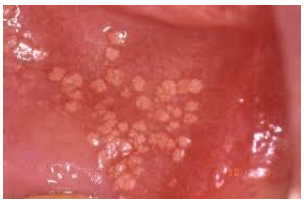
DEVELOPMENTAL DEFECTS OF THE ORAL AND MAXILLOFACIAL REGION

2- Developmental Defects of the Oral Mucosa

1- FORDYCE'S GRANULES

They represent ectopic sebaceous glands which are present in the oral mucosa in at least 80% of adults, particularly in elderly people. They grow in size with age and appear in the oral mucosa as soft, symmetrically distributed, creamy spots a few millimeters in diameter. The buccal mucosa is the main site, but sometimes the lips and rarely, even the tongue is involved.

These glands are sometimes mistaken for disease but patients can be reassured that they are of no significance. If a biopsy is carried out it shows a normal sebaceous gland with two or three lobules.

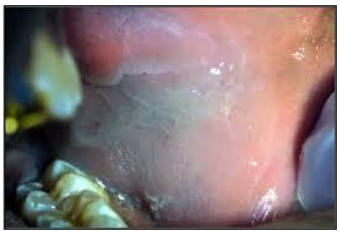


2- LEUKOEDEMA

Leukoedema is a bilateral, diffuse, translucent greyish thickening, particularly of the buccal mucosa. It is a variation of normal, present in 90% of blacks and variable numbers of whites.

Histologically, there is thickening of the epithelium with intracellular edema of the spinous layer

Treatment is unnecessary but reassurance may be required.



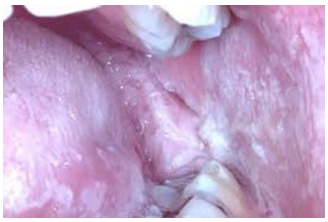
3- WHITE SPONGE NAEVUS

A developmental anomaly inherited as an autosomal dominant trait.

Clinical features:

The affected mucosa is white, soft and irregularly thickened. The abnormality is usually bilateral and sometimes involves the whole oral mucosa. There are no defined borders and the edges fade into normal tissue.

No treatment is required only reassurance.



3- Developmental defects of the tongue

1- Macroglossia

It is an abnormally large tongue, it could be congenital or acquired.

Congenital macroglossia e.g. Down's syndrome, Congenital hemangioma or lymphangioma.

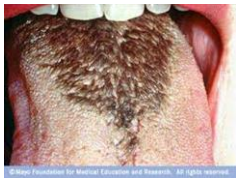
Acquired macroglossia e.g. Cretinism, Acromegaly, Amyloidosis, Lingual thyroid, Cancer.

2- Microglossia

It is an abnormally small tongue. It is uncommon, but mostly associated with a group of overlapping conditions known as (oromandibular-limb hypogenesis syndrome) which is characterized by limb abnormalities like absence of digits.

3- Hairy tongue

The filiform papillae can become elongated and hair-like forming a thick fur on the dorsum of the tongue. The filaments may be up to half a centimeter long and pale brown to black in color. Adults are affected but the cause is unknown. Heavy smoking, excessive use of antiseptic mouth washes and defective diet has been blamed, but their effect is questionable. The discoloration is probably caused by pigment-producing bacteria and fungi but not Candida albicans.



Treatment

It is difficult. The measure most likely to succeed is to persuade the patient to scrape off the hyperplastic papillae and clean the dorsum of the tongue with a firm toothbrush. This removes large numbers of microorganisms mechanically and also, by removing the overgrown papillae, makes conditions less favorable for their proliferation.

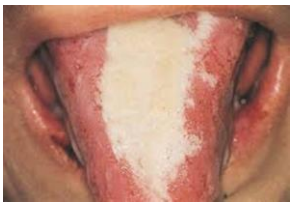
4- Black tongue

The dorsum of the tongue may sometimes become black without overgrowth of the papillae. This may be staining due to drugs such as iron compounds used for the treatment of anemia, but is

then transient. Occasionally the sucking of antiseptic lozenges causes the tongue to become black, and this may be due to pigment producing organisms, particularly Bacteroides strains.

