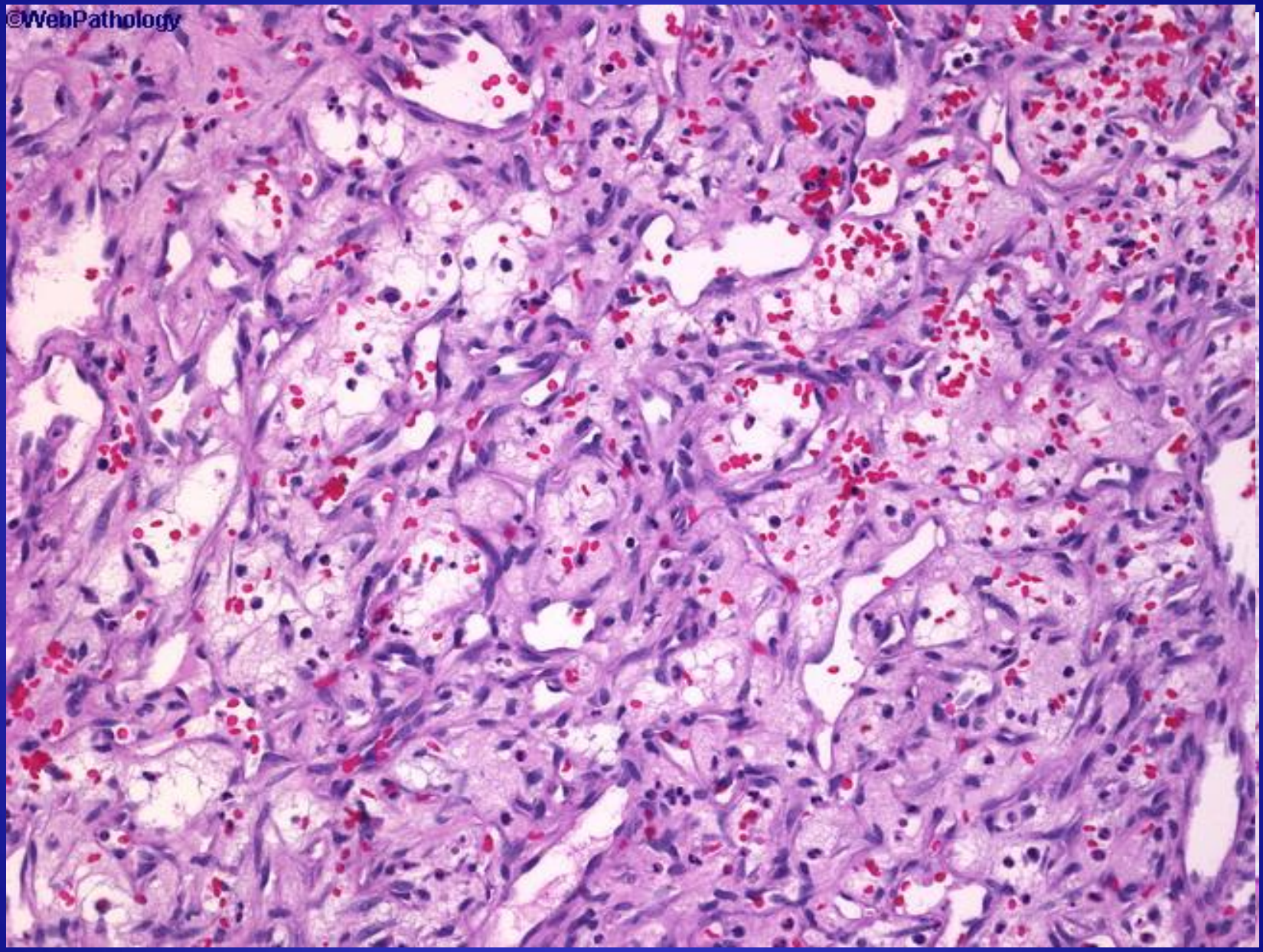
Proliferating endothelial cells

Forming lumen

Juvenile hemangioma



Lobular type of capillary hemangioma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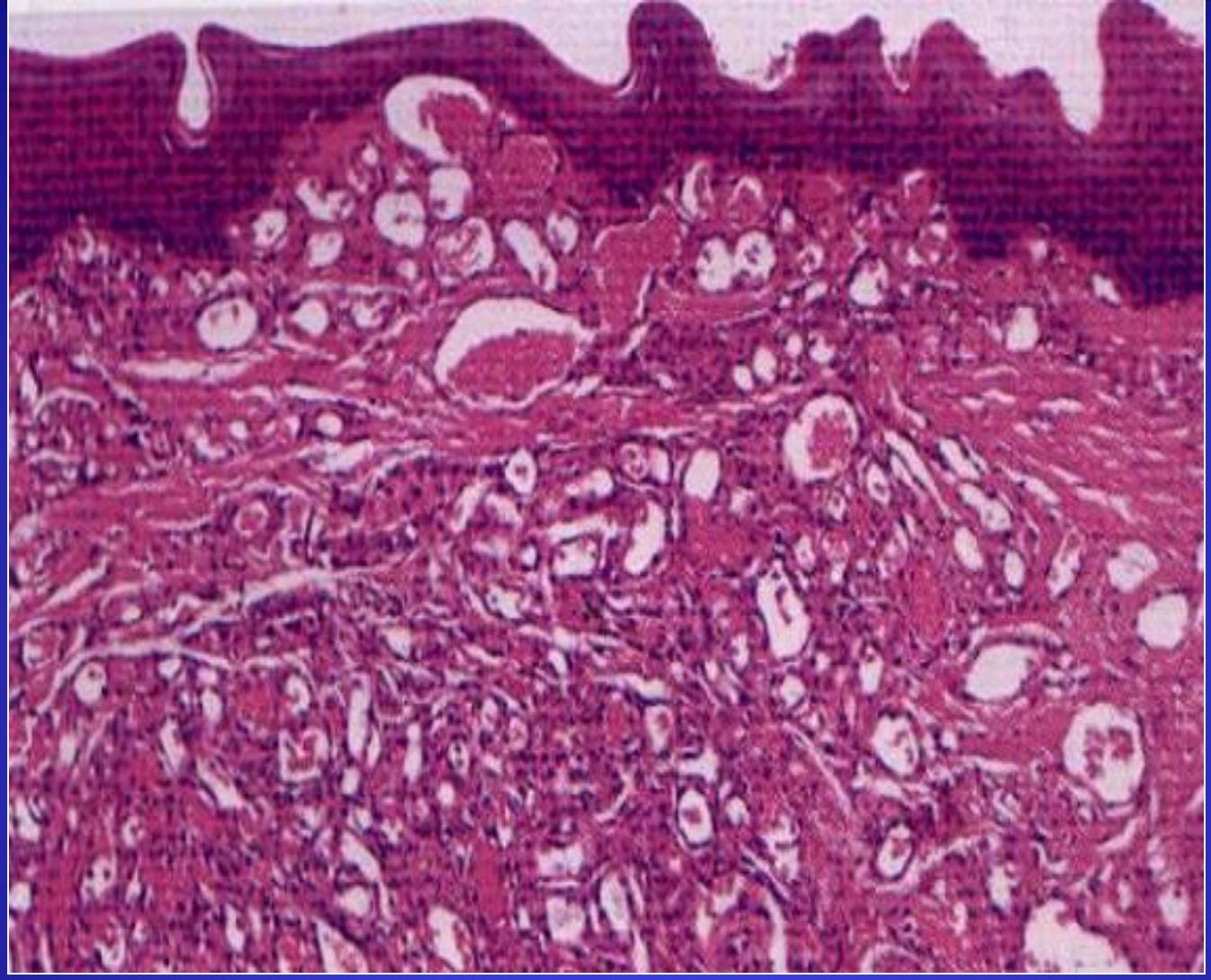
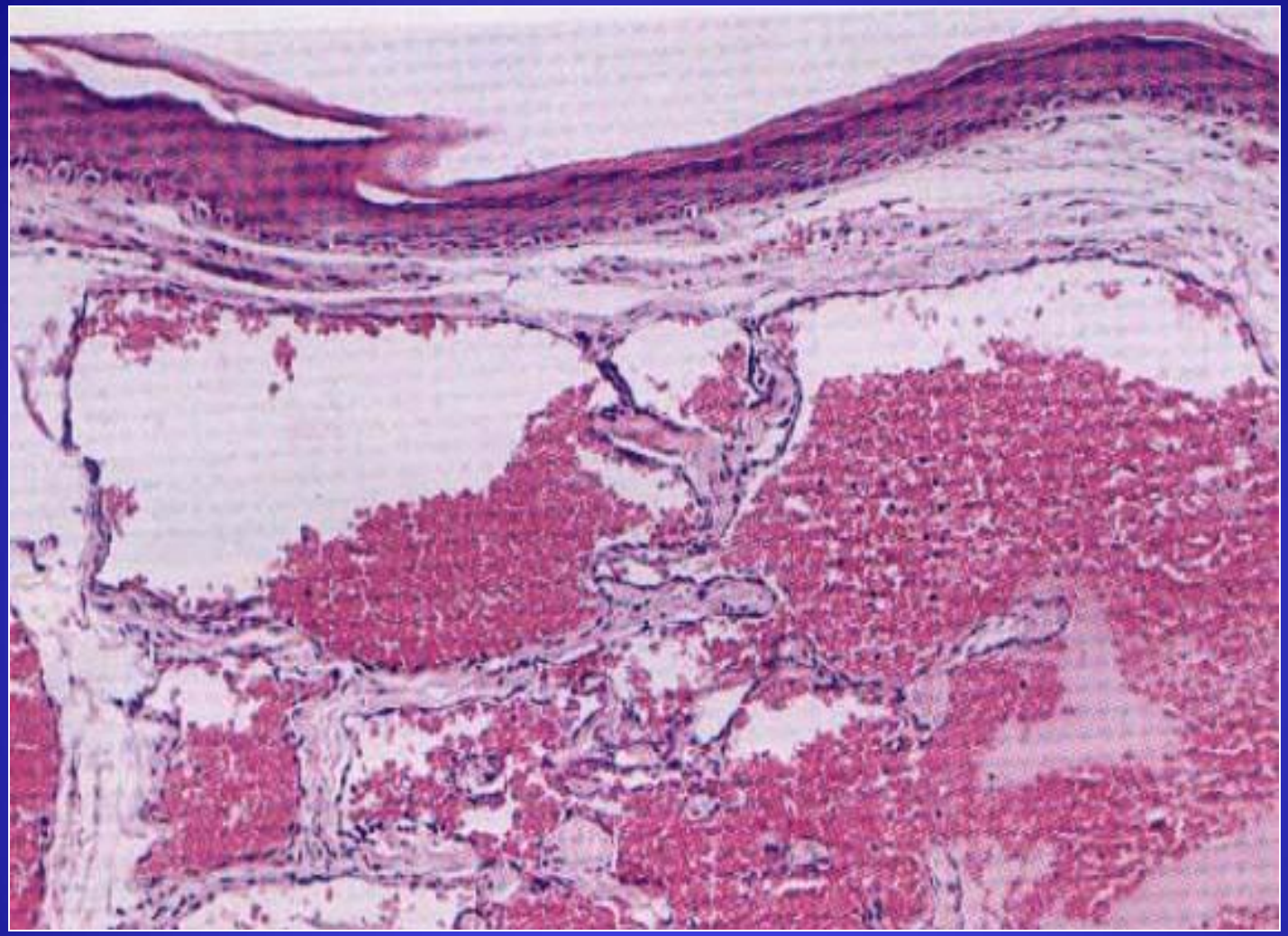


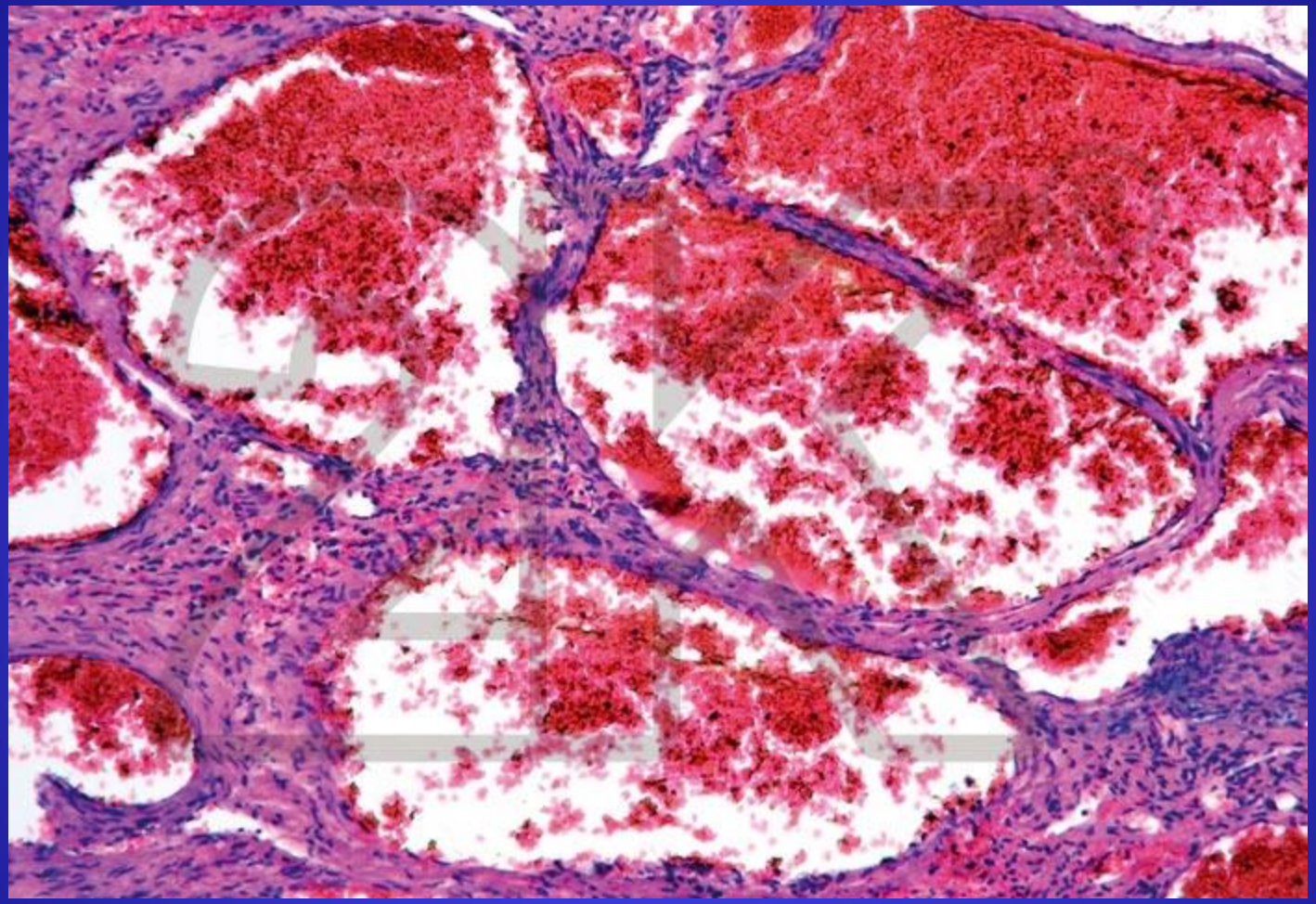
CAPILLARY HEMANGIOMA

Numerous small dilated capillaries filled with blood and lined by a single endothelial layer.



CAVERNOUS HEMANGIOMA

Large dilated blood sinuses filled with blood, having thin walls of endothelial lining



CAVERNOUS HEMANGIOMA

Treatment :

Many congenital hemangiomas have been found to undergo spontaneous regression at a relatively early age.

Cases which do not show such remission have been treated by :

- Surgery.
- Radiation therapy.
- Sclerosing agents, such as sodium morrhuate or psylliate, injected into the lesion.
- Carbon dioxide snow.
- Cryotherapy.
- Compression.
- Prognosis is excellent since it does not become malignant or recur.


Hemangioma
is removed

LYMPHANGIOMA

- A benign hamartomatous hyperplasia of lymphatic vessels, with three-fourths of all cases occurring in the head and neck region.
- Thought to be a developmental malformation of vessels which have poor communication with the normal lymph system.
- Classification suggested by Watson and McCarthy

(1) simple lymphangioma,

(2) cavernous lymphangioma,

(3) cellular or hypertrophic lymphangioma,

(4) diffuse systemic lymphangioma, and

(5) cystic lymphangioma or hygroma.

Clinical Features :

- Majority of cases of lymphangioma are present at birth.
- The most common head and neck location is the lateral neck, where this lesion typically contains large cystic spaces and is commonly called **cystic lymphangioma or cystic hygroma.**

Oral Manifestation :

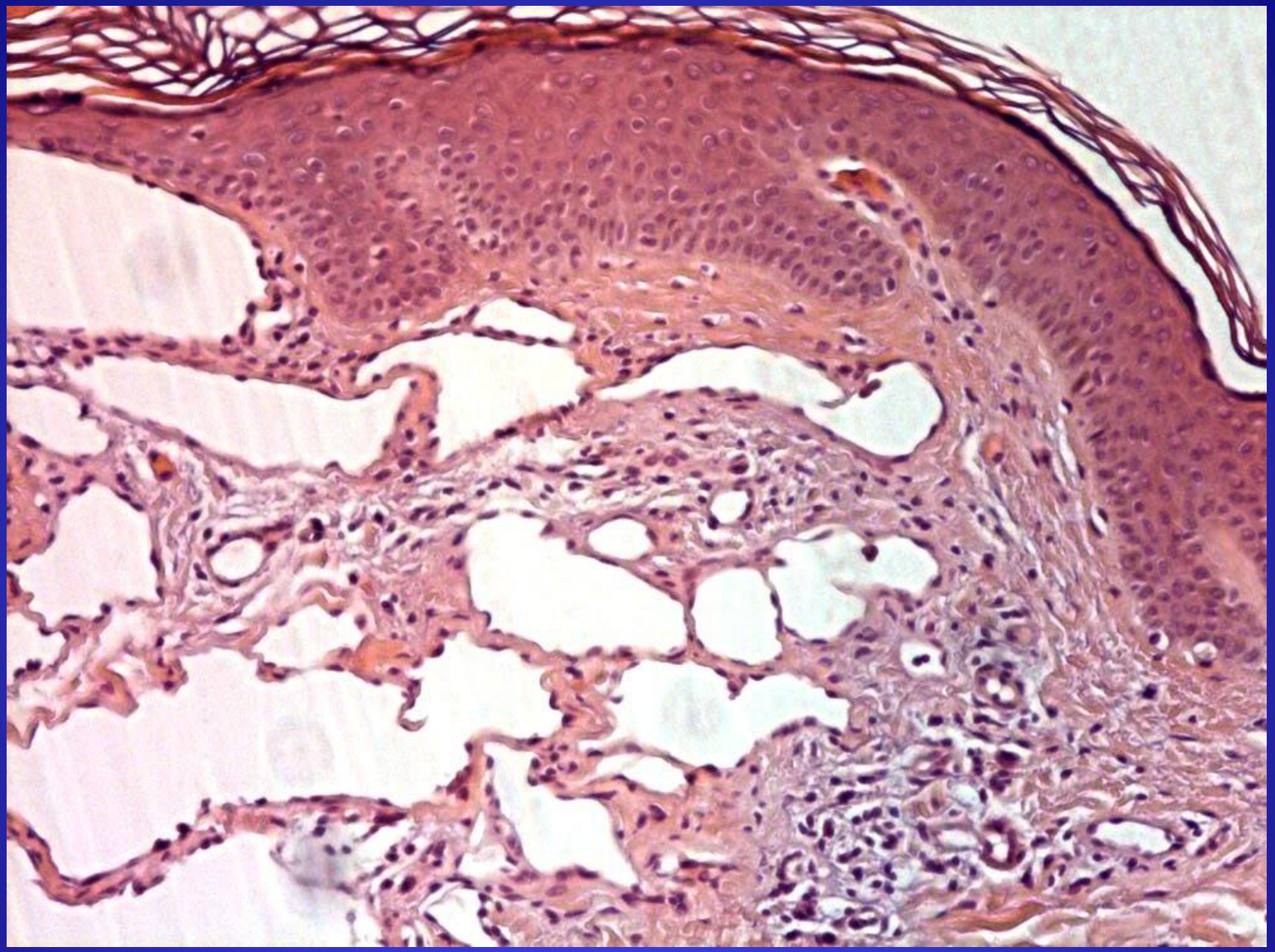
- The intraoral form most commonly occurs on the tongue.
- Superficial lesions are manifested as papillary lesions which may be of the same color or of a slightly redder hue.
- Deeper lesions appear as diffuse nodules or masses without any significant change in surface texture or color.
- The irregular nodularity of the surface of the tongue with gray and pink projections is the commonest sign of the disease, and when associated with macroglossia, is pathognomonic of lymphangioma.
- Lip involvement referred to as **macrocheilia.**
- An unusual form of lymphangioma termed **lymphangioma of the alveolar ridge in neonates** has been reported which exhibits small blue-domed fluid-filled lesions on the alveolar ridges of black newborns.

Cystic hygroma

Lymphangioma of alveolar ridge

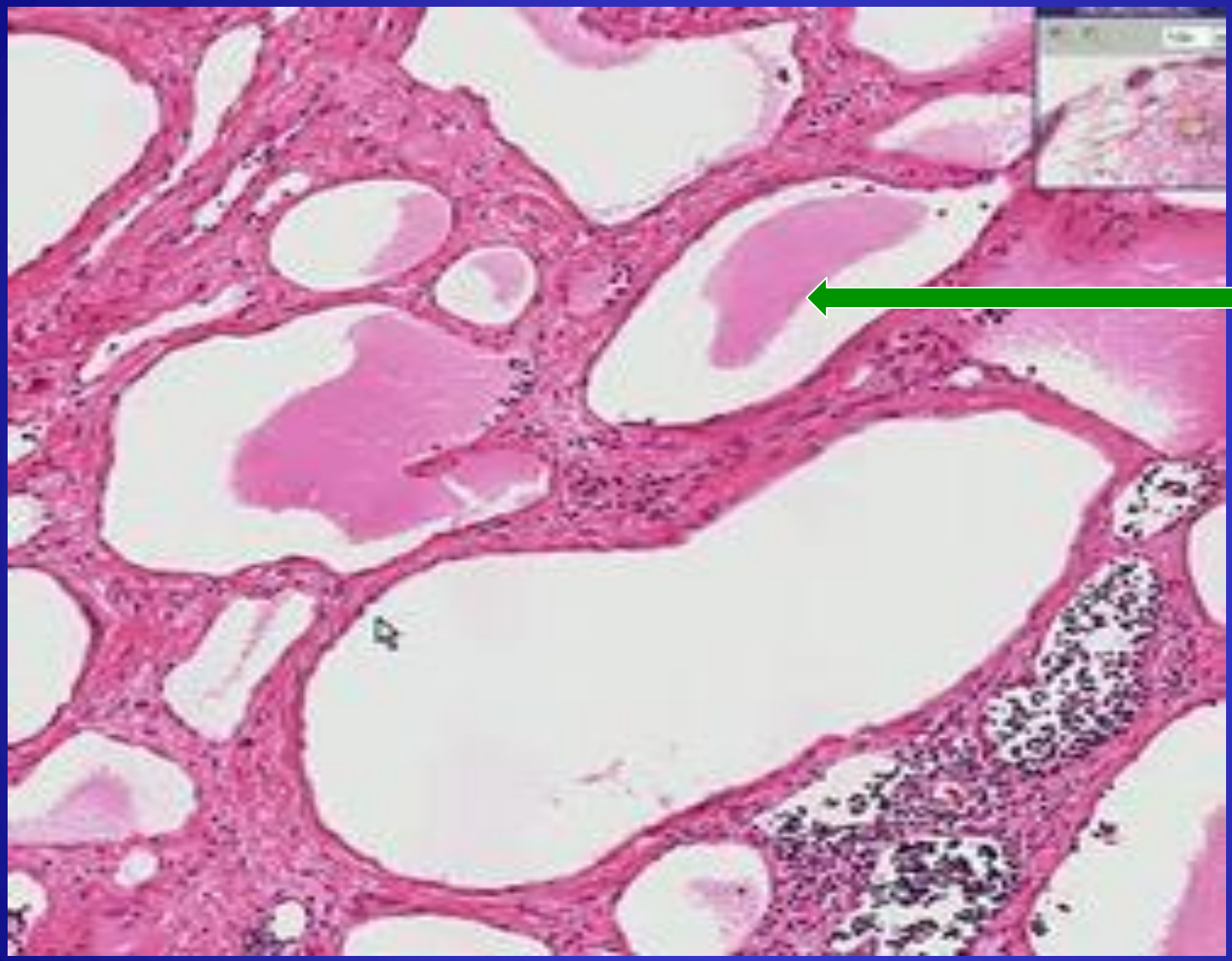
Histologic Features :

- Consists of multiple, intertwining lymph vessels in a loose fibrovascular stroma.
- Cavernous type consists of numerous dilated lymphatics, single layer of endothelial cells with flattened, occasionally plump, nuclei and containing lymph.
- Those vessels just beneath the surface epithelium tend to fill or replace the connective tissue papillae, perhaps producing a papillary surface change.
- Occasional channels may be filled with blood, a **mixed hemangiolymphangiona.**
- Occasional lesions demonstrate proliferation of lymphatic channels with smooth muscle cells called **lymphangiomyoma.**
- No encapsulation of even the tumors which appear well-circumscribed clinically.



LYMPHANGIOMA

Numerous dilated lymphatics lined by single endothelial layer and filled with lymph.


Lymph

Lymphangioma can be subclassified into four categories.

