A Case Report of Hamartomatous Bifid Tongue: A Rare Congenital Anomaly

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Abstract

Congenital bifid tongue is a rare malformation, which is usually present in association with other oral findings such as cleft palate and tongue mass. The authors have found a rare case of congenital bifid tongue together with absence of lingual frenulum, and a hamartomas mass in a 9-month-old female infant. Mother complains of difficulties in sucking breast milk and dribbling milk during breast feeding, as well as some episodes of positional dyspnea and choking with cyanosis during feeding. The mass was excised, along with an excess portion of the left tongue, and the bifid tongue was surgically repaired in layers after excision. The histopathological report was used to confirm the diagnosis of hamartoma. The review of literature, description of the lesion, diagnosis, and management of this finding are all outlined.

Keywords

- Hamartoma, Bifid Tongue
- Oral-facial-digital syndrome (OFDS)
- Lingual leiomyomatous hamartoma (LLH)

Introduction

Tongue forms in the primitive oral cavity's floor during the fourth week of pregnancy. There are two lateral and one midline lingual swellings. The anterior 2/3rd of the tongue's body is made up of tuberculum impar from the first pharyngeal arch. The lateral lingual structures quickly expand and cover the tuberculum impar, forming the tongue's anterior 2/3rd. The tip of the tongue is divided longitudinally with some length and when this process is disrupted, it results in a cleft tongue or bifid tongue. They can occur on their own or as a part of a clinical condition that includes oral-facial-digital abnormalities. [1,2,3,4]

An extremely unusual abnormality is a non-syndromic congenital bifid hamartomatous tongue. A hamartoma is a tumor that develops as the proliferation of normal tissues that are indigenous to the place of occurrence. Hamartomas' tissues

are typically disordered and ill-defined, blended with the surrounding tissues. Vessels, nerves, lymphatics, skeletal muscle, fat, and salivary gland tissue are all endogenous factors that could cause a hamartoma within the tongue.[5] Angiomatous and lymphangiomatous hamartomas are frequent in the oral cavity. Leiomyomatous hamartoma in the oral cavity is quite uncommon.

This case report is about a lingual leiomyomatous hamartoma (LLH) with a bifid tongue, which is a rare occurrence. Despite its rarity in the general community, hamartoma is widespread in those who have the oral-facial-digital syndrome (OFDS).[6]

Case Report:

The patient was a 9-month-old female infant who born with a swelling on the dorsum of the anterior 1/3rd of the tongue and the tongue was completely bifid in its anterior 2/3rd. The baby was suffering from choking episodes during feedings and was reluctant to feed with dribbling of milk during sucking. Other developmental milestones were normal, and the baby was otherwise healthy. The tongue was anteriorly bifid (V-shaped defect) with the line of division more towards the left side on examination.

The routine oral examinations showed a 10 mm long fissure located at the left laterally from the midline of the tongue, the fissure extended from the tip to the body of the tongue, a 10 x 3 x 2 mm mass with a pedicle protruded from the dorsum of the tongue and was not linked to the lip and existed on the apex of the fissure at the body. The lingual frenulum was absent. The swelling was fitted into the oral cavity when she tried to close her mouth. The mass showed intact and smooth surface with rubbery consistency. (**Figure 1** A-D). The baby was born with normal delivery, neither of the parents had a family history of cleft palate or bifid tongue among first-degree relatives, and the rest of the physical examination of the patient showed no abnormalities.

A single stage correction was performed under general anesthesia. The oral mass was excised and sent to histological examination. Excision of a portion of the smaller left tongue segment and the lingual muscles were sutured precisely to achieve a near-normal tongue contour with a midline tip and good lingual extension. Because there is no PICU in the remote location, the kid was observed in the OT

room for 24 hours to look for any postoperative respiratory difficulty due to tongue edema. The child recovered without any complications and was symptom-free. The child is currently getting speech therapy.

The histopathological examination of the mass was done using Hemotoxylin and Eosin stains. The findings were compatible with a diagnosis of lingual leiyomyomatous hamartoma (LLH).

