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Tied to the Top: A Case Report on an Isolated Ankyloglossia Superior

ABSTRACT

Objective: To report a case of isolated ankyloglossia superior in a one-month-old boy.

Methods:

Design: Case Report
Setting: Tertiary Government Training Hospital
Patient: One

Results: A one-month-old boy with failure to thrive, feeding difficulties and a palatoglossal band that limited mouth opening and anterior posturing of the tongue was diagnosed to have non-syndromic ankyloglossia superior and underwent surgical transection of the fibrous ankylosis under intravenous sedation due to difficulty of insinuating an endotracheal tube orally. Direct latch breastfeeding was successfully adapted from the second to tenth postoperative day before they were lost to follow up.

Conclusion: Despite a complicated pediatric airway, coordinated surgical and anesthesia management successfully restored tongue function and mouth opening to allow effective feeding.

Keywords: *ankyloglossia superior; palatoglossal ankylosis; palatoglossal adhesion, tongue-palate fusion; oral synechia; tongue-tie.*

The term ‘ankyloglossia’ most often refers to a tongue-tie due to a congenitally short or thick lingual frenulum; more correctly termed ankyloglossia inferior.¹⁻² Ankyloglossia superior is the reverse anomaly – the tongue is congenitally adherent to the palatal mucosa instead.³⁻⁴ To the best of our knowledge, based on a search of HERDIN Plus, the ASEAN Citation Index (ACI), WHO Global Index Medicus (Western Pacific Region Index Medicus, WPRIM and Index Medicus of the South East Asia Region, IMSEAR), Directory of Open Access Journals (DOAJ), MEDLINE (PubMed) and Google Scholar using the search terms “ankyloglossia,” “ankyloglossia superior,” “palatoglossal ankylosis,” “palatoglossal adhesion,” “tongue-palate fusion,” “maxillary ankylosis,” and “oral synechia,” we found no published cases of isolated ankyloglossia superior in English. We report one such case.

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CASE REPORT

A 30-day-old boy was born at 40 weeks age of gestation to a 29-year-old G3P3 (3003) mother outside our institution. The prenatal period included one bout of urinary tract infection at the 8th month for which prescribed medications were not complied with but followed an uncomplicated course until birth. Soon after birth, an attachment of the tongue to the palate was noted with restricted tongue movement and breastfeeding difficulties. The baby was subsequently admitted to a hospital for 14 days for sepsis neonatorum and was advised to consult an otorhinolaryngologist upon discharge.

On examination, he weighed 2.4kg (weight-for-age at the 3rd percentile) with a height of 46cm (length-for-age below the 3rd percentile) which classified him with failure to thrive. An orogastric tube through which he was fed expressed breastmilk (because of breastfeeding difficulties due to poor suckling) was in place. Examination of the oral cavity showed restricted mouth opening with maximal interalveolar distance of 12mm, and a mucosal band extending from the dorsal surface of the oral tongue to the midline and left hard palate mucosa. An orogastric tube was positioned lateral to this palatoglossal band. (Figure 1 A, B) There was no cleft of the lip, maxillary and mandibular alveolar ridges, and no anomalies of the limbs or other regions. Aside from leukocytosis on a complete blood count and pneumonia on a chest radiograph, other diagnostics (including an electrocardiogram, echocardiogram and cranial ultrasound) were unremarkable. There were no known cases of ankyloglossia, cleft lip and palate, limb anomalies, cardiac anomalies nor any other congenital malformation in the family.

On his 33rd day of life, the baby underwent surgery to transect and excise the palatoglossal band under intravenous sedation because a transoral endotracheal tube could not be easily insinuated. The surgery commenced with ligation of the palatoglossal band at its lingual end, and then the palatal end was dissected away from the palatal mucosa using electrocautery. The palatoglossal band was noted to be fibrous tissue, while examination of the overlying bony palate ruled out a cleft palate. There was minimal blood loss and the patient tolerated the procedure well. A 7-day course of Amoxicillin syrup was given and breastmilk feeding via orogastric tube was resumed.

