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Case Report

Palatoglossal fusion with cleft palate and hypoplasia of cerebellar vermis

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ABSTRACT

A new-born male presented within 12 h of birth with respiratory distress. On examination and workup, he had palatoglossal fusion, cleft palate and hypoplasia of the cerebellar vermis. A 2.5 Fr endotracheal tube was inserted into the pharynx through nostril as a nasopharyngeal stent, following which his respiratory distress improved. Once child was optimised, then feeding was started by nasogastric tube and feeds were tolerated well. Elective tracheostomy and gastrostomy were done, followed by release of adhesions between the tongue and palate at a later stage. Review of literature suggests that palatoglossal fusion is uncommon and presents as an emergency. Mostly, these oral synechiae are associated with digital and/or cardiac anomaly. Other disorders associated with intra-oral synechiae include congenital alveolar synechiae, van der Woude syndrome, popliteal pterygium syndrome and oromandibular limb hypogenesis syndrome. The authors report a hitherto undescribed association of palatoglossal fusion with cleft palate and hypoplasia of the cerebellar vermis.

KEY WORDS

Cleft palate; intra-oral synechiae; nervous system anomaly; palatoglossal fusion

INTRODUCTION

Palatoglossal fusion is a rare anomaly with only a few documented cases in the literature.[1] It presents with respiratory distress soon after birth or later with feeding difficulty. Common associations with this condition include cardiac and digital anomalies.[2-5] Management includes emergency attention to the airway access, evaluation for the associated conditions and proper imaging before definitive surgical correction. Here, we discuss a case of palatoglossal fusion presenting with an aberrant associated anomaly, along with the challenges in its management.

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CASE REPORT

A 12-hour-old male baby presented in emergency with respiratory distress and had fusion anomaly of the oral cavity. He was full-term (birth weight = 3.5 kg), born via

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normal vaginal delivery to a 19-year-old primigravida at a primary health centre. Mother had received regular antenatal care. Antenatal sonograms were essentially normal. On physical examination, the child had obstructive breathing (respiratory rate = 68/min) with severe chest retractions. He maintained a saturation of 90% with oxygen supplementation but was irritable. Examination of the oral cavity revealed fusion of palate with dorsum of the tongue. Mucosa of the floor of the mouth was in continuity with the palate [Figure 1]. It was not possible to examine the oral cavity further. No other obvious congenital anomaly was noted. Baby was admitted to Neonatal Intensive Care Unit and respiratory distress was managed with immediate placement of nasopharyngeal airway (2.5 Fr endotracheal tube). Nutrition was established through nasogastric tube feeds on day 1, which the baby tolerated very well. Further evaluation was done after the baby was stabilised. Child underwent magnetic resonance imaging (MRI) of the head and neck region, two-dimensional echocardiography (2D ECHO) and ultrasonography (USG) of the abdomen for evaluation of the associated conditions. USG of the abdomen and 2D ECHO were normal. MRI was suggestive of cleft palate with tongue protruding into the nasal cavity and hypoplasia of inferior cerebellar vermis [Figure 2]. No chromosomal (cytogenetic) analysis was done as child was not showing features of any named syndrome or external dysmorphism warranting this.

The definitive corrective surgery was planned at 3 months of age in consultation with plastic surgery colleagues. In view of unstable airway and risk of aspiration, elective tracheostomy and gastrostomy were done on day 10 of life. Thereafter, the child was discharged on day 18 of life. A definitive release of the tongue from palate [Figure 3] was done in the second stage at 3 months of life (tongue adhered to palate in the lateral half; the central half was pushed up into the nasal cavity and nasopharynx through the cleft palate). Flexible laryngo-bronchoscopy revealed tracheomalacia proximal to tracheostomy. The child was discharged with gastrostomy feeds and tracheostomy in place. During the follow-up period, baby thrived well and was on tracheostomy care at home. At 6 months of age, tracheostomy decannulation was done successfully and oral feeds were started along with gastrostomy feeds. Child's weight and height (WHO growth charts) were within normal range; at the last follow-up (age = 8 months), the child was on complete oral feeds and free of tracheostomy/gastrostomy tubes and still awaiting the cleft palate repair.