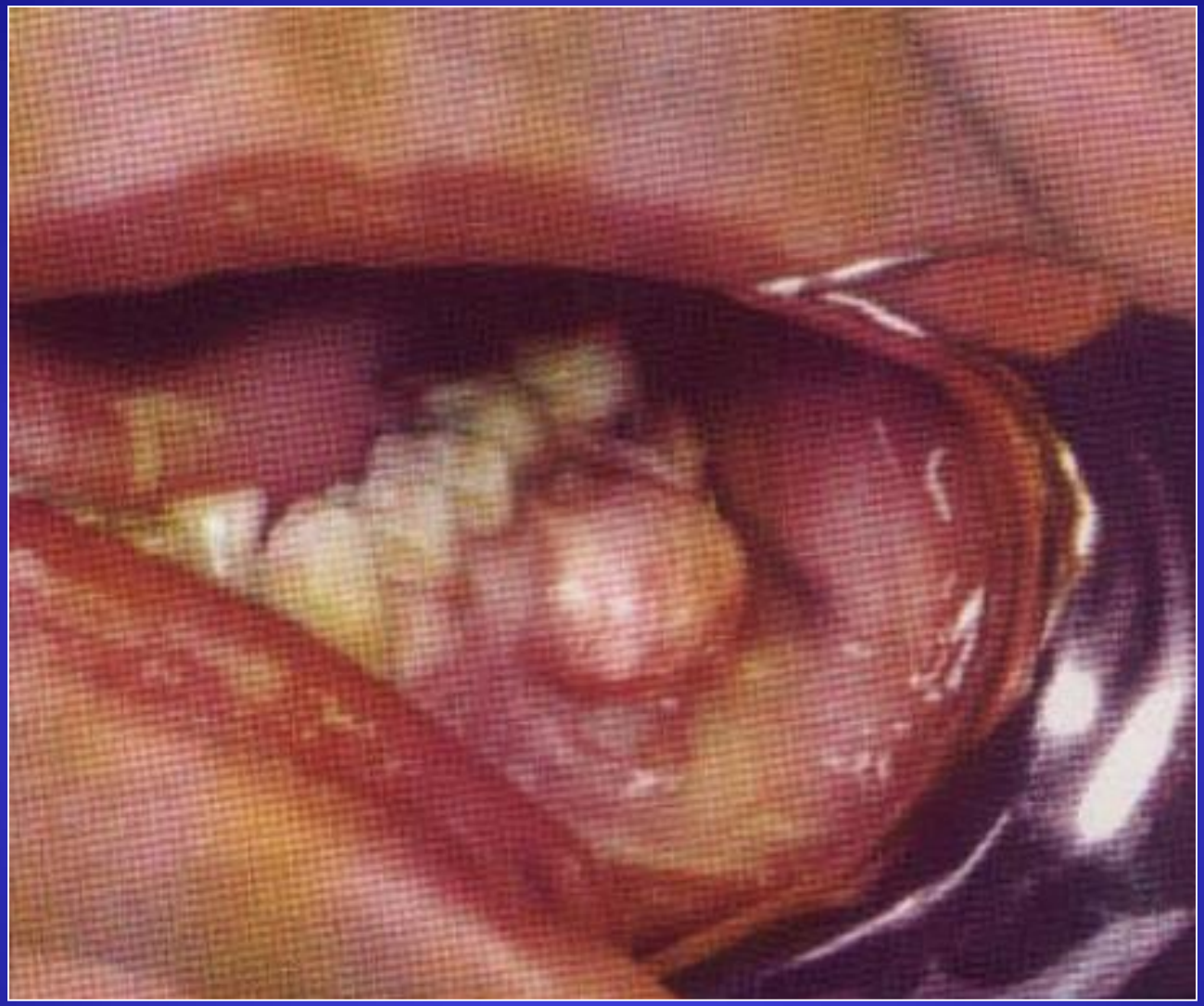
- **Lymphangioma simplex (capillary lymphangioma)** - composed of small, thin-walled lymphatics.
- **Cavernous lymphangioma-** comprised of dilated lymphatic vessels with surrounding adventitia.
- **Cystic lymphangioma (cystic hygroma)-** consisting of huge, macroscopic lymphatic spaces with surrounding fibrovascular tissues and smooth muscle.
- **Benign lymphangioendothelioma-** lymphatic channels appear to be dissecting through dense collagenic bundles.

OSTEOMA

- A benign neoplasm characterized by a proliferation of either compact or cancellous bone, usually in an endosteal or periosteal location.

Clinical Features :

- May arise at any age, somewhat more common in the young adult.
- The lesion of periosteal origin manifests itself as a circumscribed swelling on the jaw producing obvious asymmetry,
- A slow-growing tumor ; osteoma of endosteal origin is slower to present clinical manifestations, since considerable growth must occur before there is expansion of the cortical plates.
- Seldom any pain associated with this tumor.
- Multiple osteomas of the jaws, as well as of long bones and skull, are a characteristic manifestation of Gardner's syndrome.



PERIPHERAL OSTEOMA

Soft-tissue osteoma of the oral cavity is a relatively uncommon lesion, also known as ‘osteoma mucosae’, analogous to the well-recognized dermal lesion ‘osteoma cutis’ and as ‘osseous choristoma’.

- Occur almost exclusively in the tongue, although occasional cases are found in the buccal mucosa.
- Occur at any age and present as a firm nodule
- Bone itself is normal, well-circumscribed lamellar bone, usually compact but sometimes showing fatty marrow.

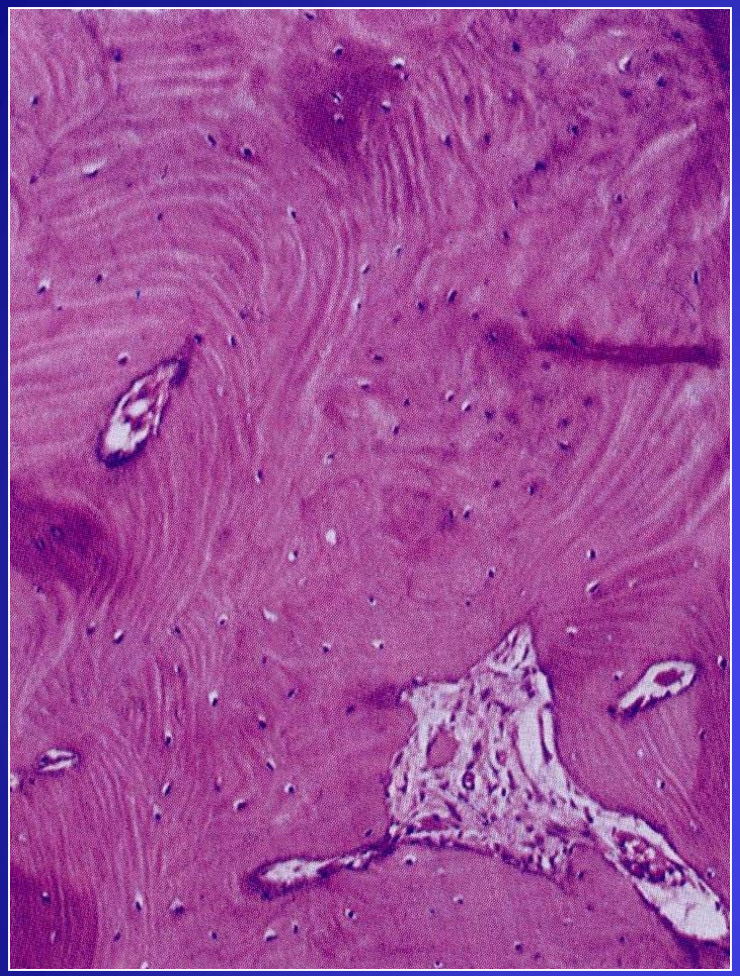
Roentgenographic Features :

- Central lesion appears as a well circumscribed radiopaque mass which is indistinguishable from scar bone.
- Sometimes this osteoma is diffuse, but it must be differentiated from chronic sclerosing osteomyelitis.
- Periosteal form is manifested as a sclerotic mass.

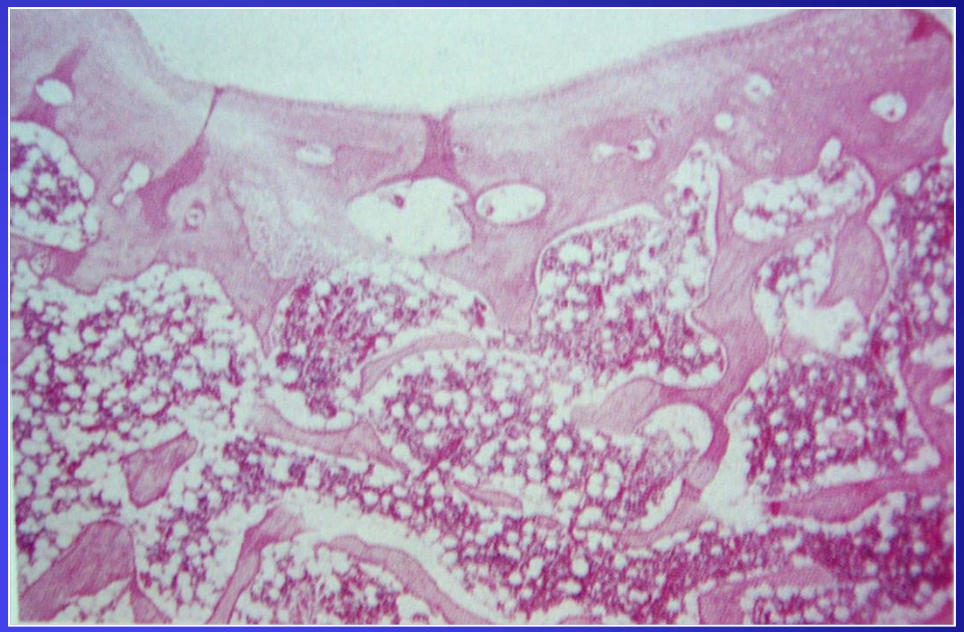
PERIPHERAL OSTEOMA

Histologic Features :

- Composed either of extremely dense, compact bone or of coarse cancellous bone.
- In any given area the bone formed appears normal
- Lesion is most often well circumscribed, but not encapsulated.
- In some tumors foci of cartilage or myxomatous tissue may be found.



COMPACT OSTEOMA



CANCELLOUS OSTEOMA

TORUS PALATINUS

The torus (bulging projection of bone) palatinus is a slowly growing, flat based bony protuberance or excrescence which occurs in the midline of the hard palate.

Both the torus palatinus and torus mandibularis are hereditary conditions thought to follow a mendelian dominant pattern.

CLINICAL FEATURES :

Women are affected more frequently than man, ratio being 2:1.

Though it may occur at any age, it Is more commonly found just before the age of 30yrs.

The torus palatinus presents itself as an outgrowth in the midline of the palate and may assume a variety of shapes.

Treatment and Prognosis :

- Surgical removal if the lesion is causing difficulty or if a prosthetic appliance is to be constructed.

It has been classified clinically as flat, spindle shaped, nodular or lobular.

The mucosa overlying the torus is intact but occasionally appears blanched. It may become ulcerated if traumatized.

The torus may be composed of either dense compact bone or of a shell of compact bone with a centre of cancellous bone and thus it is often visible on an intraoral palatal radiograph.

TREATMENT AND PROGNOSIS :

It is benign and never becomes malignant.

The torus is usually not treated although occasionally it may be of such size and shape that it is impractical to construct a full or partial denture over the structure because of undercuts, the probability of trauma to overlying mucosa or inability to sit the denture owing to rocking.

In such cases torus must be removed surgically.

TORUS MANDIBULARIS

A torus mandibularis is an exostosis or outgrowth of bone found on the lingual surface of the mandible.

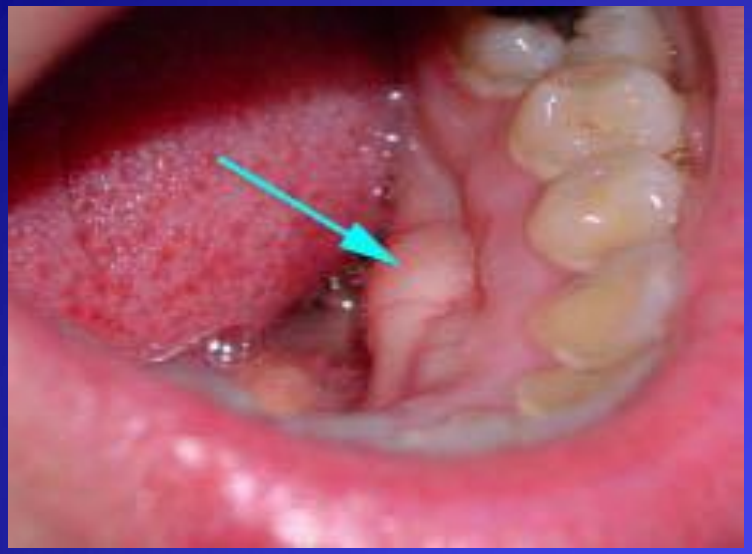
CLINICAL FEATURES :

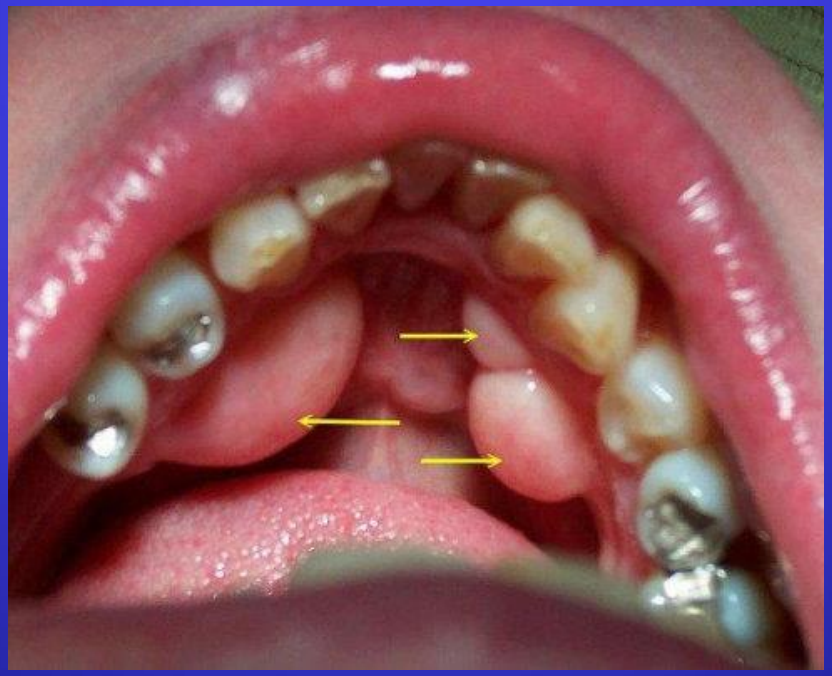
Occurs above the mylohyoid line opposite the bicuspid teeth usually bilaterally.

It may be single or multiple and frequently visible on dental radiographs.

It may be lobed or multiple.

Onset is by the age of 30yrs.





MULTIPLE EXOSTOSIS

MALIGNANT TUMOURS OF THE CONNECTIVE TISSUE ORIGIN

FIBROSARCOMA

- Composed of malignant fibroblasts in a collagenous background.

- Fibrosarcoma, particularly of the head and neck area, is a quite uncommon neoplasm.

- Sarcomas as a group differ from malignant epithelial neoplasms by their typical occurrence in relatively younger persons and greater tendency to metastasize through blood stream rather than the lymphatic, producing more widespread foci of secondary tumor growth.

- Two main types of fibro sarcoma of bone exist, **primary** and **secondary**:

✓ ***Primary type*** produces variable amounts of collagen. It is central, arising within the medullary canals, or peripheral, arising from the periosteum.

✓ ***Secondary type*** arises from a preexisting lesion or after radiotherapy to an area of bone or soft tissue. This is more aggressive tumor with poorer prognosis.

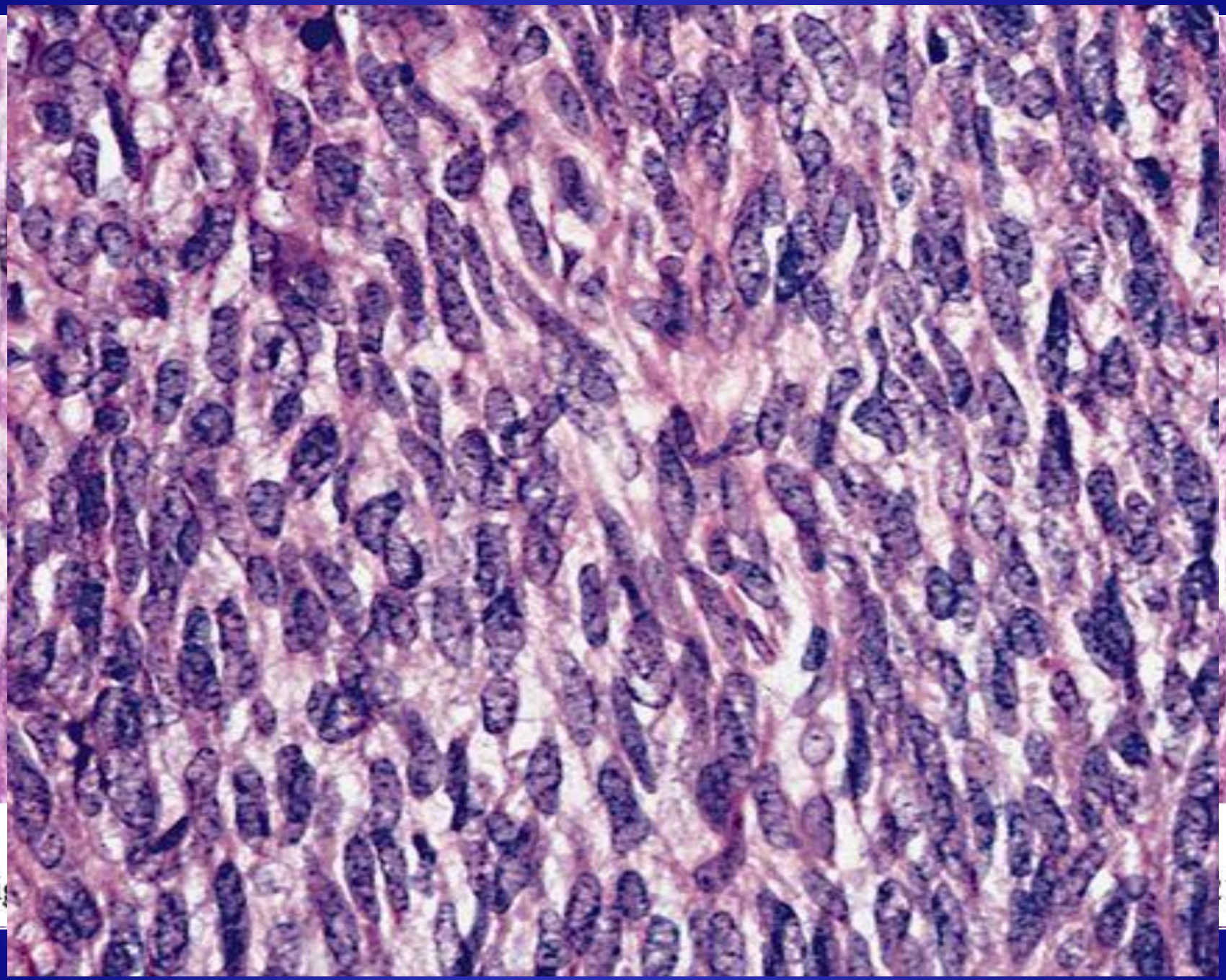
ETIOPATHOGENESIS

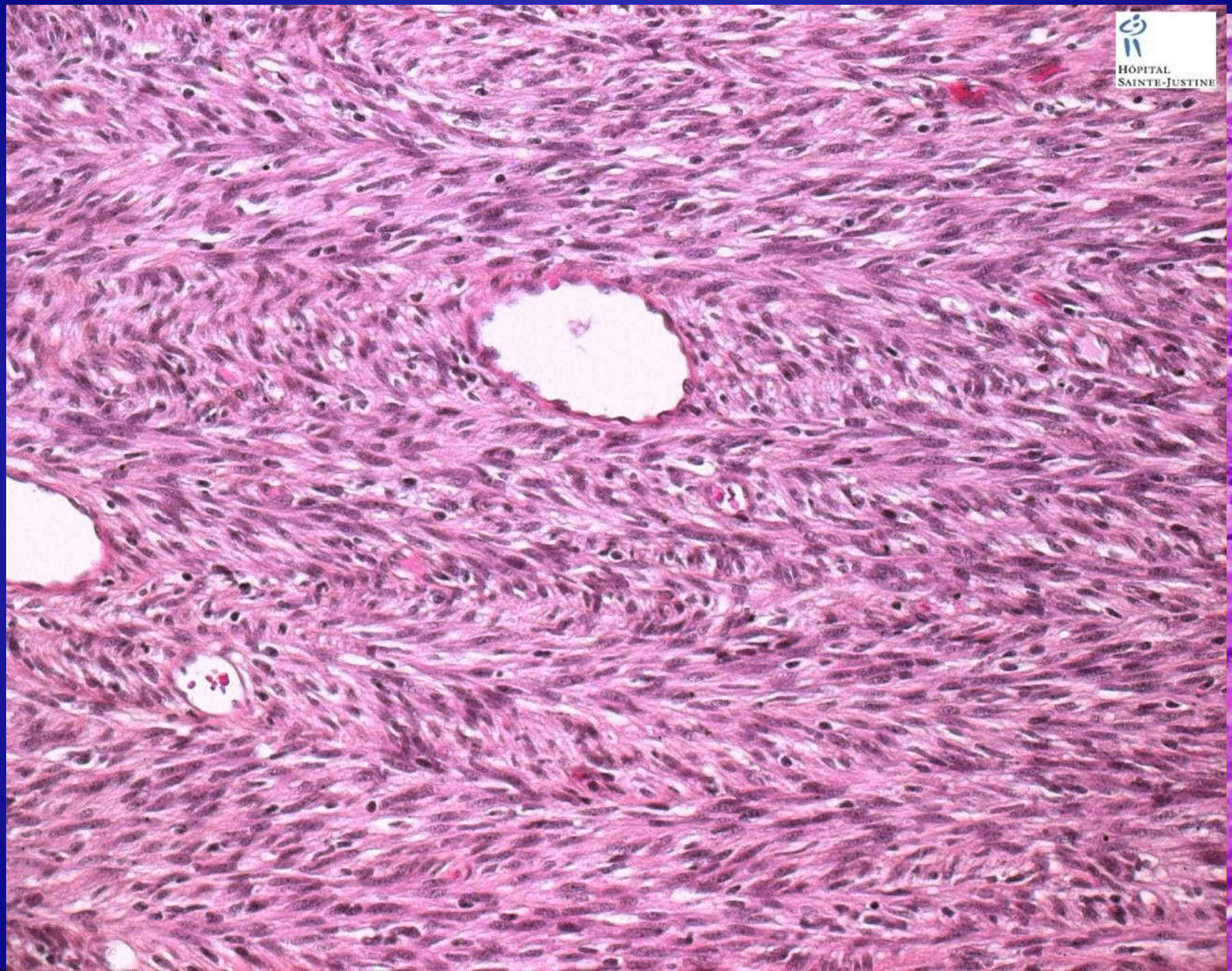
- It has no definite cause.
- Several inherited syndromes like multiple neurofibromas may have higher risk.
- It has also been noticed to arise from preexisting lesion, such as fibrous dysplasia, chronic osteomyelitis, bone infarcts, pagets disease, and in previously irradiated areas of bone. These are very aggressive and are associated with much poorer outcome than the primary fibrosarcoma of bone

Clinical features

- Occurs slightly more commonly in males.
- Fibrosarcoma of bone is most common in the fourth decade of life and in the lower extremities, especially femur and tibia.
- Fibrosarcoma of soft tissue usually affects a widened age spectrum (35-55yrs) and is seen in the soft tissues of thigh and posterior knee.
- Bone tumors often present with pain and swelling after a long duration of symptoms. They may even grow large enough to threaten the structural integrity of the bone and cause pathologic fracture.
- Soft tissue sarcomas mostly present as painless masses and may become extremely large prior to diagnosis.
- It is rarely involves the oral cavity.

Fibrosarcoma involving buccal mucosa

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Treatment :

- Radical surgery.
- Radiation therapy is used in conjunction with surgery for soft tissue fibrosarcomas.
- They seldom metastasize except late in their in the clinical course but when they do the secondary deposits are found in distant sites, especially lungs, liver and bones.

KAPOSI'S SARCOMA

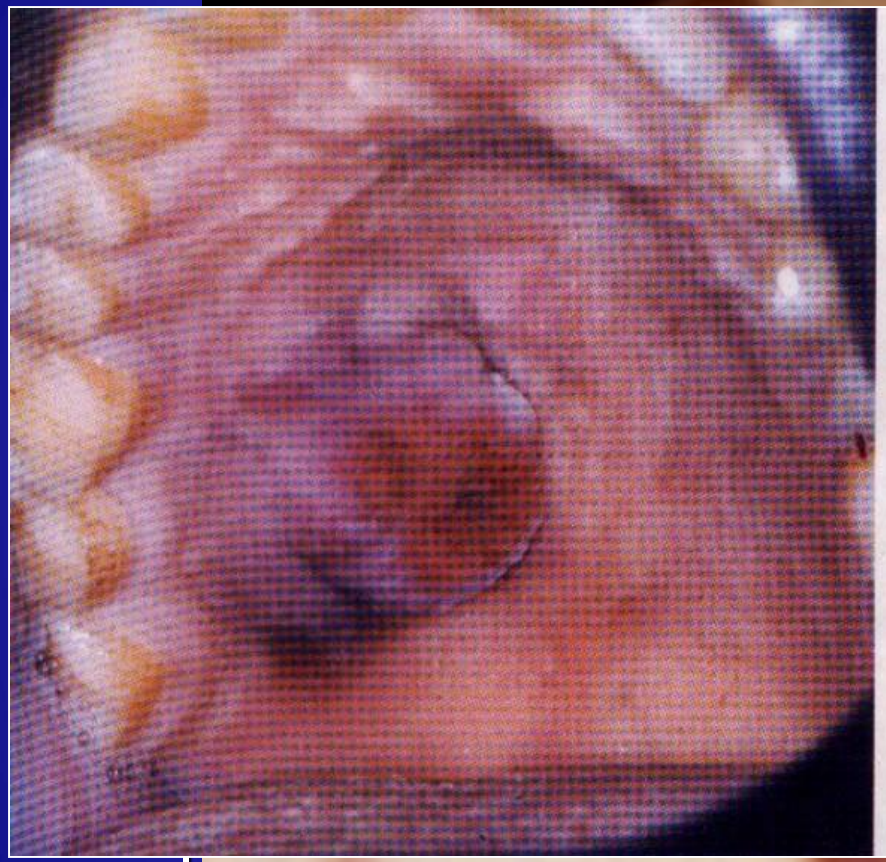
- A multicentric proliferation of vascular and spindle cell components.
- Now considered as a viral associated tumor, it is not clear whether it is a true neoplasm or a simple hyperplasia.
- It is currently incriminated with HIV/AIDS, though HIV does not seem to be the direct cause.

Etiology

- Is unknown. It has been suggested that the combined effect of various infectious agent, host factors and environmental factors encourage Kaposi's sarcoma's proliferation.
- The evidence also suggests that the disease is promoted by the effects of immonosuppression and immune activation, possible combined with a sexually transmissible infectious agent.
- Also herpes virus like DNA sequences, HHV 8 or KSHV has been isolated from lesions and in Kaposi's sarcoma-derived cell cultures.

Clinical features :

Four



d numbers,
re forming
on adjacent or distant skin.

- Oral involvement is quite unusual.

KAPOSI'S SARCOMA

The classic variant in an older man presenting as multiple purple papules and plaques on the lower leg.

Lymphadenopathic Kaposi's sarcoma

- It is endemic to young African children and presents as a localized or generalized enlargement of lymph node chains, including the cervical nodes.
- The disease follows a fulminant course with visceral involvement and minimal skin or mucous membrane involvement.
- In the head and neck region, salivary glands may be affected.
- This variant does not appear to be HIV related.

Transplantation associated Kaposi's sarcoma

- Is seen in 1-4 % of renal transplant patients.
- The extent and progression of the disease correlated directly with the loss of cellular immunity of the host.
- Oral lesions are rare.

