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Case Report

Congenital intra-oral adhesions: A surgical approach to cleft palate lateral synechia syndrome☆

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ABSTRACT

An array of genetic syndromes has been associated with intra-oral adhesions in neonates. The primary medical issues arise from airway obstruction, feeding difficulties and poor oral development, specifically with cleft palate lateral synechia syndrome (CPLSS). Despite this, a paucity of data exists for the clinical management of intra-oral adhesions in this population. We report the cases of a father and daughter diagnosed with CPLSS who presented with respiratory and feeding difficulties at birth undergoing surgical correction. Early surgical ligation of intra-oral bands allows for a stabilization of the airway, improved feeding and oral development with a good long-term outcome.

1. Background

Congenital anomalies of the oral cavity are not infrequent findings at birth often requiring otolaryngologic consultation. The incidence of such lesions has been reported as high as 1 in 100 live births, with the majority being ankyloglossia [1]. Despite an often self-limited nature, several oral cavity anomalies can lead to significant airway compromise and difficulty with nutrition [2,3]. An array of genetic syndromes has been associated with intra-oral adhesions with tissue composition ranging from mucosal to osseous bands [3,4]. In particular, Fuhrman et al. first described a series of patients having both a cleft palate and multiple cord-like bands which restricted mouth opening [5]. The majority of these patients presented with respiratory or feeding difficulties at birth. They were later diagnosed with cleft palate lateral synechia syndrome (CPLSS).

A paucity of data exists on the surgical management of patients with intra-oral adhesions, irrespective of the associated syndrome [3,6,7]. This is likely due to the low incidence of patients with intra-oral bands, often leading the otolaryngologist to proceed on a case-by-case basis. Controversy exists on the extent and timing of surgery necessary to correct oral cavity malformations due to an incomplete understanding of the disease processes [8,9]. The goal of surgery is to divide adhesions to allow normal mouth opening and oral development. Moreover, this intervention aids to secure the airway as well as the ability to feed. In this case report, we describe a father and daughter who presented with cord-like adhesions in the mouth and were diagnosed with CPLSS.

2. Case presentation

We report the case of a term baby girl named A.B. who was born in respiratory distress to a 23-year-old G_2P_2 French Canadian mother. Although the patient was not stridorous at birth, she did have significant oxygen desaturations as she was unable to open her mouth due to intra-oral adhesions. Specifically, the patient had several thick cord-like bands joining the alveolar ridge of the maxilla to the floor of the mouth just lateral to the tongue. She also had a median adhesion at the incisor region. The patient's 1 and 5 min APGAR scores were 5 and 8, respectively. She was noted to have an incomplete, 'U-shaped' secondary cleft palate and enlarged base of tongue. Additionally, the patient had a retrognathic mandible which was also noted on computed tomography (Fig. 1). No other anomalies were identified on radiologic imaging. The patient did not have webbing of skin, abnormal digits or genitalia. Given the neonate's inability to open her mouth, several of the fibrovascular synechia were divided on her first day of life in the neonatal intensive care unit. Additionally, the patient needed minimal amounts of supplemental oxygen to maintain adequate oxygen saturation. On her second day of life, the patient had worsening respiratory distress which was thought to be due to a

☆ The study was completed at the Montreal Children's Hospital, Montreal, Quebec, Canada.

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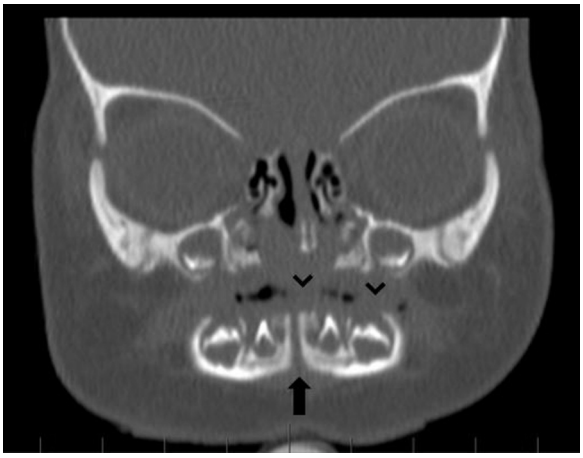

Fig. 1. Mandibular cleft and intra-oral adhesions on CT scan in patient A.B. The arrow corresponds to the midline mandibular cleft. The arrowheads indicate areas of intra-oral bands.

fixed upper airway obstruction from her remaining intra-oral bands, retrognathism, enlarged base of tongue and large omega-shaped epiglottis. Consequently, she underwent tracheostomy and excision of the remaining intra-oral adhesions in the operating room on her second day of life. Upon excision of the adhesions, the patient was able to fully open her mouth. She initially required naso-enteral feeding and was gradually decannulated after 2 months of age. The patient's cleft palate was repaired at 1 year of age with no peri-operative complications. Genetic testing revealed no abnormality; she did not possess an interferon regulatory factor 6 gene (IRF6) mutation and had a normal 46 XX karyotype. Furthermore, there was no evidence of consanguinity. However, the patient's mother did admit to taking phenytoin during her 1st trimester for a seizure disorder. In discussion with consultant geneticists, the combination of the patient's intra-oral adhesions, 'U-shaped' cleft palate, retrognathism and absence of systemic anomalies with unremarkable IRF6 gene testing suggested the diagnosis of CPLSS. On the patient's 2 year follow-up visit, her feeding and speech were unremarkable.

