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Developmental Anomalies of Tongue-A Review

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Abstract:- The development of tongue is from the pharyngeal arches during the intrauterine life. There are various developmental anomalies of tongue like aglossia, microglossia, macroglossia, fissured tongue, benign migratory glossitis, median rhomboid glossitis, hairy tongue, lingual varices and cleft tongue. The syndromes associated with each and every developmental anomaly of tongue is described. The clinical features along with the histopathological features of the various developmental anomalies of tongue are discussed here. Along with this the treatment criteria for each anomaly is also discussed in detail.

Keywords:- *Tongue, Development of Tongue, Developmental Anomalies, Clinical Features, Histopathological Features, Treatment.*

I. INTRODUCTION

The tongue is a complex muscular structure and an organ which is most sensitive. The predominant functions like speech, deglutition and mastication of the stomatognathic system involves the active role of tongue[1].

➢ *Development of Tongue:*

The development of tongue starts around fourth week of the gestation period. The pharyngeal arches that contribute to the development of various parts of the tongue are first, second, third and fourth[1].

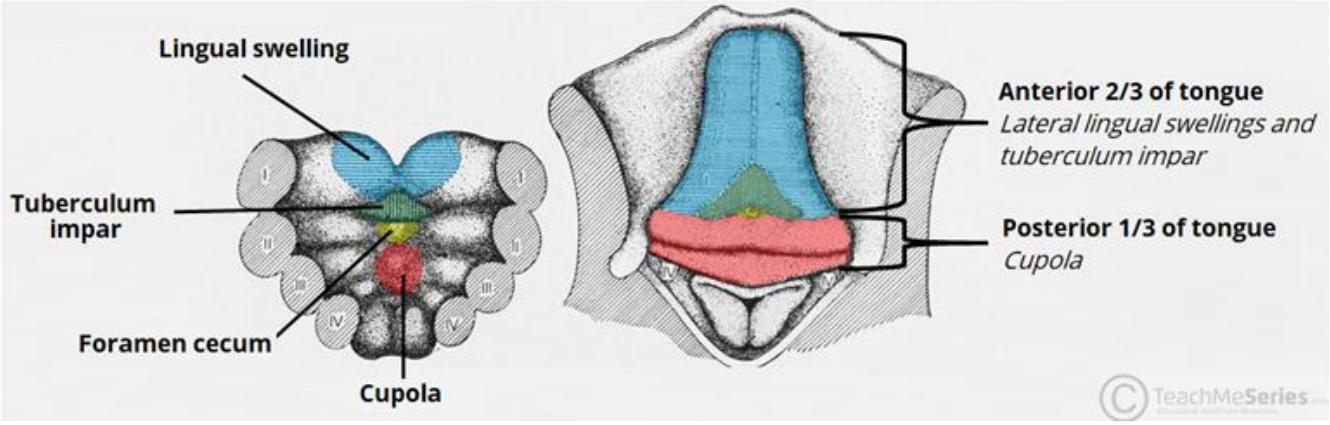

Fig 1 Development of Tongue from Pharyngeal Arches[2].

➢ *Anterior Two-Third of the Tongue:*

The anterior two-third of the tongue begins with the growth of the medial swelling from first pharyngeal arch called as tuberculum impar. Later two lateral lingual swellings develop from the same arch during the fifth week of the intrauterine life. Gradually the lateral swellings increase in size and fuse together which overlaps the tuberculum impar leading to the formation of the anterior two-third of the tongue. The sensory innervation of this part of tongue is from the mandibular branch of the trigeminal nerve (refer figure 1) [1].

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➢ *Posterior One-Third of the Tongue:*

A median swelling known as the hypobrachial eminence starts to develop from the mesoderm of the second, third, and fourth pharyngeal arch to form the posterior one-third of the tongue. The mucosa of this part gets its sensory innervation from the glossopharyngeal nerve.

The posterior most portion of the tongue develops from the third median swelling that arises from the fourth pharyngeal arch. It is innervated by superior laryngeal nerve (refer figure 1) [1].

➢ *Muscles of the Tongue:*

The intrinsic and extrinsic muscles of the tongue are derived from the myoblasts which originates from the occipital somites[1].

➢ *Taste Bud:*

During the 8th week of gestation the first sign of development of taste bud on the lingual epithelium occurs. And many taste bud primordia develop during ninth and eleventh week of the intrauterine life which further differentiates into different cell types from eleventh to thirteenth postovulatory week[1].