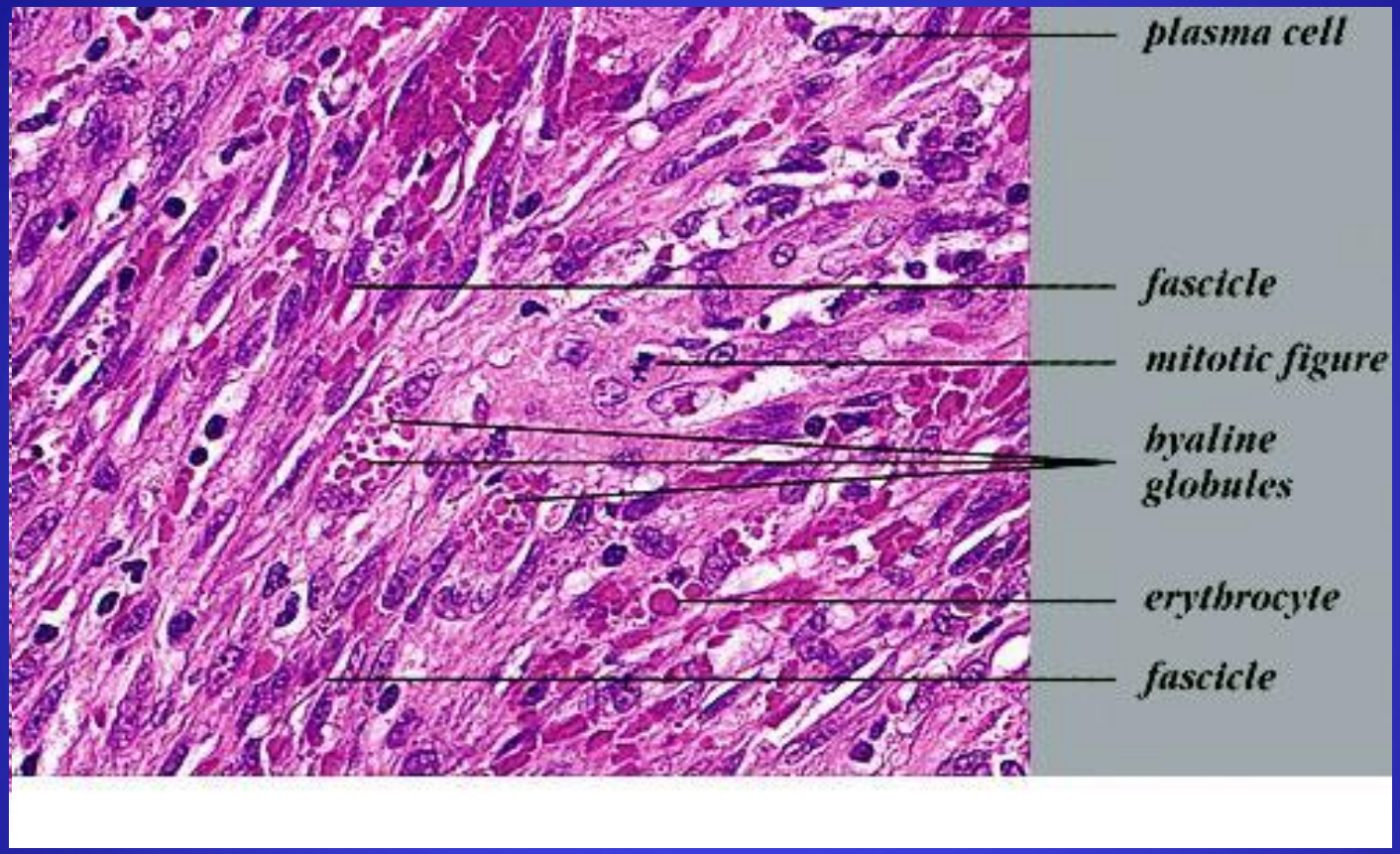
Figure 4 - HIV associated Kaposi´s sarcoma

- Cervical lymph nodes and salivary gland enlargement may also be seen.
- Patient may have oral candidiasis and AIDS related gingivitis as well.
- These lesions may interfere with eating and speaking, cause tooth loss or compromise the airways.

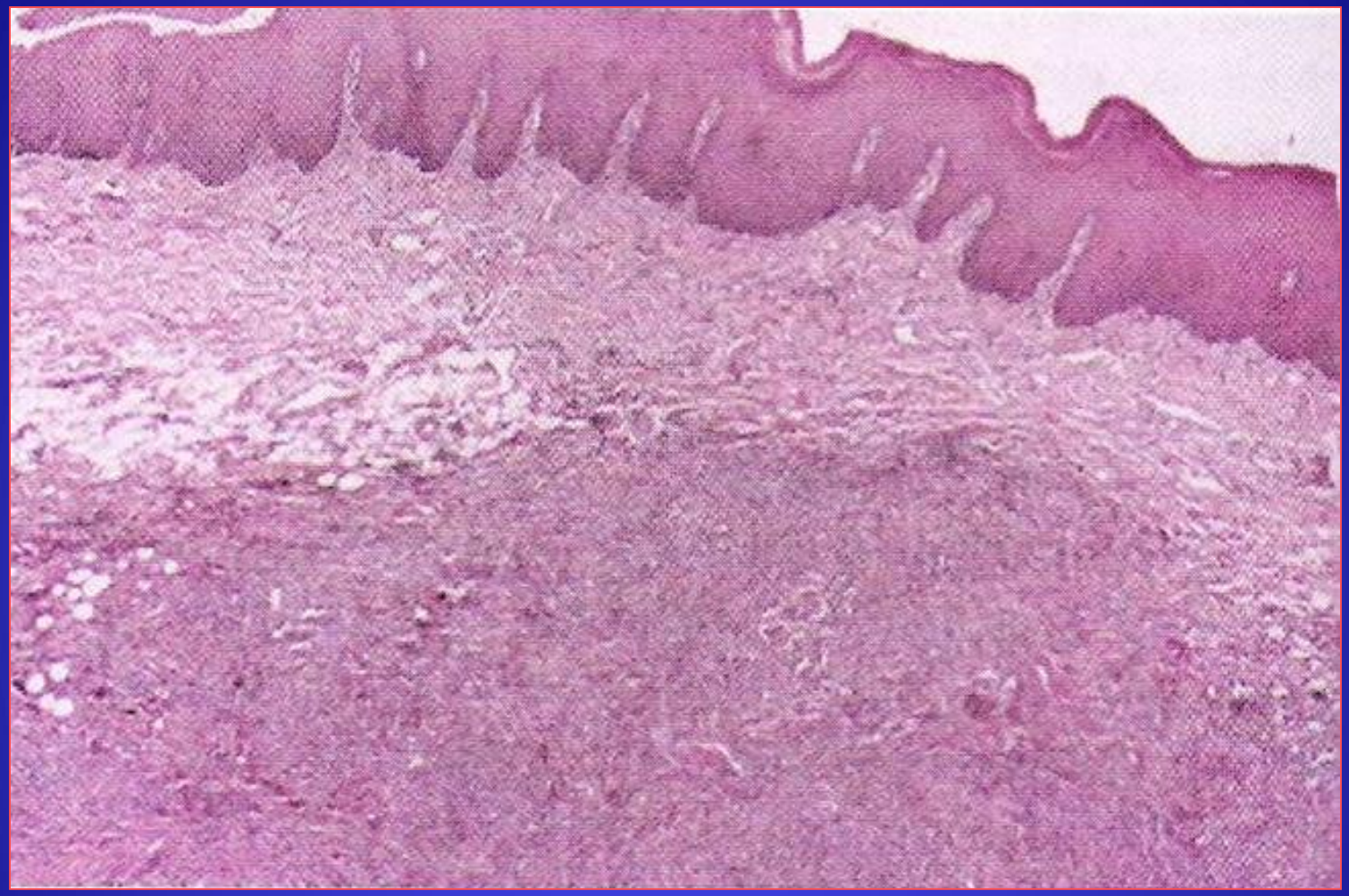
Histological features :

- Has similar histologic features in all of its clinical subtypes.
- The **early lesion (patch stage)** is characterized by a proliferation of small veins and capillaries around one or more preexisting dilated vessels, which appears slit like and are lined by plump, mildly atypical epithelial cells.
- Features resemble granulation tissue. A pronounced mononuclear inflammatory cell infiltrate, scattered erythrocytes, and hemosiderin deposits may be present.
- Inconspicuous perivascular proliferation of spindle cells, but cellular atypia is minimal.

- More **advanced lesions (plaque stage)** are nodular and show increased numbers of small capillaries or dilated vascular channels interspersed with proliferating sheets of sarcomatous or atypical spindle cells, extravasated erythrocytes and abundant hemosiderin deposition.
- Slit like vascular channels without a visible endothelial lining are typically interspersed with the spindle cells.
- Infiltration by chronic inflammatory cells is also variable.
- In the **nodular stage**, all the histological features are more prominent than the plaque stage.

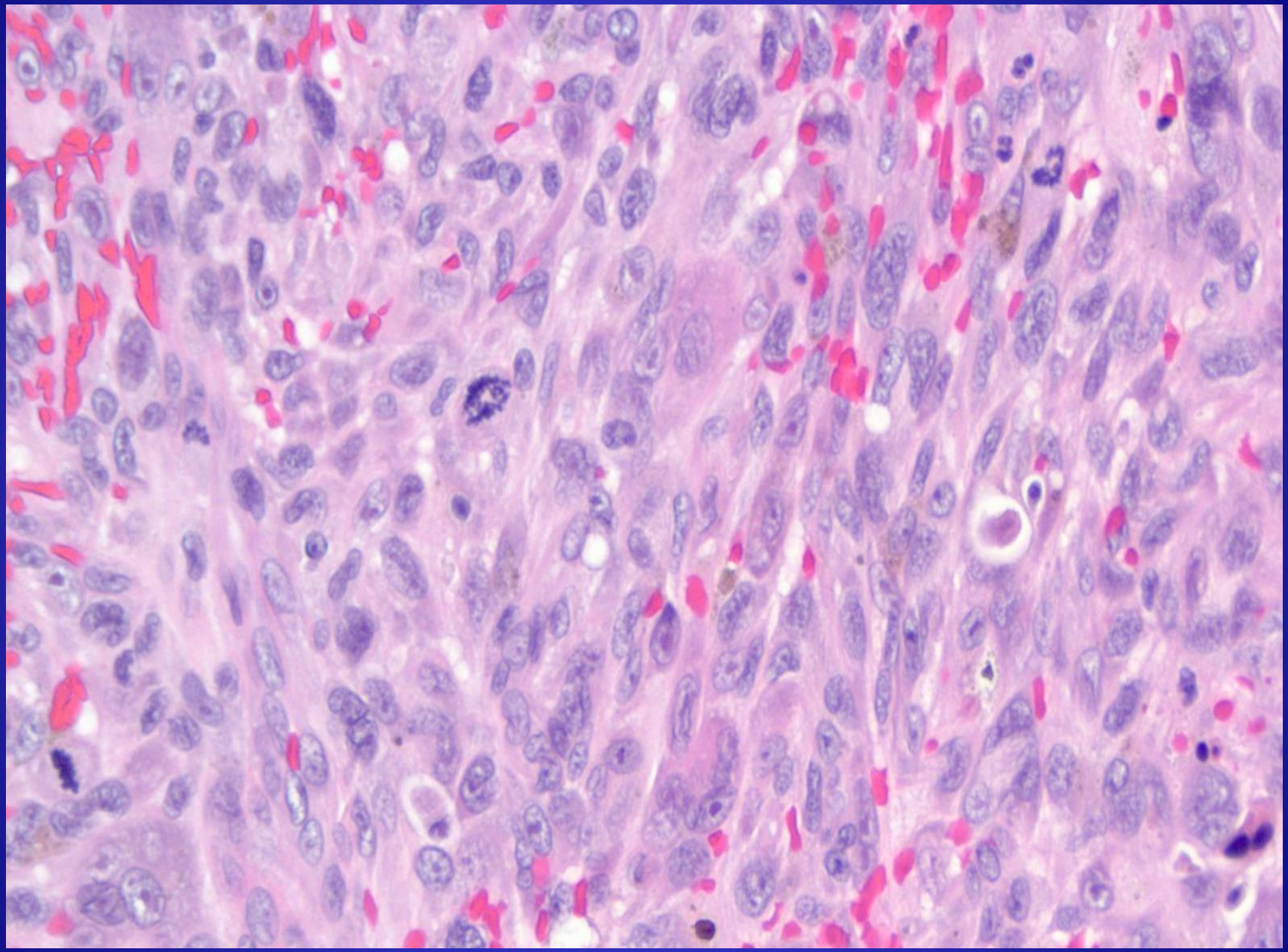


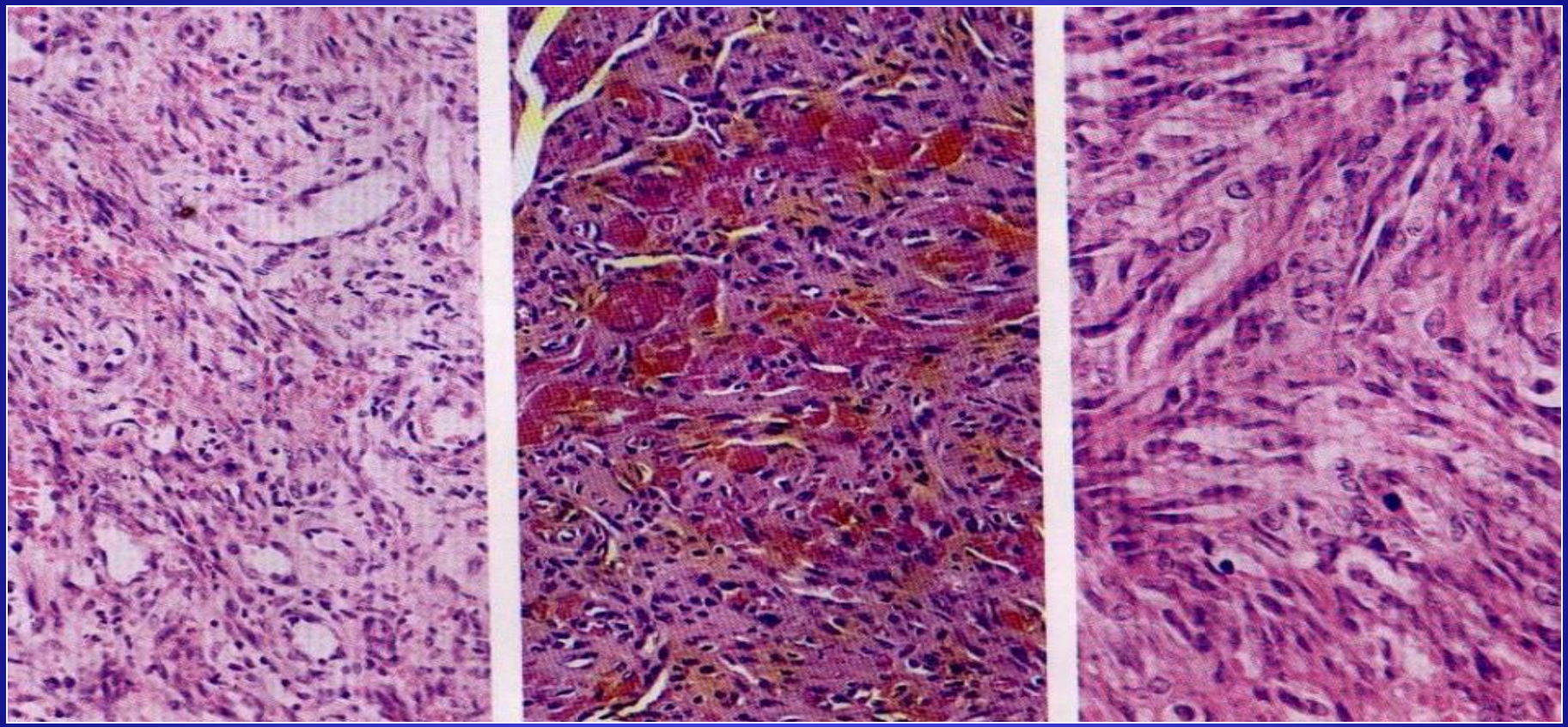
Photomicrograph of the histopathology of Kaposi's sarcoma showing fascicles of vasoformative spindle-shaped tumour cells



KAPOSI'S SARCOMA

Low power view showing the nodular stage, where the spindle cell form a tumor-like mass within the connective tissue





KAPOSI'S SARCOMA

(L)Early stage ; (M) advanced stage ; (R) nodular stage.

Treatment :

- Small or localized lesions can be surgically excised with a small surrounding healthy margin, but more recent therapies have concentrate on low dose irradiation and intralesional chemotherapy and sclerosing solutions.
- For larger and multifocal lesions, systemic chemotherapy is often effective.

OSTEOSARCOMA

- It is the third most common cancer in adolescence, occurring less frequently that only lymphomas and brain tumors.
- Though to arise from a primitive mesenchymal bone –forming cell and is characterized by production of osteoid.

Clinical features :

- Most commonly occurs in the long bones of the extremities near metaphyseal growth plates, in the femur, tibia, and humerus.
- Usually seen between 10-25 years.
- Swelling and pain, especially with activity of the involved bone, are the early features of the neoplasm.
- Complain of a sprain, arthritis, or so called growing pain.
- The patient has a history of trauma, though pathological fractures are not particularly common, except in the Telangiectatic type of osteosarcoma.
- Extremity involvement may result in a limp.

Predisposing factors :

- Rapid bone growth appears to predispose patients to oesteosarcoma.
- Exposure to radiation is the only known environmental risk factor.
- A genetic predisposition may exist.
- Bone dysplasias including Paget's disease and fibrous dysplasia.
- Regional lymphadenopathy is unusual.

Oral manifestations :

- Swelling of the involved area, often producing facial deformity and pain, followed by loose teeth, paresthesia, toothache, bleeding, nasal obstruction and a variety of other manifestation.
- Median age of patients at the time of appearance of the first symptom is 27 years.
- Has a predilection for occurrence in the mandible, than in the maxilla and in males than in females.
- It develops with considerable frequency in bone affected by osteitis deformans or Paget's disease, & fibrous dysplasia.
- It has been seen that bone that has been subjected to therapeutic x-ray radiation may undergo malignant transformation.
- Surprisingly, In nearly all cases of osteosarcoma of the jaws, there is no preceding history of trauma or of Paget's disease.

OSTEOSARCOMA OF THE MAXILLA

OSTEOSARCOMA :

This patient shows a firm painful swelling of the maxilla of recent onset.

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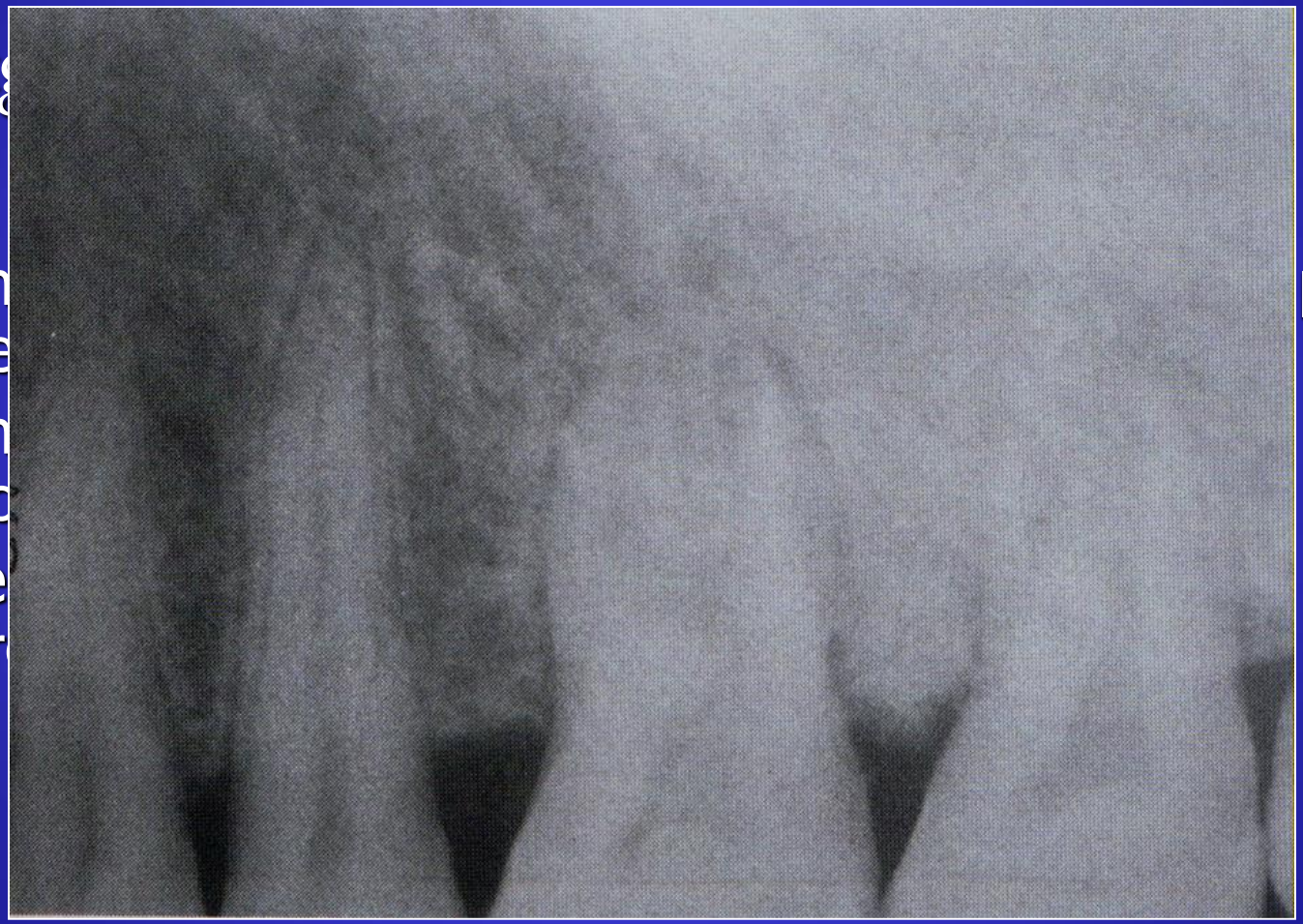

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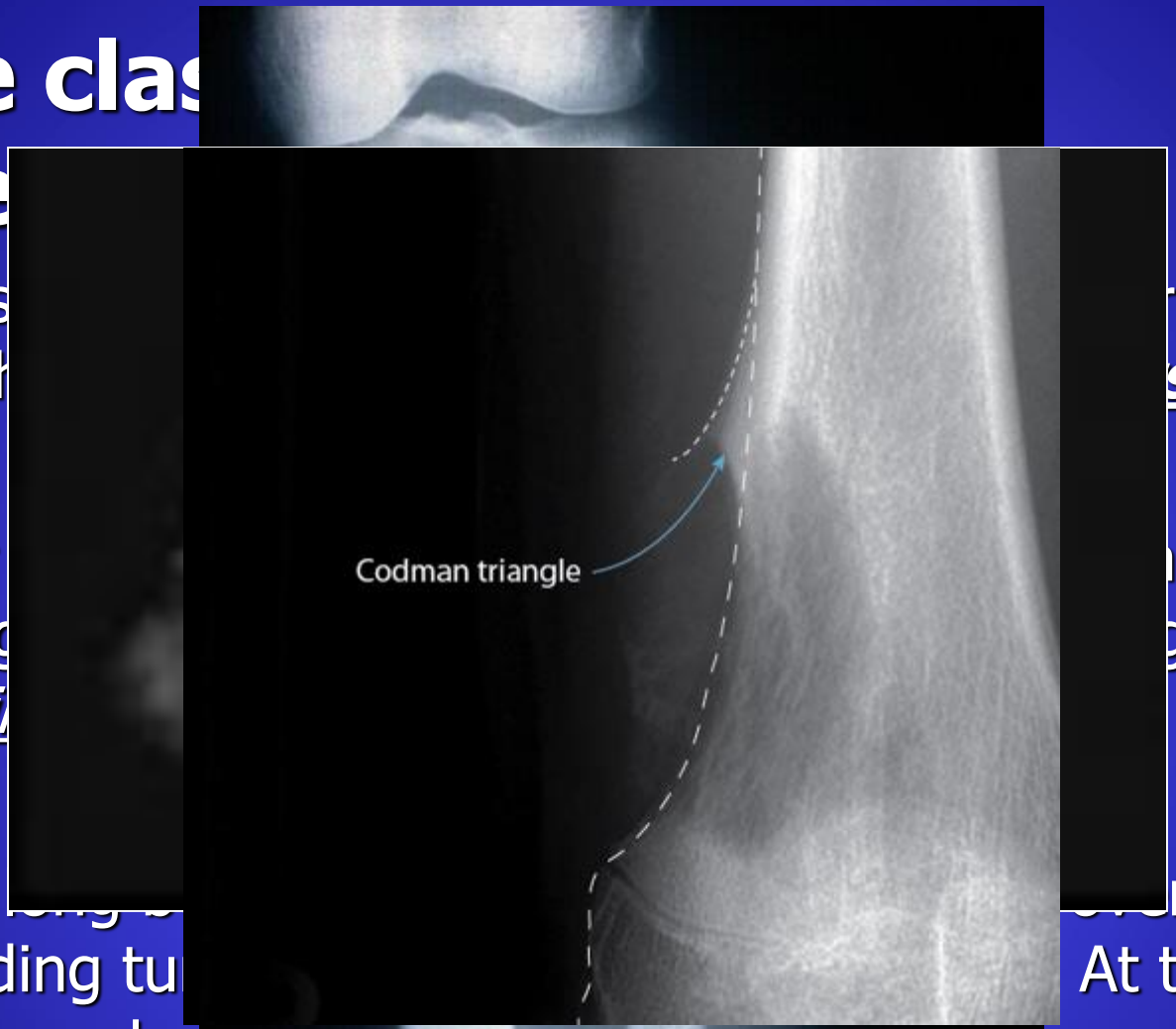
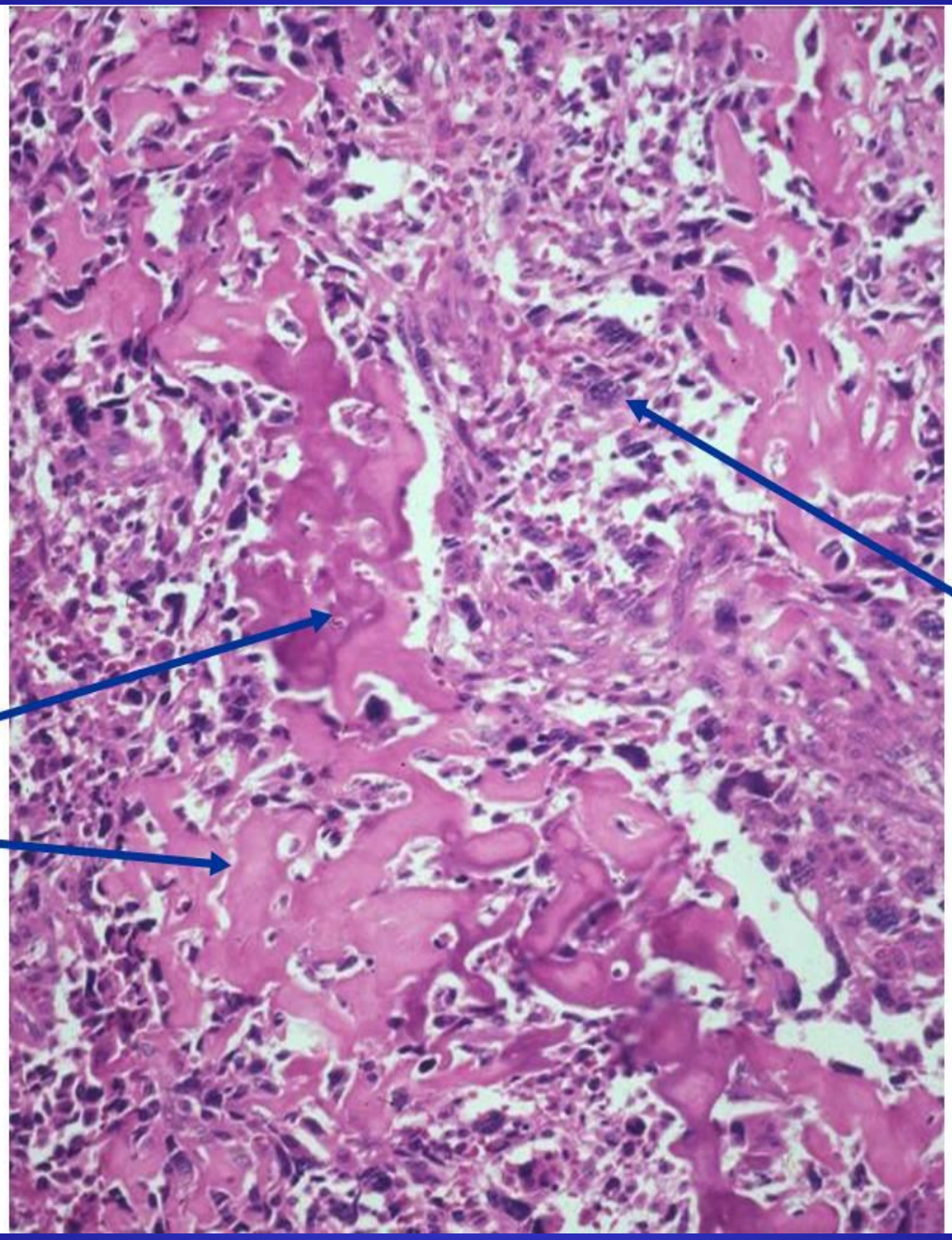


Histological features

- Characterize by the proliferation of both atypical osteoblasts and their less differentiated precursors.
- Characteristic feature is the presence of osteoid formed by malignant osteoblasts in the lesion, even at sites distant from bone.
- Stromal cells may be spindle shaped and atypical with irregularly shaped nuclei.
- In the osteoblastic subtype, atypical neoplastic osteoblasts exhibit considerable variation in size and shape, show large hyperchromatic nuclei and are arranged in a disorderly fashion about the bony trabeculae.

