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Congenital Maxillomandibular Synechia with Multiple Malformations in a Very-Low-Birth-Weight Infant: A Case Report

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Abstract

Congenital maxillomandibular synechia is a rare malformation that is characterized by a fusion of the maxilla and mandible. The fusion is fibrous or bony and prevents mouth opening, which causes difficulties in feeding and occasionally in breathing. Although extremely rare, neonatologists must understand the disease because it can be fatal and require emergency treatment after birth. We report the case of a very-low-birth-weight (VLBW) infant with congenital maxillomandibular synechia and other malformations, including cleft palate, syndactyly, and cryptorchidism. The patient presented with extremely limited mouth opening, and endotracheal intubation seemed impossible; fortunately, the patient did not have respiratory distress syndrome. The patient underwent surgical release of the fibrous bands on days 10 and 17, and good mouth opening was achieved. The patient was able to consume breast milk orally and was discharged home at a corrected gestational age of 1 month without recurrence of difficulty in mouth opening or any sequelae. This is the first reported case of a VLBW infant with congenital maxillomandibular synechia who required more complicated management of feeding, surgical intervention, and anesthesia.

Keywords

- congenital maxillomandibular synechia
- fibrous or bony fusion
- difficulty in feeding and breathing
- popliteal pterygium syndrome

A boy was born at 32 weeks of gestation to a gravida 2 para 1 mother via emergency cesarean section due to fetal growth restriction and nonreassuring fetal heart tracings. There was no significant family history or consanguinity. The patient's birth weight was 1,229 g (standard deviation: −2.6), and the Apgar scores were 4 and 9 at 1 and 5 minutes, respectively.

At birth, the patient had extremely limited mouth opening, with an interalveolar space of approximately 2 mm (►**Fig. 1**). However, during resuscitation, we were able to provide positive pressure ventilation through a face mask, and the patient's respiration stabilized. After admission to the neonatal intensive care unit, he underwent nasal continuous positive airway pressure. Fortunately, he did not have respiratory distress syndrome and did not need immediate endotracheal intubation, which seemed technically impossible due to difficulty in mouth opening.

On physical examination, the patient had several facial deformities, including a prominent occiput, bilateral cryptotia with ear canal stenoses, cutaneous syndactyly affecting both hands and toes, and bilateral cryptorchidism (►**Figs. 2**, **3**).

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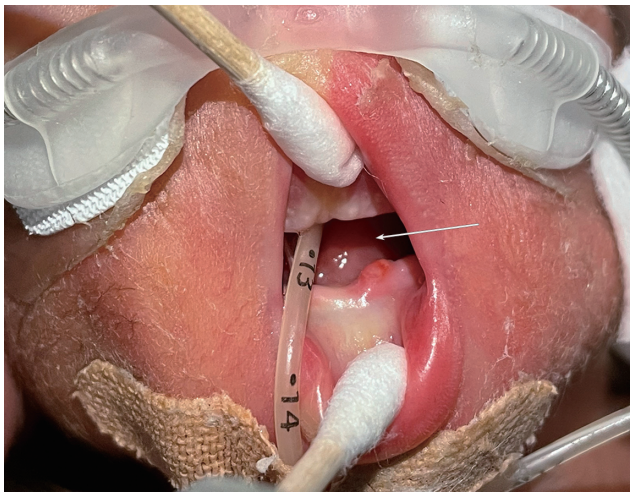

Fig. 1 The patient's mouth maximally opened with cotton swabs. Interalveolar space (*arrow*).

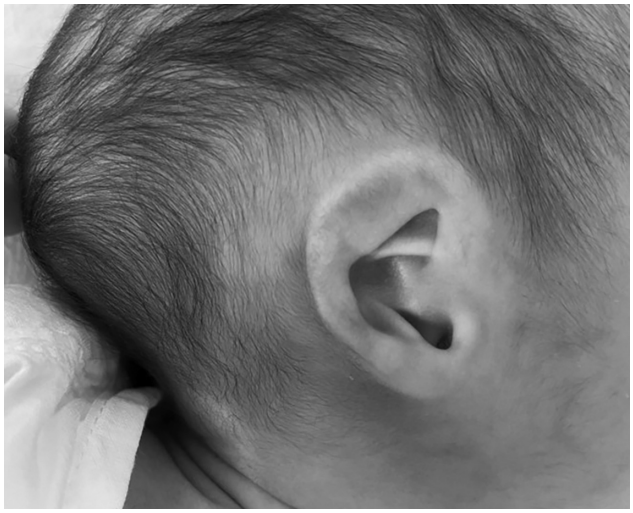

Fig. 2 Cryptotia with ear canal stenosis and prominent occiput.