Table 1 Syndromes Associated with Developmental Anomalies of Tongue[3]:

Anomalies of tongue	**Associated syndromes**
1.Aglossia	1. Occulo-auriculo-vertebral syndrome or Goldenhar's syndrome
	2. Aglossia-Adaktyla syndrome
2.Microglossia	1. Microglossia-glossoptosis syndrome or Robin's syndrome or Robin's-Lenstrup syndrome or Pierre Robin syndrome
	2. German's syndrome
	3. Congénital facial displegia or Moebius syndrome
	4. Oromandibular-limb-hypogenesis syndrome
	5. Hypoglossia-Hypodaktylia syndrome
	6. Faciocardiomelic dysplasia
	7. Peromelia and Micrognathia (Hanhar's syndrome)
3. Tongue hemiatrophy	1. Parry-Romberg syndrome
4. Tongue hemihypertrophy	1.Congenital hemifacial hyperplasia
5.Macroglossia	1. Exomphalos-macroglossia-gigantism syndrome or Beckwith's syndrome or Beckwith-Wiedemann syndrome
	2. Neurofibromatosis syndrome
	3. Generalized gangliosidosis (GMj)
	4. Mucopolysaccharidosis I-H or Hurler's syndrome
	5. Mucopolysaccharidosis I-S
	6. Mucopolysaccharidosis II or Gargoylism or Hunter's syndrome
	7. Mucopolysaccharidosis III
	8. Mucopolysaccharidosis VI A&B or Maroteaux - Lamy syndrome
	9. Hyalinosis cutis et mucosae or Lipoid proteinosis or Urbach Wiethe syndrome
	10. Mannosidosis
	11. Pycnodysostosis or Osteopetrosis
	12. Pud's syndrome
	13. 4p + syndrome
	14. 7p + syndrome
	15. Cloverleaf Skull or Tri-Lobed Skull or Kleeblattchader syndrome
	16. Trisomy 21 syndrome or Down's syndrome or Mongolism
	17. Soto's syndrome
6.Long tongue	1. Ehlers-Danlos syndrome
7.Ankyloglossia	1. Murray's syndrome or Van der Wonde's syndrome 2.Cryptophalmos syndrome
8.Cleft or bifid tongue	1. Oculo-auriculo-vertebral dysplasia or Goldenhar's syndrome
	2. OFD II syndrome or Mohr's syndrome
	3. Cleft-palate-lateral synechiae syndrome or CPLS syndrome
	4. OFD I syndrome or oro-facial-digital syndrome or Gorlin-Psaume syndrome
	5. Focal dermal hypoplasia
9.Non classified syndrome	1. Melkersson-Rosenthal syndrome
	2. Coffin-Lowdry syndrome
	3. Klippel-Trenaunay-Weber syndrome
	4. Riley-Day syndrome
	5. Fissured tongue syndrome

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II. CLINICAL FEATURES OF THE ANOMALIES

➢ *Aglossia:*

The complete absence of tongue since birth is described as aglossia[4]. These abnormalities are probably thought to be the disruptive consequences of hemorrhagic lesions during fetal development (vascular disruption around the fourth embryonic week) [5]. Patients with aglossia usually encounter difficulty in eating and speaking[6].

It is a rare and sporadic congenital malformation seen in children. It may be present as an isolated disorder or may be seen in association with other congenital deformities which include cranio-facial anomalies like partial or complete anodontia, microstomia, cleft palate, micrognathia, eye-lid defects, facial asymmetry, and cranial nerve palsies[5].

➢ *Fissured Tongue:*

It is clinically presented as a numerous grooves and furrows on the dorsal surface of the tongue which originates from the central groove and pass to the lateral border[7].

It is common to refer to more serious fissuring as "lingua plicata." This idiopathic condition is more prevalent in older people and is believed to be a reactive process [8,9,10]. Geographic tongue sufferers and people with dermatitis are more likely to have FISSURED tongue [11]. It is the most frequent tongue finding found in up to one-third of psoriasis individuals[12]. Acromegaly, Psoriasis and Sjogren syndrome are most commonly seen in association with fissure tongue[13].

➢ *Microglossia:*

It is a very rare condition with about an approximate of 50 cases reported in literature till date[14]. Jussieu first explained this anomaly in 1718. The main feature is abnormal condition of having small tongue due to developmental disturbance. This disorder is a frequent finding in association with limb abnormalities where it is grouped as a hypoglossia-hypodactilia syndrome. Microglossia frequently is known to be linked with hypoplasia of the mandible, and the lower incisors may be missing[15]

➢ *Geographic Tongue:*

The lesions of geographic tongue are characterised by loss of filiform papillae with patchy,sharply demarcated, irregular areas of surface erosion.