35 years earlier, the abovementioned patient's father (E.B.) had undergone a similar but less severe experience. He was born with the inability to open his mouth secondary to three fibrovascular intra-oral adhesions. Although he did not have any respiratory distress at birth, he was found to have an incomplete, 'U-shaped' secondary cleft palate. The three adhesions, located in the same position as his daughter, were ligated in the operating room within his first week of life, allowing the patient to have normal oral functioning (Fig. 2). The patient's genetic work-up was unremarkable, with a normal 46 XY karyotype. Advanced genetic locus testing for popliteal pterygium syndrome (PPS) and Van der Woude Syndrome (VWS) were not available at that time. The cleft palate was repaired at 11 months of age and there were no sequela after surgery (Fig. 3). E.B. was later diagnosed with CPLSS after the birth of his daughter A.B., where his past medical records and IRF6 genetic testing were reviewed.

3. Discussion

The varied clinical presentation of oral cavity anomalies can bring about a challenging differential diagnosis. Given that multiple cranio-facial syndromes often present with respiratory distress, use of a multidisciplinary team consisting of a pediatrician, otolaryngologist, geneticist and nutritionist is essential for optimal care [2]. At present, only a handful of syndromes have been identified to involve intra-oral adhesions as a hallmark of disease. Of particular note, these include VWS, PPS, Congenital Alveolar Synechiae Syndrome, Fryn's Syndrome and CPLSS [2,3,9]. Children affected with VWS can have a cleft lip or palate, hypodontia, syndactyly and heart malformations [10]. Similar to VWS, PPS includes features of skin webbing and genital abnormalities [11]. However, these syndromes are genetically distinguished by the resulting effect of

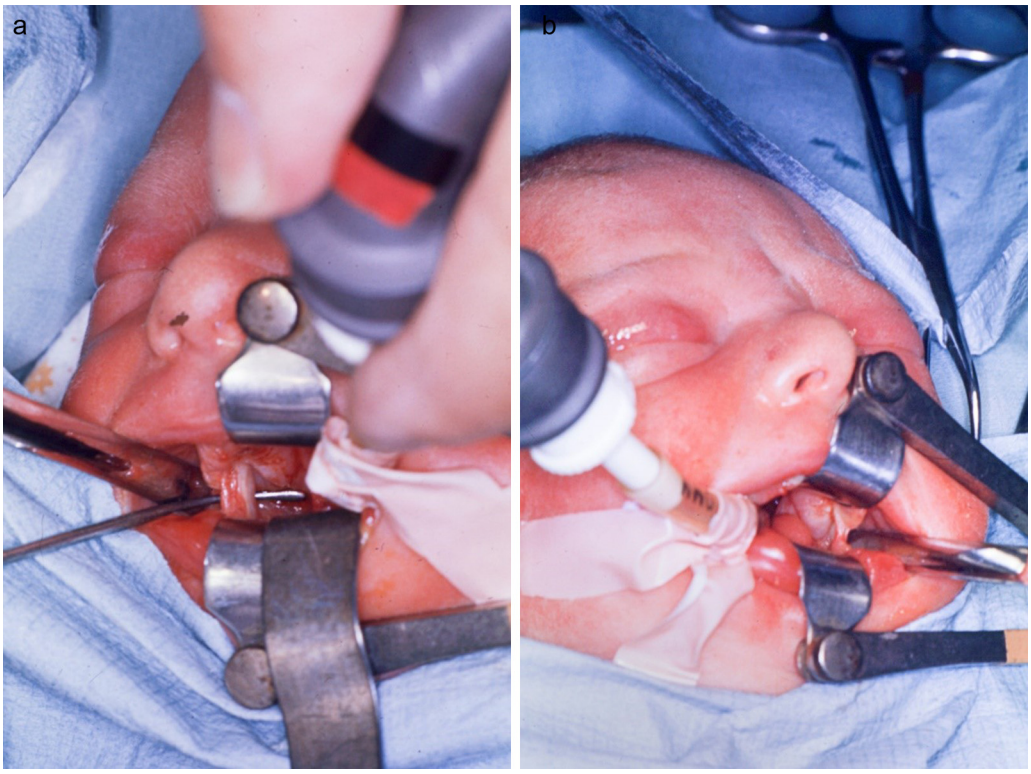

Fig. 2. Intra-oral adhesions as seen in the operating room in patient E.B. A lateral intra-oral band is noted in front of the probe in (a) and (b).