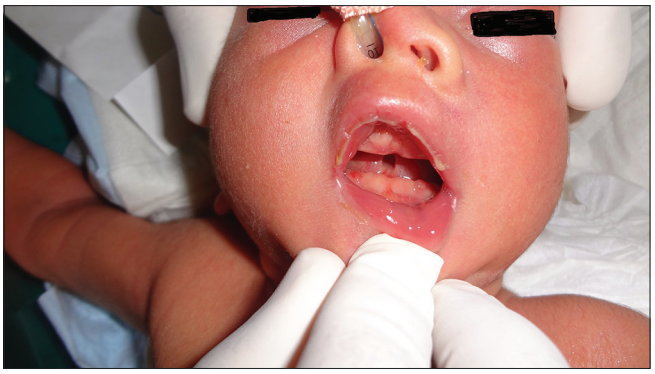

Figure 1: First presentation, note the midline tongue fusion with bifid frenulum and endotracheal tube as nasopharyngeal stent

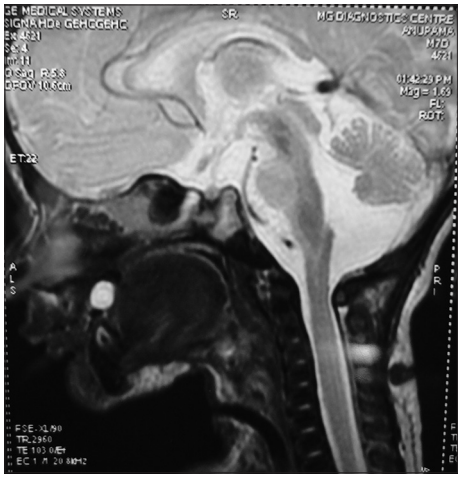

Figure 2: Magnetic resonance imaging; sagittal view showing tongue into the nasopharynx and hypoplasia of inferior cerebellar vermis

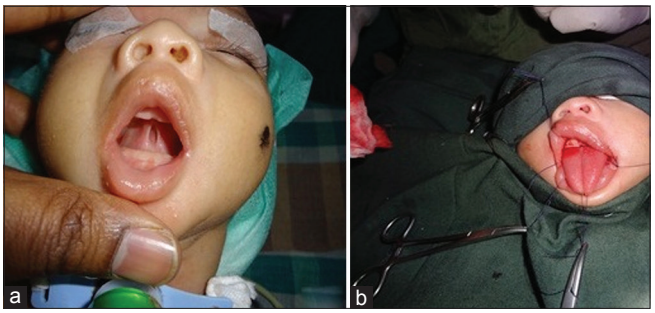

Figure 3: (a) Pre-operative image, tracheostomy *in situ* present. (b) Immediate post-operative image, complete release of tongue

DISCUSSION

Palatoglossal synechia is a rare anomaly and its cause is still unknown. During the 7th week of embryological development, normal oral development depends on the downward and forward movement of the tongue to allow for the palatal shelf fusion in the midline. The tongue protrudes through the oral cavity and as a result prevents fusion of the oral components. With the absence of tongue protrusion, prolonged contact between the

alveolar arches results in intra-oral synechiae.[1] Genetic, teratogenic or mechanical insult during this critical stage predisposes to abnormal fusion.[6] Goodacre and Wallace's experimental work on the aetiology suggested persistence of buccopharyngeal membrane, amniotic constriction bands in the region of developing the first branchial arch, environmental insults, drugs such as meclizine and large doses of Vitamin A as possible causes; however, they failed to prove any association for the same. Dawson *et al*. reviewed five cases of intra-oral synechiae and found no evidence of any familial tendency, history of drug and toxin exposure or consanguinity.[7] Intra-oral synechiae are commonly associated with a variety of syndromes and include oromandibular-limb hypogenesis spectrum (comprised sporadic disorders, varying degrees of extremity abnormalities, cardiac anomalies and microstomia).[3] Other associations mentioned in the literature that exhibit overlapping clinical features are Moebius syndrome, hypoglossia–hypodactylia syndrome, Hanhart syndrome, glossopalatine ankylosis syndrome, splenogonadal fusion syndrome and Charlie M. syndrome.[8,9]

The present case had no associated syndrome but had hypoplastic cerebellar vermis which was not reported earlier. The other important observation is management of acutely obstructed airway by simple nasopharyngeal airway, which is easy, effective and does not require special training. In our case, we had placed 2.5 Fr endotracheal tube into the pharynx through the nostril.

Review of literature suggests a role for adhesion release in emergency, under local anaesthesia. Release under local anaesthesia was not tried in this case as it was thought to be risk with unprotected airway. We suggest complete evaluation of the child before major surgical intervention; because depending on the severity, these patients may present formidable anaesthetic challenges. The conventional orotracheal intubation or the use of a laryngeal mask may not be possible due to the presence of the synechiae. Fibre-optic nasotracheal intubation may not be feasible due to technical difficulties of finding a bronchoscope small enough for an infant.[10]

CONCLUSION

Palatoglossal fusion is a rare anomaly with varied associations. Guiding principles for management are emergency airway care, nutrition and proper evaluation. Team approach and staged reconstruction result in satisfactory outcome.

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Conflicts of interest
There are no conflicts of interest.

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