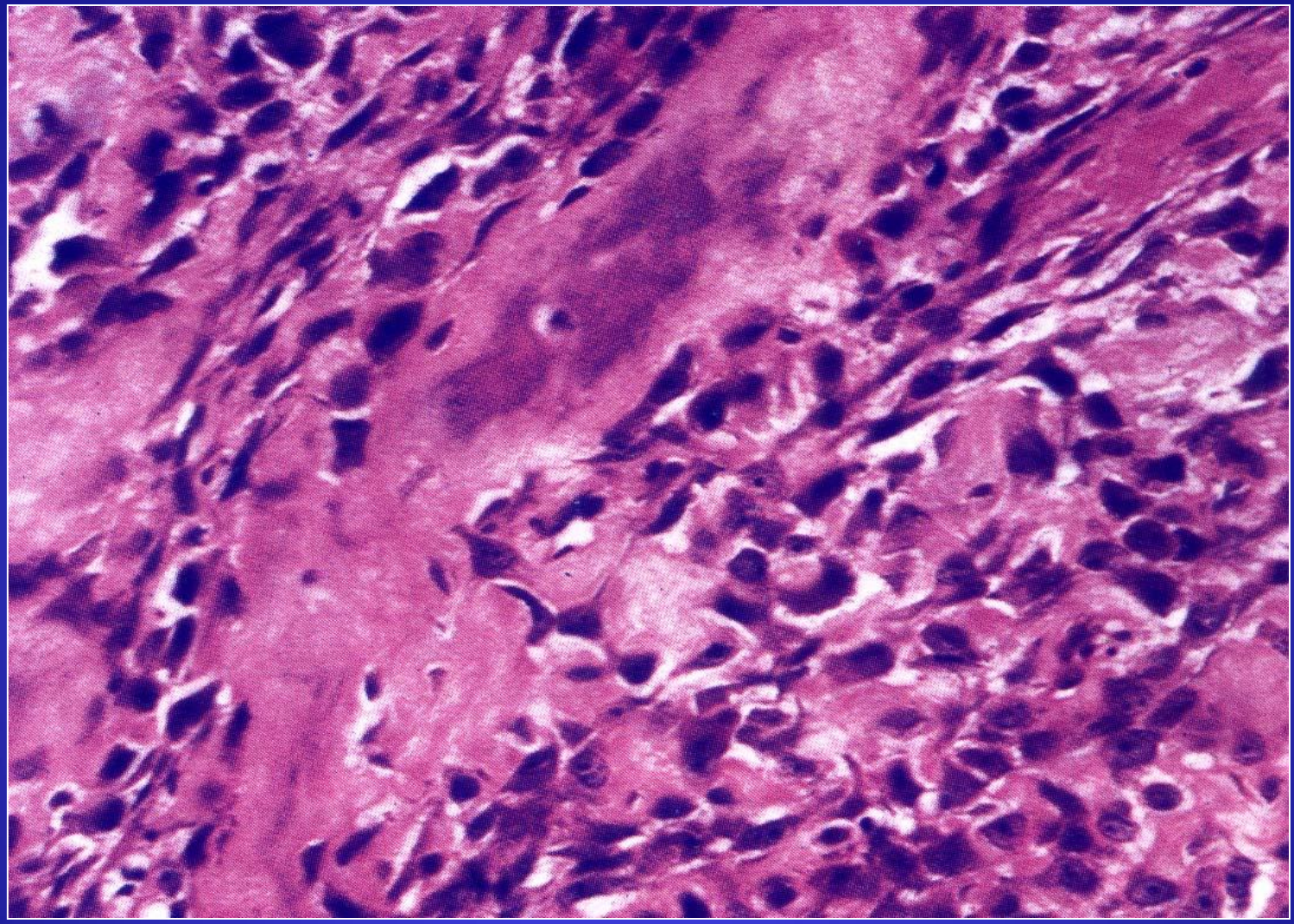
- In addition there is a great deal of new tumors, osteoid and bone formation, mostly in an irregular pattern and some times in solid sheaths rather than in trabeculae.

- Varying degrees of proliferation of anaplastic fibroblasts are also found and in the absence of significant amounts of tumors, osteoid or bone, when these cells predominate, the lesion is designated as a fibroblastic type of osteosarcoma.



Osteoid Production

High Grade Anaplastic Spindle Cells with Large Hyperchromatic Nuclei



OSTEOSARCOMA

Treatment :

- In case of long bone involvement, amputation is the prime requisite.
- In other sites it must be treated by radical resection but especially in the jaws it is difficult to excise completely.
- Neoadjuvant (preoperative) chemotherapy has been found to facilitate subsequent surgical removal by shrinking the tumor.
- At least 50% of jaw lesions show metastasis usually to the lungs.

MALIGNANT LYMPHOMA

It is a neoplastic proliferative process of the lymphopoietic portion of the reticuloendothelial system that involves cells of either the lymphocytic or histiocytic series in varying degrees of differentiation and occurs in an essentially homogeneous population of a single cell type. The process is basically multicentric in character.

NON-HODGKIN'S LYMPHOMA

- **They are a heterogeneous group of lymphoproliferative malignancies which can involve lymph nodes and lymphoid organs as well as extranodal organs and tissues.**

Etiology

- Genetic abnormalities.
- Environmental factors – pesticides and herbicides, solvents and organic chemicals and wood preservative. Patients who receive cancer chemotherapy and radiation therapy are at increased risk.
- Viruses including the EBV in Burkitt's lymphoma, Sino nasal Lymphoma, and lymphomas in immunocompromised patients; HTLV -1 in adult T- cell lymphoma/leukemia; and HHV-8 in body cavity- based lymphomas in patients with HIV infections.

- Immunodeficiency states that seem to predispose to NHL include congenital immunodeficiency states as well as acquired immunodeficiency states.
- Connective tissue disorders, including Sjogren's syndrome, rheumatoid arthritis, chronic lymphocytic thyroiditis, and SLE are also associated with increased risk of NHL.

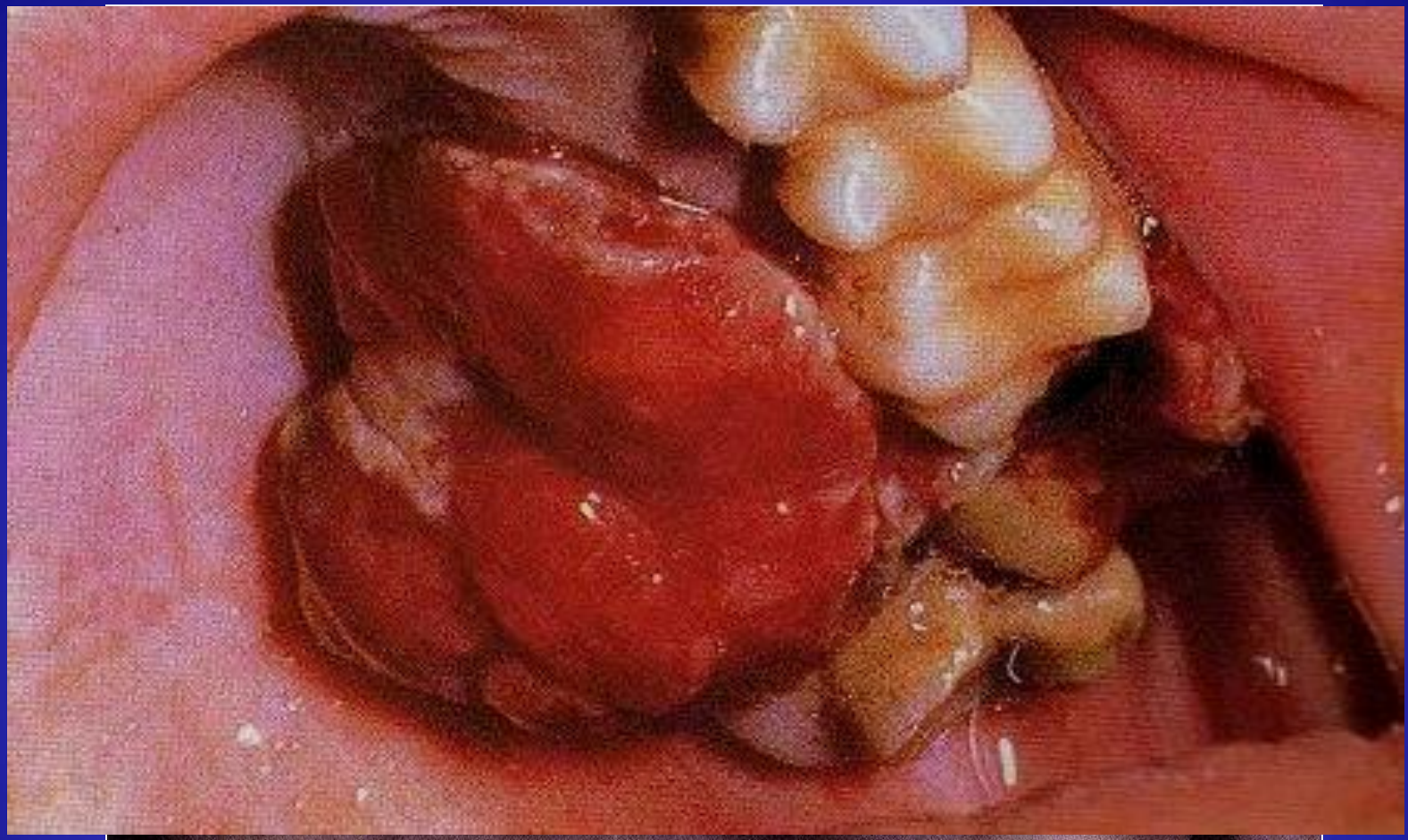
Clinical features :

- Mean age is more than 50 years. More common in males.
- Lymphadenopathy is the most common manifestation of lymphoma.
- Systemic symptoms like fever, night sweats, weight loss, and fatigue, pruritus are noticed. Sometimes waxing and waning lymphadenopathy may be seen.

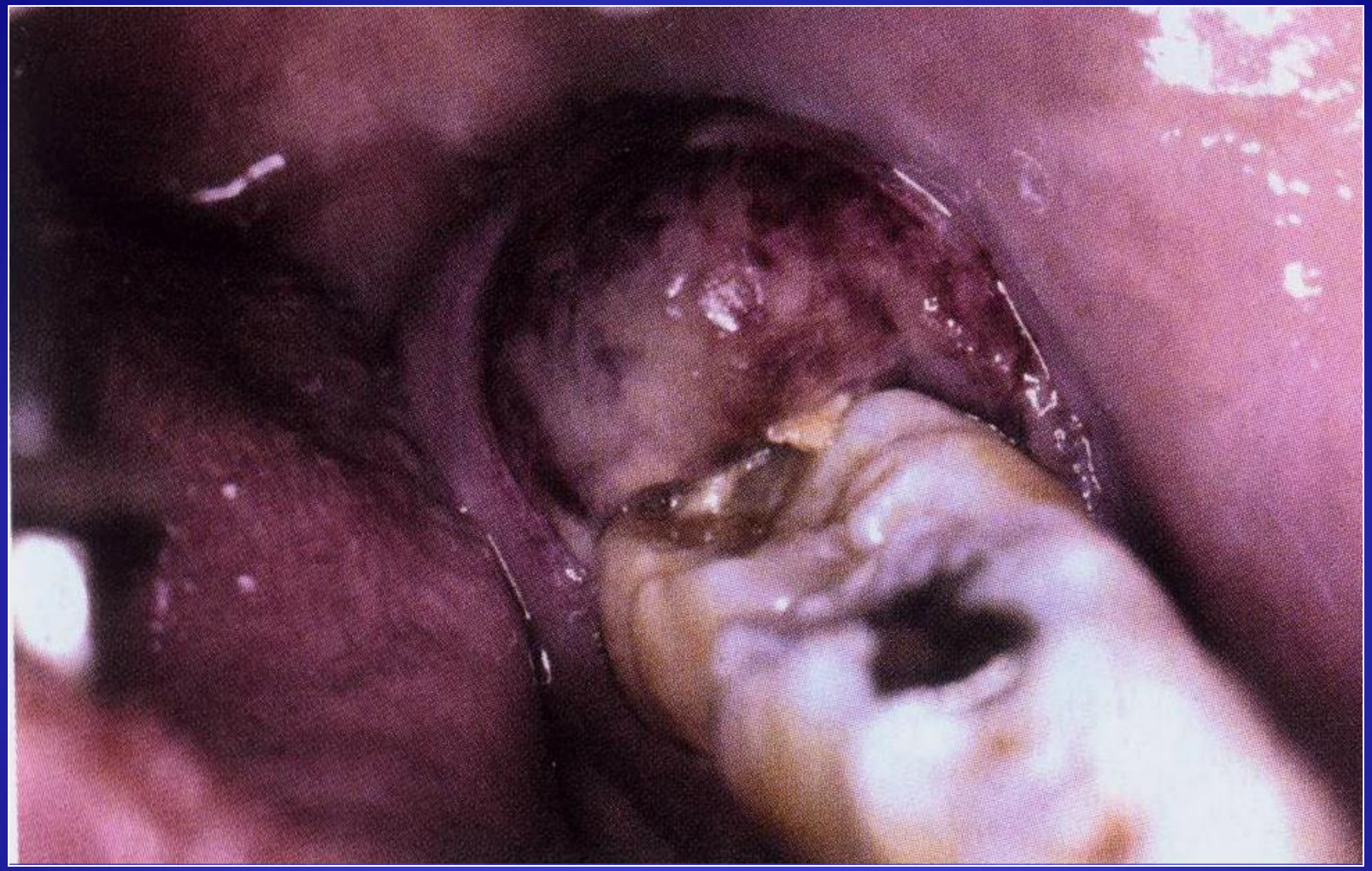
- Some times shortness of breath, chest pain, abdominal pain or bone pain will be seen.
- Neurological symptoms are important because CNS involvement may occur with aggressive lymphomas.
- Oral lesions are characterized by swelling which may grow rapidly and then ulcerate.
- When underlying bone is involved, tooth mobility and pain may develop. Also paresthesia of mental nerve may be seen.
- Reported as **lymphoproliferative disease of the hard palate**, these lesions proved to be NHL. occurred primarily in elderly. They were soft fluctuant swellings which were occasionally bilateral and may be ulcerated or discolored.

NON-HODGKIN'S LYMPHOMA

The matted non tender lymph node enlargement in the lateral cervical region is a common presentation of NHL.



NON-HODGKIN'S LYMPHOMA : palate is the most common location of presentation, where the tumor appears as a non tender, boggy swelling. Note the overlying telangiectatic blood vessels, a common feature of malignancy.

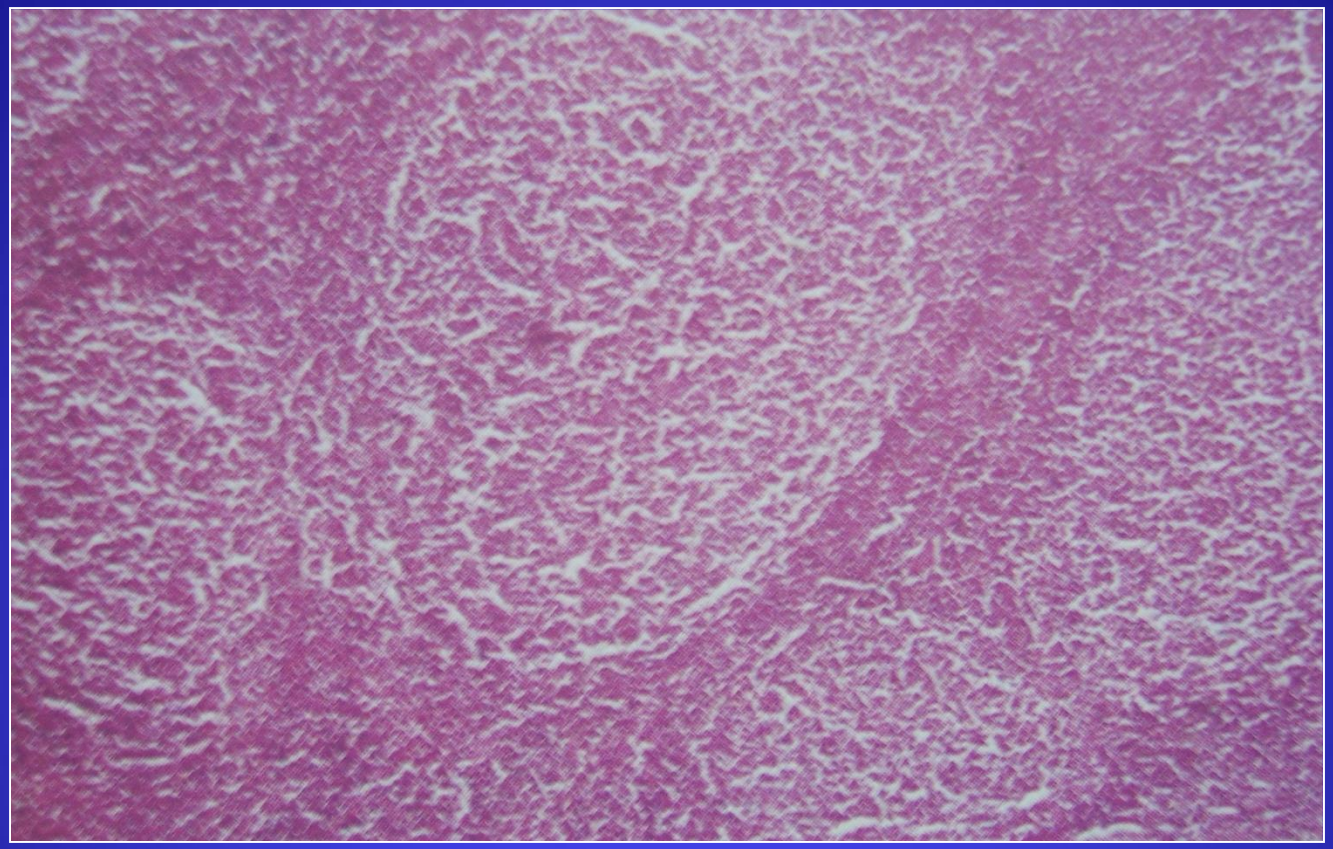


NON-HODGKIN'S LYMPHOMA

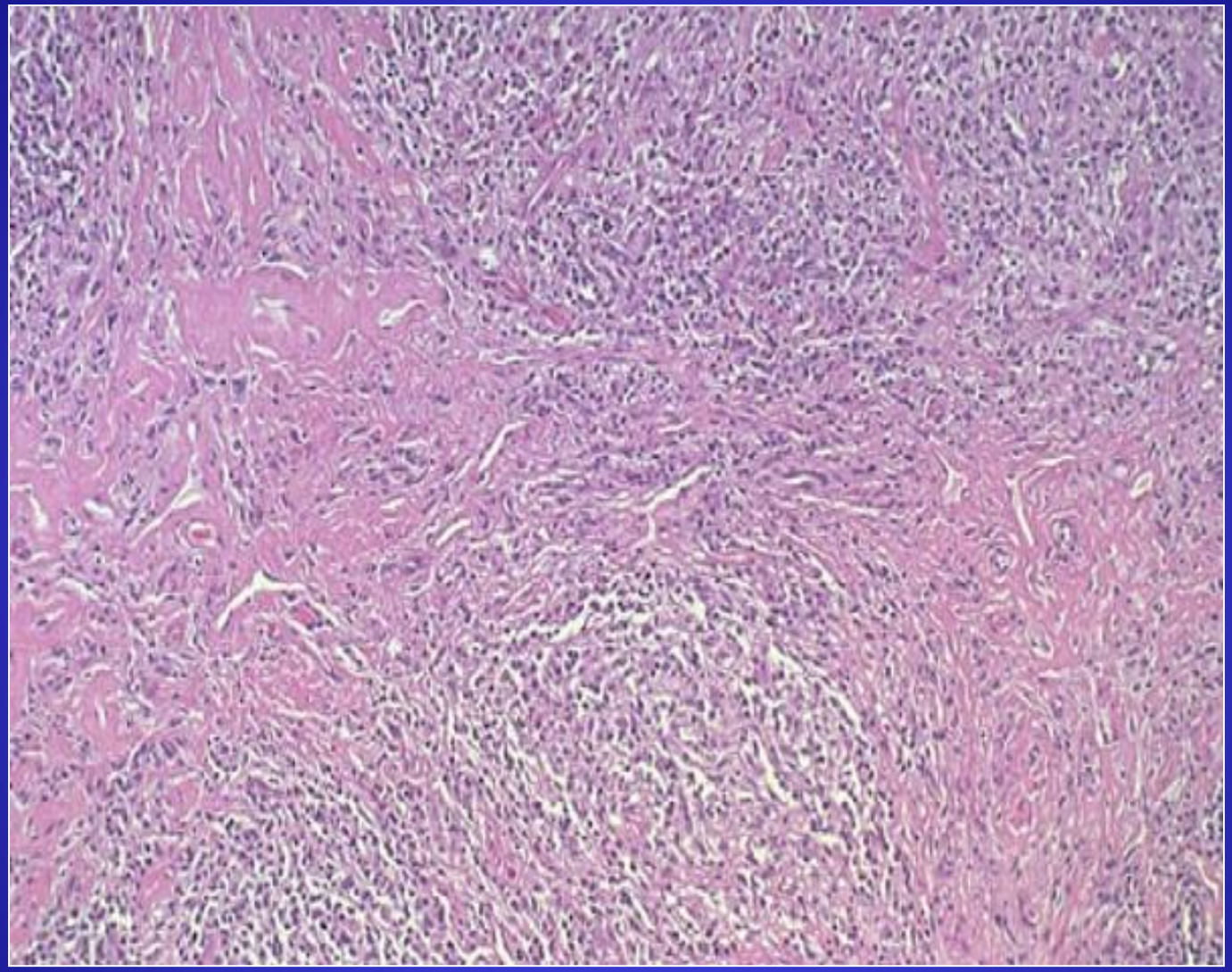
The ulcerated mass of the retromolar region represents extranodal lymphoma, which originated in bone and now involves the oral soft tissues.

Histological features :

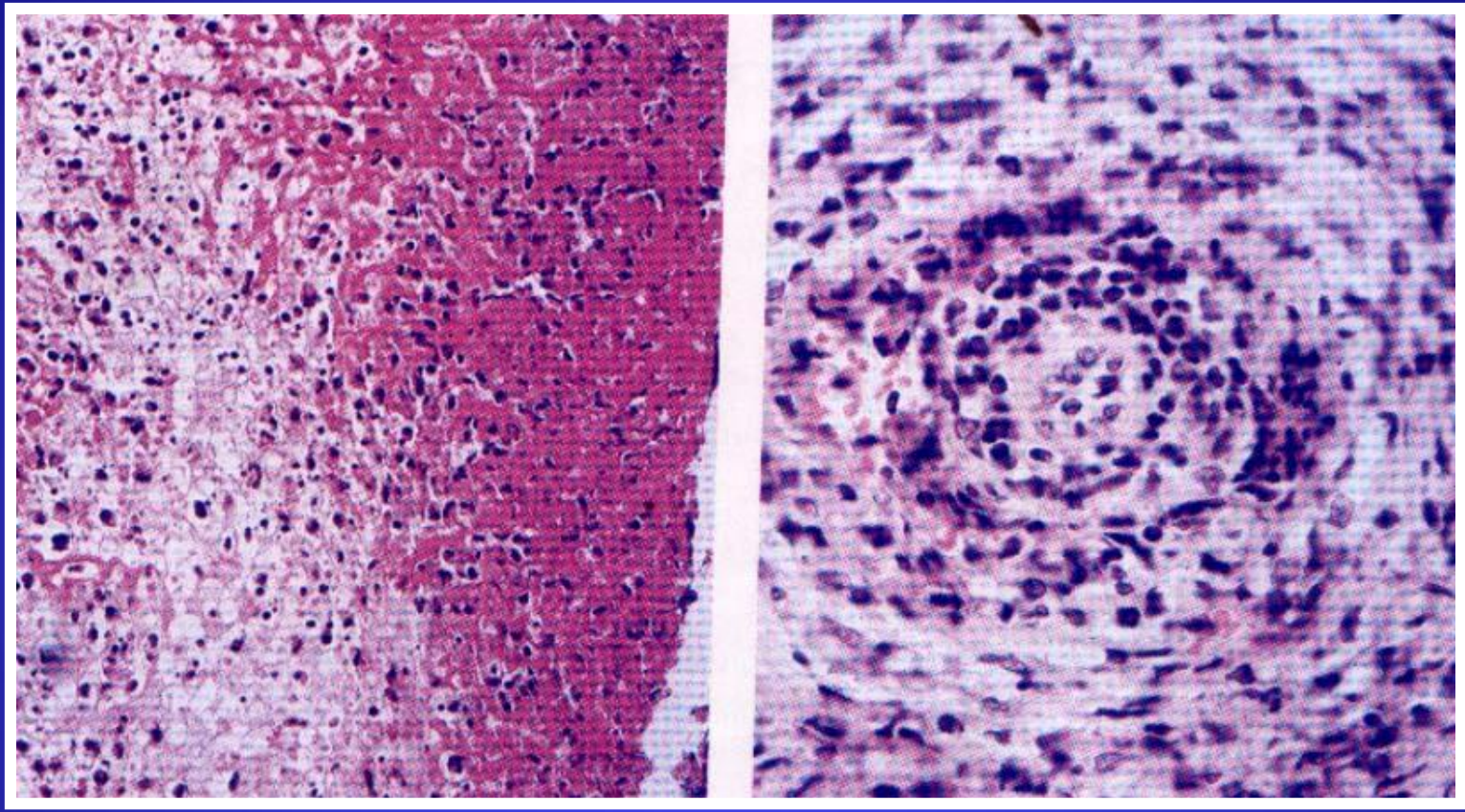
- Histologic pattern may be either nodular or diffuse.
- In the nodular pattern, neoplastic cells tend to aggregate in large clusters.
- Diffuse pattern is characterized by a monotonous distribution of cells with no evidence of nodularity or germinal centers.
- The histological pattern has a definite clinical significance. Since the nodular pattern is seen more commonly in lymphoma in adults, and exhibit a more favorable prognosis.
- Nodular lymphomas are of B cell origin whereas the diffuse type may be of B cell of T-cell origin.
- **Nodular (follicular) lymphoma** – 2 principal cell types are observed : small cells with irregular or cleaved nuclear contours & scant cytoplasm called **centrocytes** (small cleaved cells), and larger cells with open nuclear chromatin, several nucleoli, and modest amounts of cytoplasm called as **centroblasts.** Usually centrocytes comprise the majority.