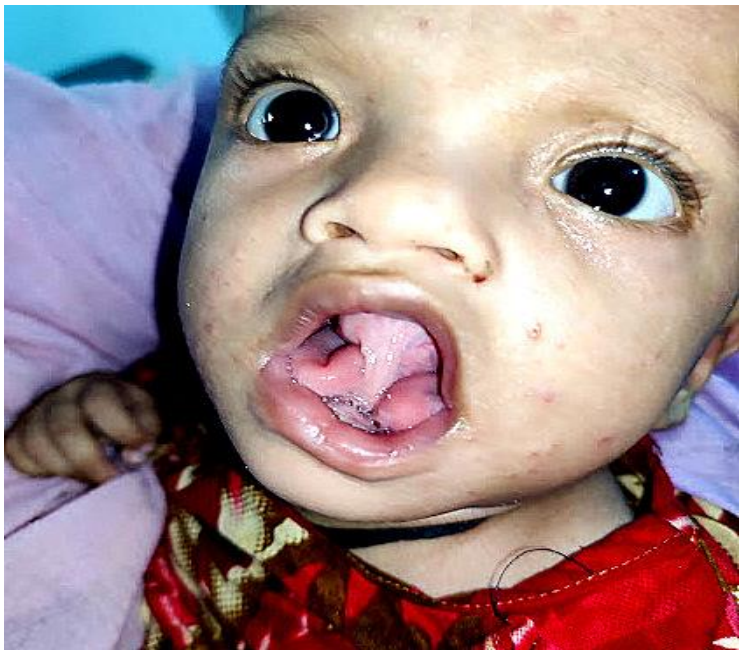

Figure 1: The mass was covered with normal stratified squamous epithelium with mature adipose tissue and salivary glands exhibited in the lamina propria of mucosa.

Three attempts at hydrostatic reduction under general anesthesia were unsuccessful and an exploratory laparotomy was performed. Intraoperatively, there was ileocolic intussusception which was reduced manually by milking the ileal intussusceptum from the intussuscipiens. On manual reduction of ileocolic intussusception, another ileo-ileal intussusception measuring about 8 cm