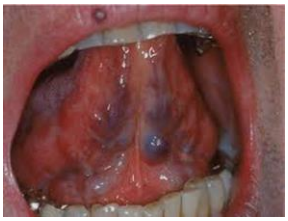
5- Furred tongue

The tongue becomes coated with desquamating cells and debris, in those who smoke heavily, in many systemic upsets, especially of the gastrointestinal tract, and infections in which the mouth becomes dry and little food is taken. A furred tongue is often seen in the childhood fevers, especially scarlet fever.



6- Lingual varicosities

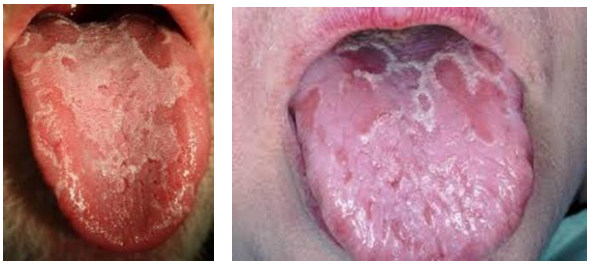
Dilated tortuous veins may be seen along the ventral surface of the tongue and tend to become more prominent with age. They may be noticed by patients who need to be reassured that they are not abnormal.



7- Geographical tongue

It is the recurrent appearance and disappearance of red areas on the tongue. The cause is unknown but sometimes there is a clear family history of its presence in several generations. In many patients, geographical tongue seems to be a developmental anomaly but there also appears to be an association with psoriasis.

Clinically: an irregular, smooth, red area appears, usually with a sharply defined edge. It extends for a few days, and then heals, only to appear again in another area. Sometimes the lesion is annular with a slightly raised pale margin, and several of these areas may coalesce to form a scalloped pattern. Most patients have no symptoms but some adults complain of soreness.

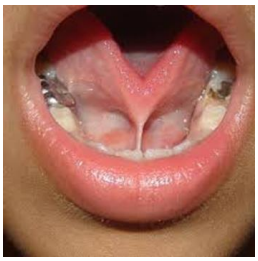


Histologically: there is thinning of the epithelium in the center of the lesion with mild hyperplasia and hyperkeratosis at the periphery, there are chronic inflammatory cells in the underlying connective tissue. Sometimes the changes are the same as those of psoriasis.

The Condition is considered important, because it can be confused with more serious form of glossitis and even premalignant or malignant lesions.

8- Ankyloglossia

It is characterized by a short, thick lingual frenum resulting limitation of tongue movement. The frenum sometime extends forward and attach to the tip of the tongue and there may be a slight clefting of the tongue. Occasionally, high mucogingival attachment of the lingual frenum may lead to local gingival and periodontal diseases in the regional frenal attachment.



9- Lingual thyroid nodule

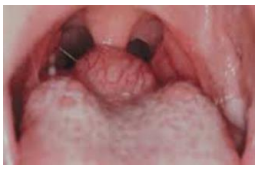
1- Accessory accumulation of thyroid tissue within the body of posterior tongue.

2- It represents a thyroid remnant in the region of the thyroid gland origin.

3- More common in females apparent during puberty and adolescence.

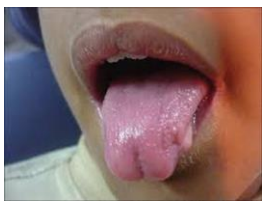
4- 2-3 cm, smooth, sessile mass on mid —posterior dorsum of the tongue in the region of foramen caecum.

5- Symptoms include dysphagia, dysphonia and hypothyroidism.



10- Cleft tongue

disunion of tongue usually occurs due to failure of fusion of the two lateral part of the tongue (mainly anteriorly) and this will lead to bifid tongue or cleft tongue.



4- DEVELOPMENTAL DEFECTS OF THE LIPS AND PALATE

1-Orofacial clefts:

A- Cleft lip and palate:

Clefts can form in the lip or palate alone or in both. The etiology is unknown but there is a genetic component in approximately 40% of cases. The risk of having such defects is greatly increased if one, and particularly if both, of the parents are affected.

Cleft lip: Developing defect usually of the upper lip characterized by a wedge-shaped defect resulting from the failure of two parts of the lips to fuse into single structure. Cleft lip (with or without a palatal cleft) is more common in males, while cleft palate alone is approximately twice as common in females. The incidence of cleft lip is about 1 per 1000 live births, while that of isolated palatal clefts is about 1 per 2000 live births.