NON-HODGKIN'S LYMPHOMA
(nodular type)



Diffuse large B-cell lymphoma



PERIPHERAL T-CELL LYMPHOMA

Treatment :

- Can be treated with radiotherapy, chemotherapy (forms the cornerstone of therapy) or biologic therapy (interferons and monoclonal therapy).
- NHLs can be divided into 2 prognostic groups :

1. The ***indolent lymphomas*** have a relatively good prognosis but are not curable in advanced stages. Most of these are nodular in morphology.
2. The ***aggressive lymphomas*** have a shorter natural history but usu.they can be cured with intensive combination chemotherapy.

AFRICAN JAW LYMPHOMA (BURKITT'S LYMPHOMA)

- It is a childhood tumor but also occurs in adults.
- It is one of the fastest growing malignancies in humans with a high growth fraction.
- It is a high grade B-cell neoplasm and has two major forms: the **endemic form** and the **nonendemic (sporadic) form.**

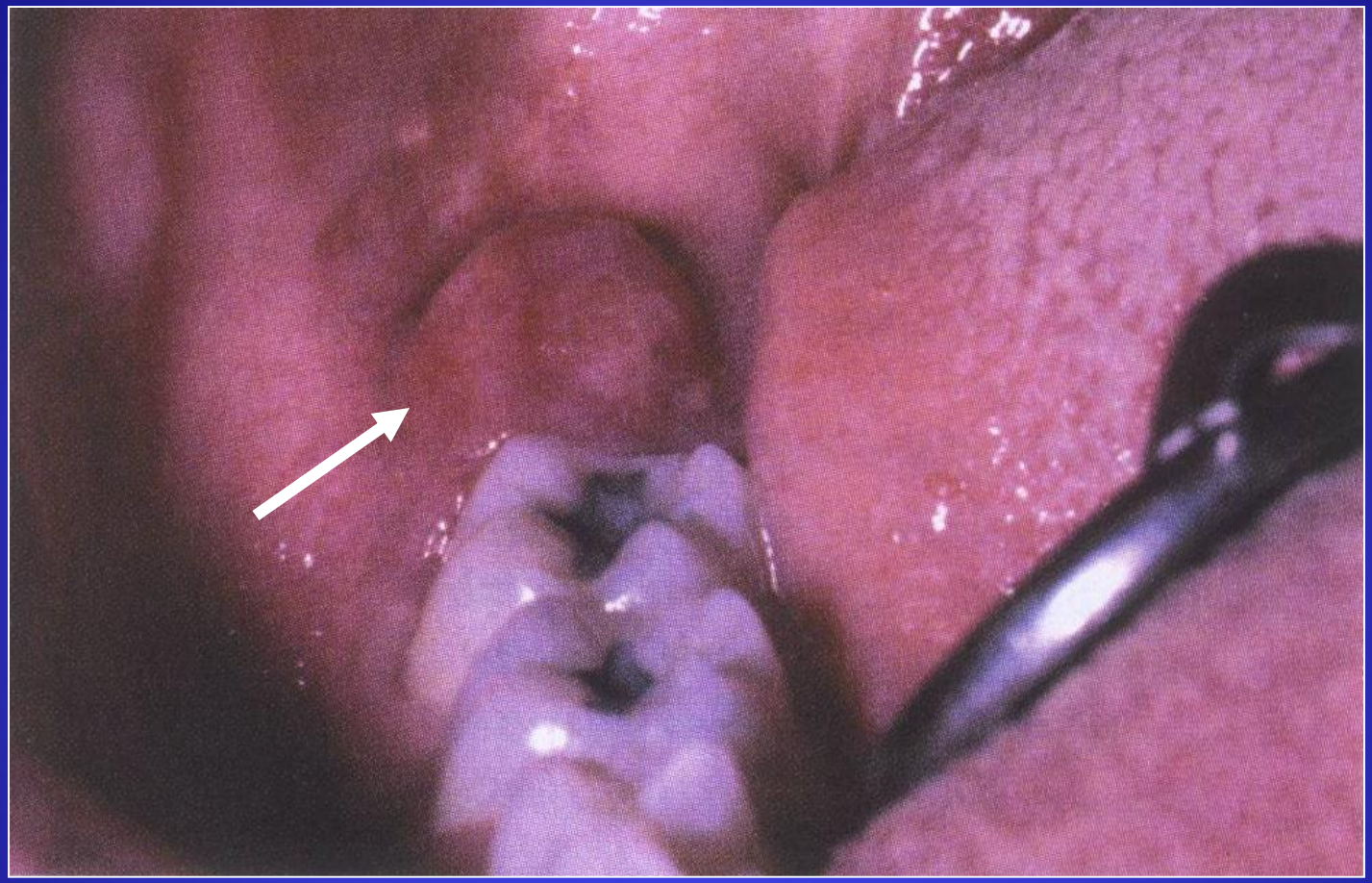
Clinical features :

- The endemic form most often involves the maxilla or mandible, while the sporadic form most often involves abdominal organs, pelvic organs, and facial bones.
- EBV is closely related with the endemic form.

- In the ***endemic form***, patients most often present with swelling of the affected jaw or other facial bones, loosening of the teeth, and swelling of the lymph nodes, which are non tender and rapidly growing, in the neck or below the jaw.
- In the ***sporadic form***, most common presentation is the abdominal tumors causing swelling and pain in the affected area.
- Because of rapid growth of BL, patients may quickly manifest significant metabolic derangement and renal function impairment.
- Major signs of BL include a soft tissue mass associated with the involvement of the jaw or other fascial bones, enlarged cervical lymph nodes, abdominal masses, and ascites.

Burkitt's lymphoma

Burkitt lymphoma
Figure 2-87
Burkitt lymphoma presenting as a large tumour of the jaw in an African child.
(Courtesy of: WHO, World Cancer Report.2003) p. 255

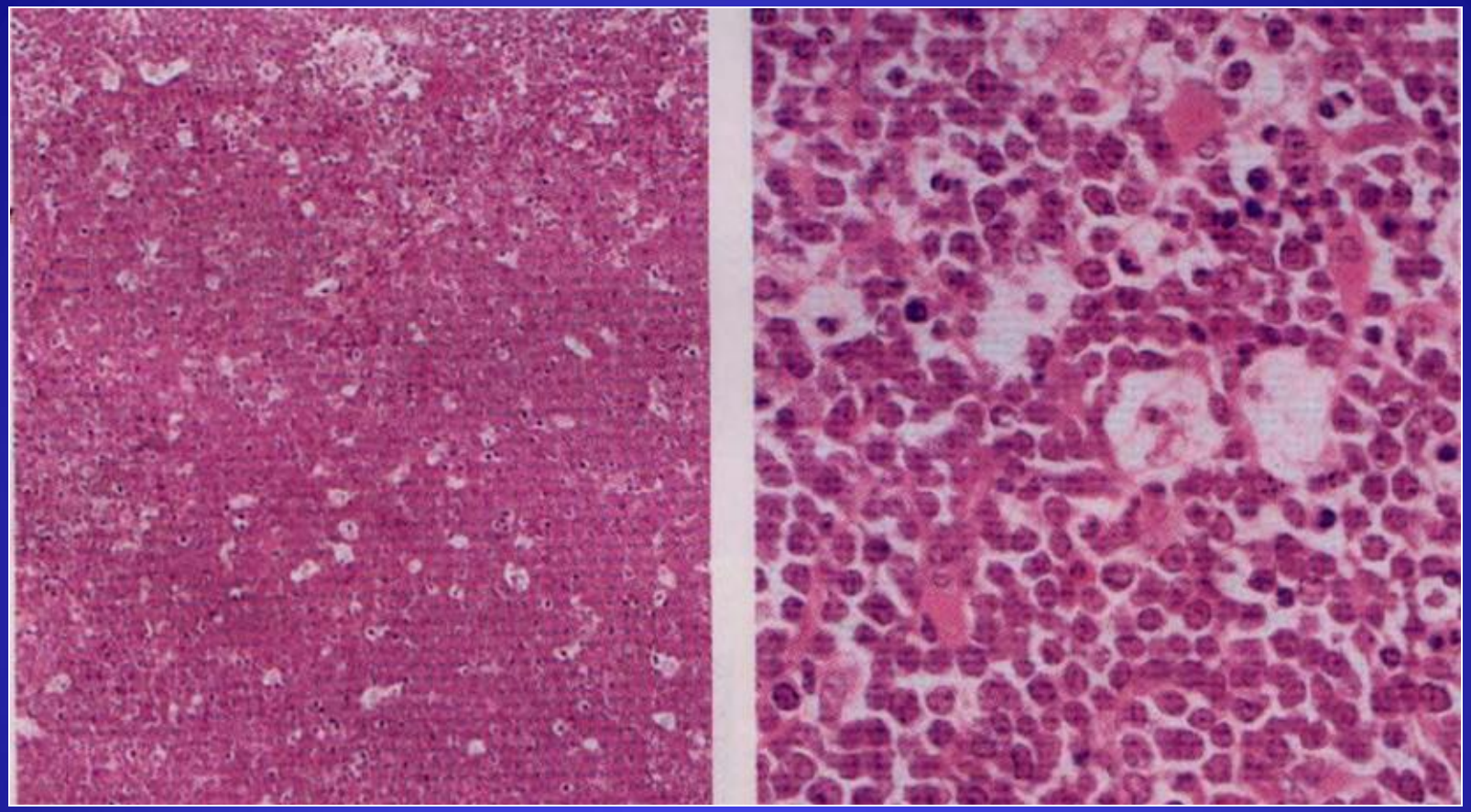


BURKITT'S LYMPHOMA

The retromolar swelling represents oral involvement in a case of sporadic form, seen commonly in the abdominal region.

Histologic features

- It is a monoclonal proliferation of B lymphocytes characterized by small non cleaved cells that are uniform in appearance and that produce a diffuse pattern of tissue involvement.
- Burkitt's cells are homogenous in size and shape, with round to oval nuclei and slightly coarse chromatin, with multiple nucleoli, and with intensely basophilic vacuolated cytoplasm that contains neutral fat.
- Frequent mitotic figures are observed.
- A characteristic ***starry sky*** appearance is imparted by scattered macrophages with an abundant clear cytoplasm, often containing phagocytic cellular debris.



Burkitt's lymphoma
(L) shows the typical starry sky appearance of this neoplasm; (R) shows homogenous cells with round nuclei and multiple nucleoli and intensely basophilic cytoplasm.

Treatment :

- With combination chemotherapy and CNS prophylaxis (intrathecal chemotherapy), the survival rate is now 60%
- Patients with limited disease have a good prognosis.
- Adults with the disease, especially those in advanced stage, do more poorly than affected children.

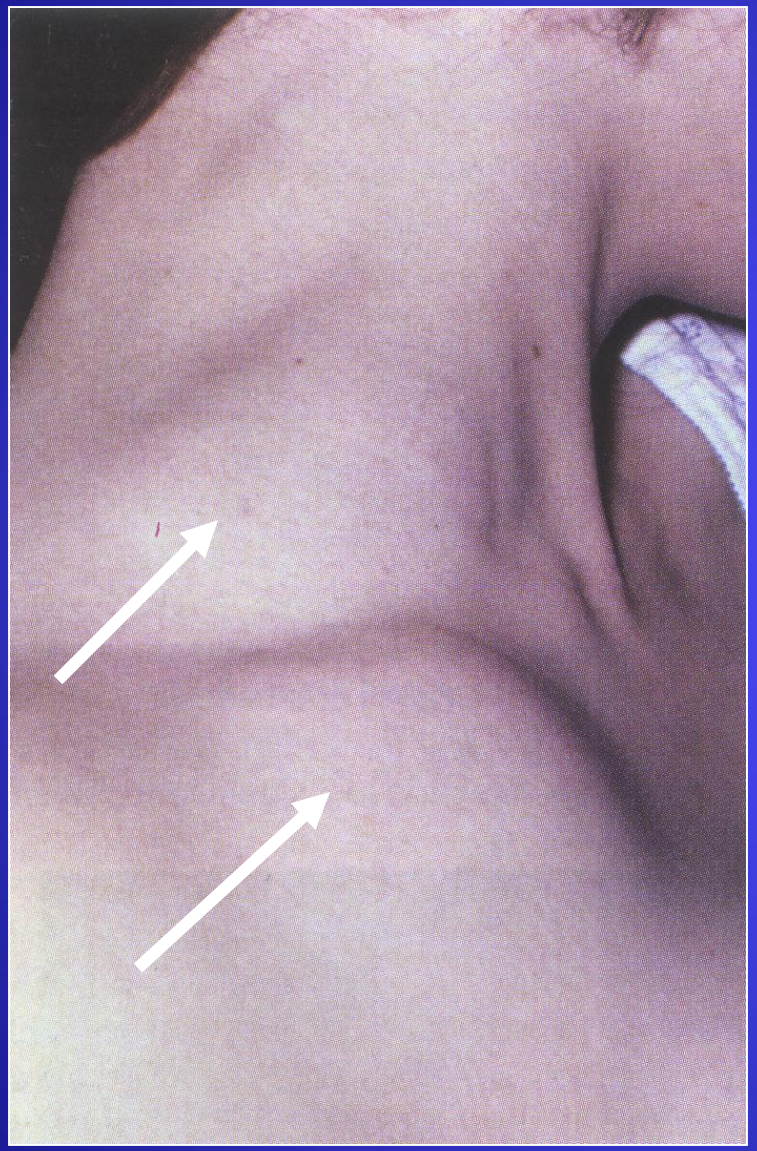
HODGKIN'S DISEASE

- It is a potentially curable malignant lymphoma
- Etiology is unknown. Infectious agents, especially the Epstein-Barr virus (EBV), may be involved in the pathogenesis.
- Patients with HIV infection have a higher incidence of HD
- Genetic predisposition may play a role in the pathogenesis.

Clinical Features :

- Age-specific incidence rates have a bimodal distribution in both genders, peaking in young adults (aged 15-34 years) and older individuals (>55 years).
- More common in males
- more common among whites.
- In developing countries, The incidence of the mixed-cellularity subtype in children is higher. On the other hand, in developed countries, young adults have the highest incidence of the nodular sclerosis subtype.

- First manifestation is painless enlargement of one or more cervical lymph nodes, axilla, inguinal area and occipital nodes. Nodes are usually firm and rubbery in consistency.
- Constitutional symptoms such as unexplained weight loss, fever, night sweats are present in about 40 per cent of patients. Chest pain, cough, and/or shortness of breath may be present due to a large mediastinal mass or lung involvement.
- Alcohol induced pain at sites of nodal disease is specific for HD patient may present with pruritus or intermittent fever.
- Back, abdomen or bone pain may occur rarely due to splenomegaly, hepatomegaly, pressure from enlarged lymph nodes, involvement of bone or vertebrae.



HODGKIN'S DISEASE

The prominent supraclavicular and cervical masses represent hodgekin's lymphoma.

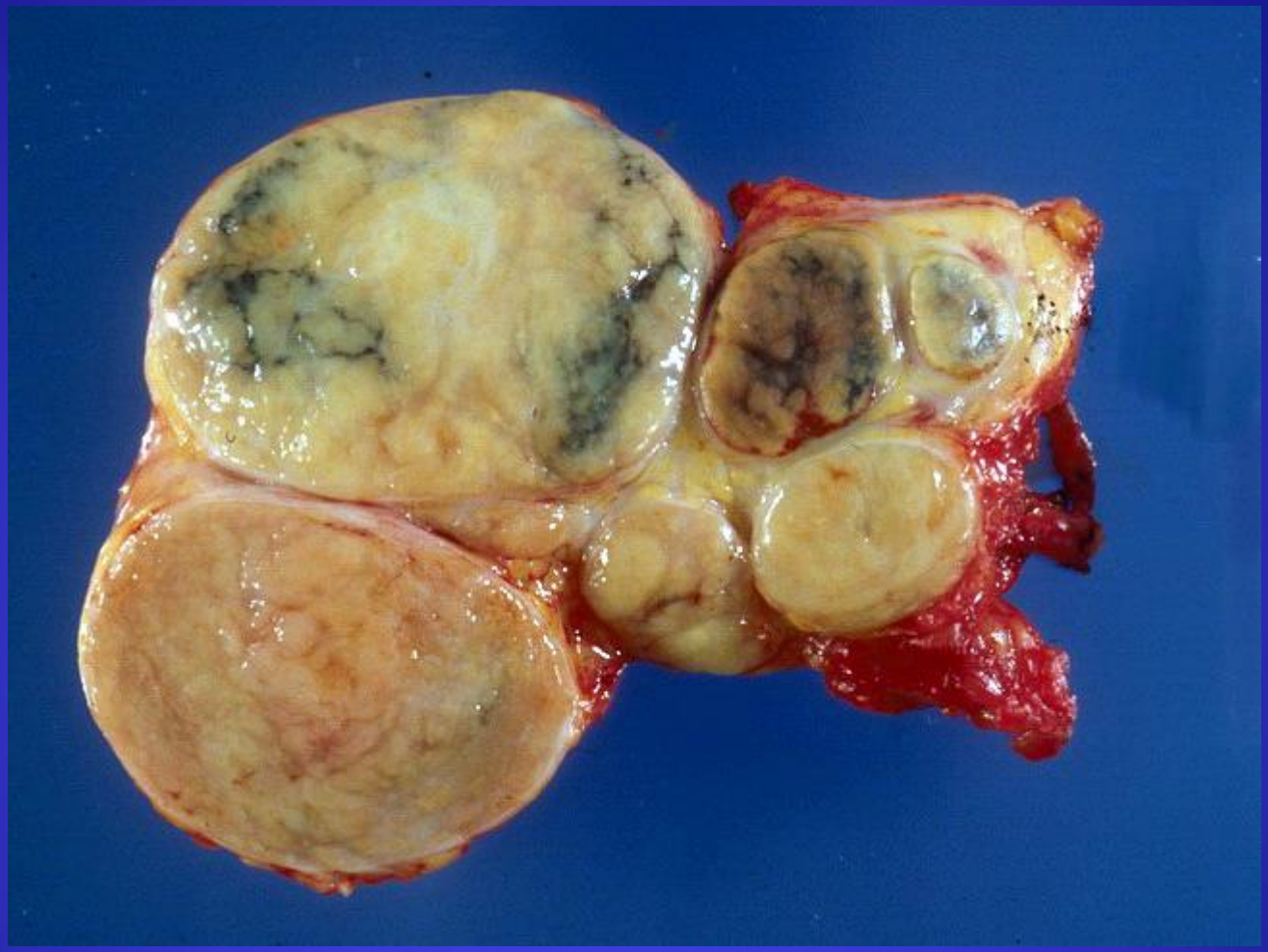
Histologic Features :

- **Nodular Sclerosis Hodgkin's disease :**

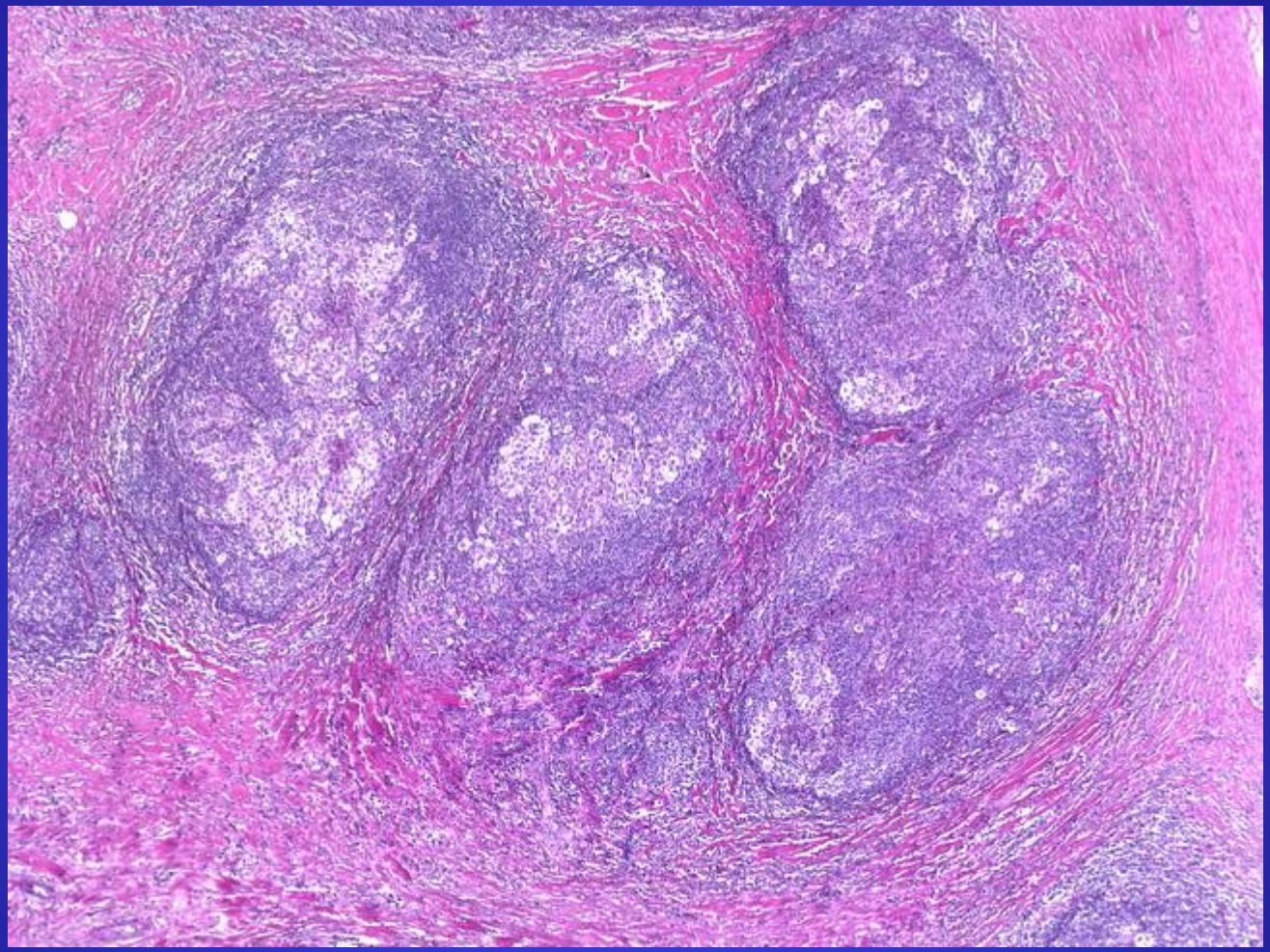
✓ Shows nodular pattern. The broad bands of fibrosis divide the node into 'nodules'. The capsule is thickened.

✓ The characteristic cell is the lacunar-type **Reed-Sternberg (RS) cell**, which has a monolobated or multilobated nucleus and a small nucleolus.

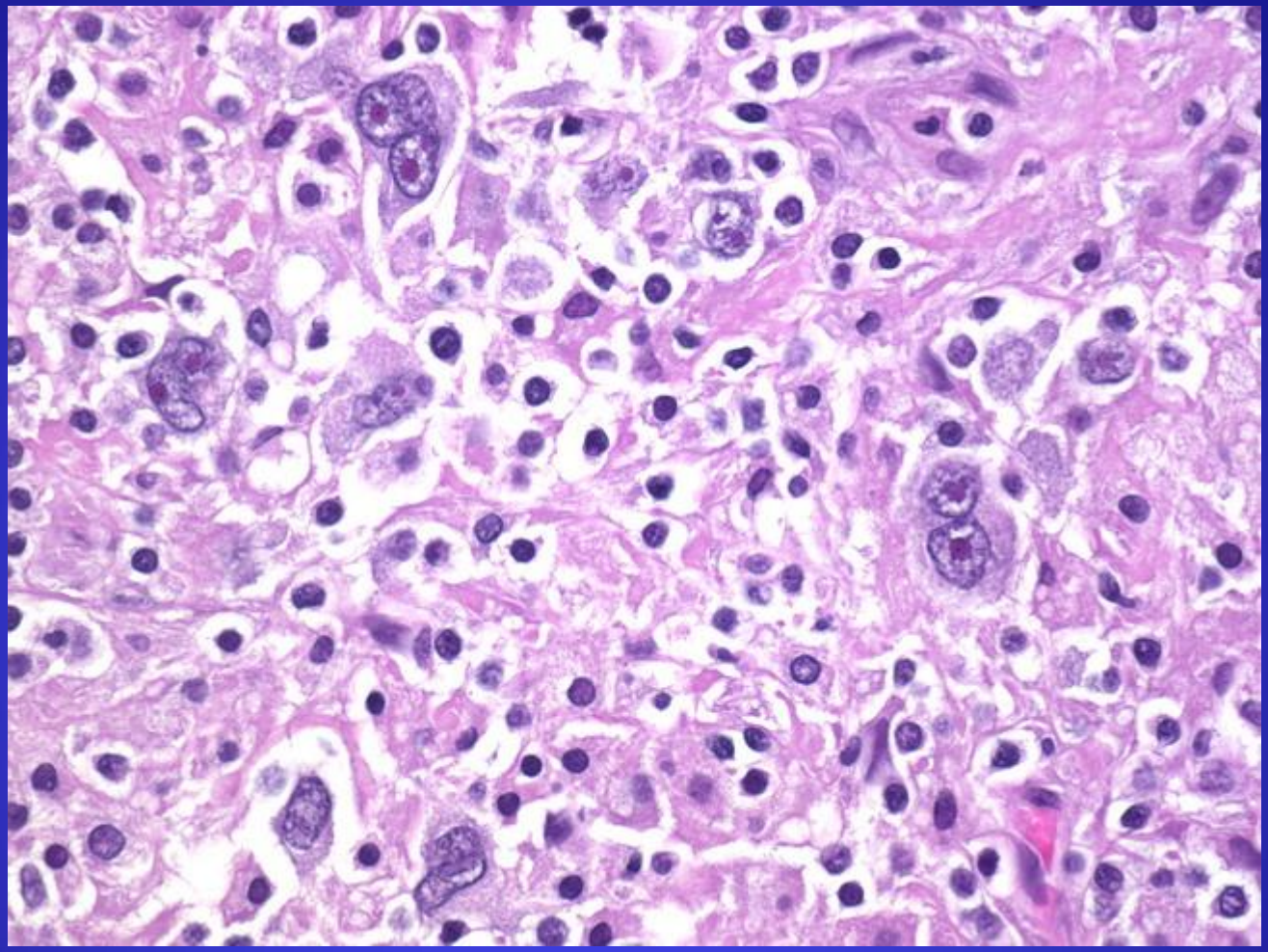
✓ Observed in adolescents and young adults.