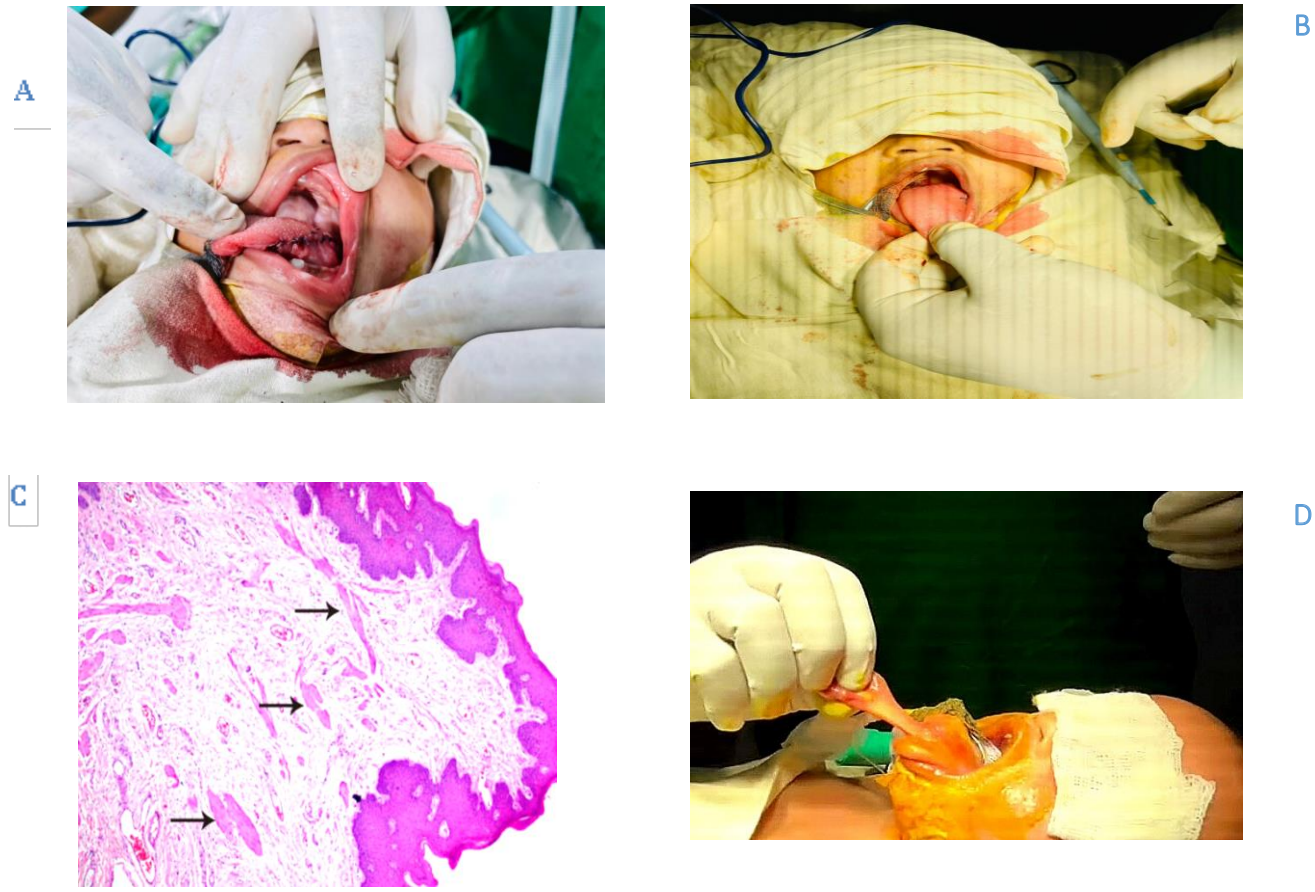

Figure 2: The swelling on the dorsum of the tip of the bifid tongue (A). The swelling was excised and frenulum was divided to allow the tip of tongue to reach beyond the lips. Glossoplasty was completed in three layers (B). The immediate post-operative appearance (C) (D)

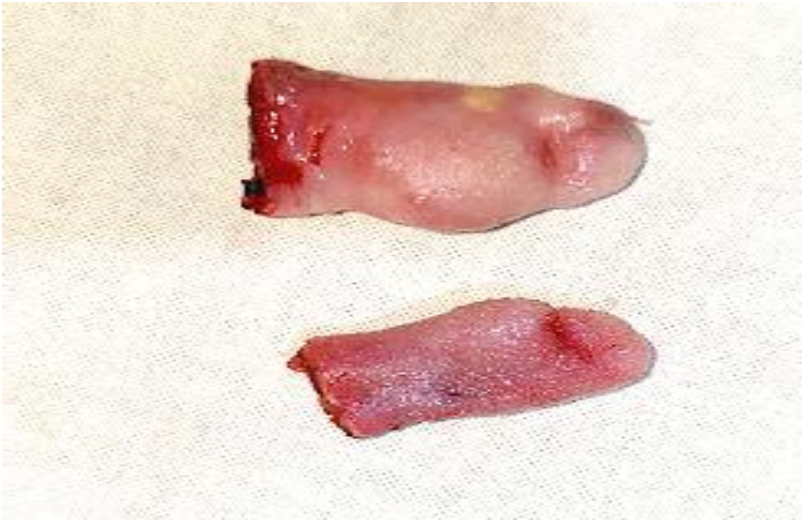

Figure 3: Excised hamartoma (A) and Part of left tongue (B)

Figure 4: Appearance at 7th POD.

Discussion

Bifid tongue is rare and has often been reported as associate features in oral-facial-digital syndrome.[7] However, in the present case, the patient presented with no other symptoms associated with the oral– facial– digital syndrome other than bifid tongue and a hamartomatous tongue mass, thus we defined this case as a non-syndromic condition.

Lingual hamartomas are uncommon, with only a few case reports have been found in the literature. Hamartomas are tumor-like malformations that have abnormally arranged normal tissue components. Fibrous tissue, fat, salivary gland acini, lymphatics, blood vessels, and smooth muscle make up lingual hamartomas.[8, 9] Hamartomatous growth of the lingual striated muscle or herniations through the underdeveloped lamina propria are proposed as explanations histologically.[10] Tongue hamartomas coexisting with OFDS and Dandy-Walker malformation, as well as isolated leiomyomatous hamartomas of the tongue without associated anomalies, can all occur.[11, 12] Glial tissue can occasionally be found in the tongue, causing choristoma.[13] Choristoma is a histologically normal ectopic mass. A 3-month-old female was diagnosed with an unusual tongue hamartoma, as well as ectrodactyly, ectodermal dysplasia, and cleft lip and palate syndrome. The lesion was made up of a haphazard collection of salivary glands, adipose tissue, smooth and skeletal muscles.[14] Stome et al.[13] emphasized the importance of intraoperative electrodiagnostic monitoring in preserving tongue neuromuscular function. These benign lesions can be life threatening as they can lead to positional dyspnoea, cyanosis, and bradycardia. In this case, there were episodes of positional dyspnoea and choking with cyanosis at the time of feeding. It seems that the long pedicle of the mass with increased mobility added to the problem. Excision of the lesion is simple but careful preservation of the neuromuscular function of the tongue using intra-operative electro-diagnostic monitoring is important.[15]

Conclusion

Bifid tongue is also known as cleft tongue/diglossia, and it is associated with a number of orofacial syndromes. Our case does not fit into any well-defined syndrome and is not linked to a history of postnatal

trauma, a genetic predisposition, or tongue piercing.

Ethical Consideration

Written consent for participation was obtained from the parent or guardian of the participant in the study**.** This study was approved by Palong General Hospital.

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Conflict of interests

There is no conflict of interest

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