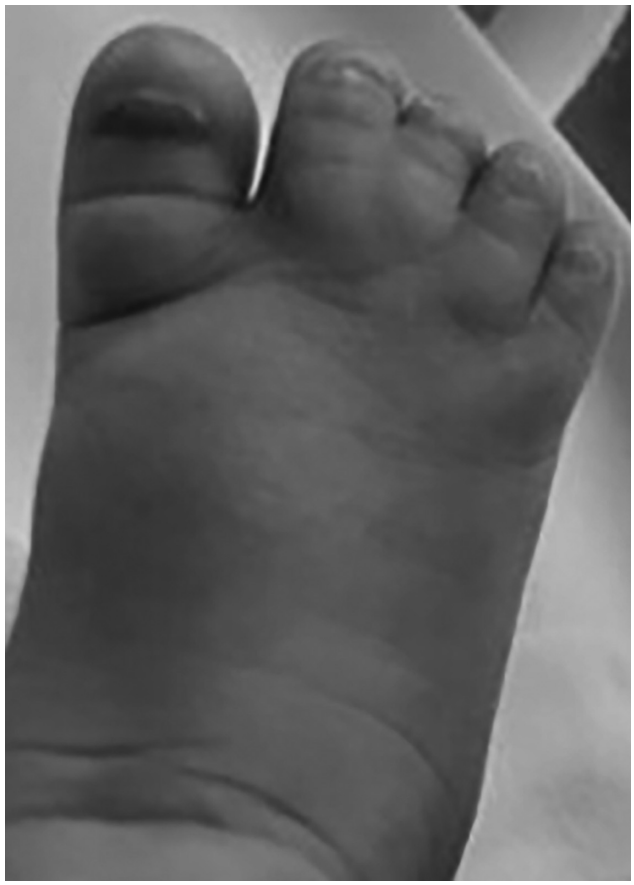

Fig. 3 Syndactyly in the right foot.

Radiography and echocardiography of the head, heart, and abdomen did not show any congenital malformations, except for patent ductus arteriosus.

We observed the oral cavity through a narrow space approximately 48 hours after birth when mouth opening slightly improved and found three alveolar fibrous bands (►**Fig. 4A, B**). Although the extent of the adhesions could not be assessed completely, they apparently interfered with

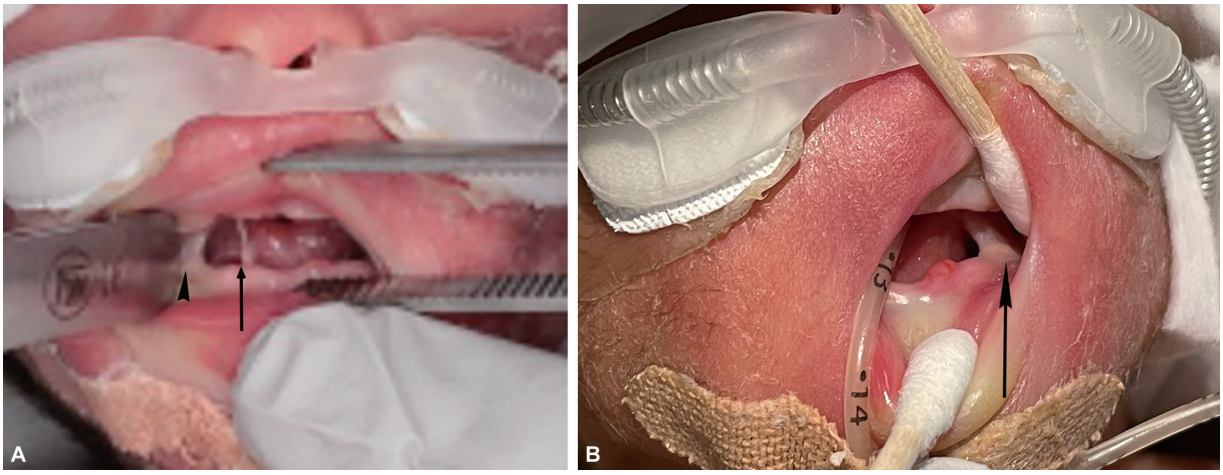

Fig. 4 Three alveolar fibrous bands. (**A**) A very thin band between the hard palate and the floor of the mouth (*arrow*). A thick band between alveolar crests on the right side (*arrowhead*). (**B**) A thick band between the lower crest and hard palate on the left side (*arrow*).