Classical Hodgkin's Lymphoma, Nodular Sclerosis: In more advanced cases of Hodgkin's Lymphoma, several lymph nodes from the same group may become matted together, as seen in this group of mediastinal lymph nodes.



In Classical Hodgkin's Lymphoma, Nodular Sclerosis type, broad collagen bands separate the lymphoid tissue into nodules.

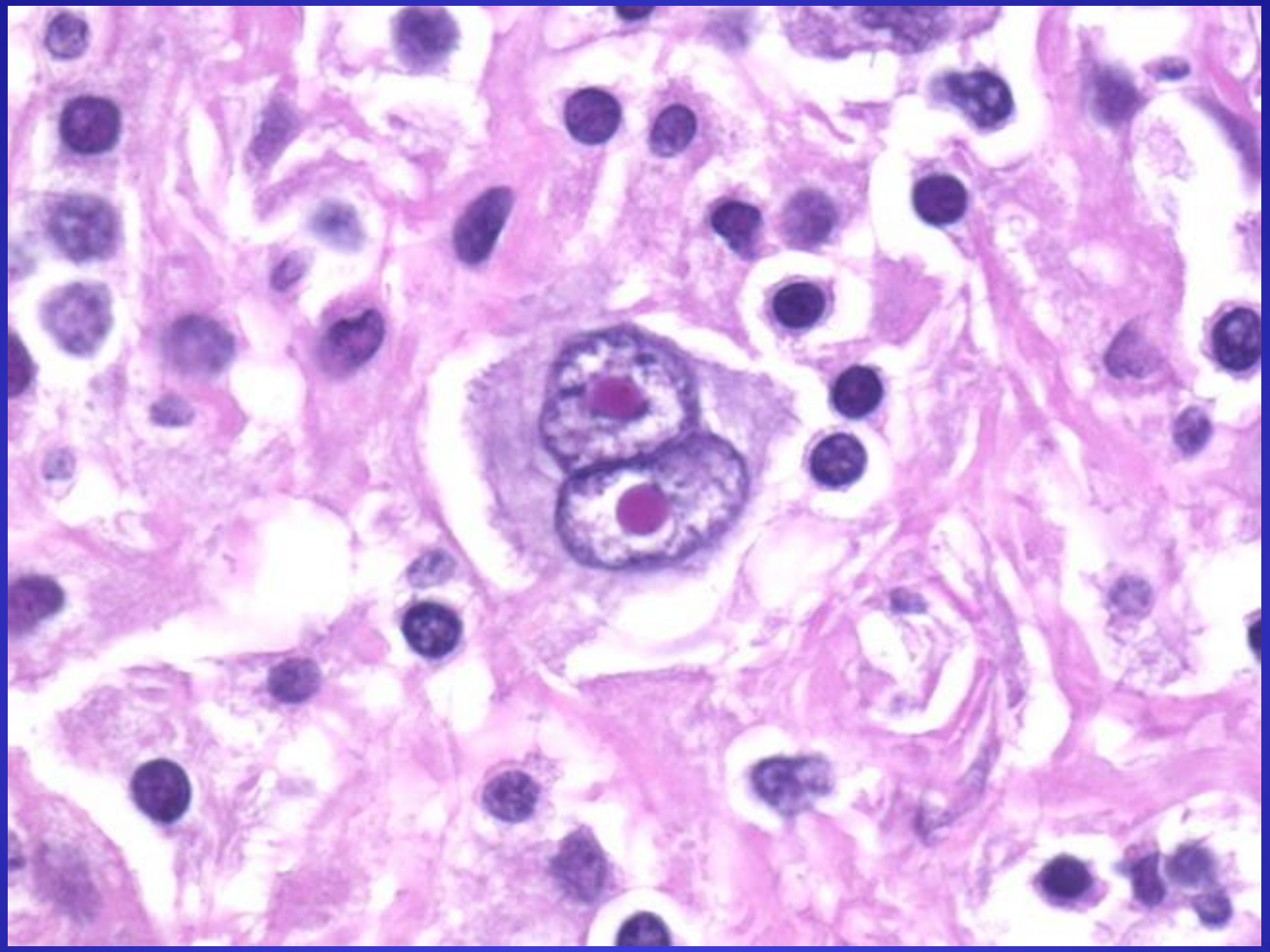


Hodgkin's Lymphoma, Nodular Sclerosis, Reed-Sternberg Cells: The conventional definition of Hodgkin's lymphoma requires the presence of Reed-Sternberg cells (many are seen in this image) in a characteristic background infiltrate composed of eosinophils, lymphocytes, plasma cells, and histiocytes. It lacks the monomorphic appearance of non-Hodgkin's lymphomas. Variable degree of fibrosis is also present (depending upon the subtype).

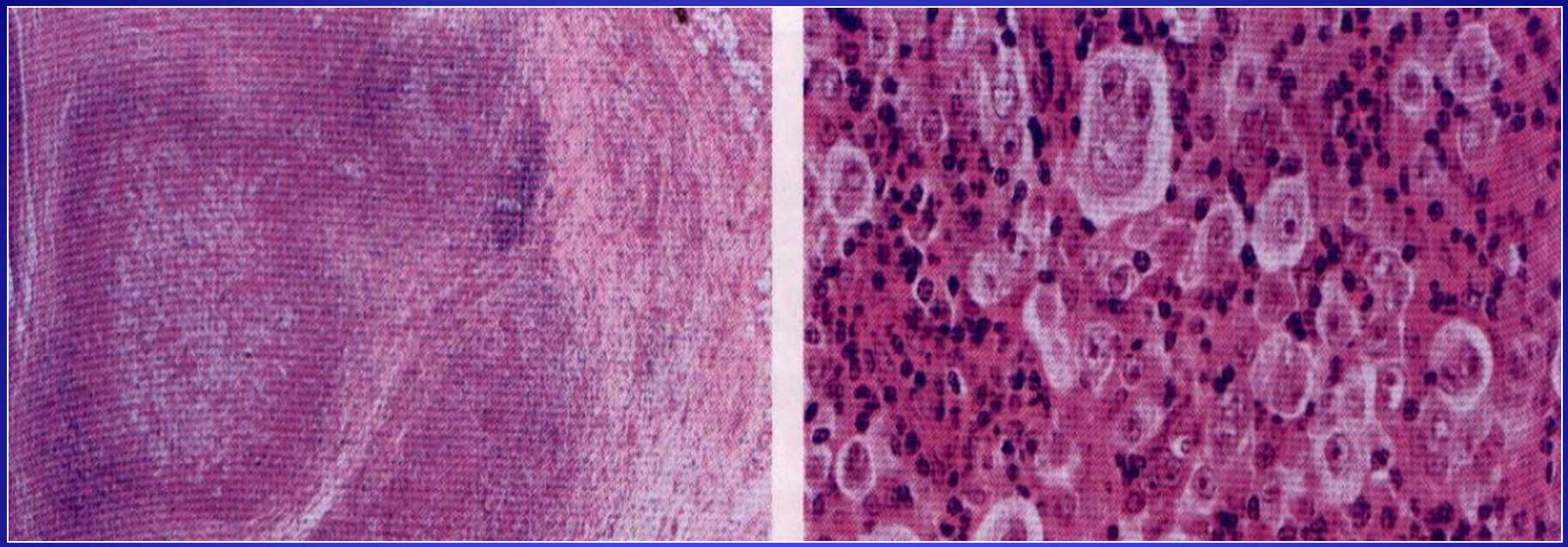
Characteristics of typical Reed-Sternberg cell :

- Characteristic malignant cells of Hodgkin's disease are large cells known as Reed-Sternberg cells abundant, amphophilic, finely granular/homogenous cytoplasm.

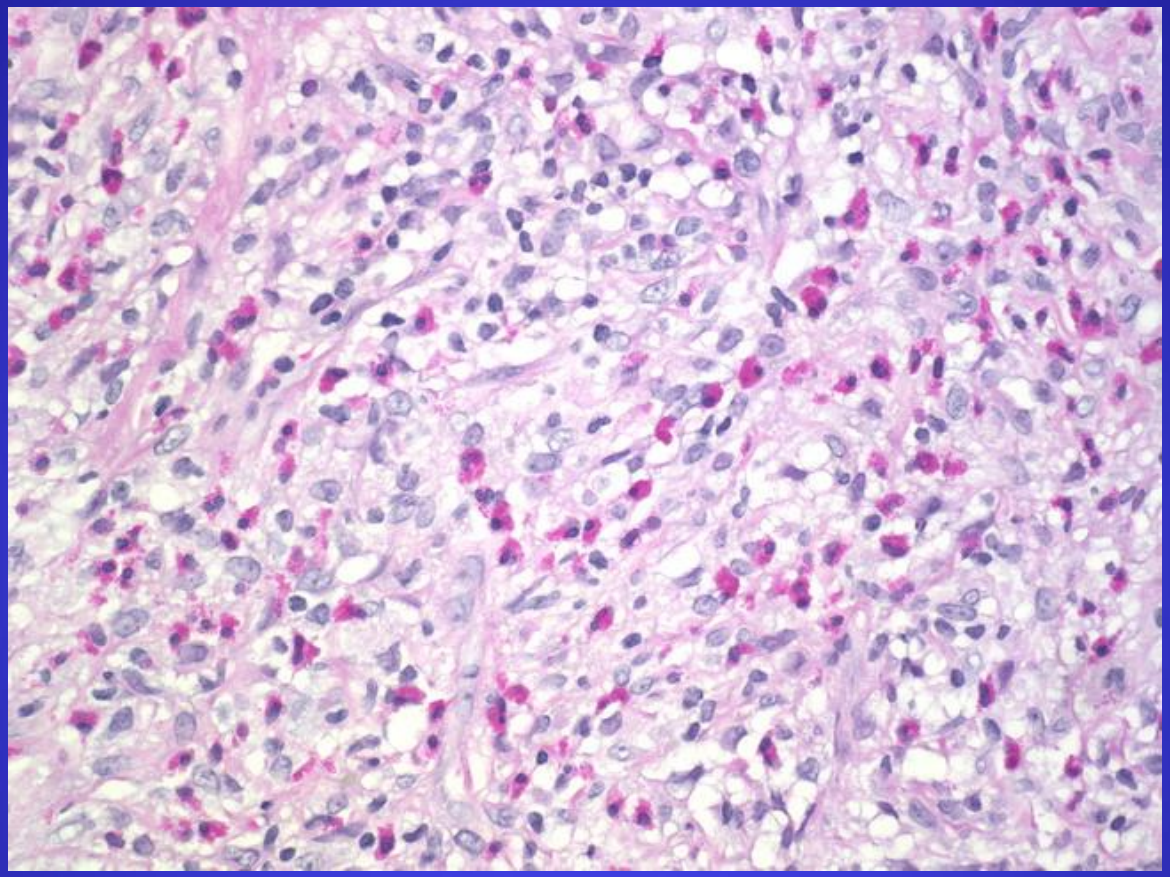
- Two mirror-image nuclei (owl eyes) each with an eosinophilic nucleolus and a thick nuclear membrane (chromatin is distributed at the cell periphery).



Classic Reed-Sternberg cell is 20 to 50 microns in size, with acidophilic or amphophilic cytoplasm, large bilobed or multilobed nucleus with vesicular or coarse chromatin. A large, round, eosinophilic central nucleolus is always present.



HODGKIN'S DISEASE – nodular sclerosis (L) shows gross capsular thickening and fibrous tissue bands break the node into discrete nodules containing foci of pale lacunar cells. (R) high power shows that the abundant pale cytoplasm of the characteristic lacunar cells has shrunk during fixation leaving a halo around the cells



he characteristic polymorphic background seen in Hodgkin's lymphoma is better appreciated in this image. Numerous eosinophils, lymphocytes, a few plasma cells, and histiocytes are present with some background sclerosis.

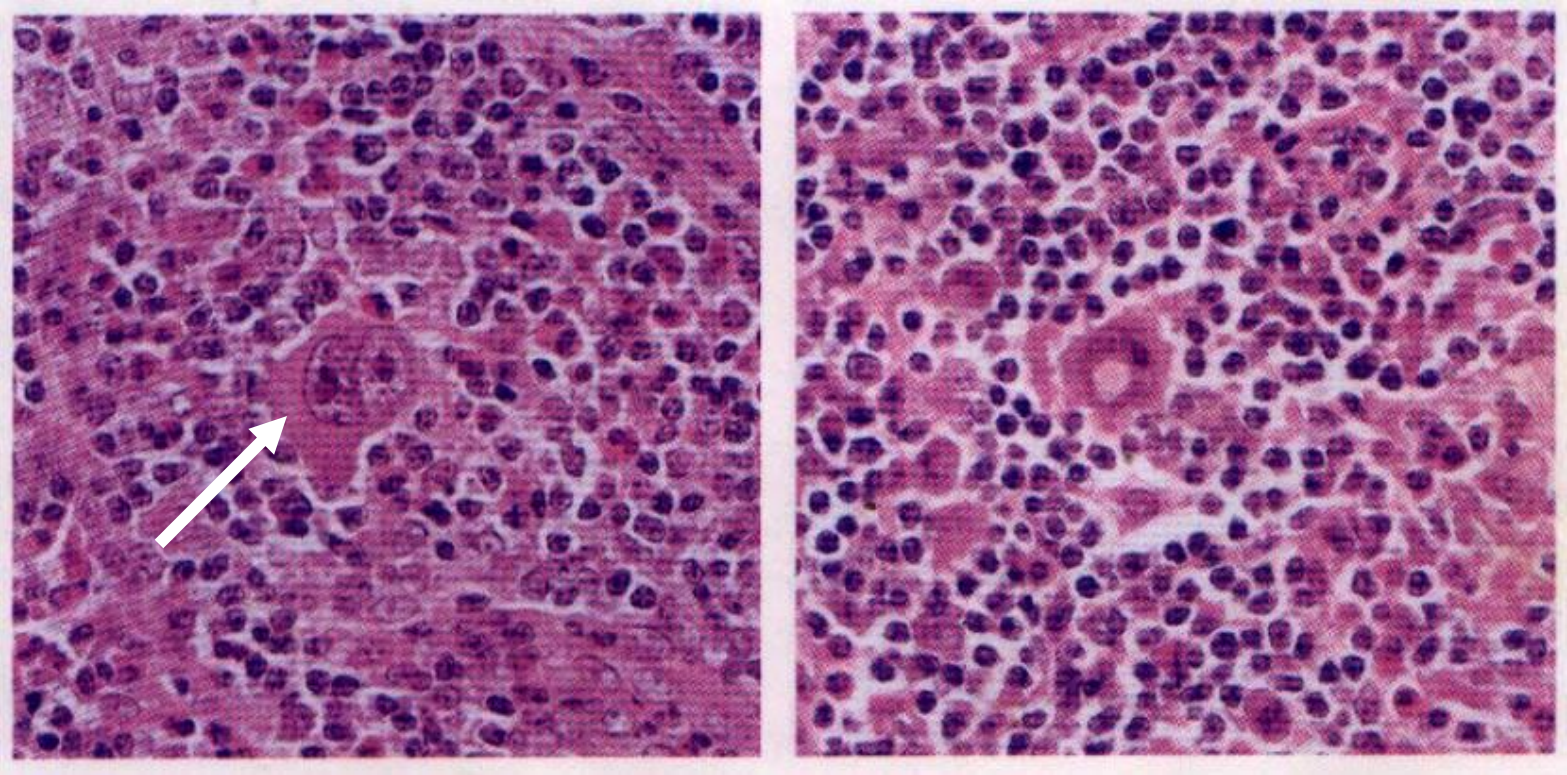
- **Mixed-cellularity Hodgkin's disease :**

✓ The infiltrate is usually diffuse.

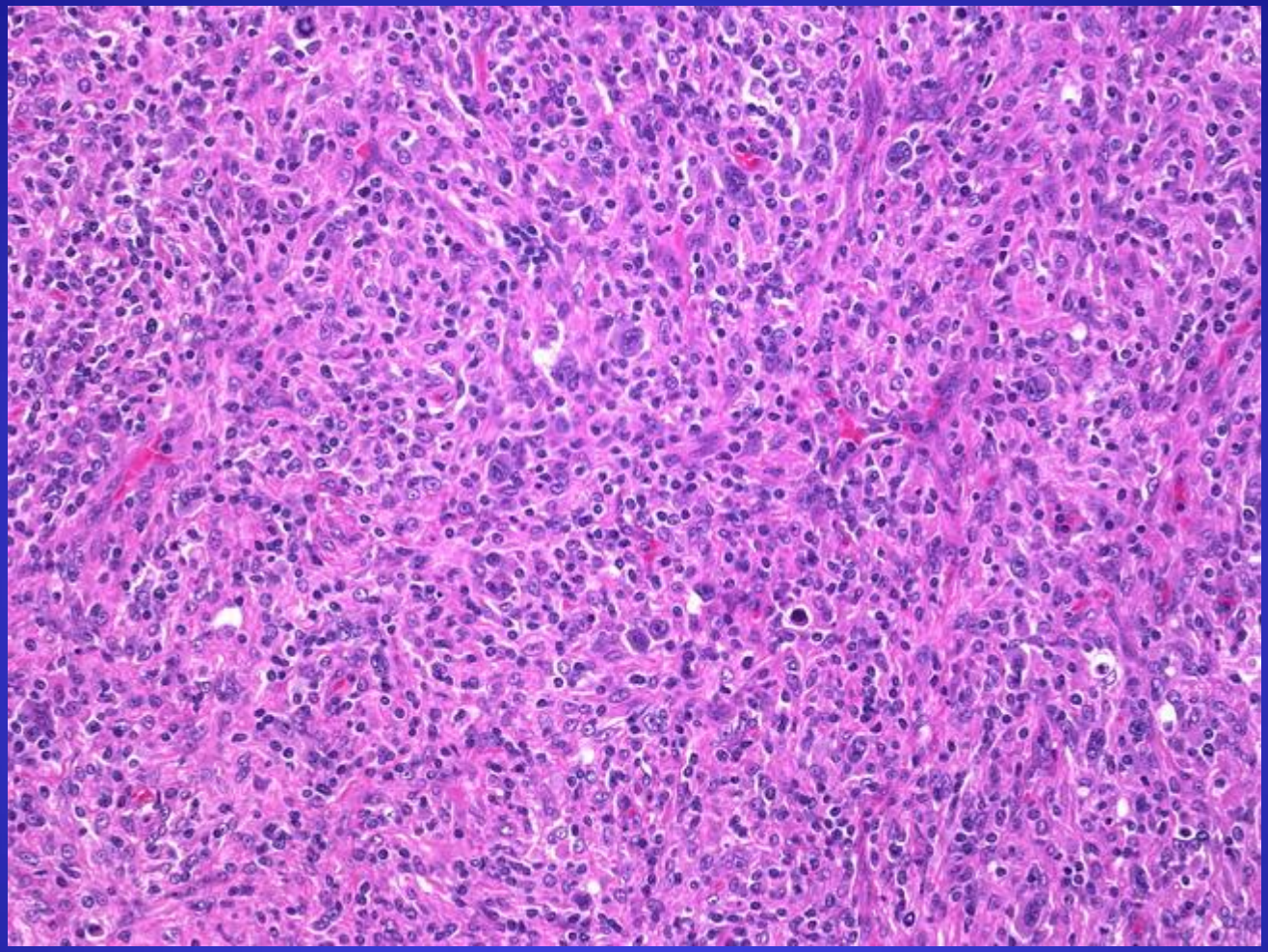
✓ RS cells are of the classic type (large, with bilobate, double or multiple nuclei, and a large eosinophilic inclusion like nucleolus).

✓ It commonly affects the abdominal lymph nodes and spleen.

✓ Patients with this histology typically have advanced-stage disease with systemic symptoms and immunodeficiency.



HODGKIN'S DISEASE – mixed cellularity (L) shows abundant lymphocytes, plasma cells, & eosinophils ; arrow shows a typical Reed-sternberg cell with bilobed nucleus (R) shows a multinucleated RS cell in the centre of the field.



Mixed Cellularity type accounts for 20% to 25% of cases of Classical Hodgkin's Lymphoma. Reed-Sternberg cells are present in a background of eosinophils, plasma cells, lymphocytes, and atypical mononuclear cells. Fibrosis is usually absent.

- **Lymphocyte-depleted Hodgkin's disease :**

✓ The infiltrate is diffuse and often appears hypocellular.

✓ Large numbers of RS cells and bizarre sarcomatous variants are present.

✓ Associated with older age and HIV positivity.

- **Lymphocyte-rich classic Hodgkin's disease :**

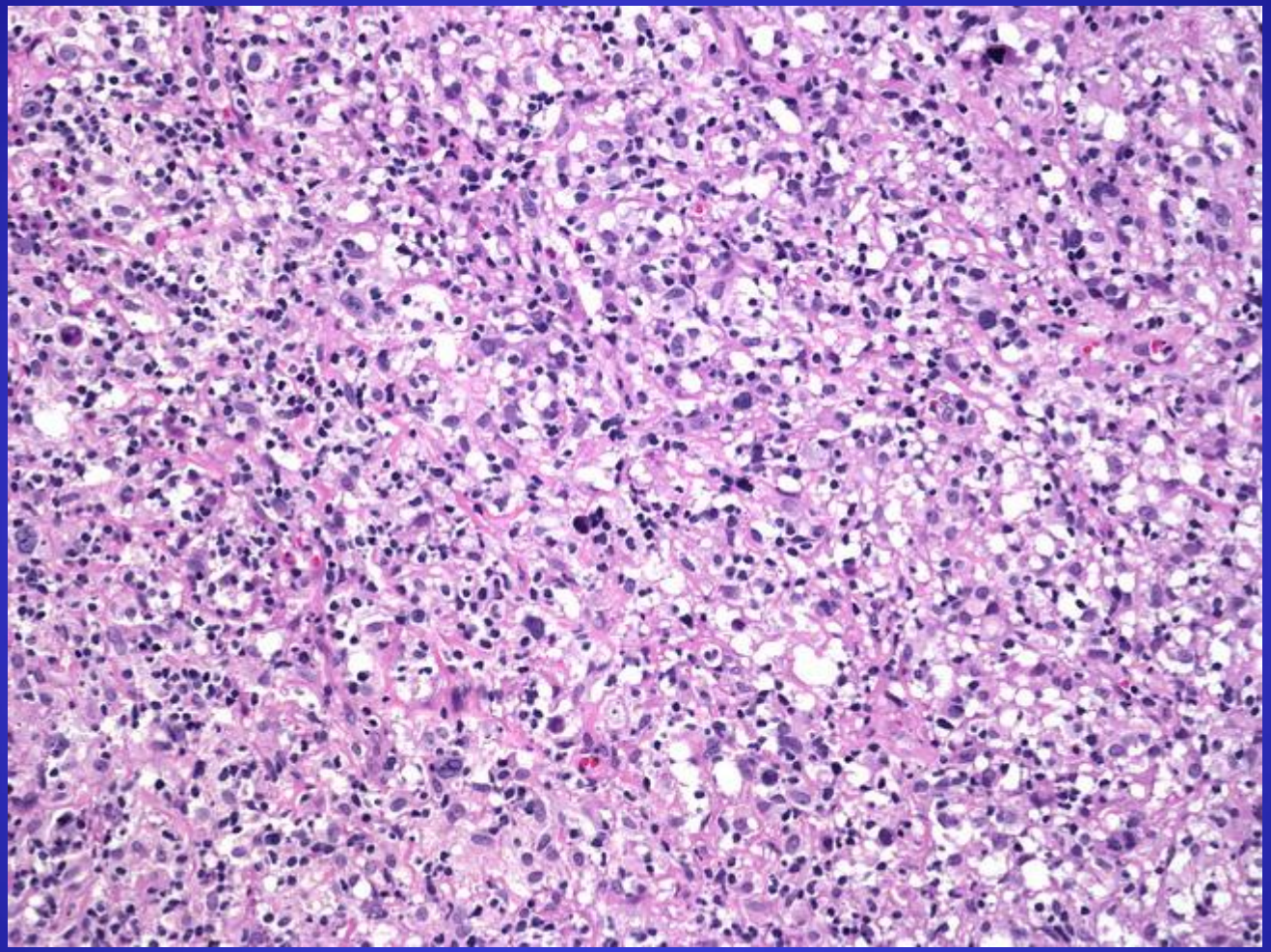
✓ RS cells of the classic or lacunar type are observed, with a background infiltrate of lymphocytes.

✓ Clinically, the presentation and survival patterns are similar to those for mixed-cellularity Hodgkin's disease.

- **Nodular lymphocyte-predominant Hodgkin's disease :**

✓ The typical RS cells are not observed or appear infrequently.

✓ Instead, a variant of RS cells, the lymphocytic and histiocytic cells (L &H), or ***popcorn cells*** (their nuclei resemble and exploded kernel of corn), are seen within a background of inflammatory cells, predominantly benign lymphocytes.



Hodgkin's Lymphoma, Lymphocyte-depleted

Treatment and Prognosis :

- Proper treatment can lead to long-term remission and even cure.
- Radiation therapy and combination chemotherapy effective lymphocyte predominant type has the most favorable prognosis, followed by nodular sclerosis, mixed cellularity and lymphocyte depletion, the least favorable.
- Male gender, older age and systemic symptoms also are associated with poor prognosis.

