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Management of a Neonate with a Rare Congenital Palatosubglossal Synechia in Pierre Robin Sequence

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ABSTRACT

Development of the human face begins in the fourth week of gestation, as a series of several complicated and well-organized sequential intrauterine events, disruption of which causes some sort of facial dismorhogenesis.

Oral synechiae is a rare congenital anomaly usually recognized at birth secondary to airway or nutritional compromise. They help to understand the intrauterine developmental steps of the facial region.

In this particular case with congenital palatosubglossal synechia, anterior soft palate of the patient with Pierre Robin sequence was repaired with the oral mucosal flap elevated from the floor of the mouth by using the synechial band as the pedicle of the flap.

Keywords: congenital palatosubglossal synechia, Pierre Robin Sequence, surgical repair

PİERRE ROBİN SENDROMLU BİR YENİDOĞANDA NADİR GÖRÜLEN PALATOSUBGLOSAL YAPIŞIKLIĞIN TEDAVİSİ

ÖZET

Yüzün gelişimi hamileliğin 4. haftasında başlar. İyi organize, bazı komplike intrauterin olayların ardısıra cerayan etmesi sonucu yüzün gelişimi tamamlanır. Bu süreçleri bozacak olaylar ise çeşitli fasiyal dismorfogenetik olaylara neden olabilir.

Ağız içinde sineşi görülmesi nadir bir konjenital durumdur. Genelde doğum sonrasında nefes problemleri ve beslenme problemlerine neden olabileceği için tanı konulur. Bu gelişimsel bozukluklar bize yüzün embriyoljik olarak nasıl geliştiği konusunda da ip ucu verir.

Bu vaka sunumumuzda Pierre Robin Sekanslı bir hastada olan palatosubglossal sineşi bandının pedikül olarak kullanılarak, dilin alt tarafından kaldırılan mukoza flebi ile onarılan anterior palatal yarığın tedavisini sunduk.

Anahtar sözcükler: konjenital palatoglosal sineşi, Pierre Robin Sekansı, cerrahi tedavi

Pierre Robin sequence is characterized by the sequence of clinical events associated with a small mandible (micrognathia). The tongue is enlarged and retracted (glossoptosis), obstructing the airway, which results respiratory symptomatology, that can make oral feeding difficult or sometimes impossible in severe cases. Patients with Pierre Robin sequence may also present with a cleft palate, which is not a diagnostic criteiron. It is thought that a relatively prominent tongue in such micrognatic patients leads to mechanical interference of the lateral palatine processes during palatal fusion. Therefore cleft palate might result from a delay in the transition of the palatine shelves from a vertical to horizontal orientation (1).

Development of the human face begins during the fourth week of gestation when migrating neural crest cells from the dorsal region of the anterior neural tube (cranial neural crest, [CNC]) combine with mesodermal cells to form the facial primordia. Then the maxillary prominences enlarge and grow towards each other and the nasal prominences.

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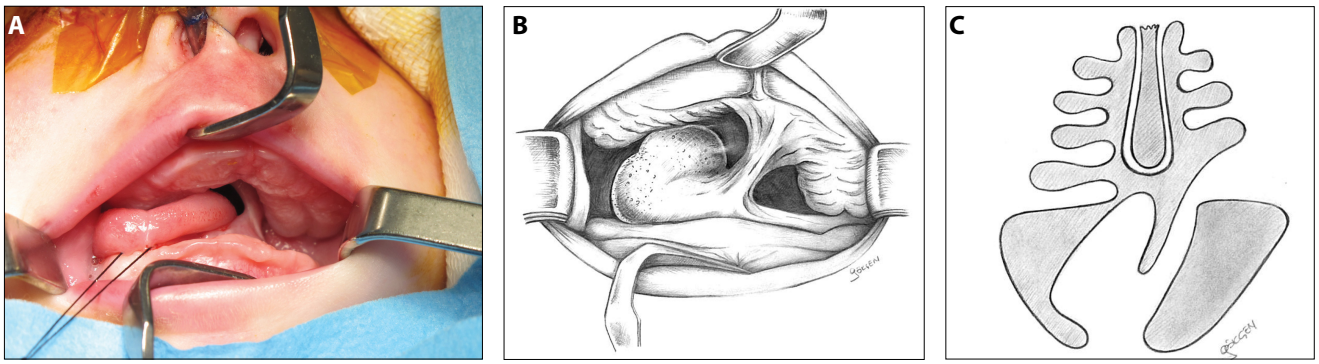