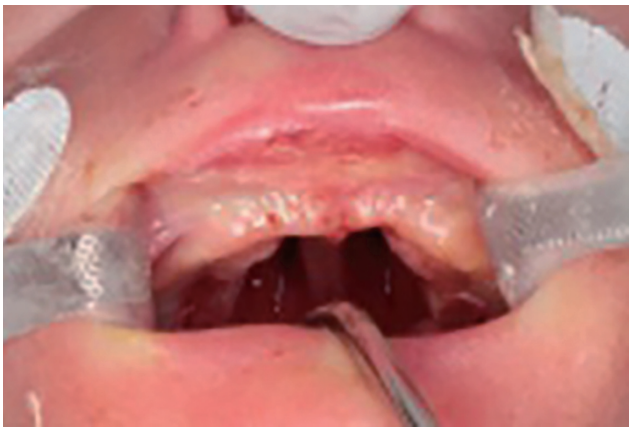

Fig. 5 Cleft palate.

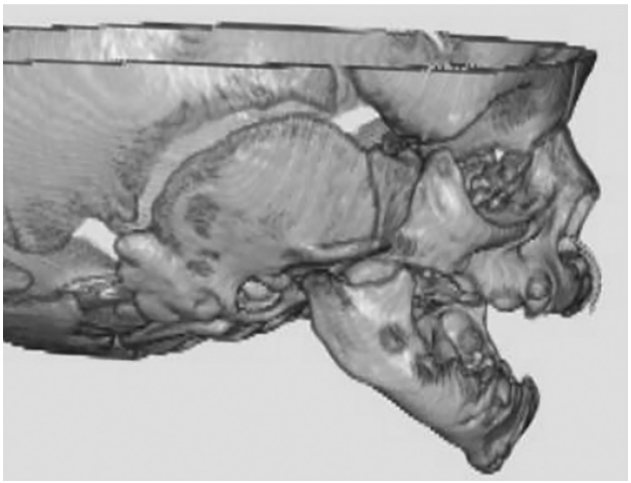

Fig. 7 Computed tomography images showing no bony fusion.

mouth opening, and the infant was diagnosed with congenital maxillomandibular synechia.

We started enteral feeding via a nasogastric tube, although we were concerned about potential emesis and pulmonary aspiration because vomiting from the mouth seemed difficult for the patient, and he could easily aspirate breast milk. However, enteral feeding progressed without any problems, and he tolerated full enteral feeding.

On day 10, the patient underwent surgery, which was performed by an oral surgeon in our facility. We had no choice but to use local anesthesia because airway management, such as orotracheal or nasotracheal intubation, which is inevitable for general anesthesia, was not possible. Following the injection of the anesthetic, the bands were resected using surgical scissors, although the band on the left side was left partly unreleased because of its extensibility. After the operation, mouth opening improved to 10 mm, and a cleft palate was noted for the first time (►**Fig. 5**).

One week later, additional intervention to resect the remnant of the left band was performed under local anesthesia because limited mouth opening persisted. The surgeon used a bipolar electrosurgical knife instead of scissors because of its greater accessibility. After the band was successfully resected, mouth opening improved to 15 mm (►**Fig. 6A, B**). In case orotracheal intubation was needed, laryngoscopy could be performed with a laryngoscope blade.

A head computed tomography scan to check for associated synostosis between the mandible and maxilla or zygoma showed no bony fusion (►**Fig. 7**). The patient underwent physiotherapy (manual jaw-opening exercises), and there was no recurrence of limited mouth opening.