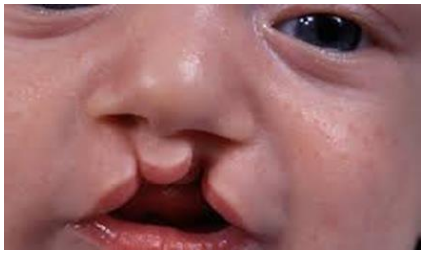
Classification

The main types of cleft lip and palate are:

1- Cleft lip

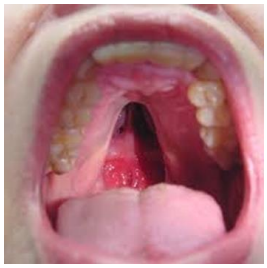
-Unilateral (usually on the left side), with or without an anterior alveolar ridge cleft

-Bilateral, with or without alveolar ridge clefts, complete or incomplete

2- Palatal clefts

Bifid uvula, Soft palate only, both hard and soft palate



3- Combined lip and palatal defects

Unilateral, complete or incomplete

Cleft palate with bilateral cleft lip, complete or incomplete

In the worst cases, there is complete separation of the anterior palate, which projects forward with the center section of the lip and is attached only by the nasal septum.

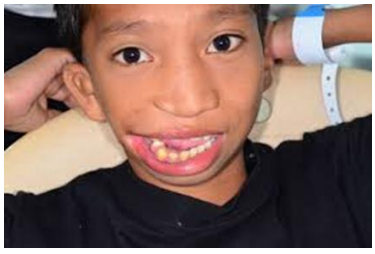
B- Oblique facial cleft

It represents failure of fusion of the lateral nasal process with the maxillary process. It extends from the upper lip to the eye and always associated with cleft palate.

C- Lateral facial cleft

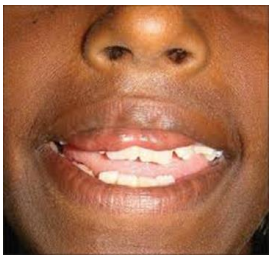
It results from lack of fusion of the maxillary and mandibular processes.

Occurs as isolated defects or may be associated with other disorders as mandibular dysostosis. It is either unilateral or bilateral extending from the commissures toward the ear resulting in macrosomia.



2- Double lip

this anomaly characterized by a horizontal fold of redundant mucosal tissue that is usually located on the inner aspect of the upper lip. Most often congenital in nature, but it may be acquired later in life.



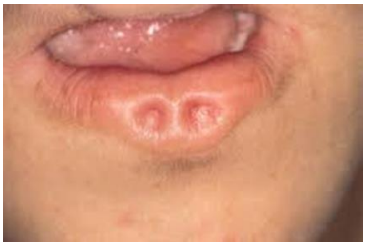
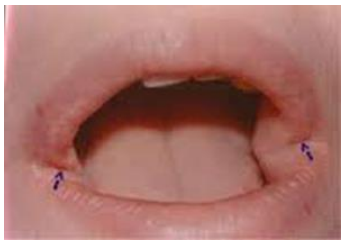
3- Congenital lip pits

developmental defects that may involve the Para median portion of the vermilion of the lower and upper lip (Para median lip pit), or the labial commissural area (commissural lip pit).

Para median lip pit: present as bilateral and symmetric fistulas on either side of the midline of the vermilion of the lower lip. It occurs as an isolated condition or may be associated with cleft lip or cleft palate.

Commissural lip pits: A small mucosal invagination that occur at the corner of the mouth on the vermilion border. It may represent a failure of fusion of the maxillary process and mandibular process. It is either unilateral or bilateral.

Clinically it represents as blind fistula that may extend to a depth of 1-4 mm or it may be present as dilated ectopic salivary gland tissue.