Figure 1: **A)** Operative view of the congenital palatosubglossal synechia located at the anterior part of the left palatal shelve of the cleft palate of the patient with Pierre Robin sequence, **B)** Illustrative drawing of the same pathology, demonstrating the synechial connection between the edge of the cleft palate and base of the tongue, **C)** Illustrative drawing of the same pathology, demonstrated as coronal cross-section.

During gestational weeks 6-7, the nasal prominences merge to form the intermaxillary segment resulting in both the filtrum and primary palate. This region then fuses to the maxillary prominences, which form the lateral parts of the upper lip (2). The secondary palate is also CNC derived and forms the palatal shelves, which grow out from the maxillary prominences (2). Although there are several theories on how the palatal shelves elevate, the mechanism remains unclear. The palatal shelves initially make contact at the midpoint and then palate closed towards both the primary palate and the uvula. The resulting epithelial seam rapidly disappears via a combination of programmed cell death, epithelial cell migration and transdifferentiation (3).

Oral synechiae is a rare congenital anomaly usually diagnosed in newborns secondary to airway or nutritional compromise. They arise between the upper and lower alveolar ridges (syngnathism) or between the tongue and margins of the palate or maxilla (glossopalatal ankylosis). Synechiae can also arise from the lower lip, floor of the mouth, or the oropharyngeal isthmus and may also consist of membranes or bands supported by connective tissue and even muscle or bone (3).

Case report

Herein we present a neonate with Pierre Robin sequence in which the anterior soft palate was repaired with the oral mucosal flap elevated from the floor of the mouth via use of a rare congenital palatoglossal synechiae as the pedicle of the flap.

A 35 day-old, malnourished, female weighing 2.9 kg was referred to our center because she could not be fed orally due to a congenital band in her mouth. Physical examination showed abnormal facial morphology, with a small mandible and palatal cleft, which were consistent with Pierre Robin sequence. Additionally, she had palatosubglossal synechia extending from the anterior part of the left cleft shelf to the base of the tongue (Figure 1). She had a nasogastric tube that had been used to feed her since birth.

She was scheduled for surgery in order to release the present band and to initiate oral feeding but the synechia was observed to extend between the anterior 1/3 of the left side of the cleft shelf and the base of the tongue. As such, instead of simply releasing the synechia, it was used as a pedicle to transfer the soft tissue from the oral base to the palatal defect. Mucosal soft tissue surrounding the attachment of the synechial band at the oral base was elevated like an inverted umbrella (Figure 2). The defect that remained was closed primarily. Then, the nasal and oral mucosae of the right cleft shelf were dissected as 2 layers. The mucosal edges of the oral mucosa that were elevated as a flap on the synechial band pedicle were sewn to the mucosa free edges of the nasal and oral layers of the right cleft shelf. Use of this technique facilitated closure of the anterior 1/3 of of the palatal defect with the soft tissue transferred from the oral base.

The patient was bottle-fed with breast milk immediately following surgery and began breast-feeding a few days later.

The soft tissue transferred from the oral base was still in place at the anterior 1/3 of the palatal cleft 3 weeks post surgery (Figure 3), but the patient did not come for subsequent follow-up visits. It was learned from her parents that she died due to lower respiratory track infection 3 months post surgery, before the definitive palatal repair was performed.