The rest of the course was uncomplicated considering the patient's very-low-birth-weight (VLBW) status. The patient did not have chronic lung disease. His patent ductus arteriosus spontaneously closed without treatment. Magnetic resonance imaging of the brain at corrected full-term age showed no periventricular leukomalacia or other abnormalities. On ophthalmologic examination, he showed no abnormality, including retinopathy of prematurity. He passed the automated auditory brainstem response (AABR) test on the right ear but failed on the left. The ABR test results showed an elevated V-wave threshold of 90 dB on the left ear.

From the corrected gestational age of 35 weeks, he was fed orally using a special nipple for patients with a cleft palate that is soft and has a crosscut with a squeezable bin to make it easier for the patients to draw milk into the mouth. He was able to consume breast milk orally and was discharged home at a corrected gestational age of 1 month, 45 weeks without any sequelae.

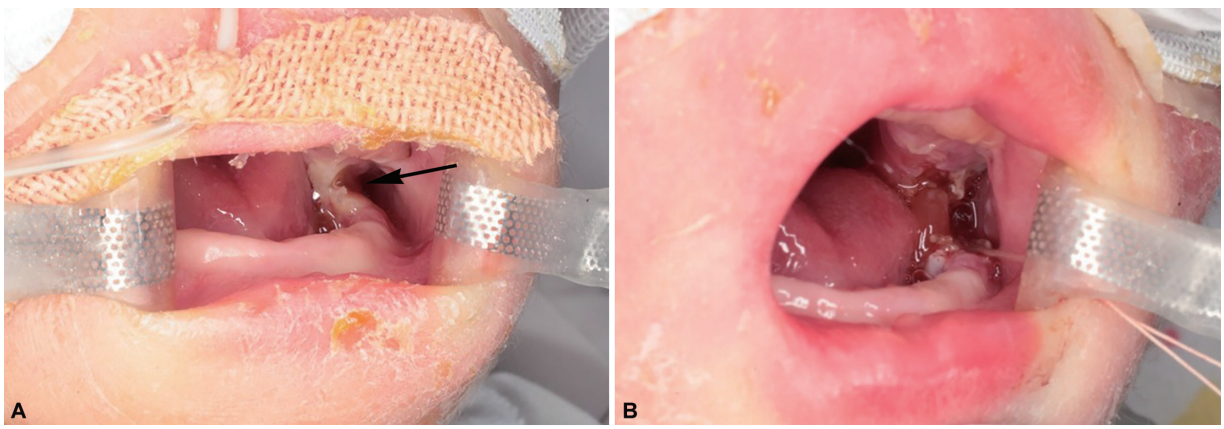

Fig. 6 (A) Remnant of the left band before the second operation (*arrow*). (B) After the second operation, the mouth opening improves to 15 mm.

Discussion

Congenital maxillomandibular synechia is a rare anomaly characterized by gingival mucosal fusion, bony fusion, or both. It limits mouth opening ability to varying extents, which can lead to difficulty in feeding and swallowing and can critically affect the airway.

The first case was published in 1907 by Kettner,[1] who described a bony-gingival adhesion of the upper and lower jaws with a cleft palate, macroglossia, and anomalous extremities. Since then, there have been sporadic case reports or case series and several comprehensive reviews.[2–4] Olusanya and Akadiri reviewed 153 (110 articles) patients between 1907 and 2017, and approximately half of the articles ($n = 52$, 47.3%) were from Asia,[3] which is where the infant in the present case was also born. To our knowledge, this is the first reported case of congenital maxillomandibular synechia in a VLBW infant.

Olusanya and Akadiri classify the disease into two types: fibrous fusion as type 1 and bony fusion as type 2. According to Olusanya and Akadiri, type 1 accounts for 53% of patients and type 2 for 47%. In a review article, Kumar et al evaluated 118 patients, of whom 62 (53%) had a bony fusion, 48 (34%) had a fibrous fusion, and 8 (13%) had a combined fusion.[4]

Several reviews have proposed additional classifications depending on the extent and site of fusion. The Olusanya classification system comprises three subtypes:

Subtype a: Anterior fusion that does not extend beyond the canine region
Subtype b: Posterior fusion that does not extend beyond the retromolar/retromaxillary region
Subtype c: Fusion involving the ascending ramus/zygomatic complex.

