Ankyloglossia Superior Associated With Moebius Syndrome: A Case Report

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Ankyloglossia superior is an exceedingly rare congenital condition that consists of a connection between the tongue and hard palate. This abnormality is considered part of the ankyloglossia superior syndrome when found with other malformations such as limb deformities, gastrointestinal malformation, and cleft palate. Ankyloglossia superior can also be associated with other known syndromes. We have presented the case of a female infant born with multiple malformations, including partial agenesis of the feet and hands, micrognathia, a lack of expression of the facial muscles, convergent strabismus, mouth opening limitation, and tongue–palate adhesion. The patient's presenting diagnosis was ankyloglossia superior associated with Moebius syndrome. Computed tomography revealed the extent of the ankyloglossia superior and the loss of integrity of the palatal shelf. Surgical release of the ankyloglossia superior was performed with the patient under general anesthesia. Multiple management challenges were encountered postoperatively. To the best of our knowledge, ankyloglossia superior presenting in conjunction with Moebius syndrome had not been formally described in a case report.

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Tongue hypomobility will typically be due to the anterior insertion of the lingual frenulum to the floor of mouth. However, atypical etiologies of tongue hypomobility exist. An extremely rare cause is ankyloglossia superior, also known as glossopalatine ankylosis, which is a congenital anomaly characterized by the attachment of the lingual frenulum to the palate, instead of its usual insertion to the anterior part of the floor of mouth.

When ankyloglossia superior is accompanied by other malformations, such as limb deformities, gastrointestinal malformation, and cleft palate, it is referred to as ankyloglossia superior syndrome.[1-4] Investigators have reported an association of ankyloglossia superior with other syndromes (eg, Charlie M syndrome, aglossia-adactylia, Hanhart syndrome).[1-6] However, to the best of our knowledge, no case report has described its presentation in

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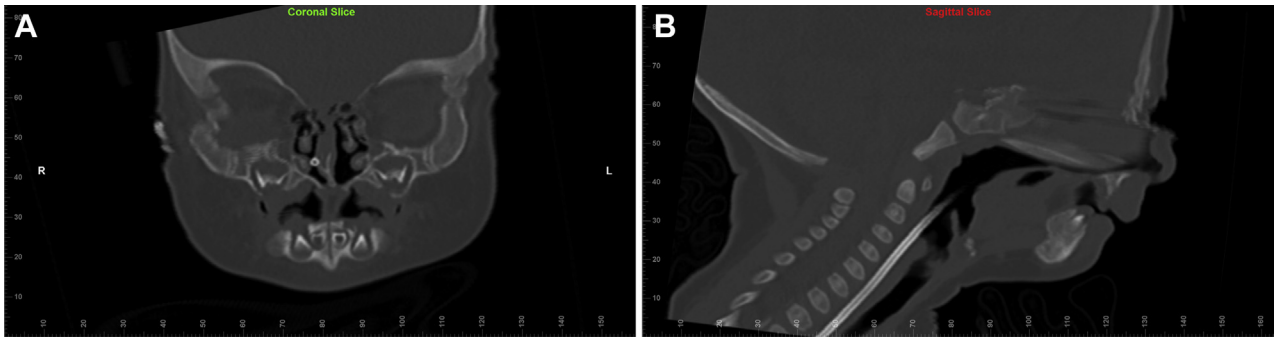

FIGURE 1. Computed tomography scans showing the extent of tongue adherence to the palate and loss of continuity of the palatal shelf. *A,* coronal view; and *B,* sagittal view.

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association with Moebius syndrome. Moebius syndrome is a rare congenital neurological disorder characterized by palsy of cranial nerves VI and VII.[7]

The present report illustrates the management challenges encountered in a rare case of ankyloglossia superior associated with Moebius syndrome. Our report was prepared in accordance with the CARE guidelines (available at: http://www.care-statement.org). The patient's legal guardian provided written informed consent authorizing publication of the treatment images for scientific purposes. The study was performed in accordance with the Declaration of Helsinki, and the institutional review board of the University of Cuiabá provided ethical approval as a part of a greater project titled "the use of tridimensional technologies in the management of facial deformities" (approval no. 2.421.462).