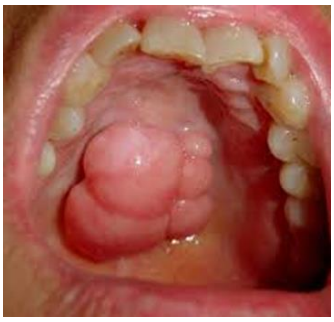
5- DEVELOPMENTAL DEFECTS OF THE JAW BONES

1- BONY OVERGROWTHS (Bony exostosis)

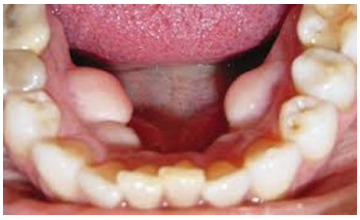
Localized overgrowths of bone that arises from normal cortical plate (exostoses) are more common.

Small exostoses may form irregularly on the surface of the alveolar processes and specific variants are torus palatinus and torus mandibularis. They differ from other exostoses only in that they develop in characteristic sites and are symmetrical.

Torus palatinus commonly forms towards the posterior of the midline of the hard palate. The swelling is rounded and symmetrical, sometimes with a midline groove. It is not usually noticed until middle age.



Torus mandibularis form on the lingual aspect of the mandible opposite the mental foramen. They are typically bilateral, forming hard, rounded swellings. The management is the same as that of torus palatinus.

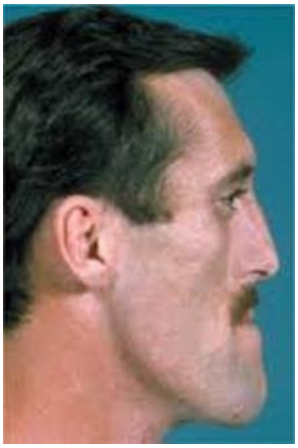


2- Agnathia (nathia= jaw, Ag = Agenesis).

It is developmental congenital absence of one of the jaws; it is a rare condition and mostly occurs as part of the mandible is absent.

3- Macrognathia

It is abnormally large jaw, sometimes called prognathism. This defect occurs either due to local cause, e.g. fibrous dysplasia of bone, reactive or neoplastic bone tumor, odontogenic cysts and tumors or associated with systemic diseases as Acromegaly and Paget's disease of bone.



4- Micrognathia: very small jaw

It is a developmental disturbance affecting one of the jaws and lead to abnormally small jaw. The condition gives rise to numerous dental problems.

Micrognathia may be associated with other developmental defect like in Pierre Robin's syndrome which is characterized by cleft palate, micrognathia and glossoptosis (posterior displacement of the tongue, lack of support of tongue musculature and airway obstruction).

5- Coronoid hyperplasia

It is rare developmental anomaly, which results in limitation of mandibular movement. The condition may be unilateral which result from osteoma and osteosarcoma or bilateral which may result from endocrine influence during puberty.

6- Condylar hyperplasia

Excessive growth of one condyle is of unknown cause but local circulatory problems, endocrine disturbances and trauma have been suggested as possible etiological factors.

7- Condylar hypoplasia

Congenital: - associated mandibulofacial dysostosis and hemifacial macrosomia.

Acquired: - result from disturbance of center of the developing condyle secondary to trauma, radiation or rheumatoid arthritis.

8- Bifid condyle

Double-headed mandibular condyle of uncertain cause.

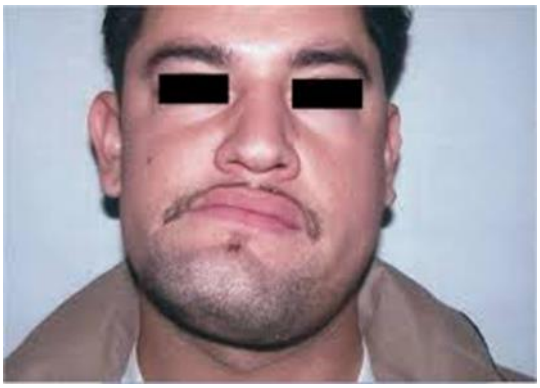
Anteroposterior bifid condyle may be traumatic in origin during childhood.

Mediolateral bifid condyle may result from abnormal muscle attachment.

9- Hemifacial hypertrophy

Significant unilateral enlargement of the face as a result of an increased neurovascular supply to the affected side of the face.

Unilateral enlargement of the facial tissues, bones and teeth is usually present resulting in asymmetry of the face with malocclusion and deviation of the affected side of the face to the unaffected side of the face.