In this review, type 1a accounted for 4.2% of all patients, type 1b for 39.4%, and type 1c for 7.0%. Our patient had three mucosal bands bilaterally and no bony fusion; therefore, our case was classified as Olusanya type 1. The patient's fibrous band on the left side extended beyond the retromolar region; therefore, the case could be classified as type 1c, which is infrequent in soft tissue fusion.

Congenital maxillomandibular synechia can cause difficulty in feeding and breathing, and early diagnoses and interventions are necessary because in long-standing cases, temporomandibular joint ankylosis often occurs owing to immobility and lack of function, which can necessitate more complicated surgical interventions.[5] However, no definitive management protocol is available for this disorder because of its rarity and varied presentation.

Patients with this anomaly are at a risk of aspiration, which might be fatal because they have difficulty in expelling vomitus through the mouth; therefore, clinicians must be on the alert for aspiration when feeding starts. Olusanya and Akadiri reported that 61% of type 1 (fibrous fusion) patients tolerated oral feeding, whereas 39% required tube or parenteral feeding. They also reported that the mean interalveolar distance was 5.3 mm in the oral feeding group and 2.3 mm in the tube or parenteral feeding group.[3] In our case, we considered whether to feed the patient orally or via tube feeding. As our patient was born at 32 weeks of gestation, when the deglutition reflux was still immature, we chose to feed via a nasogastric tube. We commenced feeding 48 hours after birth, and fortunately, feeding progressed successfully.

General anesthesia is the preferred surgical intervention for fusion. Olusanya and Akadiri documented that 68% of patients with type 1 and 98% with type 2 (bony fusion) required general anesthesia.[3] Local anesthesia is only recommended in patients with types 1a and 1b where quick release is possible.[6,7] However, this anomaly poses an anesthetic challenge because inability to open the mouth prevents laryngoscopy for orotracheal intubation. In Olusanya and Akadiri's review, among patients with type 1 in whom general anesthesia was employed, 34.5% underwent orotracheal intubation, 31% underwent nasotracheal intubation, and 6.9% underwent tracheostomy.[3]

In the present case, general anesthesia would have been challenging because the patient was a VLBW. Although we considered nasotracheal intubation and tracheostomy, we concluded that they were technically impossible and performed surgical release under local anesthesia.

Congenital maxillomandibular synechia occurs either in isolation or with associated anomalies. Olusanya and Akadiri reported that 34% of patients had isolated synechia and 66% had associated anomalies, including cleft palate (27.7%), ocular anomaly (11.7%), musculoskeletal anomaly (8.7%), and digits anomaly (8.2%).[3] Commonly associated syndromic conditions include van der Woude syndrome and popliteal pterygium syndrome (PPS). Both syndromes share manifestations, such as a cleft lip or palate and lip pits and are caused by mutations in *IRF6* gene.[8] PPS is a rare autosomal dominant disorder with an incidence of 1 in 300,000 live births and is characterized by popliteal pterygia, syndactyly, abnormal external genitalia, intraoral adhesions, ankyloblepharon, and pyramidal skin on the hallux in addition to cleft lip/palate and lip pits. Our patient, who had a cleft palate, lip pit, syndactyly, cryptorchidism, and maxillomandibular fusion, met the diagnostic criteria for PPS as proposed by Leslie et al.[9]

To our knowledge, this is the first reported case of congenital maxillomandibular synechia in a VLBW infant. The patient needed more complicated management than what is usually required for term infants with the disease. He underwent surgical resection of the bands under local anesthesia, whereas type 1c cases usually require general anesthesia. Although general anesthesia is safe in many patients, for patients in whom performing tracheal intubation or tracheostomy is difficult, such as LBW infants with severely limited mouth-opening ability, clinicians should consider the surgical release of bands under local anesthesia.

Conclusion

Congenital maxillomandibular synechia is a malformation that limits mouth opening, leading to difficulty in feeding or breathing. As it can be lethal, neonatologists must diagnose and classify this condition correctly and appropriately

manage feeding and airway. The standard treatment is surgical resection under general anesthesia in many cases. However, if airway management such as tracheal or nasotracheal intubation is difficult to perform in type 1 cases, clinicians must consider surgical resection of bands under local anesthesia.

Conflict of Interest

None declared.

Acknowledgments

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