Organization Logo **Photograph**

Non- syndromic bifid tongue: a case report

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Introduction

A bifid or cleft tongue (glossoschissis) is a tongue where a groove or split runs lengthwise along the tip of the tongue. They can be syndromic or non-syndromic and may occur along with other oral abnormalities. Disturbance in the organogenesis of tongue can lead to malformations like tongue tie, bifid tongue and hairy tongue.

Case Report: History

51-year-old women reported with a chief complaint of difficulty in pronunciation during talking from her childhood. History revealed that she had a consanguineous marriage and that her mother had no history of abortion or drug intake during pregnancy. There was also no history of hereditary predispositions and postnatal trauma. Surgical correction of the defect was planned under local anesthesia. Past medical history revealed that she was hypertensive and was on medication for the same for the past 15 years.

Case Report: Examination

General examination revealed that she was obese with a body mass index of 29kg/m2.Extra oral examination revealed -slanted palpebral fissures, alopecia, dry scalp with coarse hair, prominent nose tip, excessive facial hair with no orofacial defects. Intraoral examination revealed a split in the tip and anterior 2/3rd of the tongue with the tip diverted to the left There was absence of any tongue piercings. The upper frenum was hypertrophic and the cleft between the upper lateral incisors and canines. Intraoral examination also revealed the presence of bilateral deciduous canines.(fig.1)

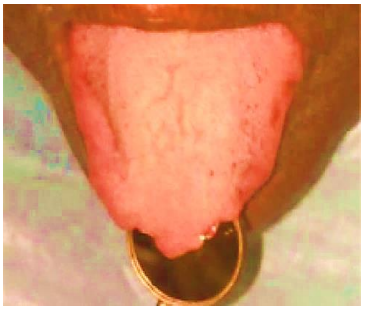

Fig.1 – Bifid tongue

Case Report: Investigations

Laboratory investigations revealed an abnormal glucose leading test and abnormal 3-hour glucose tolerance test.

Case Report: Diagnosis
History, systemic, extraoral and intra oral examination suggests that this is a case presentation that cannot be categorized into any well-defined syndrome- non-syndromic bifid tongue

Case Report: Management
Surgical correction of the defect was done under local anesthesia median parts of the defect were reconstructed by suturing the muscles in layers.Post-operative medications consisting of analgesics and anti-inflammatory medicines were prescribed. Patient was called after 10 days for a follow-up. Post-surgical healing was uneventful and no speech therapy was needed.

Case Report: Discussion
Bifid tongue is a rare clinical feature caused by failure of fusion of the branchial swellings during organogenesis. It is associated commonly with a syndrome. However, non-syndromic cases have also been reported. [1,2]

Development of tongue
During the fourth week of intrauterine life, the tongue is formed by the fusion of tuberculum impar and two lateral lingual swellings which originate from the first pharyngeal arch. The posterior part of the tongue is formed by a median swelling called Copula or hypobranchial eminence, originating from the second, third, and part of the fourth branchial arch.When the process of lingual structures rapidly growing to cover the tuberculum impar to form the anterior two-thirds of the tongue occurs, the tongue may divide longitudinally to form bifid tongue or cleft tongue in the presence of any disturbance[3].(fig.2, fig.3,fig.4, fig.5)