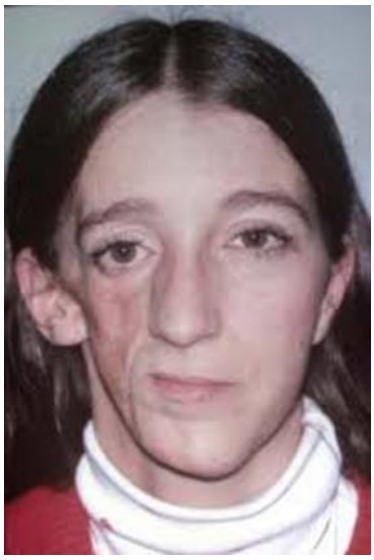
10- Hemifacial atrophy

Uncommon poorly understood degenerative condition, characterized by:

1- Atrophic changes affecting one side of the face.

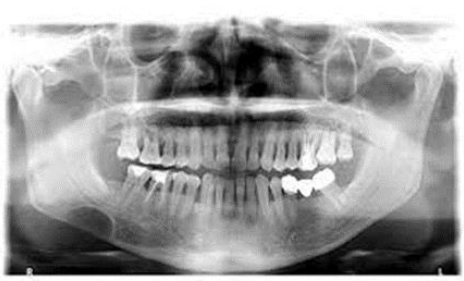
2- The mouth and nose are deviated toward the defective side.

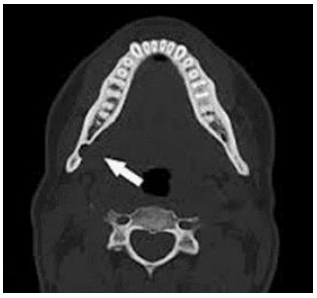
3- The covering skin often exhibit dark pigmentation.



11- Lingual mandibular salivary gland depression (Stafne defect)

Developmental concavity of the cortex of the mandible in the molar area, that forms around an accessory lateral lobe of submandibular gland which has radiographical appearance of a well-circumscribed cystic lesion within the bone usually below the inferior alveolar canal.





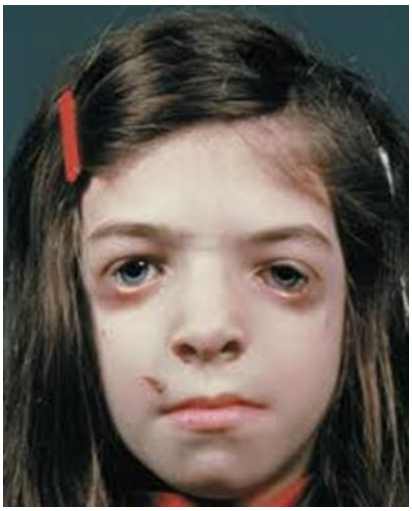
In most cases biopsy revealed histologically normal salivary gland tissue suggesting that these lesions represent developmental defects containing portion of the submandibular gland.

12- Mandibular Dysostosis (Treacher-Collins syndrome)

Autosomal dominant disorder characterized by:

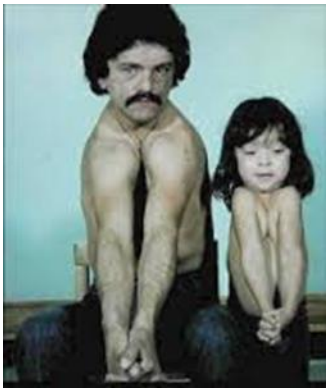
1- Hypoplastic zygoma, resulting in narrow face with depressed check and downward slanting of palpubral fissures.

2- Underdeveloped mandible with retruded chin and cleft palate may be seen.



13- Cleidocranial Dysplasia or Dysostosis

It is rare familial disorder characterized by defective formation of the clavicles, delayed closure of fontanelles and sometimes retrusion of the maxilla. Partial or complete absence of clavicles allows the patient to bring the shoulders together in front of the chest.



This disorder is one of the few recognizable causes of delayed eruption of the permanent dentition. Many permanent teeth may remain embedded in the jaw and frequently become enveloped in dentigerous cysts. Supernumerary teeth may be seen radiographically.

