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Subglossopalatal Membrane With Associated Cleft Palate, Cardiovascular, and Neurologic Anomalies

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Abstract: Subglossopalatal membrane (or subglossopalatal synechia) is a rare clinical entity that can lead to respiratory distress and feeding difficulty due to oral obstruction. Here, the authors present a case of subglossopalatal membrane with associated cleft palate and cardiovascular and neurologic anomalies that was treated with surgical excision and lip-tongue adhesion. Etiology of these membranes is believed to be intrauterine fetal insult. Membranes should be treated with excision, whereas taking care to ensure patency of the airway. Presence of a subglossopalatal membrane should prompt thorough examination for additional congenital anomalies.

Key Words: Buccopharyngeal, cleft palate, lip-tongue adhesion, membrane, subglossopalatal membrane, subglossopalatal synechia

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Subglossopalatal membrane (SGPM) (or subglossopalatal synechia) is a rare clinical entity that can lead to respiratory distress and feeding difficulty due to oral obstruction. It was first described in the literature by Nakajima et al.[1] The etiology is unknown, and whereas it has been categorized as a buccopharyngeal membrane (BPM) remnant, Zalzal et al[2] suggests that it may be due to an abnormal fusion event in the 6th or 7th week of gestation. Eight cases in the literature describe subglossopalatal membranes,[1–8] with 5 of them being associated with additional congenital anomalies (Supplementary Digital Content, Table 1, http://links.lww.com/SCS/D433). Here, we present a case of subglossopalatal membrane with associated anomalies that was treated with surgical excision.

Clinical Presentation

A 1-day old female infant was transferred from an outside hospital for intraventricular hemorrhage, hydrocephalus, respiratory distress, hypoglycemia, and possible ankyloglossia. Delivery was via Cesarean section due to preeclampsia at 33 weeks; brain anomalies had been noted prenatally as she had experienced selective growth restriction in a monochorionic-diamniotic twin pregnancy. The other twin was healthy. By day 1, the patient was hemodynamically stable, breathing comfortably on room air, euglycemic, and receiving total parental nutrition. Physical exam noted a thin midline fibrous band extending from the ventral surface of the tongue and floor of the mouth to the incisive foramen (Fig. 1). It was approximately 0.5 cm thick. Plastic surgery and otolaryngology were consulted. There was no acute airway obstruction. Although there appeared to be micrognenia, the alveolar relationship was a very mild class II with a deep bite. Associated anomalies were cleft hard and soft palate, hydrocephalus, and mild rocker-bottom feet. There were no apparent abnormalities of the ears, eyes, lips, or spine.

Investigations

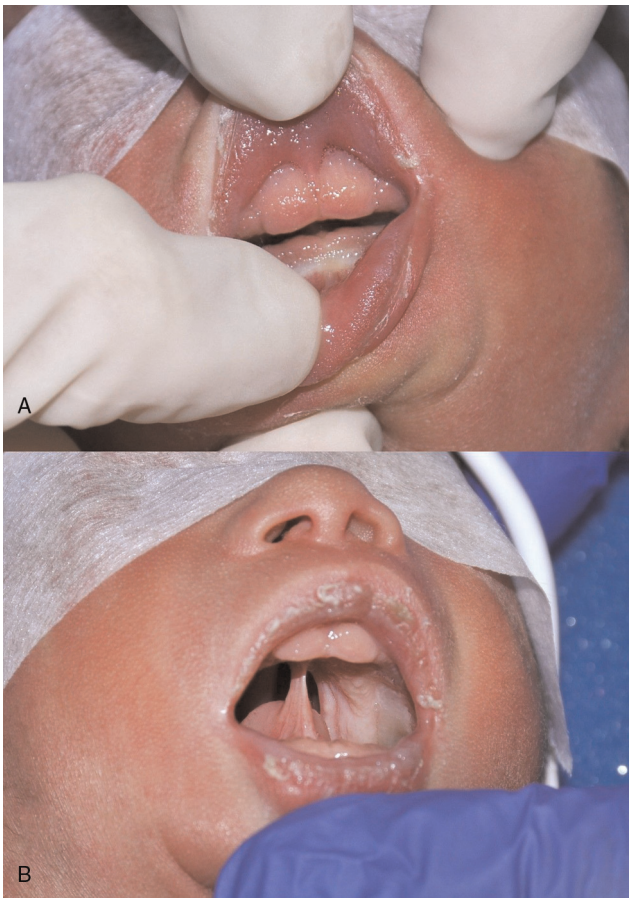

FIGURE 1. (A) The patient at rest demonstrating mild Class II alveolar relationship with deep bite, (B) with jaw opening, the subglossopalatal membrane extends from the floor of the mouth to the incisive foramen, with the tongue displaced posteriorly.

