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Case Report

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Congenital trifid tongue with macroglossia: a rare orofacial anomaly

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ABSTRACT

Trifid tongue is a rare congenital structural anomaly in which the anterior portion of the tongue is divided into three segments. It is most often associated with syndromic conditions such as orofacial digital syndrome. Neonates with a trifid tongue may present with airway obstruction, feeding difficulty, and drooling, necessitating early intervention. A 10-day-old full-term male neonate was admitted with feeding difficulty, drooling of saliva, choking episodes, and transient apnea. He had macroglossia and was unable to breastfeed. Surgical correction was performed using excision of the central tongue lobe and trimming of the medial aspects of the lateral lobes, followed by layered closure with absorbable sutures, preserving lingual neurovascular structures. The patient achieved effective sucking and feeding postoperatively. Early surgical management in congenital trifid tongue can result in excellent functional outcomes. A comprehensive evaluation to exclude associated syndromes is recommended.

Keywords: Trifid tongue, macroglossia, cleft soft palate, neonate, tongue reduction surgery

INTRODUCTION

Trifid tongue is a congenital and structural defect of the tongue in which its anterior end is divided longitudinally for a greater or lesser distance into three parts. The prevalence of trifid tongue associated with oro-facial digital syndrome type 1 is 1 in 50,000 to 1 in 250,000 live births.[1] But due to the rarity of the trifid tongue, the prevalence is not given in the literature. We present a rare case of trifid tongue accompanied by a soft cleft palate, with involvement of the anterior one-third of the tongue

Incomplete or abnormal development of structures derived from the lower half of the first branchial arch is rare; it may present as a complete or incomplete cleft of the lower lip, mandible, and tongue.[2]

The tongue is one of the main embryological structures that is derived from the pharyngeal apparatus. Towards. By the 5th to 6th gestational week, the (oral part) anterior 2/3rd develops from the fusion of two lateral lingual swellings and a median tongue bud called tuberculum impar.[3] The trifid tongue is formed by the failure of the fusion of lingual swellings with each other and with the tuberculum impar.[4]

A patient with an enlarged tongue may have symptoms of sleep apnea, respiratory distress, drooling, difficulty speaking, swallowing, and sucking. In the first two years of life, the airway symptoms in infants usually improve with supportive care. Urgent tongue reduction is recommended in infants with severe airway obstruction secondary to an enlarged tongue.[1]

CASE

A 10-day-old male baby weighing 3 kg was admitted to the pediatric ICU at Shaikh Zayed Hospital, Lahore. Examination at birth revealed an enlarged tongue. The patient was born at full term via cesarean section and passed meconium after 36 hours of his birth. He was unable to breastfeed because of an enlarged tongue since birth, and was being fed with a dropper.

He presented to the emergency department with complaints of transient apneic spells, though not quantified with SpO_2 values, choking, difficulty in sucking his mother's breast, drooling of saliva, constipation for 1 week, and abdominal distention for 1 day. On admission, he maintained an oxygen

saturation (SpO_2) of 94%-98% on room air. However, during episodes of apnea, transient desaturation was noted, with SpO_2 levels decreasing to approximately 88%–90%. In view of these recurrent apneic spells and airway compromise secondary to macroglossia, the patient was supported preoperatively with supplemental oxygen at 1 L/min via nasal cannula. There were no other antenatal and postnatal events. There was no history of any congenital anomaly in the family. He was the first child born to consanguineous parents.

On detailed examination after admission, the tongue was hypertrophied. Adequate examination of the soft and hard palate was not possible due to macroglossia. The mandible was normal, and the patient was unable to close his mouth due to an enlarged tongue. The abdomen was distended, with no palpable viscera. The spine and external genitalia were normal, and the anus was in its normal position. The remainder of the systemic examination was unremarkable. A workup for constipation was planned, along with examination of the oral cavity under anesthesia.

The patient was kept nil per os, and a microenema was given. He passed a small amount of stool, and abdominal distention was reduced. Complete blood count and serum biochemistry were within normal limits. A plain X-ray of the abdomen showed dilated bowel loops. Barium studies and ultrasound examination of the abdomen and pelvis were reported to be normal. The patient was operated on the 12th day of life, and a detailed examination of the oral cavity was done under general anesthesia; findings were noted, the tongue was hypertrophied, W-shaped, having three projections (**Figure 1**), and a soft cleft palate. The hard palate and mandible were normal. There was no evidence of ankyloglossia. Surgery was planned to correct the trifid tongue after detailed examination.

