PALATE-TONGUE FUSION IN A NEONATE WITH PIERRE ROBIN SEQUENCE AND BILATERAL INCOMPLETE CLEFT PALATE

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ABSTRACT

Educational objective: To learn about palate-tongue fusion in a neonate and how this may be managed in a patient with Pierre Robin sequence.

Introduction: During palatogenesis, the bilateral palatal shelves first grow vertically and flap the developing tongue. They then elevate and fuse in the midline to form the secondary palate above the tongue. Palate-tongue fusions are rare but have been reported.

Study design: Case report and literature review.

Setting: Tertiary care pediatric hospital.

Methods: The case of a full term newborn male who presented to the neonatal intensive care unit for evaluation of palate-tongue fusion and cleft palate is reviewed (Figure 1). On exam, he demonstrated fusion between the ventral tongue and cleft palate edges (nearly circumferentially), as well as a thin adhesion between the maxillary and mandibular alveolar ridges on the left side. Moderate micrognathia was also present. A comprehensive literature review of similar cases was performed and compared to the current patient.

RESULTS

The patient underwent surgical lysis of the adhesions at 2 days of age (Figure 2). There were no postoperative complications. However, he subsequently developed glossoptosis and intermittent, positional airway obstruction, thus diagnosing Pierre Robin sequence. A maxillofacial CT scan without contrast was obtained around 10 days of age, which showed micrognathia and normal temporomandibular joint anatomy (Figure 3). He later required mandibular distraction osteogenesis to assist with alleviation of his obstructive symptoms. This procedure was successful; he was able to orally feed, and his obstructive symptoms resolved (Figure 4).

INTRODUCTION

Abnormal adhesions between structures in the oral cavity/oropharynx are called intraoral synechiae.1 Palate-tongue adhesions and other intraoral synechiae are rare but have been reported. Many are associated with cleft palate.1-2 During normal palatogenesis, the bilateral palatal shelves first grow vertically and flap the developing tongue. They then elevate and fuse in the midline to form the secondary palate above the tongue. Palate-tongue fusions are rare but have been reported.

METHODS

The case of a full term newborn male who presented to the neonatal intensive care unit for evaluation of palate-tongue fusion and cleft palate is reviewed (Figure 1). On exam, he demonstrated fusion between the ventral tongue and cleft palate edges (nearly circumferentially), as well as a thin adhesion between the maxillary and mandibular alveolar ridges on the left side. Moderate micrognathia was also present. A comprehensive literature review of similar cases was performed and compared to the current patient.

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DISCUSSION

Various intraoral synechiae have been described in the literature, though all of them are quite rare. In most cases, these intraoral synechiae are associated with cleft palate, such as in our current case. Depending on the location of the synechiae, patients may exhibit airway obstruction, trismus, dysphagia, and/or difficulty with oral endotracheal intubation.1-2 The exact etiology of intraoral synechiae is unclear, but many attribute them to remnants of the buccopharyngeal membrane. Some hypothesize that the synechiae prevent forward and inferior movement of the tongue, thus hindering fusion of the palatal shelves.1 This may explain their association with cleft palate, though in the current case, micrognathia may have been the inciting event of the sequence. Most described cases of intraoral synechiae involve interalveolar adhesions, and there are few documented reports of cases similar to the current patient.4-5

CONCLUSIONS

Palate-tongue adhesions and other intraoral synechiae are rare anomalies. Surgical intervention is usually indicated for lysis of the adhesions, thus allowing the commencement of oral feeds and alleviation of airway obstruction if present. In patients with Pierre Robin sequence, these adhesions may function as natural lip-tongue adhesions. When lysed, classic glossoptosis and intermittent airway obstruction may develop, thus requiring further intervention.

REFERENCES