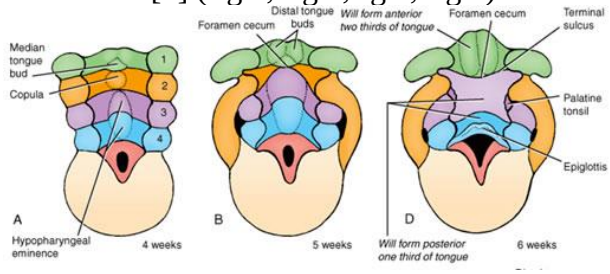

Fig.2 – Branchial arches

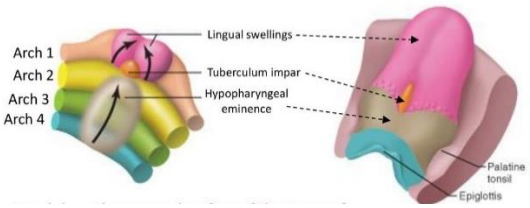

Fig.3 – Development of tongue

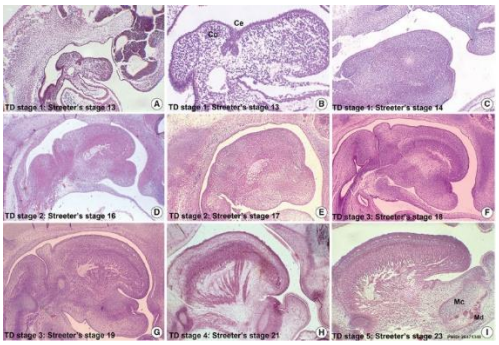

Fig.4 – Development of tongue- histology

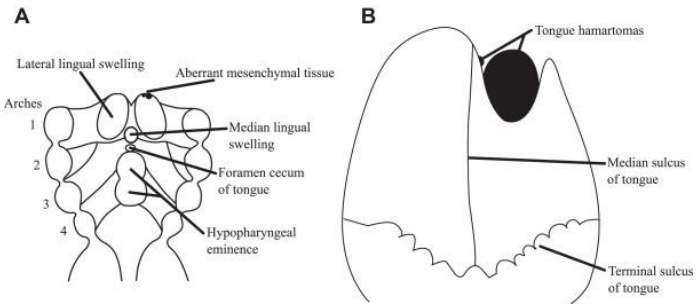

Fig.5 – Development of bifid tongue

Classification of tongue anomalies(fig.6)

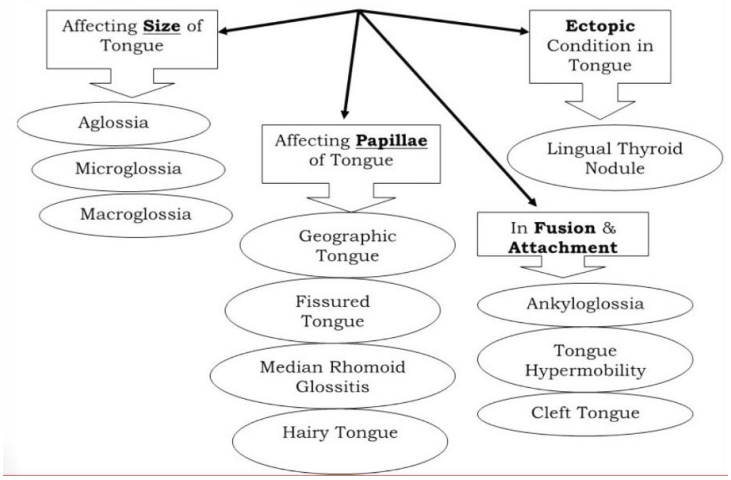

Fig.6 – Classification of tongue anomalies

Syndromes associated with bifid tongue

Oro-facial-digital syndrome I (OFDS I)(Papillon-League and Psaume syndrome)- mutations in the OFD1 gene leading to abnormal developmental signalling pathways critical to cellular development. Oro-facial-digital syndrome II (OFDS II) , also called Mohr's syndrome is an autosomal recessive disease with unknown molecular genetic cause.

OFDS I is characterized by bifid tongue in comparison to OFDS II where the induvial exhibits tongue nodules. OFDS I is characterized by bifid tongue in comparison to OFDS II where the induvial exhibits tongue nodules. OFDS I is also characterized by cleft lip, cleft palate, hypodontia, micrognathia and polycystic kidney disease.(fig.7, fig.8)