Case Report

The present patient was a female infant born via cesarean section at 8 months' gestation, with a birth weight of 2.840 kg. The mother of the infant had reported 3 previous pregnancies, resulting in 2 deliveries without malformations and 1 miscarriage. Her social history was negative for smoking or drinking habits. However, she did report exposure to unspecified agrotoxic agents during the gestational period. She had developed pregnancy-induced hypertension, which had required pharmacologic control. The father of the child reported controlled systemic hypertension and a family history of cleft lip and palate.

Multiple malformations were observed after the patient's delivery. These anomalies included partial agenesis of the feet and hands, micrognathia, a lack of expression of the facial muscles, and convergent strabismus. The intraoral assessment was limited secondary to her restricted mouth opening (<10 mm). However, it was possible to visualize a small and posteriorly displaced tongue that was adherent to the hard palate by a band of fibrous tissue. Instinctive suckling movements were not noted. Within the limitations of the examination, the tongue did not appear bifid nor was a cleft palate visible.

From birth, the patient had required a nasoenteral feeding tube owing to the inability to perform the suction required for feeding and to prevent repetitive bronchoaspiration. At 5 months of age, the patient was referred to the County Hospital of Cuiabá for evaluation by the maxillofacial and pediatric surgical teams. On the basis of the clinical features, the diagnosis of ankyloglossia superior associated with Moebius syndrome was established. A computed tomography (CT) scan was obtained to assess the extent of the tongue–palate adhesion and to evaluate the integrity of the hard palate. The combined decision was to release the ankyloglossia superior with the patient under general anesthesia and to provide a thorough assessment of the tongue and any potential palatal defect, because the CT assessment displayed signs of loss of continuity of the palatal shelf (Fig 1).

Because of our concern for tongue collapse after its release, the surgical intervention began with a tracheostomy to establish a definitive airway. The tongue adhesion was then removed from the hard palate via a dorsal lingual frenectomy (Fig 2). This resulted in mild improvement of the mouth opening, and the absence of an oral–nasal communication was confirmed. However, atypical swallowing remained after the surgery, along with a continued risk of bronchoaspiration. Even after a multidisciplinary effort to improve the swallowing patterns, no significant improvements were observed, and the patient underwent gastrostomy. However, the patient died before reaching 1 year of age of a reported cardiac arrest.

Discussion

Ankyloglossia superior is a congenital condition characterized by adhesions of the dorsal aspect of