It is caused due to recurrent acute inflammatory diseases[16] and most commonly seen in patients with insulin dependent diabetes mellitus due to presence of tissue type HLA-B15[17].

➢ *Benign Migratory Glossitis:*

All age groups are affected by benign migratory glossitis (BMG), also known as geographic tongue and annular transient spots of the tongue. 1 to 2% of people have BMG, which is more prevalent in youthful patients and frequently gets worse as people get older to 25-28 years. An annular pattern of alternating raised, hyperkeratotic plaques and smooth, atrophic areas defines BMG[18].

The dorso-lateral aspect of the tongue is more frequently affected by the lesions, which are dynamic and alter over hours to form a "migratory pattern. BMG frequently experiences ups and downs. Geometric stomatitis, or ectopic BMG, is uncommon[19].

Up to 1 in 4 people with BMG experience a burning sensation or food sensitivity. BMG is frequently asymptomatic[18].

➢ *Macroglossia*:

In general, the word "Macroglossia" refers to tongue enlargement that may cause both functional and aesthetic issues (also known as "tongue hypertrophy," "prolapsus of the tongue," or "enlarged tongue" or "pseudo Microglossia"). Virchow once referred to it as a type of elephantiasis.

Butlin and Spencer ascribed it to inflammation, muscle hypertrophy, or lymphatic dilation in the past 100 years[20].

Depending on the precise cause, Macroglossia can look differently under the microscope.

Macroglossia has been linked to neurofibromatosis, angioedema, hypothyroidism, Down syndrome, TB, sarcoidosis, amyloidosis, multiple myeloma, infections (syphilis), and other conditions[21] .An abnormal protein material is deposited in the tongue of an amyloidosis sufferer.

➢ *Median Rhomboid Glossitis:*

It is a congenital anomaly often present in association with Candida albicans[7]. It is commonly observed on the tongue's dorsum surface. A well-defined central papillary atrophy of the tongue called medial rhomboid glossitis (also known as central papillary atrophy of the tongue) occurs prior to the circumvallate papillae.

- It is flat, nodular, and barely elevated.
- The absence of filiform papillae makes it stick out clearly from the rest of the tongue.
- This atrophic region typically has no symptoms.
- Three times as many men are impacted as women[22].
- Although there are rarely any symptoms, it is possible to experience scorching or itching[23].
- It frequently results from a fungal infection and can be treated with antifungals (such as nystatin, clotrimazole, and fluconazole) given orally or as a suspension[24,25].

➢ *Hairy Tongue*:

A filiform papillae retention hyperkeratosis on the anterior 2/3 of the dorsal part of the tongue causes hairy tongue (HT), also known as a furred tongue[26]. Rates of HT are inconsistently described in the literature, ranging from 0.5% to 11.3%[27]. As much as three to one seems to favour men, and the frequency is higher in older people—40% in those over 60—than in younger people[27]. Papillae

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have a typical length of 1 mm, but in people with HT, the central column of the filiform papillae has defective cell desquamation, which results in an increase in length of 10–20 times that of a normal papillae[28].

➢ *Lingual Varices:*

A vein, artery, or lymph channel that is enlarged and tortuous is said to have varicosity. Orally, varicosities are perceived as purple to crimson clusters with shooting edges. Varices are uncommon in children but prevalent in older adults, suggesting that age may play a significant role in their etiology[15].

Oral varicosities are thought to most frequently manifest as sublingual varix. Sublingual varicosities are typically visible as numerous blue-purple, elevated, or noticeable blebs on the tongue's lateral and ventral sides. Less frequent locations include the lips and buccal mucosa[29]. With the exception of a few rare cases where secondary thrombosis develops, the lesions are typically asymptomatic. Given their characteristic feature of multiple, rounded little masses of purplish blue colour, they have been given the common term "caviar tongue"[30].

➢ *Cleft Tongue:*

A completely cleft or bifid tongue is an extremely uncommon disease that is thought to result from the lateral lingual swellings of this organ not merging. More frequently, partial tongue clefting causes a deep furrow to appear in the middle of the dorsal surface[20].

III. HISTOPATHOLOGIAL FEATURES OF ANOMALIES OF THE TONGUE[31]

➢ *Macroglossia*

- In case of tumour, proliferation of a specific tissue type that has undergone neoplastic changes for example, lymphatic vessels, blood vessels and neural tissue.
- In case of amyloidosis, deposition of abnormal protein material in the tongue occurs.

➢ *Fissured Tongue:*

- On the surface of the filiform papillae there is loss of keratin and the rete pegs undergo hyperplasia and there are grooves in between the papilla separating them from one another.
- Presence of microabscess in the upper epithelium is evident.
- Migration of the polymorphonuclear leukocytes into the epithelium is also present.