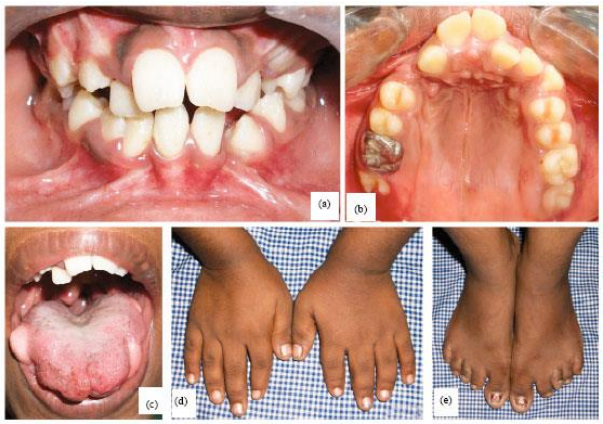

Fig.7 – Clinical features of OFDS

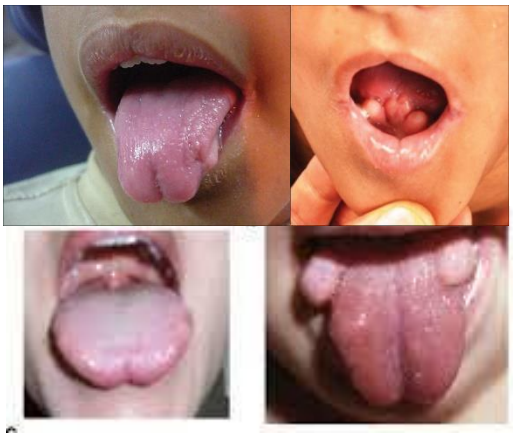

Fig.8 – Tongue nodules seen in OFDS

Tessier type 30 craniofacial cleft also known as "median mandibular cleft is a rare congenital anomaly characterized by a cleft through the tongue, lower lip and mandible. Bifid tongue with ankyloglossia and double tongue has been reported to be associated with this syndrome.(fig.9)

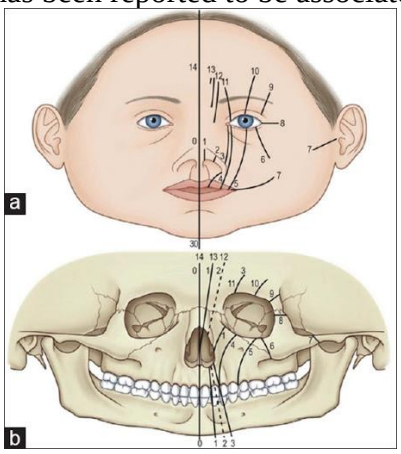

Fig.9 – Tessier type 30 craniofacial cleft features

Mandibular cleft, cleft of the lower lip, hyoid split, sternum split, split of the tracheal cartilages, thyroglossal cyst and dermoid cyst have also been reported to be associated with this syndrome.(fig.10)

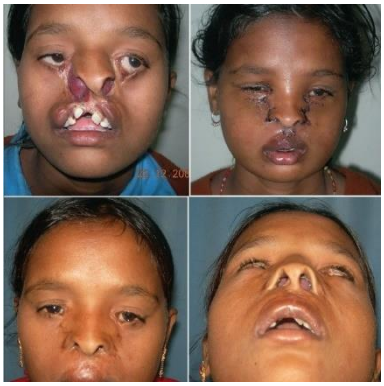

Fig.10 – Clinical features of mandibular cleft

Opitz G BBB syndrome is a rare genetic disorder which can be either autosomal dominant of X linked recessive. This disorder affects structures along the midline of the body. This is characterized by hypertelorism, laryngo-tracheo-esophalgeal and hypospadias.Cases with bifid tongue have also been reported.(fig.11, fig.12)

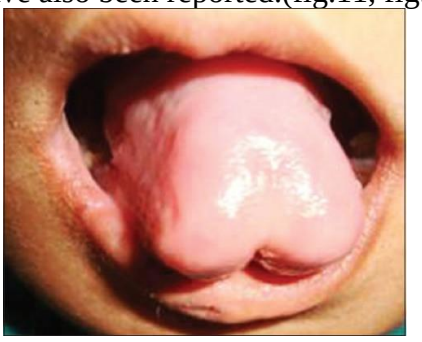

Fig.11 – Bifid tongue seen in Opitz syndrome

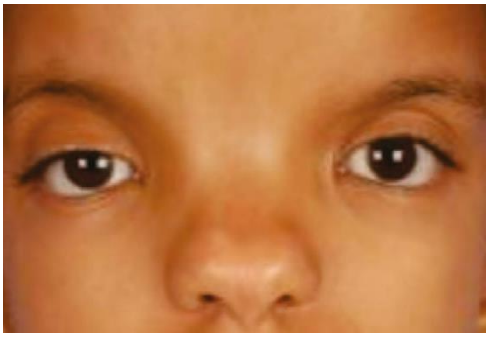

Fig.12 – Hypertelorism

Klippel–Feil anomaly syndrome is a rare congenital condition also called as cervical vertebral fusion syndrome reported to be associated with mutations of the genes GDF6, GDF3 and MEOX1. This leads to abnormal fusion of the bones in cervical vertebrae.It is commonly characterized by restricted movement of the neck.The presence of bifid tongue has also been reported in a few cases.(fig.13)