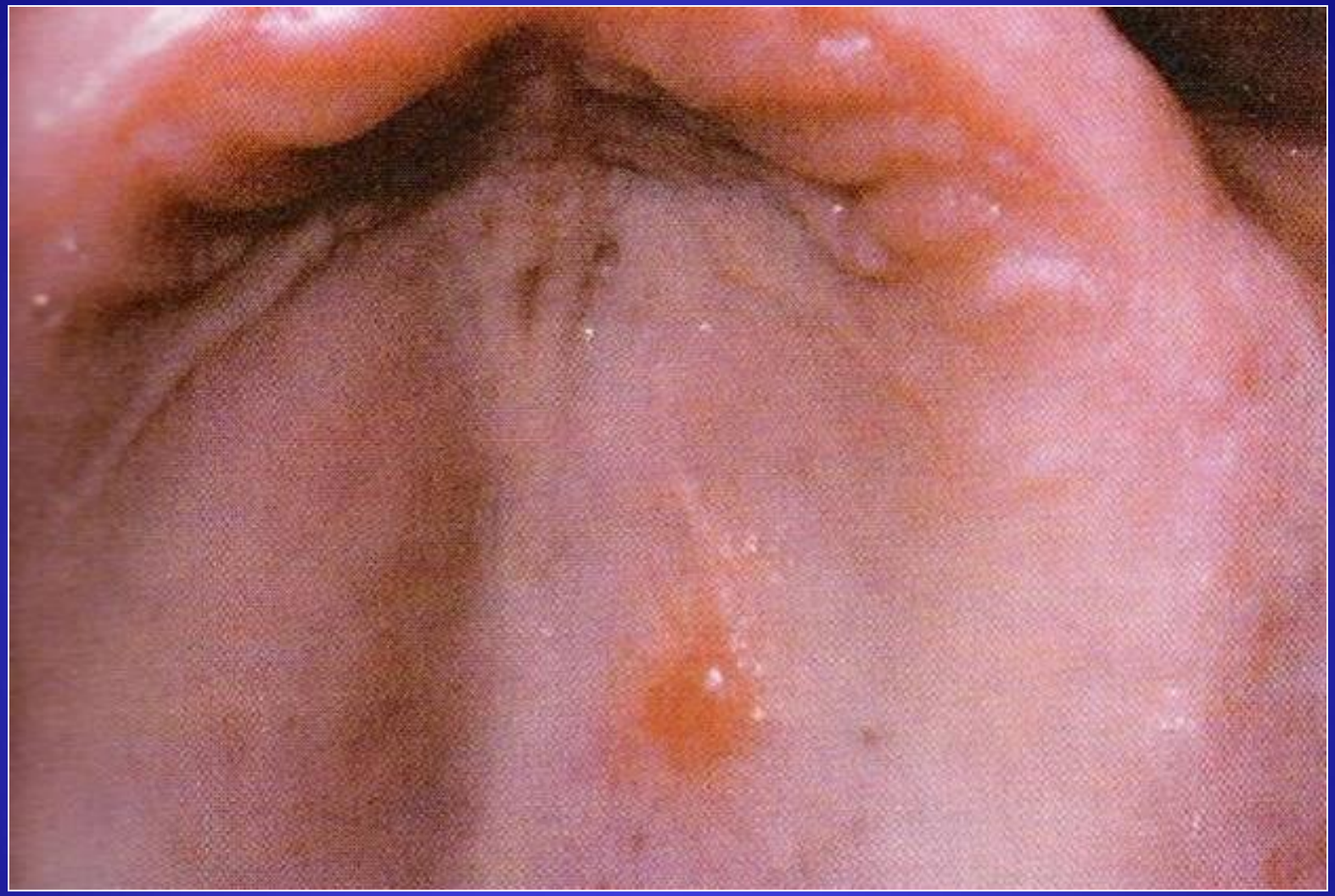
BENIGN TUMORS OF MUSCLE TISSUE ORIGIN

LEIOMYOMA

- A benign tumor derived from smooth muscle. Clinically ,that are soft tissue tumors that present with pain. they are uncommon in the oral cavity.

Clinical features-

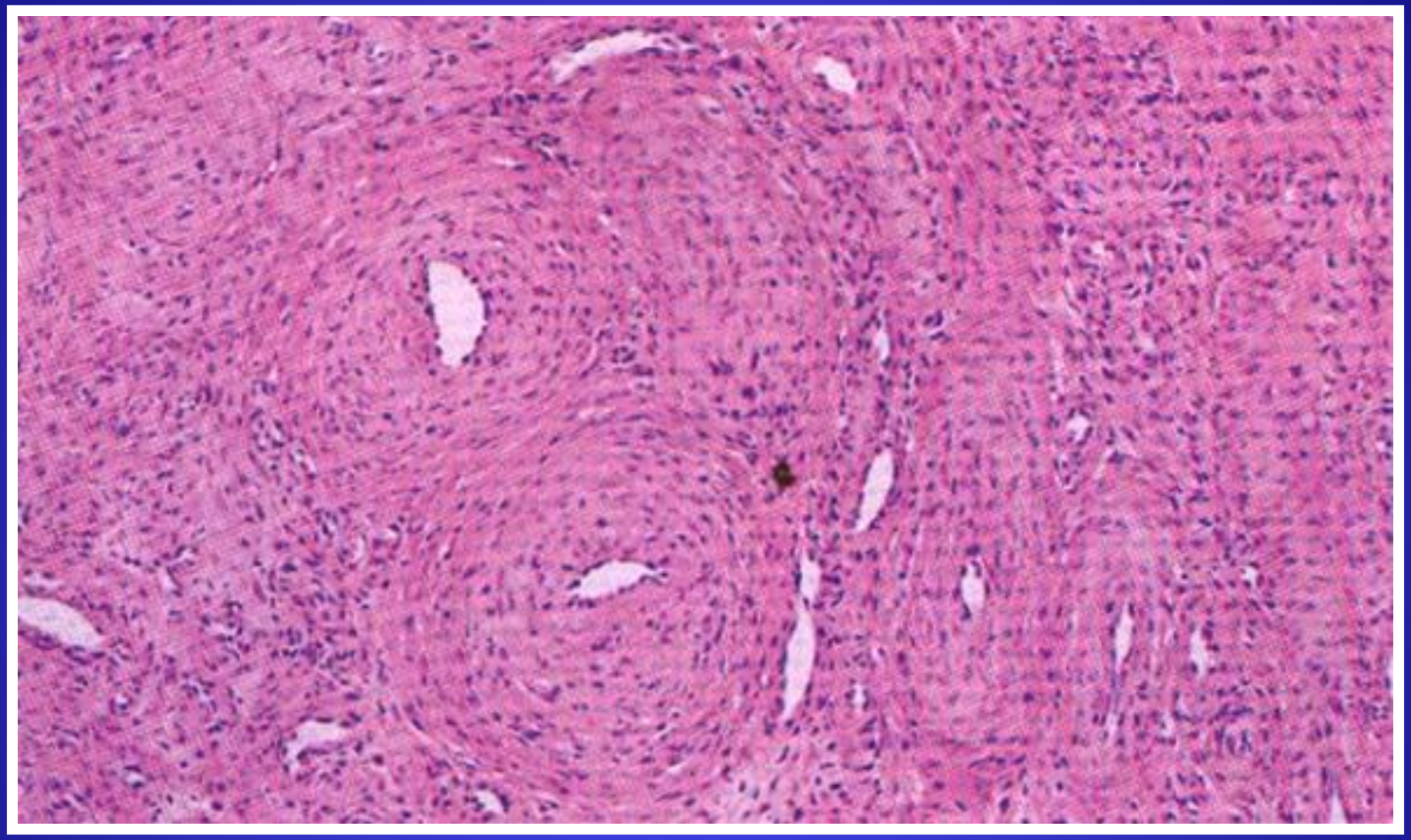
- Majority of the cases occur on the posterior portion of the tongue.
- Usually they occur in adults in the middle decades of life.
- It is a slow-growing painless lesion that is superficial and often pedunculated.
- Presenting symptoms may be a 'sore throat' or a 'tumor in throat'.



LEIOMYOMA : small pinkish red nodule on the posterior hard palate, lateral to the midline.

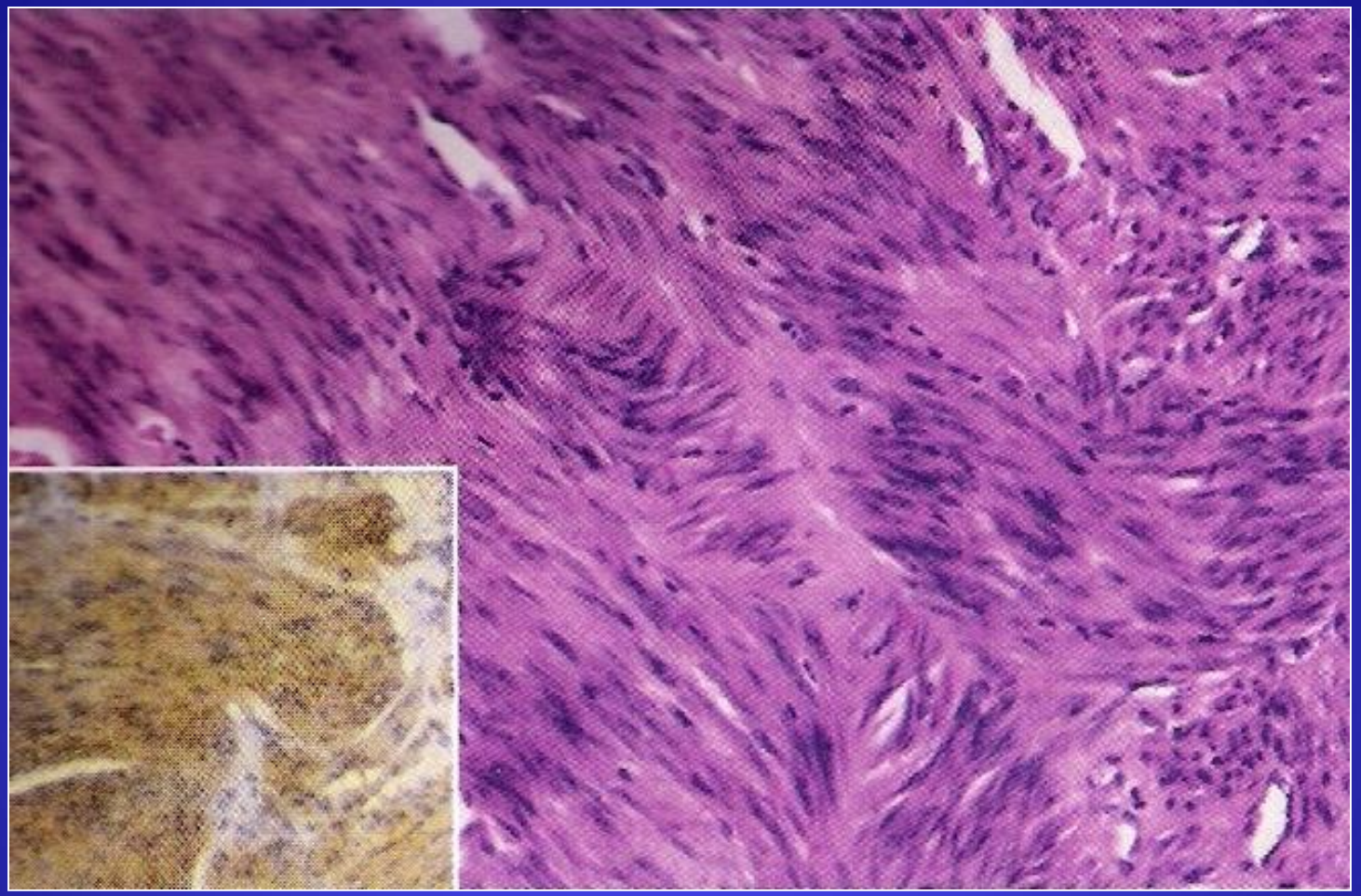
Histologic features-

- Composed of interlacing bundles of smooth muscle fibers, interspersed by varying amounts of fibrous connective tissue.
- Muscle nuclei are typically spindle –shaped with blunt ends and quite vesicular. fiber bundles appear to form whorls because of their fascicular arrangement in varying planes.



Leiomyoma

low power view shows a cellular mass of spindle shaped smooth muscle cells.



LEIOMYOMA

high power view showing spindle shaped cells with blunt-ended nuclei. Immunohistochemical analysis shows strong positivity for smooth muscle actin (inset).

- TREATMENT :

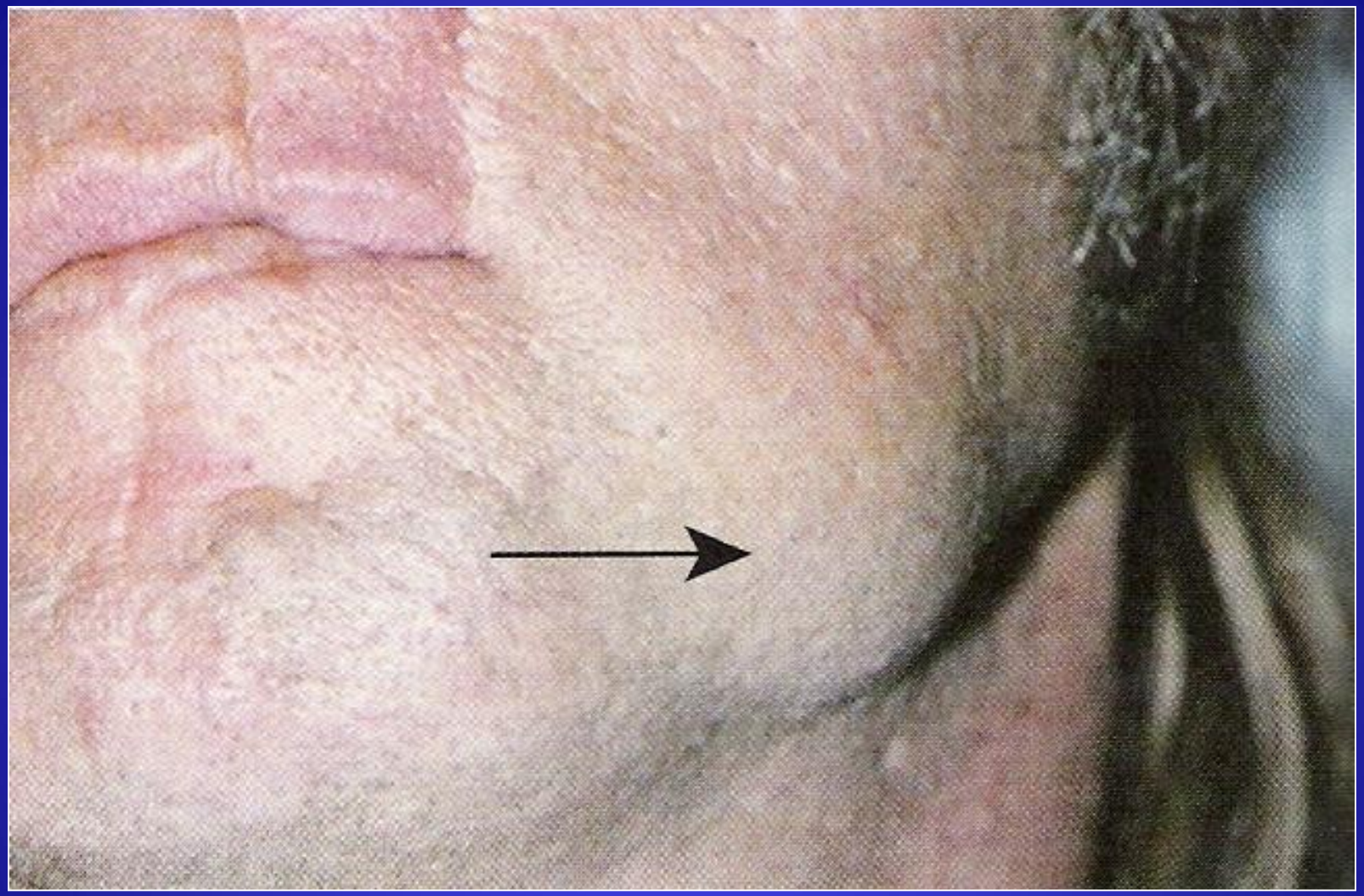
Conservative surgical excision

RHABDOMYOMA

- It is a benign neoplasm of striated muscle tissue, consisting usually of polygonal vacuolated glycogen containing cells, with a fine granular deeply acidophilic cytoplasm, resembling a myofibril in cross-section.
- It usually is seen in the cardiac muscle,but is a rare neoplasm of maxillofacial region.
- It is subdivided into adult , fetal and genital histological subtypes.

Clinical features-

- Adult form occurs primarily in the middle aged.
- Marked male predominance of 5:1
- Most common sites of involvement are the pharynx (usu.base of tongue and soft palate) and oral cavity (usu.oral floor).
- Fetal type usu.occurs in newborns and young children.
- Most common sites are post- or preauricular region, or face, but not mouth.
- Both types present as a nodule which may grow larger. Multinodular tumors have been described with 2 or more nodules,adjacent to one another.



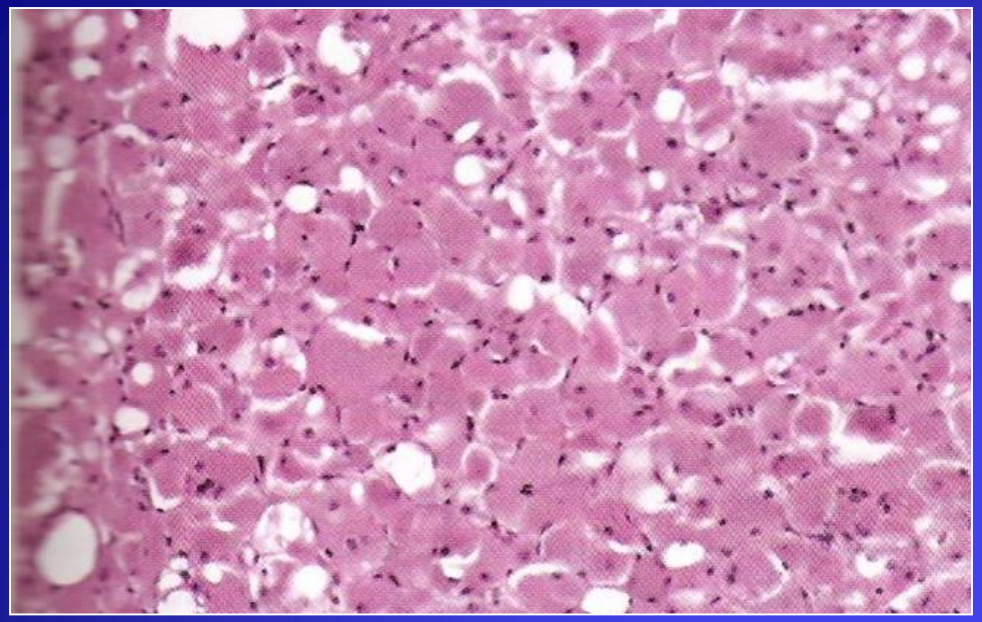
RHABDOMYOMA : nodular mass in the left cheek

Histological features-

- Large round cells which have a granular, eosinophilic vacuolated cytoplasm and show irregular cross-striations.
- Cytoplasm is rich in glycogen.
- Stroma is fibrous but occasional degeneration vacuoles or clear spaces between the tumor cells may be seen.
- Fetal type shows less mature,pleomorphic ,polygonal muscle cells admixed with spindle shaped cells.they are typically more cellular than the adult type and often has a myxoid stroma.

Treatment-

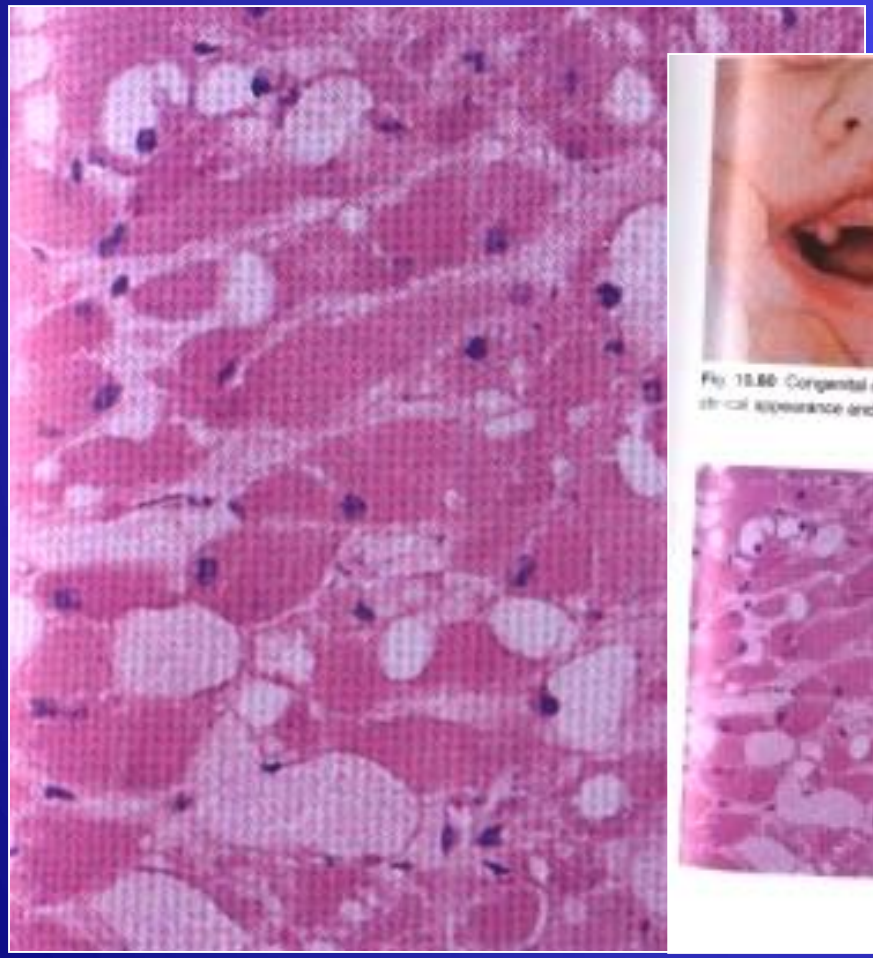
conservative surgical excision.

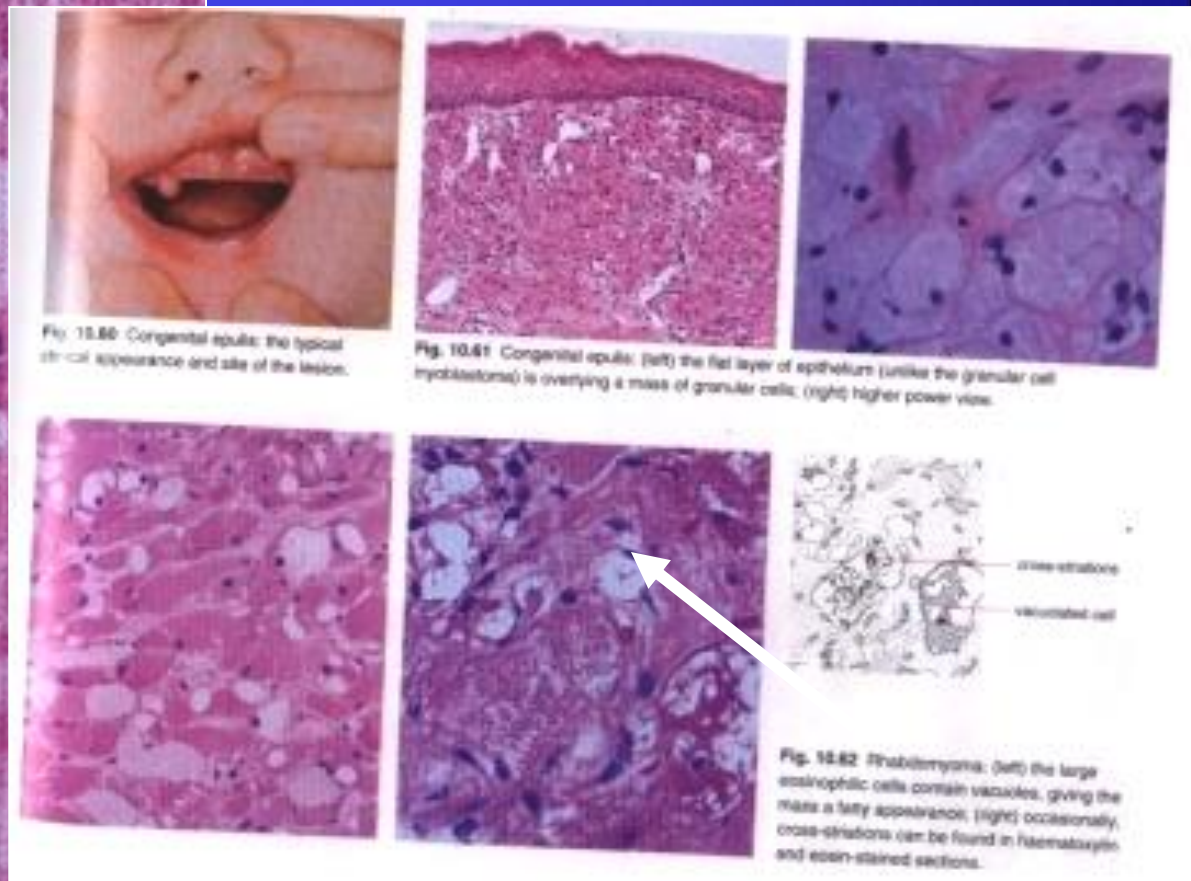


RHABDOMYOMA :
a uniform tumor composed of rounded and polygonal cells with focal vacuolizaton.

RHABDOMYOMA :
PTAH stain shows focal cross striations in some cells.

RHABDOMYOMA – (L) large eosinophilic cells contain vacuoles, giving the mass a fatty appearance ; (R) sometimes cross-striations are evident in H & E stain.





NEUROFIBROMA (neurofibromatosis, von Recklinghausen's disease of skin, fibroma molluscum)

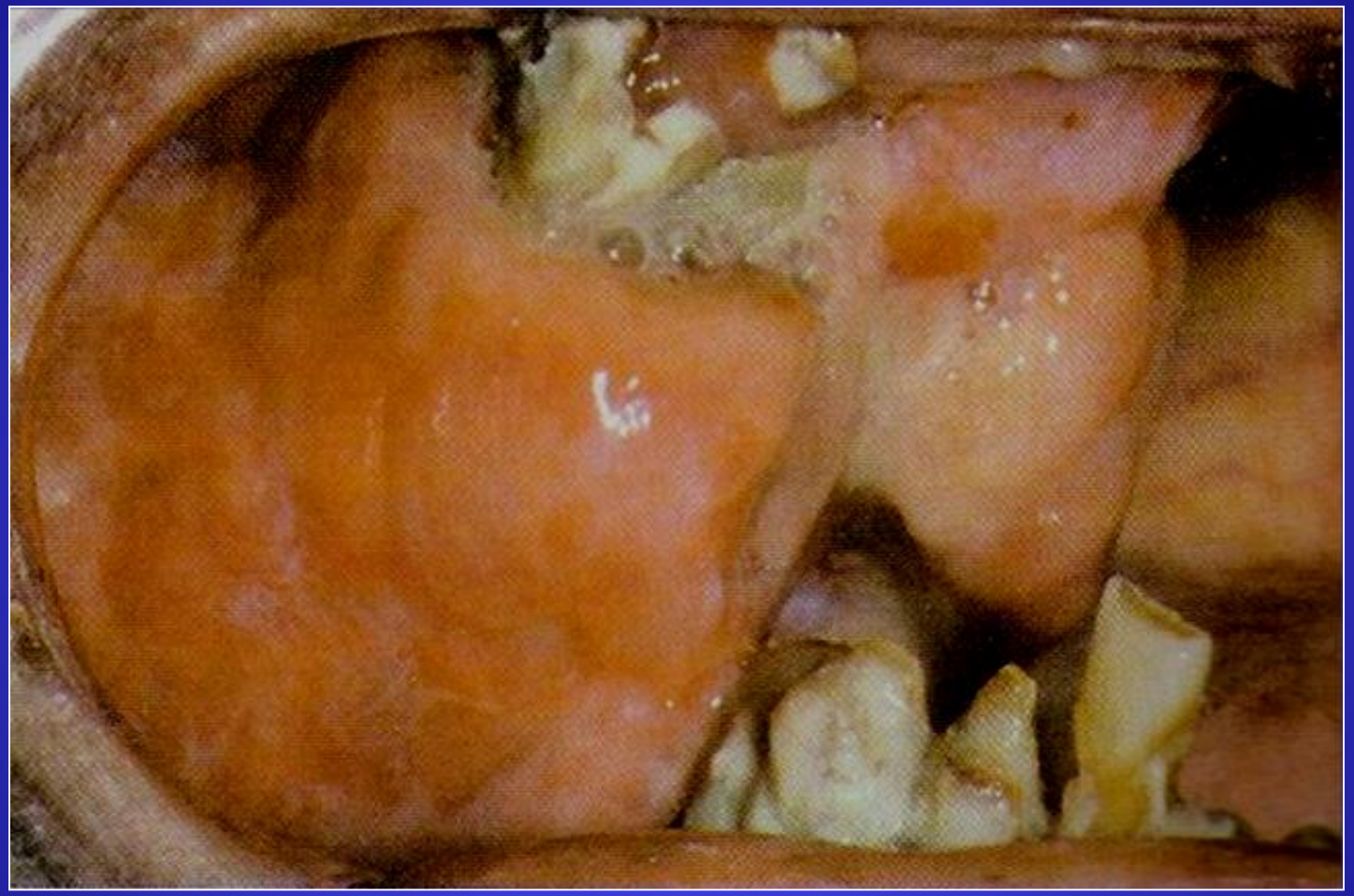
- A benign tumor of nerve tissue, derived from cells that constitute the nerve sheath.
- Neurofibroma is seen either as a solitary lesion or as a part of the generalized syndrome of neurofibromatosis (von Recklinghausen's disease).
- Recently 2 subsets have been defined - neurofibromatosis type I & type II.