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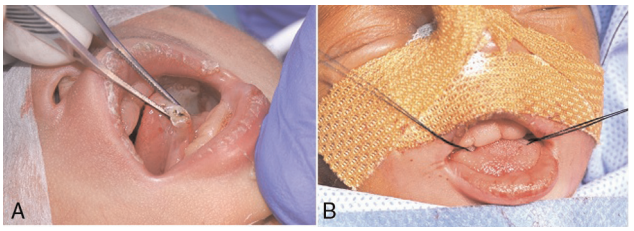

FIGURE 2. (A) Immediately after membrane excision. (B) Status-post lip-tongue adhesion with 2 looped tongue sutures placed for safety.

Ultrasound of the head and spinal cord showed callosal dysgenesis, an enlarged right lateral ventricle with concern for open lip schizencephaly, and a spinal cord terminating at L4 likely representing underlying tethered cord. Magnetic resonance imaging of the brain and spine confirmed agenesis of the corpus callosum with a large interhemispheric cyst, cerebellar hypoplasia with retrocerebellar cyst consistent with Dandy-Walker Syndrome, and tethered cord with conus terminating at the level of L3. Echocardiogram found secundum atrial septal defect, high muscular bidirectional ventriculoseptal defect, and large bidirectional patent ductus arteriosus. Ultrasound of the abdomen including the bladder showed no abnormalities. Chromosomal microarray failed to identify any clinically significant abnormalities, and cytomegalovirus, rubella, and toxoplasma Immunoglobulin M assays were all negative.

Treatment

Because of concerns for feeding and the inability to intubate if respiratory decompensation were to develop, the patient was brought to the operating room by plastic surgery and otolaryngology on day-of-life 2 for nasal endoscopy, direct laryngobronchoscopy, and possible ligation of subglossopalatal membrane. On initial nasal endoscopy, there was airway obstruction in the supine position with the jaw unsupported. A jaw thrust maneuver effectively opened the airway. In the lateral decubitus positions, there was no airway obstruction. Direct laryngobronchoscopy showed mild tracheomalacia and was otherwise normal. Next, with nasal endoscopic monitoring, the membrane was divided (Fig. 2A). Nasal endoscopy showed glossoptosis causing airway obstruction. Lip-tongue adhesion was then performed, and looped tongue stitches were placed as a safety maneuver should dehiscence occur (Fig. 2B). The patient remained nasally intubated and sedated following the procedure.

Outcome and Follow-Up

On postoperative day 7 the patient was taken to the operating room for extubation. Post-extubation nasal endoscopy showed the airway was open with the patient on her side without jaw support, and partially open in the supine position with jaw thrust. There was inspiratory collapse in both positions. A feeding tube was placed on postoperative day 13. On postoperative day 25, the patient began receiving syringe feeds per os. Postoperative computed tomography scan of the head demonstrated an open airway at the nasopharyngeal level in addition to cleft palate and micrognathia (Fig. 3)

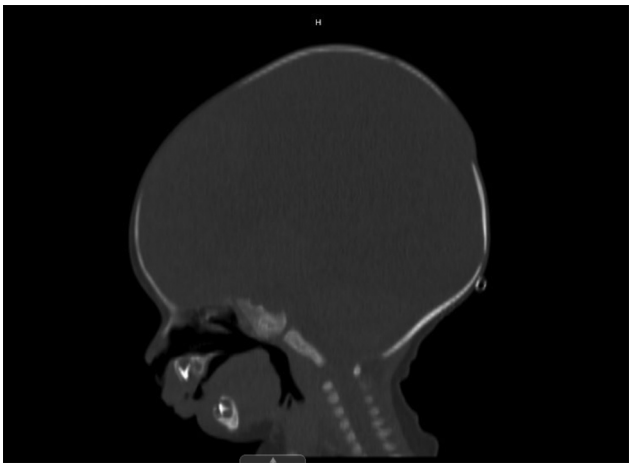

FIGURE 3. CT head demonstrating a patent airway after lip-tongue adhesion.