Discussion

The presence of oral synechiae may be a marker of additional congenital defects, such as cleft palate, cleft lip, microglossia, micrognathia, temporomandibular joint disorders, and lip

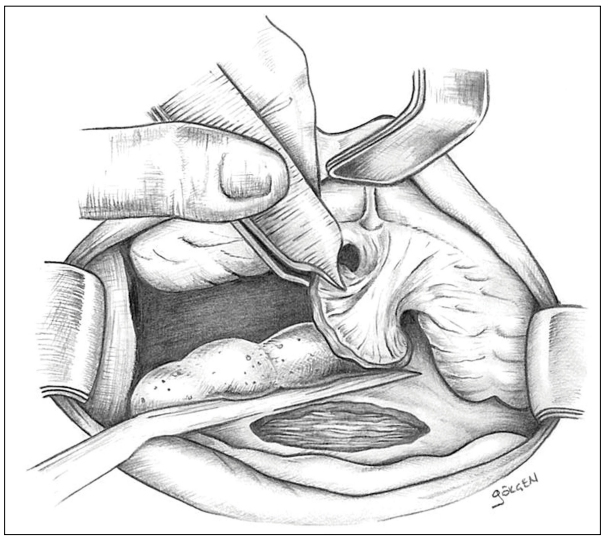

Figure 2. Illustrative drawing of the transfer of the mucosal soft tissue from the oral floor on the pedicle of the synechial band.

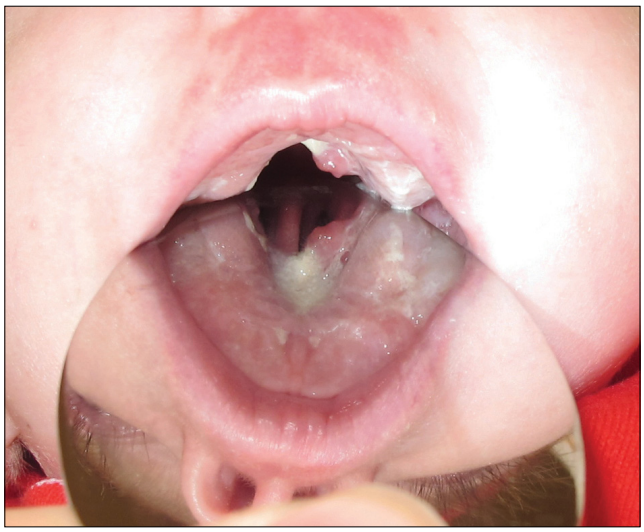

Figure 3. View of the transferred soft tissue three weeks after the operation.

anomalies. The etiology of oral synechiae remains unknown. Teratogenic or mechanical insults during gestational week 7-8 development may lead to abnormal fusion (4).

The timing and method of treatment of Pierre Robin sequence, characterized by micrognathia, glossoptosis, and/or cleft palate, are dependent on the severity of respiratory symptomatology at initial presentation. Treatment ranges from simple positioning of the baby to prevent posterior displacement of the tongue to mandibular distraction at a very early age.

The presented case supports the common belief that micrognathia leads to posterior and superior positioning of the tongue that does not allow the palatal shelves to undergo a sudden elevation to bring them into horizontal apposition above the flattening tongue, so as to fuse. Adhesion of the cleft shelf underneath the tongue shows that the palatal shelves between which the tongue is located were positioned vertically during early embryogenesis.

In the presented case the soft tissue of the oral floor on the synechial band was transferred to the anterior part of the cleft palate, instead of simply releasing the synechial band. This method facilitated repair of the anterior 1/3 of the cleft palate, decreasing the risk of the development of an anterior oronasal fistula, as per the Pittsburg fistula classification system (5). It is also benficial to elongate the vertical length of the palate by the time definitive repair is performed in order to decrease the risk of velopharengeal insufficiency.

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