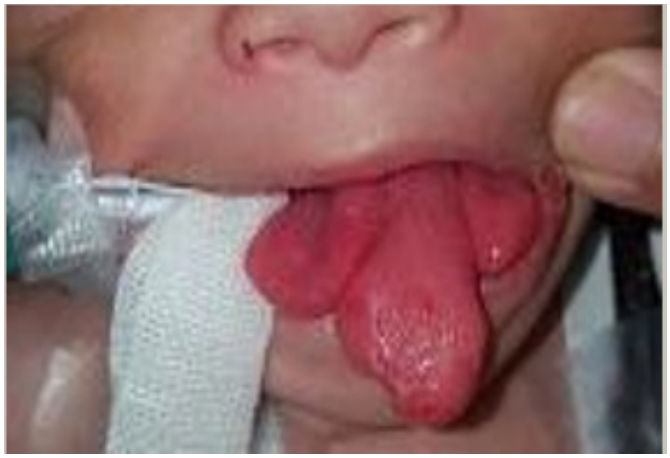

Figure 1. Preoperative intraoral photograph demonstrating a markedly hypertrophied tongue with a characteristic trifid configuration, showing three anterior lobulated projections consistent with congenital trifid tongue. The enlarged tongue occupied the oral cavity and contributed to impaired mouth closure and feeding difficulty.

Under general anesthesia, orotracheal intubation was achieved without difficulty, despite the enlarged tongue. The operative approach followed the principles of the Oji uniform tongue reduction technique, with minor modifications adapted to the trifid morphology.

Intraoperatively, the tongue was exposed using traction sutures placed at the anterior tip. A central wedge-shaped excision was designed along the midline, encompassing the entire central lobe of the trifid tongue. The excision extended from the anterior free margin posteriorly toward, but not beyond, the level of the circumvallate papillae. Cutting diathermy was used for excision to reduce tongue thickness. The resected segment measured approximately 2.0–2.5 cm in length and 1.0–1.2 cm in width at its widest anterior portion, tapering posteriorly. Additional controlled trimming of approximately 3–4 mm was performed along the medial edges of both lateral lobes to achieve symmetric contouring and adequate reduction in tongue width. Excision was limited to the intrinsic tongue musculature and performed using cutting diathermy to minimize blood loss. Particular care was taken to preserve the lingual arteries and the hypoglossal and lingual nerves by avoiding deep lateral dissection near the neurovascular bundles. Reconstruction was achieved with layered closure in three planes: approximation of the intrinsic musculature, closure of the submucosal layer to eliminate dead space, and mucosal closure using absorbable polyglactin sutures to restore a linear midline contour (**Figure 2, 3**).

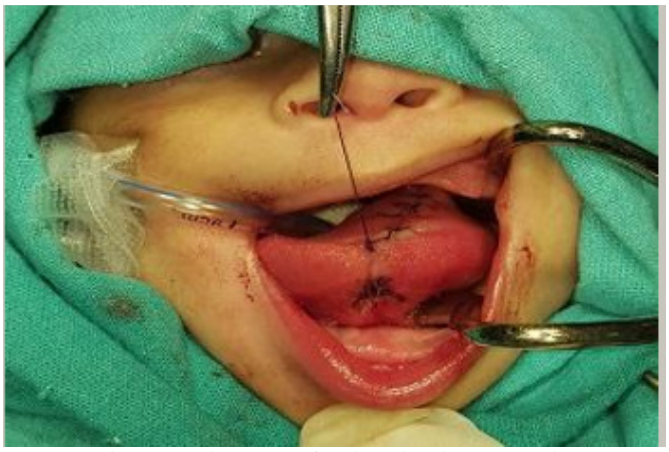

Figure 2. The image shows a uniformly reduced tongue with restoration of a linear midline contour, achieved after excision of the central lobe and controlled trimming of the medial aspects of the lateral lobes, consistent with a modified Oji tongue reduction technique

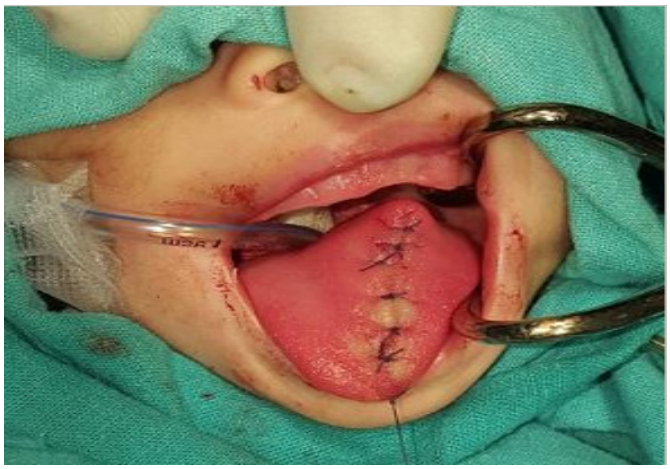

Figure 3. Immediate postoperative intraoral photograph demonstrating satisfactory tongue contour following reduction. The tongue shows reduced length and width, absence of lobulation, and adequate oral cavity space, allowing improved mouth closure. Sutured midline reconstruction is visible with preserved symmetry.