➢ *Medial Rhomboid Glossitis:*

- The stroma is moderately fibrosed, with dilated capillaries covered by atrophic stratified squamous epithelium forming a smooth or nodular surface.
- There is no presence of fungiform and filiform papillae.
- The subepithelial deeper fibrovascular tissue shows mild to moderate intense chronic inflammatory cell infiltrate.

➢ *Erythema Migrans:*

- In the keratin layer and spinous layer, micro abscesses known as Monro's abscess are produced by the inflammatory cells.
- Long and thin rete ridges are present covering the connective tissue with a thin layer of epithelium.
- Chronic inflammatory cell infiltration is present in the stroma.
- On silver or PAS staining candida hyphae or spores are present in the superficial layers of the epithelium.

➢ *Lingual Varices:*

- Presence of dilated vessels with smooth muscle and elastic fibres.
- Lumen of the vessels may be occluded by platelets and erythrocytes due to secondary thrombosis.

➢ *Lingual Thyroid Nodule:*

- Embroyonic thyroid tissue or normal thyroid tissue is present in incomplete or poorly defined capsule with atrophic follicular cells.

➢ *Hairy Tongue:*

- The filiform papillae are the hairs on the surface with hyperplasis of the rete ridges and loss of keratin.
- The papillae are separated by deep grooves which vary in size.
- Polymorphonuclear leukocytes migrating to the epithelium leads to formation of microabscess in the upper epithelium.
- Lamina propria shows mixed inflammatory cell infiltration.

IV. TREATMENT OF THE DIFFERENT ANOMALIES

➢ *Aglossia:*

Patients with aglossia usually suffer from difficulty in eating and speaking [6]. Such patients do not need reconstruction of the tongue; as feeding, and swallowing adaptation is often shown by the patient with increasing age [31].

➢ *Ankyloglossia:*

The superior and recommended procedure of ankyloglossia is frenectomy ("clipping" or simple release of the frenulum), especially in patients with speech defects [15].

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- *Fissured Tongue:*

Treatment is not necessary. Treatment is necessary in case of mild inflammation which is present at the base of fissures. Maintenance of good oral hygiene and brushing of the tongue, especially deep to the fissures to remove debris and halitosis [32,33].

If pain is present, there are chances of a systemic disease/infection, and treatment should be aimed at reducing inflammation or removal of the infection [34].

- *Median Rhomboid Glossitis:*

Topical fungal agents are recommended. It is found commonly in association with candida species infection and reacts well to antifungal medications (e.g., nystatin, clotrimazole, fluconazole). It can be given as a suspension or oral route [35,25].

- *Geographic Tongue:*

It is self-limited and no treatment is indicated other than reassuring the patient [36].

- *Lingual Thyroid Nodule:*

In asymptomatic patients with lingual thyroid nodule a follow up is maintained periodically. In symptomatic patients, suppressive therapy with supplemental thyroid hormone will reduce the size of the lesion [15].

- *Cleft Tongue:*

Surgical correction, in case of a partial or complete cleft of the tongue [31].

- *Hairy Tongue:*

No treatment is required as it is self-resolving [37]. Due to its appearance, patients wish to undergo treatment. First-line of treatment is the improvement of oral hygiene by brushing the tongue using dentifrice or using 1.5% hydrogen peroxide (5 to 10 strokes daily) with a hard toothbrush. Second-line treatments include topical retinoids, antifungals, and keratolytic agents[38]. Oral therapy using antifungals, antibiotics, and antivirals should be prescribed for cases with positive cultures [34].

- *Oral Varices:*

Treatment is not required. However, conservative excision is effective [34].

- *Infantile Hemangioma:*

The treatment is dependent upon the risk to vital organs, structures, and cosmesis. Now, Oral propranolol is FDA-approved and has become the gold standard treatment [39]. Positive outcome rates for propranolol are 98% with a goal dose of 2mg/kg/day and 6 months of therapy. Laser therapy is also effective but it may not be appropriate for intraoral disease [40]. Corticosteroids, interferon alpha, and vinca alkaloids are other treatment options[34].

V. CONCLUSION

There are various clinical manifestations of developmental anomalies of tongue. The histopathological features shows the clear description of the anomaly. Proper identification of the anomaly can be done from analysing both the clinical and histopathological features. The above content clearly depicts the stage of development at which the anomaly occurs and the causes which leads to the development of the anomaly. The various treatment strategies required for the various anomalies are discussed here and some may not require any of the treatment. To conclude, this article gives clear knowledge regarding the developmental anomalies of the tongue in all aspects.

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