DISCUSSION

Subglossopalatal membrane is a rare congenital oral synechia. It has been described in the literature only a handful of times[1–8] since Nakajima et al[1] first defined it in 1979 (Supplementary Digital Content, Table 1, http://links.lww.com/SCS/D433). Delineation between SGPM and BPM remnants based on embryologic anatomy and associated anomalies have been proposed.[2,9]

In the 3rd week of development, the stomodeum deepens and the BPM slopes superoposteriorly, and in the 4th week, facial growth causes it to break down. The theoretical attachments of the BPM are the soft palate, the anterior tonsillar pillars, and the junction of the anterior two-thirds and posterior one-third of the tongue.[9] So, a BPM remnant is unlikely to be attached to the floor of the mouth as in the present case and those in Supplementary Digital Content, Table 1, http://links.lww.com/SCS/D433.

Zalzal et al[2] explains that a SGPM is more likely an abnormal fusion event rather than an embryologic remnant. In the 7th week of normal development, the palatine shelves are positioned vertically. Skeletal muscle growth of the tongue combined with relative mandibular hypoplasia causes the tongue to wedge itself between the palatine shelves.[10] In the 9th week, the mandible grows down and lengthens, causing the tongue to retract inferiorly and allowing the palatine shelves to elevate and fuse at midline.[10] During week 6 or 7, aberrant fusion could take place between the inferiorly displaced palatine shelves and the floor of the mouth. If these attachments are not obliterated with vertical growth of the face, the result is a subglossopalatal membrane.[2] This membrane can keep the tongue elevated between the palatine shelves, increasing likelihood of cleft palate, as seen in our case and most of those previously mentioned. Micrognathia could also contribute to the development of a cleft palate by failing to draw the tongue inferior and anterior.[11]

As suggested by Gartlan et al,[9] the theory of subglossopalatal membrane (as opposed to BPM remnant) is strengthened by the presence of additional congenital anomalies, which may suggest a more global genetic or environmental insult occurring in the 6th or 7th week. In the present case, SGPM was associated with cleft palate, micrognathia, callosal dysgenesis, schizencephaly, possible tethered cord, atrial septal defect, ventricular septal defect, patent ductus arteriosus, and rocker bottom feet. It was also known that the patient experienced growth restriction secondary to monochorionic diamniotic twinning. This case differs from others not only in its constellation of anomalies, but also in the attachment sites: the superior attachment of the SGPM was at the incisive foramen. Alternative to the mechanism proposed by Zalzal et al, it is possible that the cleft palate was the antecedent abnormality, maintaining the tongue in a retroverted position with its underside in close proximity

to the incisive foramen, thereby promoting synechia formation.[2] The synechia in the present case is certainly more anterior than where a BPM remnant would be expected, and it is farther anterior than all aforementioned SGPM cases.

Treatment of subglossopalatal membrane involves excision of the membrane.[1,2] This may be required on an emergent basis in the event of airway obstruction. As suggested by Pandey et al,[5] outcomes may be improved by first stabilizing the patient because neonates are nasal breathers, and a non-obstructing membrane may not be the cause of respiratory distress. In our case, excision took place on day-of-life 2, when the patient was stable. Additionally, a lip-tongue adhesion was required once membrane excision caused glossoptosis and airway obstruction; it is important to be prepared for this.

CONCLUSIONS

Subglossopalatal membranes, or subglossopalatal synechiae, are rare congenital oral synechiae. Although their etiology is unknown, they are believed to be a result of intrauterine fetal insult. These membranes should be treated with surgical excision under controlled conditions, with capability to perform lip-tongue adhesion or other necessary maneuvers to ensure airway patency. The presence of a subglossopalatal membrane should prompt thorough examination for additional congenital anomalies.

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