Postoperatively, tongue range of motion restoration and mouth opening were immediately apparent. The maximal interalveolar distance post-transection was 25mm. (Figure 2 A-C) With good suckling activity and no excessive bleeding or airway compromise observed, the orogastric tube was removed and successful feeding was accomplished by direct latch on the second postoperative day. The mother and child were discharged home on the third day. Although they missed their day 7 follow-up schedule, the patient’s mother reported good

breastfeeding when telephoned on day 10. They were lost to follow up thereafter.

DISCUSSION

Ankyloglossia inferior or mandibular ankyloglossia is a common isolated malformation in infants characterized by gross restriction in tongue protrusion or elevation, or may be seen as a notched or heart-shaped tongue as it is tethered by the short frenulum. It has a prevalence of 4.2-10.7% with 21% to 50% of cases report a family history of tongue-tie.⁵⁻⁶ The most widely accepted theory of its pathogenesis is an incomplete cellular degeneration of tissues between the floor of mouth and the tongue.⁷

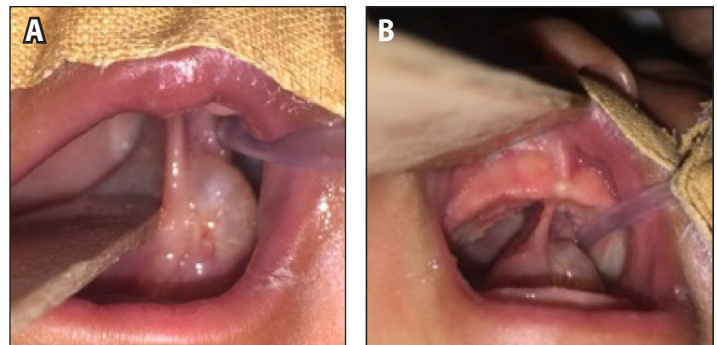


Figure 1. Palatoglossal band seen in the patient with ankyloglossia superior: **A.** palatoglossal band extending from the tongue tip to the midline and left hard palate mucosa; and **B.** orogastric tube in place lateral to the adhesion.

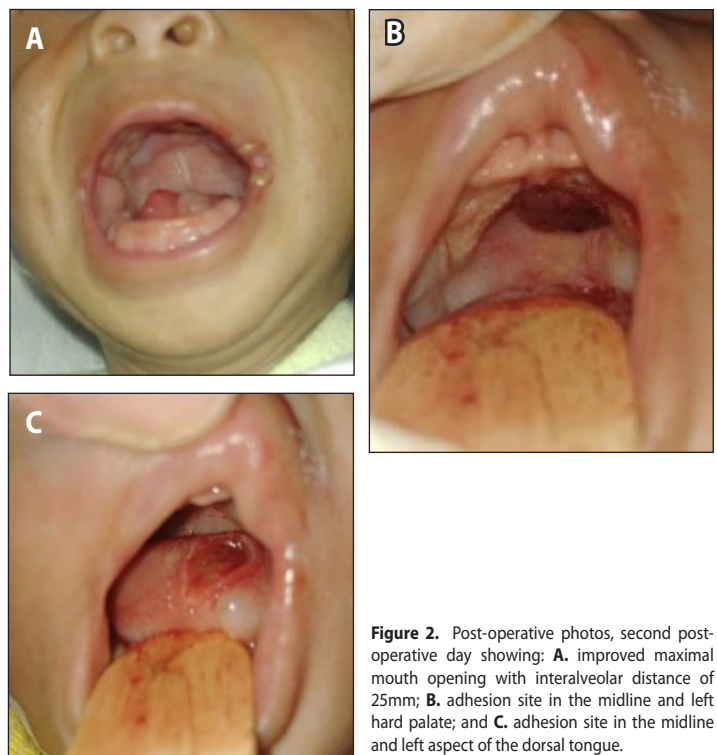


Figure 2. Post-operative photos, second post-operative day showing: **A.** improved maximal mouth opening with interalveolar distance of 25mm; **B.** adhesion site in the midline and left hard palate; and **C.** adhesion site in the midline and left aspect of the dorsal tongue.