DEVELOPMENTAL CYSTS OF THE ORAL AND MAXILLOFACIAL REGION

Also, are called fissural cysts or occlusion cysts, because they arise from embryonic epithelium that becomes entrapped during embryogenesis.

Clinically, they present as a soft or fluctuant swelling.

1-Nasolabial Cyst (Nasoalveolar cyst)

is a rare developmental soft tissue cyst that develops in the upper lip in the canine region. Etiology: Unclear. Although there are two major theories:

One theory considers this cyst to be a fissural cyst arising from epithelial remnants entrapped along the line of fusion of the maxillary, medial nasal and lateral nasal processes.

A second theory suggests that these cysts develop from misplaced epithelium of the nasolacrimal duct.

Clinical and radiographical features: - It appears as a soft-tissue swelling in the mucobuccal fold of the maxilla, lateral to the midline. Occasionally, the patient may complain of nasal obstruction, discomfort, or difficulties in wearing dentures. The cyst is more common in women, usually between 40 and 50 years of age. Because this cyst arises in soft tissues, in most cases there are no radiographic changes, but resorption of the underlying bone may occur.

Histopathological examination shows lining of the cyst by pseudo stratified columnar epithelium, often show goblet cells and cilia. The cyst wall is composed of fibrous connective tissue with adjacent skeletal muscle.

Differential diagnosis: -

Soft tissue abscess, tooth abscess, mucocele, radicular cyst, salivary gland neoplasms, and mesenchymal neoplasms.

Treatment: Surgical excision.

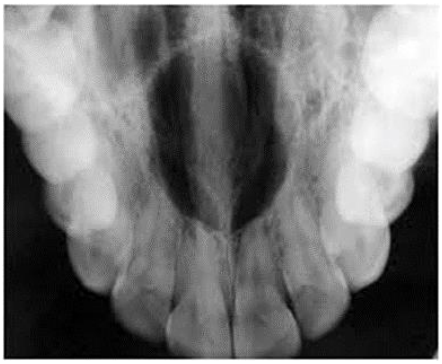
3- Nasopalatine duct cyst (incisive canal cyst)

It is the most common non-odontogenic cyst of the oral cavity.

Etiology: It arises from epithelial rests in the incisive foramen.

Clinical and radiographical features: It appears as a slow-growing soft swelling of the palatine papilla, covered with normal mucosa. The cyst, after mechanical irritation, may be inflamed and becomes painful due to local infection.

Radiographically: usually demonstrates a well circumscribed radiolucency in or near the midline of the maxilla, between and apical to the central incisor teeth. It may be difficult to distinguish a small nasopalatine duct cyst from a large incisive foramen. It is generally accepted that a diameter of (6 mm) is the upper limit of normal size for incisive foramen. Therefore, a radiolucency that is (6 mm) or smaller in this area is usually considered a normal foramen unless other clinical signs and symptoms are present. The clinical diagnosis should be confirmed by histopathological examination that showed epithelial lining composed of either:1- Stratified squamous epithelium.2- Pseudostratified columnar epithelium.3- Simple columnar epithelium. 4- Simple cuboidal epithelium.



N.B.: to distinguish between the nasopalatine cyst & P.A. cyst:

1- N.P.cyst, the tooth is vital, but in case of P.A. cyst the associated tooth is nonvital.

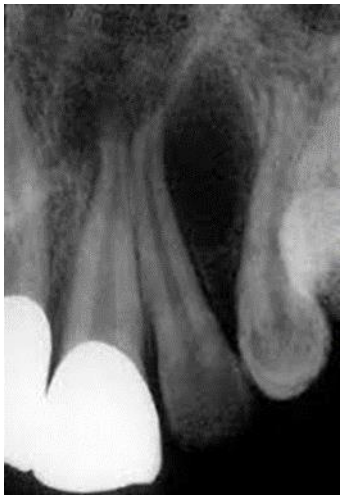
2- Because the N.P.cyst is not related (attached) to the apex of the root, so by changing the direction of the X-ray beam we see if the lesion remain attached to the apex of the root, so it mean it's radicular cyst, if not it means N.P. cyst. Treatment: Surgical removal.

3- Globulomaxillary cyst

Globulomaxillary cyst were once considered fissural cyst, located between the globular and maxillary processes. The former theory of origin related to epithelial entrapment within a line of embryologic closure with subsequent cystic change.