The surgery on the soft palate was deferred until the 3rd month of age. Excised tissue was sent for biopsy. Rectal biopsy was also done to rule out the suspicion of Hirschsprung's disease under the same anesthesia. The total operative time was approximately 40 minutes. There were no intraoperative complications, including bleeding, airway instability, or neurovascular injury. Postoperatively, the patient was maintained on prophylactic oxygen inhalation at 0.5 L/min, which was gradually tapered and discontinued on the first postoperative day without recurrence of desaturation episodes.

The patient demonstrated satisfactory tongue mobility, improved lip closure, and successful initiation of oral feeding by the third postoperative day with good sucking. Biopsy of the resected tissue showed normal tongue tissue. On rectal biopsy, ganglion cells and fine nerve fibrils were present. Ideally, genetic testing, detailed radiological studies, echocardiography, pulmonary function tests, CT scan, and ultrasound of the kidney, ureters, and bladder were indicated in this patient to rule out associations with other syndromes, but the parents refused these tests. Based on the results of the clinical and physical examinations, no additional anomalies were identified; however, the absence of advanced genetic and systemic assessments constitutes a limitation of this study.

DISCUSSION

Macroglossia with tongue lobulation has been described in association with several systemic conditions, including Beckwith–Wiedemann syndrome, vascular malformations, congenital hypothyroidism, glycogen storage disorders, and chromosomal abnormalities, most commonly Down syndrome.[1]

Trifid tongue is most frequently reported as part of the oro-facial-digital syndromes (OFDS), a heterogeneous group of disorders characterized by malformations of the oral cavity, craniofacial structures, and digits.[2] OFDS type I is the most common subtype, inherited in an X-linked dominant pattern and typically lethal in males, making live male presentation extremely uncommon.[3]

Mohr syndrome (OFDS type II), an autosomal recessive disorder, is characterized by a median cleft lip, a multilobed tongue, dental anomalies, and polydactyly.[5] However, most reported cases of trifid tongue occur in syndromic contexts rather than as isolated anomalies.

In our case, trifid tongue was associated with just a soft cleft palate and no evidence of facial dysmorphism, limb abnormalities, or systemic involvement.

Congenital tongue anomalies have also been reported in other syndromes such as Klippel–Feil syndrome and in infants of diabetic mothers, although these associations were not present in our case.[6,7]

Surgical intervention was indicated due to significant feeding difficulty, airway compromise, and inability to achieve adequate oral closure.

Unlike peripheral excision, which may leave a bulky central tongue, or wedge resection techniques that primarily reduce length while inadequately addressing tongue width, the Oji technique provides a balanced reduction in length, width, and thickness. Compared with the classic keyhole technique, the Oji method avoids creating a postoperative "square-shaped" tongue, which has been associated with adverse functional and speech outcomes as the child grows.[8-10]

The Oji tongue-reduction technique is a central-wedge glossectomy originally described for the management of diffuse macroglossia, most commonly in syndromic conditions such as Beckwith–Wiedemann syndrome. The classical indication of this technique involves diffuse tongue hypertrophy rather than focal or cleft-related malformations.

In our case, the indication was not macroglossia but congenital trifid tongue. Therefore, the Oji technique was modified to address the unique trifid morphology.

The principal modification consisted of tailoring the central wedge excision to incorporate the entire central lobe of the trifid tongue, rather than performing a symmetric volume reduction for diffuse macroglossia. A midline wedge was designed to excise the redundant central segment responsible for the trifid configuration. The excision extended from the anterior free margin posteriorly to just anterior to the circumvallate papillae, consistent with Oji principles of limiting posterior extension. However, unlike the classical Oji reduction aimed at global volumetric reduction, the resection in this case was anatomically corrective and morphology-driven.[2-11]

In our patient, tongue reduction using the Oji technique resulted in satisfactory functional and cosmetic outcomes, with early restoration of feeding and preserved tongue mobility. Histopathology confirmed normal tongue tissue.

CONCLUSION

Whenever a patient presents with such an anomaly, proper history, detailed examination, and detailed investigations should be performed to rule out associated anomalies. Genetic counseling for the family can help individuals make informed decisions about their medical condition. Molecular genetic testing enables confirmation of this disease and provides counseling to family members.

ETHICAL DECLARATIONS

Informed Consent

Informed consent was obtained from the legal guardians of the pediatric patient described in this report. Where developmentally appropriate, assent was also sought from the child. The inclusion of vulnerable populations in this study adhered to national and international ethical guidelines. Extra care was taken to ensure voluntary participation, understanding, and protection of participant dignity and autonomy.

Peer Review Process

This report underwent external peer review.

Conflict of Interest

The authors declare no conflicts of interest.

Financial Disclosure

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Author Contributions

Concept: SI; Design: SI; Control: SI; Data Collection and/or Processing: AI; Analysis and/or Interpretation: AI; Literature Review: MAS; Article Writing: SI; Critical Review: All Authors.

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