Clinical features :

- It is clinically significant as, aside from the cosmetic problem, there is a potential for malignant transformation as well. But solitary neurofibromas rarely undergo malignant transformation.

- Intraoral neurofibromas are discrete, non ulcerated nodules, same color as the normal mucosa, usually seen on the buccal mucosa and palate. other cases exhibit diffuse masses of tissue, most commonly seen on tongue as macroglosssia.
- Occasional cases are seen in the jaws, usually in mandible, associated with the mandibular nerve and roentgenographically seen as fusiform enlargement of the mandibular canal.
- Involvement of the trigeminal nerve may cause facial pain or paresthesia.

Multiple neurofibromas of the face

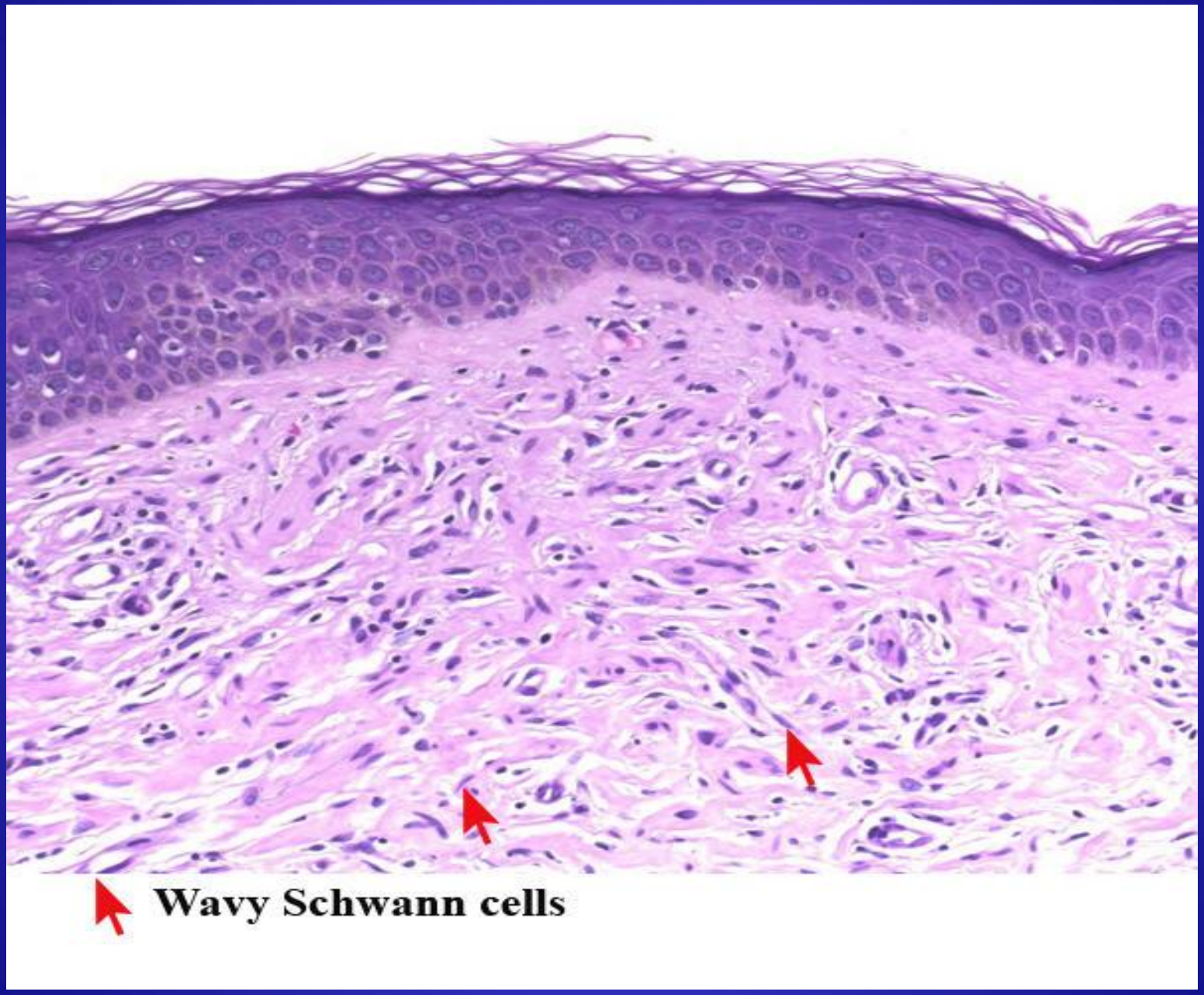


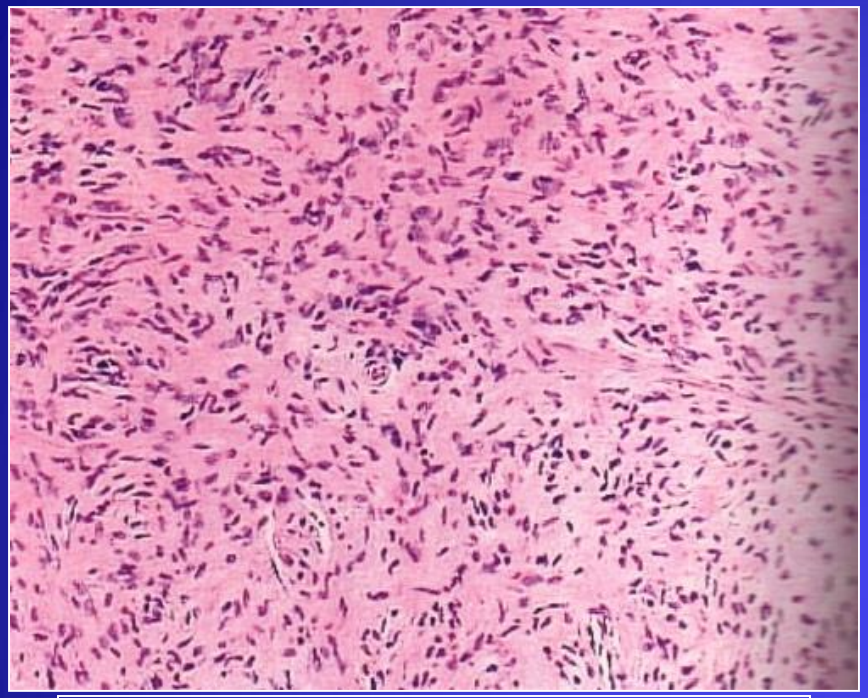
NEUROFIBROMA

huge tumor involving the maxillary gingiva and the hard palate.

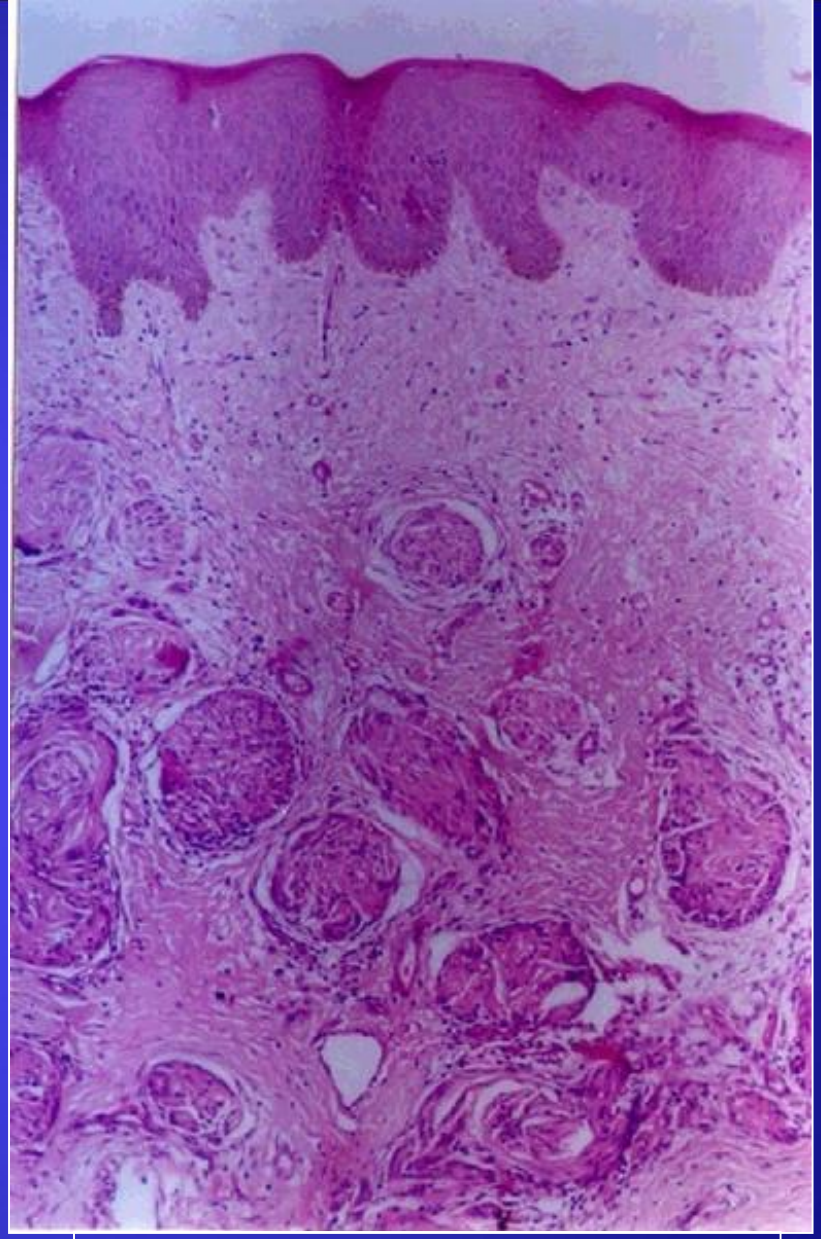
Histologic features :

- Proliferation of delicate spindle cells with thin wavy nuclei, intermingled with neurites in an irregular pattern, as well as delicate, intertwining c.t. fibrils.
- Cellular and myxoid patterns predominate.
- Melanocytes are sometimes found but mast cells are commonly found.
- In plexiform type, distorted masses of myxomatous peripheral nerve tissue still within the perineural sheath are scattered within a collagen-rich matrix. this picture is diagnostic of neurofibromatosis , even in absence of other symptoms.


Wavy Schwann cells



Neurofibroma :
Spindle shaped cells with wavy nuclei.



Plexiform neurofibroma

Plexiform neurofibroma is a benign tumor of peripheral nerves commonly seen in patients with type 1 neurofibromatosis. The tumor is characterized by tortuous proliferation of all components of peripheral nerves including axons, Schwann cells, fibroblasts and perineurial cells. This photomicrograph shows typical appearance of a plexiform neurofibroma with multiple tortuous enlargements of cutaneous peripheral nerves. In contrast to sporadic solitary neurofibroma a plexiform neurofibroma is rather circumscribed with distinct proliferative nodules.

Treatment :

- Surgery
- It may result in recurrence and multiple recurrences are associated with malignant transformation.

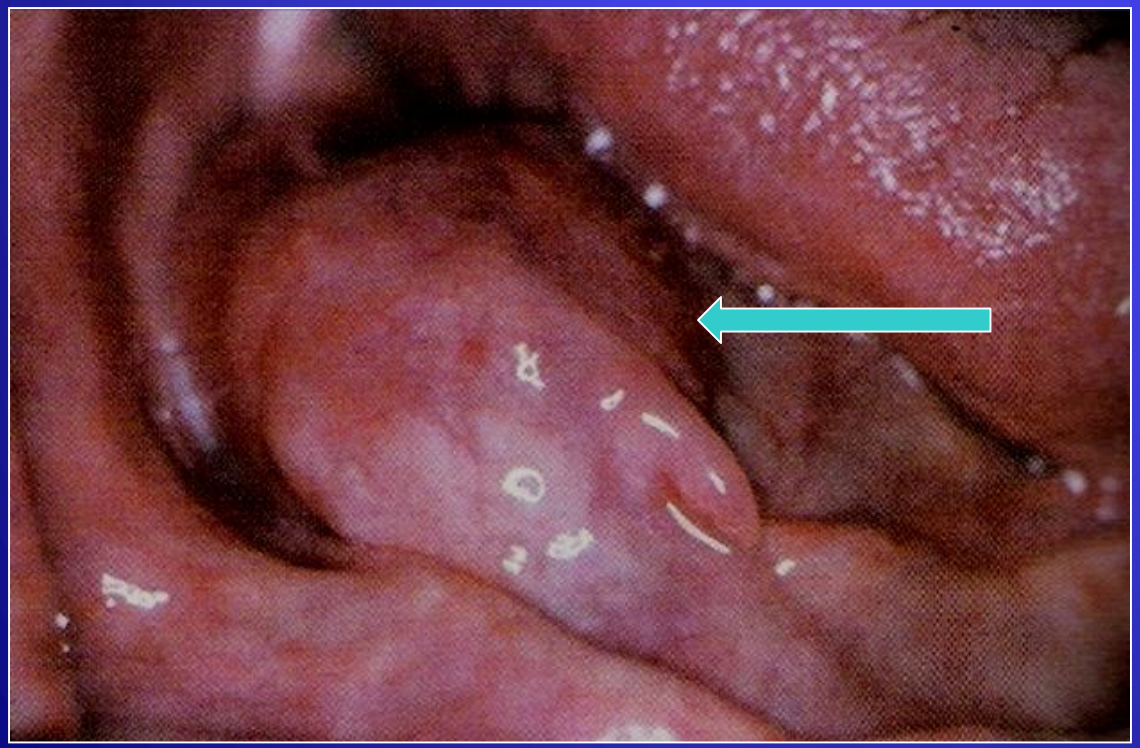
NEUROLEMMOMA (neurilemmoma, schwannoma, neurinoma, lemmoma)

- A common tumor derived from schwann cells.
- Neurites are not a component but may be found on the surface of the tumor.

Clinical features :

- A slow-growing lesion, usually of long duration, presents only as a tumor mass.
- Usually painless unless causing pressure on adjacent nerves.
- Head &neck are common sites for this tumor.

- Intraorally, most common site is tongue, followed by palate, floor of mouth and buccal mucosa. It is usually a single, circumscribed nodule of varying size, with no pathognomonic features.
- Sometimes a central lesion within bone is also found, especially in mandible arising from the mandibular nerve. It causes bone destruction with expansion of the cortical plates.



NEUROLEMMOMA
a nodular mass in the floor of the mouth.

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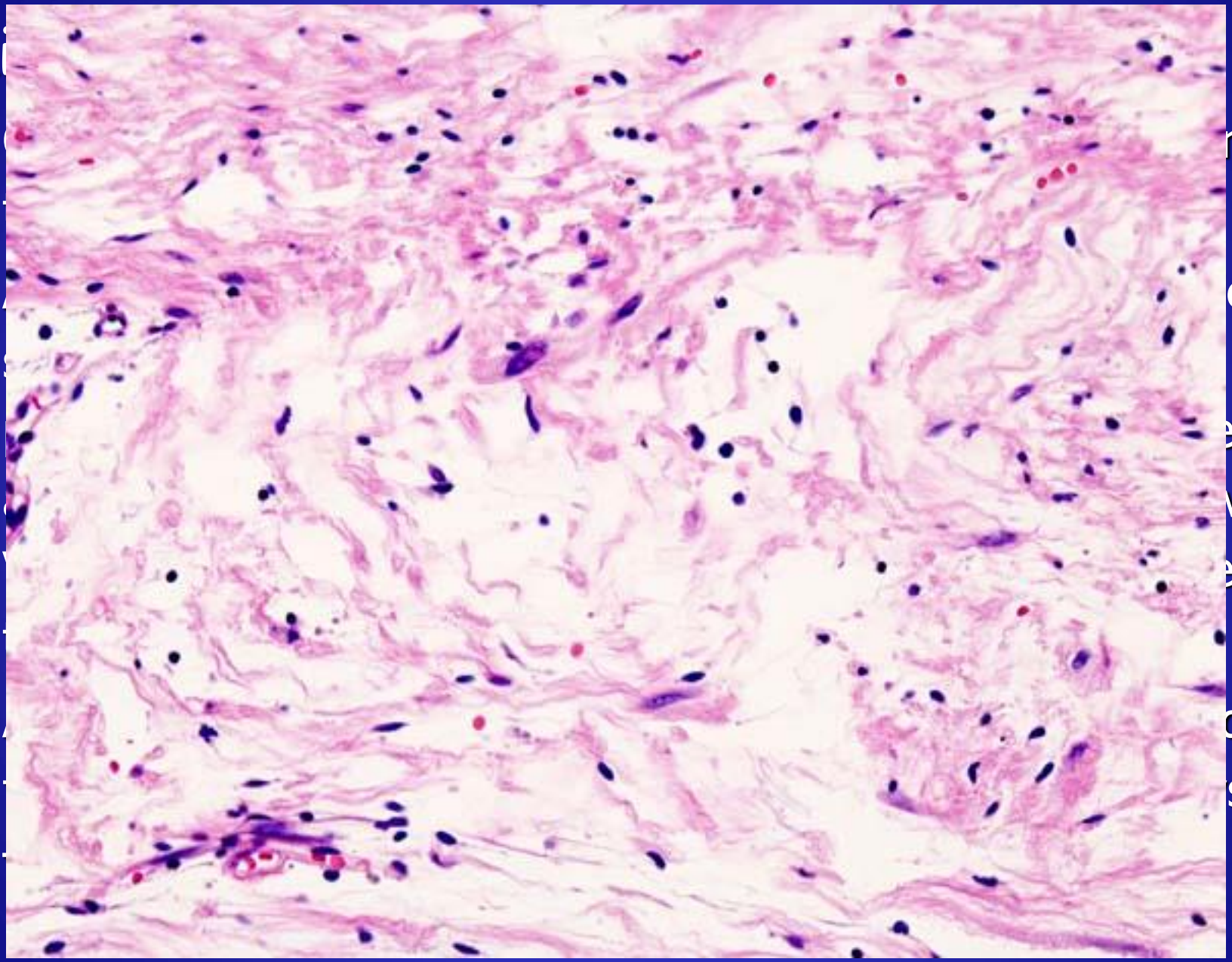

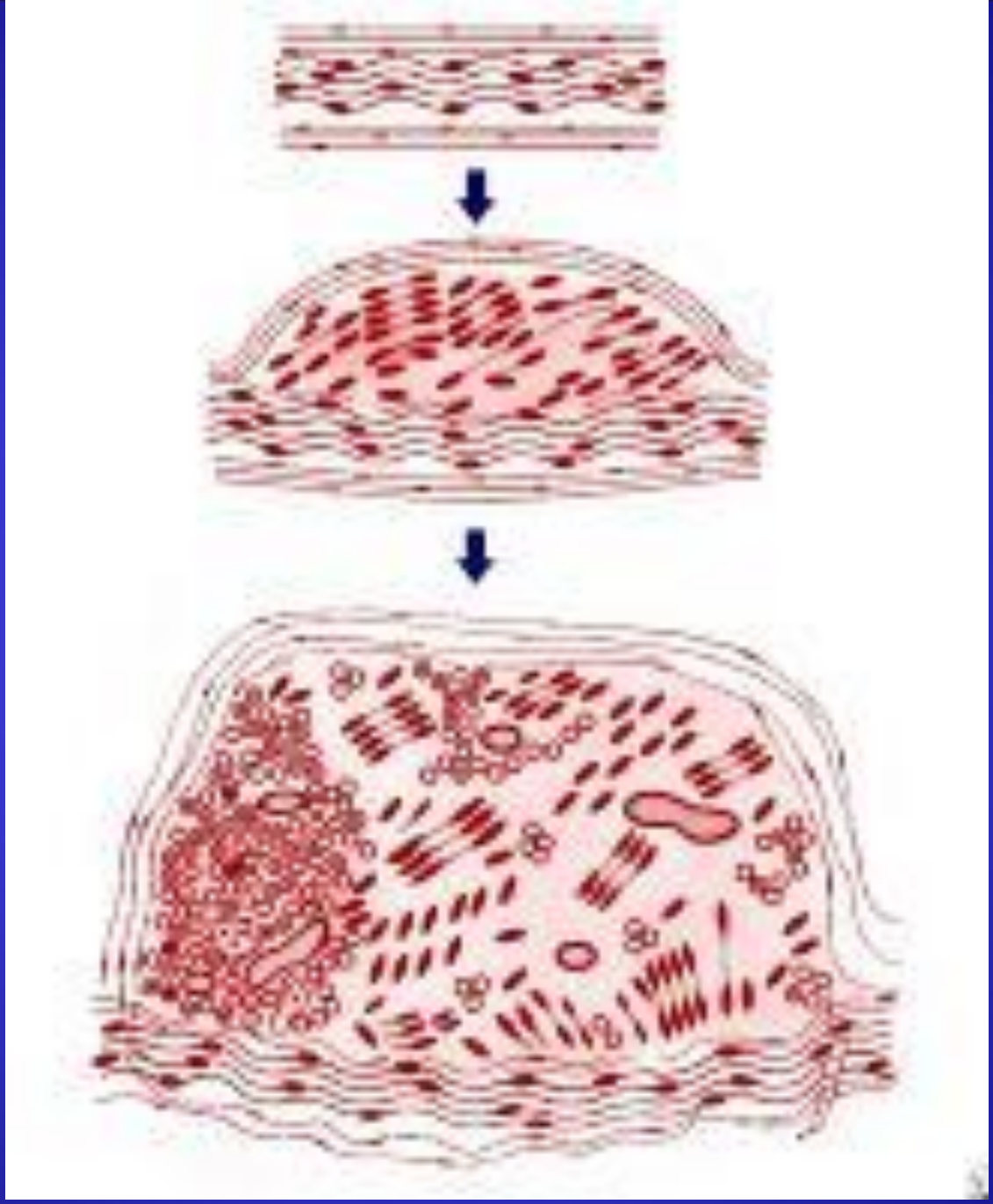

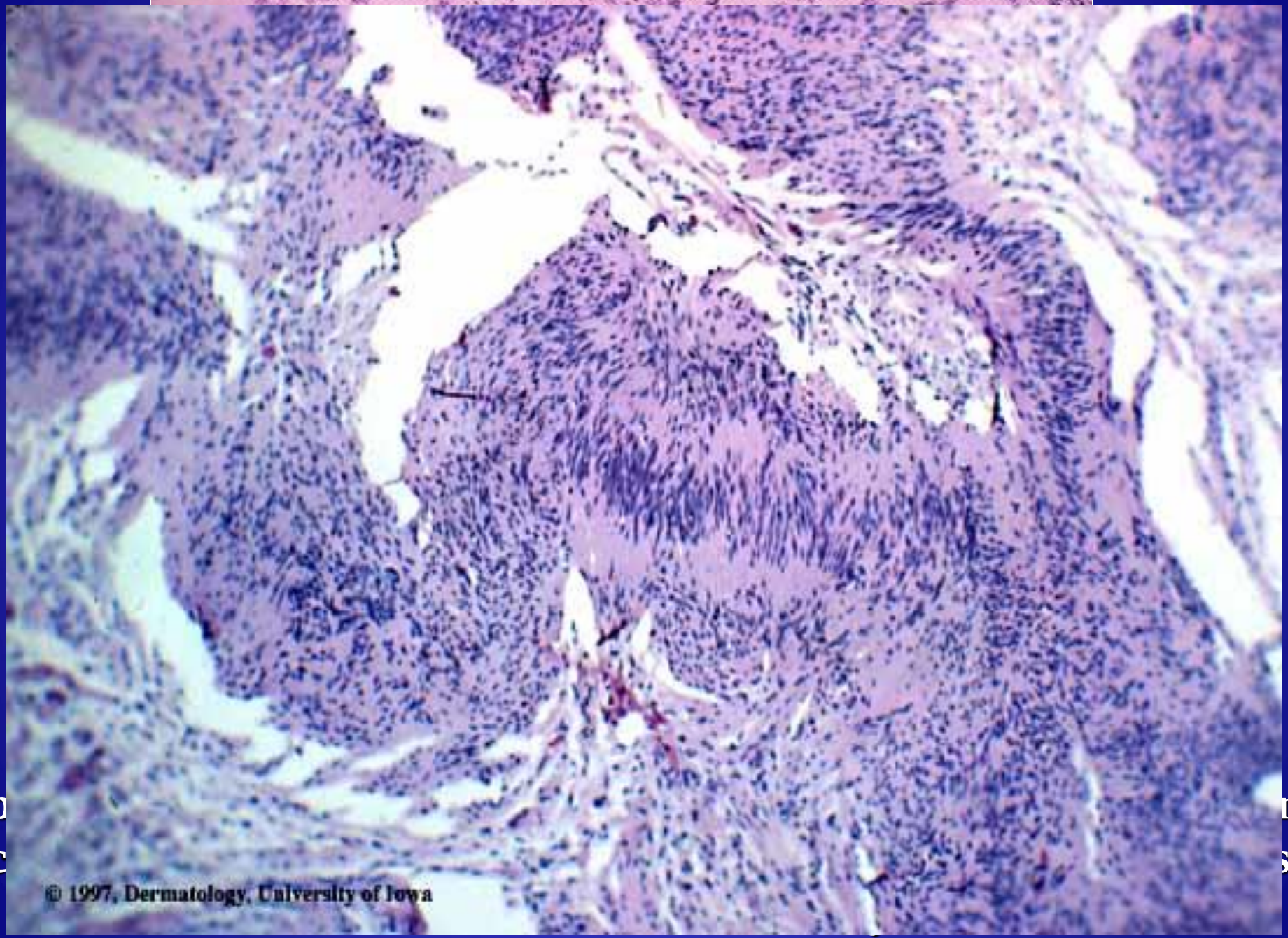
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Treatment :

- Surgery
- Not responsive to x-rays.
- Where complete removal is not possible, a portion of the tumor may be left without danger of recurrence or malignant transformation.

THANK YOU