Embryologic evidence now shows that the premaxilla and maxillary processes do not fuse in this manner, and thus there can be no fusion-related mechanism to account for a distinct globulomaxillary cyst in this location thats why a current theory holds that most of cysts that develop in the globulomaxillary area, are of odontogenic origin.

Radiolucencies in this location, when reviewed microscopically, have been shown to represent radicular cysts, periapical granulomas, lateral periodontal cysts, OKCs, central giant cell granulomas, calcifying odontogenic cysts, and odontogenic myxomas. Thus, today the term globulomaxillary can be justified only in an anatomic sense, with definitive diagnosis of lesions located in this area made by combined clinical and microscopic examination. Radiologically: a globulomaxillary lesion appears as a well-defined radiolucency, often producing divergence of the roots of the maxillary lateral incisor and canine teeth. Radicular cyst and periapical granuloma can be ruled out with pulp vitality testing.



Because of the array of potential diagnoses, the histology varies considerably from case to case.

Histologically: - lining epithelium is stratified squamous and sometimes pseudostratified ciliated columnar respiratory epithelium. Thin C.T. wall which is free from inflammation.

Treatment and prognosis are determined by the definitive microscopic diagnosis.

4- Lymphoepithelial Cyst

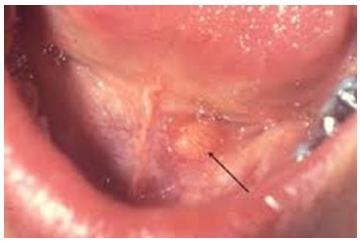
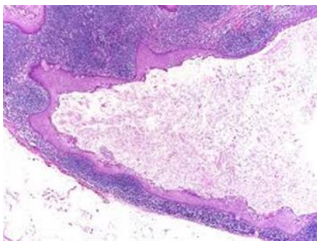
Definition: Lymphoepithelial cyst is an uncommon developmental lesion of the oral mucosa.

Etiology: Probably caused by cystic degeneration of glandular or surface epithelium entrapped in lymphoid tissue during embryogenesis.

Clinical features: It presents as an asymptomatic, mobile, well-defined nodule, usually firm on palpation and elevated, with a yellowish or whitish color. The size ranges from 0.5 cm to 2 cm in diameter. The floor of the mouth is the most frequent location, followed by the posterior lateral border and the ventral surface of the tongue.

Lymphoepithelial cysts are histologically similar to the branchial cleft cysts that develop in the lateral neck.

Histopathological examination which showed epithelial lining of stratified squamous that may or may not be keratinized. The wall of the cyst typically contains lymphoid tissue often demonstrating germinal center formation.

Differential diagnosis: lymphoid tissue aggregation, dermoid cyst, mucocele, lipoma, fibroma and other benign tumors. Treatment: Surgical removal.

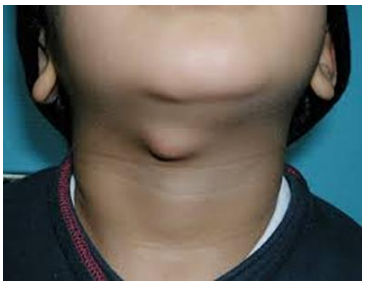
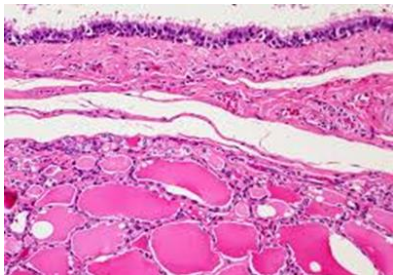
5- Thyroglossal Duct Cyst

Definition: Thyroglossal duct cyst is a rare developmental lesion that may form along the thyroglossal tract.

Etiology: Remnants of thyroglossal duct epithelium.

Clinical features: The cyst is usually located under the hyoid bone but can be located anywhere from the suprasternal notch to the foramen cecum of the dorsal tongue. Intraorally, it appears as a painless, fluctuant swelling usually 1-3 cm in diameter, located in the midline of the dorsum of the tongue close to the foramen caecum. Occasionally, a fistula may form following infection. The cyst is most often diagnosed in patients less than 20 years of age.