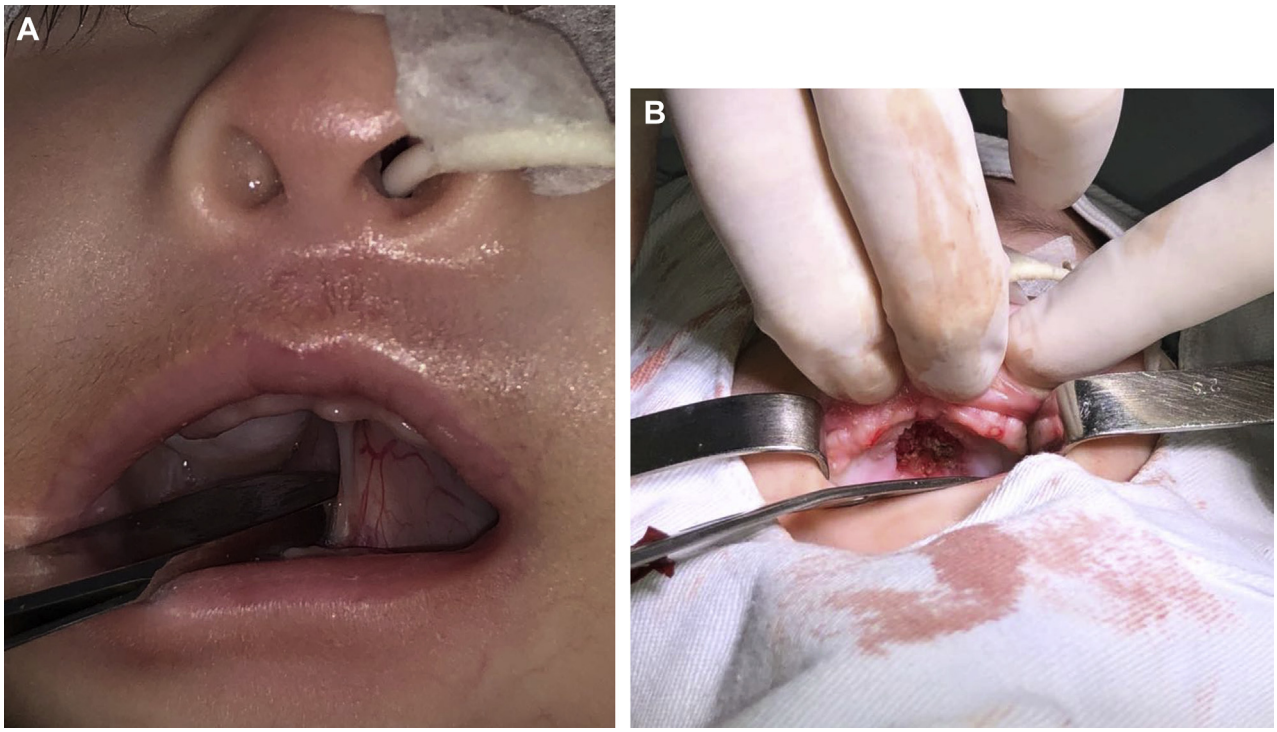

FIGURE 2. Intraoperative features showing *A*, initial presentation of ankyloglossia superior and *B*, final aspect of the palate.

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the tongue to the palatal mucosa, which can adversely affect breastfeeding and swallowing, cause serious respiratory dysfunction, and interfere with maxillofacial development. It has been suggested that this malformation will occur at 6 to 11 weeks of intrauterine life.[1,6] Ankyloglossia superior is an extremely rare condition, with few cases reported and no identified etiology. Both genetic and environmental factors have been suggested, including possible associations with drug intoxication or infection during pregnancy or with amniotic membrane disease.[1-3,5,6] Although speculative, the history of exposure to agrotoxic agents during the gestational period could have been an etiological factor in our patient.

The diagnosis of ankyloglossia superior is relatively simple when an adequate examination is possible. Although older children can easily display symptoms such as decreased tongue mobility and difficulty with phonation, the diagnosis in infants can be challenging in patients with a severely limited mouth opening.[1,8] The management of ankyloglossia superior will typically include removing the tongue–palate adhesion and any other indicated surgical corrections.[1,3] The timing of surgical intervention has varied. Some of the reported cases have described surgical removal of the adhesion with the patient under local anesthesia for those older than 1 year of age,[1] and others have treated younger patients without complications.[2,5,6,9] Most procedures were performed with the patient under local anesthesia,[1,3,5,6,9] with only a few treated under general anesthesia.[2,8] In the present patient, who had been treated under general anesthesia, special care was taken to secure the airway owing to the possibility of tongue collapse and airway obstruction after release of the adhesion. Moreover, the CT finding of loss of continuity of the palatal shelf suggested the possibility of a residual oronasal communication. Thus, a careful incision that would leave sufficient soft tissue coverage to the palate was paramount. To the best of our knowledge, no previous study has reported an ankyloglossia superior case with CT assessment in the initial evaluation.

In the present patient, the concomitant presence of convergent strabismus, malformations of the upper and lower limbs, and the absence of movement of the muscles for facial expression was consistent with the diagnosis of a coexisting Moebius syndrome. Although studies describing an association of ankyloglossia superior with Moebius syndrome have been reported,[2,3,8] no specific case report has been described to our knowledge.[10] The presence of Moebius syndrome in association with the ankyloglossia superior complicated the management of our patient because the mouth opening limitation (which was believed would persist even after tongue–palate adhesion removal) was severe enough to warrant tracheostomy.

In conclusion, although isolated ankyloglossia superior might not pose significant challenges in the treatment of the patients, syndromic associations can significantly increase the complexity.[1,5] It is important to consider some factors that could negatively affect the outcomes such as anatomic defects, the presence of other malformations, health service access, and the performance of late repairs.[1] Although the mouth opening might not improve immediately, it has been shown to increase in the long-term.[2,8] Adverse outcomes occurring after intervention have seemed to correspond with coexisting malformations. For our patient, death was likely the unfortunate result of an unknown cardiac event.[4,11]

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