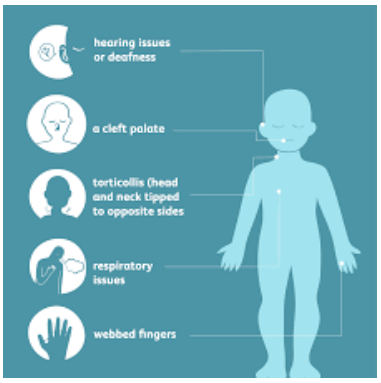

Fig.13– Clinical features of Klippel-Feil syndrome

Larsen syndrome is a congenital disorder which can either be autosomal dominant or recessive. It is caused by mutation in the gene encoding for the protein filamin B. Defects in this leads to abnormal cytoskeleton. The common orofacial features include cleft palate, malocclusion, hypodontia and microdontia. Other signs and symptoms include prominent forehead, flattened facial appearance, dislocated hips, shoulders and knees.Bifid tongue has also been reported in a few cases. [3-6] (fig.14)

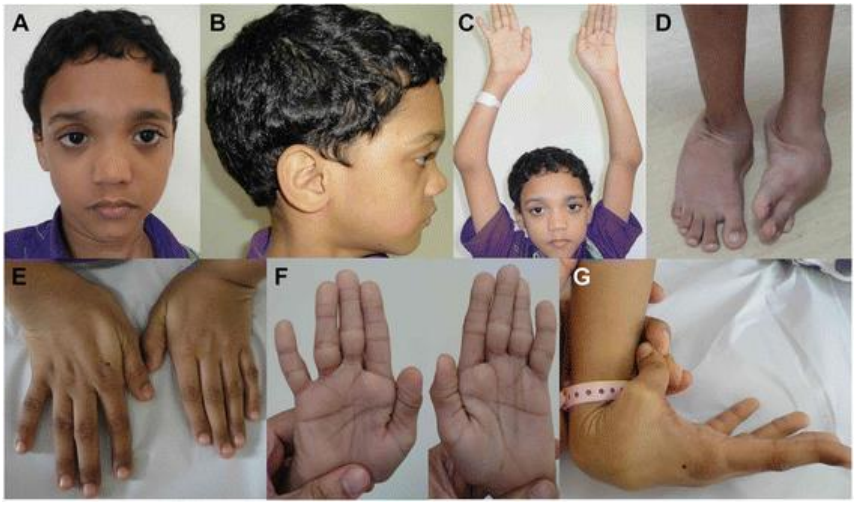

Fig.14 – Clinical features of Larsen syndrome

Diabetic mothers have higher congenital malformation rates. [7] They include cardiovascular, respiratory, gastrointestinal, genitourinary, musculoskeletal, renal and neural disorders. Bifid tongue has been reported in few cases. Rare cases of tongue piercings causing bifid tongue have also been reported. [8] (fig.15, fig.16)

Fig.15 – Tongue piercing

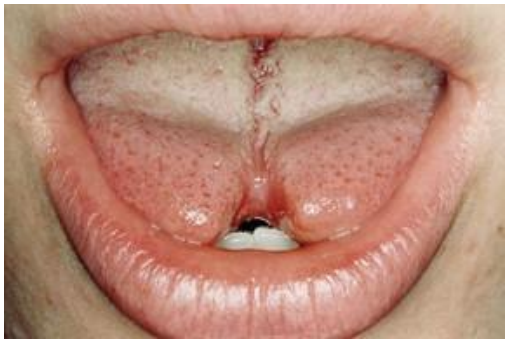

Fig.16 – Bifid tongue due to tongue piercing

This case report describes a patient with bifid tongue who has no other features suggestive of the above-mentioned syndromes. A reason for delayed finding of bifid tongue could be due to the fact that Hindus consider bifid tongue a “good fortune” since they resemble snakes which are considered holy in Hindu mythology.

Case Report: Conclusion

Bifid tongue is a rare condition which is mostly encountered as a developmental abnormality.
Non - syndromic bifid tongue is reported in this article which suggests that bifid tongue need not always be found associated with syndromes. Proper patient history and clinical examination must be ensured before it is misdiagnosed as a syndrome.

References

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