Histopathological examination, showed a lining epithelium of stratified squamous, or columnar or small intestinal epithelium, or mixture of them. The C.T. tissue wall may contain normal thyroid tissue.

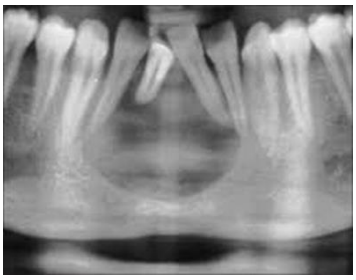
Differential diagnosis: Median rhomboid glossitis, benign and malignant tumors.

Treatment: Surgical removal.

6- Median mandibular cysts

Like globulomaxillary cysts, were once considered fissural cysts, in which a fissural origin was based on the theory of epithelial entrapment in the midline of the mandible during the "fusion" of each half of the mandibular arch. There is now embryologic evidence of an isthmus of mesenchyme between the mandibular processes that is gradually eliminated as growth continues, and therefore no evidence of epithelial fusion. So recently it is thought to be of odontogenic origin.

Clinical and radiographical features: Swelling In the midline of the mandible. In x-ray it appears as a well circumscribed radiolucency between the two lower central incisors in the midline.



Histopathology: Lining epithelium is mainly stratified squamous. C.T. wall is free from inflammation.

Treatment: surgical removal

7- Median palatal cyst (palatine cyst):

It is rare fissural cyst that develops from epithelium entrapped along the embryonic line of fusion of lateral palatal shelves. This cyst may be difficult to distinguish from nasopalatine duct cyst.

Clinical and radiographical features:

This cyst is present as firm or fluctuant swelling in the midline of the hard palate posterior to the palatine papilla. Most of these cysts are asymptomatic, but sometimes pain may be present.

X-ray: occlusal radiograph showed a well —circumscribed radiolucency in the midline of the hard palate.

A midline radiolucency without clinical evidence of expansion is probably a nasopalatine duct cyst.

Histopathology: Cyst is usually lined by stratified squamous epithelium. Areas of ciliated pseudostratified columnar epithelium may be present in some cases. Chronic inflammation may be present in the cyst wall. Treatment: surgical removal.

8- Oral lympho-epithelial cyst

It occurs intraorally and usually located in the posterior part of the tongue or in the floor of the mouth and sometime near the soft palate and the pharynx and in the tonsillar area (lymphoid tissue).

Clinically: asymptomatic swelling in the oropharynx area, lateral border of the tongue and floor of the mouth.

9- Dermoid & Epidermoid cyst

These represents a simple form of cystic teratoma derived from skin epithelium entrapped during embryonic development. Most of these cysts occur in the head & neck region, primarily in the skin around the eyes & the anterior upper neck, extending superiorly into the floor of the mouth.

Clinically: Mostly occur in young adults, present as painless swelling exhibiting a doughy consistency on palpation, & may cause elevation of the tongue & can interfere with eating & speaking.

Histopathology: The cyst lined by a layer of orthokeratinized squamous epithelium, surrounding by C. T. capsule. In dermoid cyst in addition to these, the lesion exhibiting variable numbers of dermal appendages including hair follicles, sebaceous glands. Treatment: surgical excision.

Dermoid cyst:

These cysts probably form as a result of some abnormality of development of the branchial arches or pharyngeal pouches. It is generally classified as a benign form of Teratoma.

Clinical features

Dermoid cysts develop between the hyoid and jaw or may form immediately beneath the tongue. They are sometimes filled with desquamated keratin giving them a semi- solid consistency

If the cyst develops above the geniohyoid muscle a sublingual swelling may displace the tongue upward and create difficulty in eating, speaking or even breathing. Cysts that occur below the geniohyoid muscle often produce a submental swelling with a double chin appearance.

Dermoid cyst is more deeply placed than a ranula; the latter is obviously superficial, having a thin wall and a bluish appearance. A dermoid cyst causes no symptoms until large enough to interfere with speech or eating.

Pathology:

The lining of epidermoid cysts is keratinising stratified squamous epithelium alone. Less often, cysts also have dermal appendages (sebaceous gland, hair follicle or sweat gland) in the wall and are then referred to as dermoid cysts. These cysts should be removed surgically.

Treatment - routine follow up