Ankyloglossia superior, on the other hand, refers to a tongue tied to the palatal mucosa.^{3,4} It is often accompanied by various other anomalies, and is classified an ankyloglossia superior syndrome when a fibrous or osseous connection between the tip of the tongue and the hard palate occurs with a cleft palate, gastrointestinal malformations, and deformed limbs. In a retrospective review of 4000 cases in a facial cleft deformity clinic over 30 years, there were only 6 cases of a palatoglossal synechia – 4 had cleft palate lateral synechia syndrome or orofacial digital syndrome, and only 2 were non-syndromic.⁸ Illera and Kramer described the first cases of ankyloglossia superior syndrome in 1887 and 1911, respectively, according to Gima,⁹ but documented cases are few and far in between.

Ankyloglossia superior is rarely reported, and when it is, the literature documents a palatoglossal ankylosis occurring with various combinations of malformations. One patient had concomitant cleft palate,¹⁰ another had extensive jejunal and ileal atresia leading a fatal course.¹¹ In a number of cases, the anomalies appear in the maxillofacial region and limbs. Of 17 cases reviewed by Spivack and Bennett,¹² several had micrognathia, microglossia, cleft palate, absent teeth and limb anomalies. Weiker and Sieg in 2012 reported a case of ankyloglossia superior syndrome in a five-year-old Filipino male exhibiting a palatoglossal ankylosis along with limb aplasia and dysplasia in the upper and lower extremity, and a claw-hand deformity.² In our present case, no other congenital malformations occurred in conjunction with the palatoglossal band, making it a rare isolated ankyloglossia superior.

With the scarcity of cases, the etiology of ankyloglossia superior – syndromic or non-syndromic – has yet to be established. The reported cases have been observed to occur sporadically, hence a hereditary factor is unlikely. Gartlan posits that the etiologic event occurs in utero between the eighth and 11th week of gestation, the period of development for the oromandibular region.¹³ On the other hand, Lekkas and Bruaset¹⁰ and Wilson *et al.*¹⁴ suggest that injury or de-epithelization of the tongue tip during palatal fusion in utero results in this adhesion.

Due to the lack of cases, the complications and sequelae for ankyloglossia superior is still an unstudied area. Potential sequelae for ankyloglossia inferior may then be assumed for ankyloglossia superior due to their similar tongue restricting qualities. One of these is poor articulation and speech development, although ankyloglossia inferior cases have developed normal speech without intervention.⁶ Another postulated sequela for ankyloglossia is poor feeding. Poor tongue protrusion and mobility creates an ineffective seal while latching to a nipple, then affecting the ability to suck, swallow and breastfeed overall. This was demonstrated in our present case where direct latch breastfeeding was not tolerated resulting in failure to thrive.

Also notable in our case was the challenge for airway access encountered in the preoperative planning stage. The surgical and anesthesia teams learned that with an intraoral band of tissue restricting tongue mobility and mouth opening, ankyloglossia superior complicated pediatric airway access as endotracheal

intubation was impeded. This limitation posed the possibility of more difficulties in the event of intractable bleeding during surgery. The surgical and anesthesia teams opted for intravenous anesthesia with immediate endotracheal intubation planned following ligation of the palatoglossal band, and creation of a surgical airway was considered in anticipation of complications that necessitate securing the airway. Coordination between the surgeon and anesthesiologist was vital in the surgical management of this case. Once adequate sedation was achieved, ligation of the palatoglossal band with a silk ligature and use of electrocautery were utilized to minimize bleeding. While this surgery incurred only minimal blood loss (dispensing with the need for a surgical airway), preparing for a surgical airway should be considered in similar cases. Other considerations included assuming suboptimal pulmonary function that could result in unwanted anesthetic effects and possible cardiopulmonary problems from undetected anatomical anomalies.

Indeed, the rarity of this condition and varieties in its presentation and associated malformations are challenging. Our experience highlights the importance of close coordination between the surgical and anesthesia teams. Despite a complicated pediatric airway, coordinated surgical and anesthesia management successfully restored tongue function and mouth opening to allow effective feeding. While cases occur few and far apart, we hope our experience contributes to the profile of the ankyloglossia superior syndrome, particularly isolated ankyloglossia superior.

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