Isolated Congenital Maxillomandibular Synechiae

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ABSTRACT

Introduction: Congenital maxillomandibular syngnathia is a rare condition that restricts the mouth from opening. This may lead to issues with feeding, swallowing, and respiration.

Study Design: Case report.

Discussion: We report a 1-day-old male with isolated bilateral soft tissue alveolar fibrous bands. He presented with difficulty feeding secondary to trismus. No bony or muscular involvement in the synechiae was noted. The physical exam was otherwise unremarkable. The bilateral alveolar synechiae were divided under local anesthesia using surgical scissors. The procedure was tolerated well by the patient and minimal blood loss was noted. The mouth opening was immediately improved to approximately 1.5 cm; however, a minimal amount of restriction was still present. The palate was noted to be intact and the baby had a good suck reflex. The baby was discharged later in the day after demonstrating the ability to breastfeed without difficulty.

INTRODUCTION

Congenital maxillomandibular syngnathia, first reported in 1936, is a rare deformity leading to difficulties in feeding and ability to thrive. The presentation may be unilateral, bilateral, or complete, but is most often syndromic in etiology.

Isolated cases of maxillomandibular fusion are extremely rare. The composition of the fusion can vary from mucosa only to muscle involvement to bony involvement. If the treatment is delayed, ankylosis of the temporomandibular joint may result.

A case report of a baby boy with isolated bilateral maxillomandibular fibrous bands is presented. The included review of the literature demonstrates only 11 cases worldwide of isolated maxillomandibular synechiae.

CASE REPORT

A baby boy, born via an uncomplicated vaginal delivery, was noted to have difficulty feeding due to trismus. The baby’s mouth opening was approximately 6 mm. Upon examination, it was established that the trismus was a result of bilateral posterior maxillomandibular fibrous bands at the approximate location of the primary molars (Figure 1). The bands were estimated to be 1 cm long and 2 mm wide. It was clear that the bands did not contain any bony component due to the easily compressible presentation (Figure 2).

No other craniofacial abnormalities were noted on exam; however, a complete palate exam was limited by the trismus. The mandible was palpated along with the temporomandibular joint and no obvious abnormalities were noted. For the first day of life, the baby was fed using a syringe and did not display any other difficulties. The mother had an uncomplicated pregnancy and received proper prenatal care. The family history was also unremarkable. This baby boy was the family's first child.

Following injection of local anesthetic (1% lidocaine with 1:100,000 epinephrine) into the fibrous bands, the adhesions were divided using surgical scissors. The procedure was tolerated well by the patient and minimal blood loss was noted. The mouth opening was immediately improved to approximately 1.5 cm; however, a minimal amount of restriction was still present. The palate was noted to be intact and the baby had a good suck reflex. The baby was discharged later in the day after demonstrating the ability to breastfeed without difficulty.

CONCLUSIONS

Most patients with congenital maxillomandibular syngnathia have an associated oromaxillofacial abnormality such as cleft palate, cleft lip, microglossia, micrognathia, aglossia, or TMJ anomalies. The fusion can vary in severity from simple mucosal adhesions to extensive bony fusion, with soft tissue fusion being the most common.

While the exact etiology of the adhesions is unclear, two common theories exist. During the seventh and eighth weeks of embryological development, the tongue, palatal shelves, and alveolar ridges are in close contact. For palatal closure to occur, the tongue must contract inferiorly and anteriorly. If the tongue does not protrude, then there is a predisposition for alveolar fusion. Others postulate that the adhesions are buccopharyngeal membrane remnants or amniotic constriction bands in the region of the developing branchial arches. The cause of the present case is believed to be an embryologic remnant or a formation of ectopic membranes. Given that the patient had no other head and neck abnormalities and there was no significant family history, a genetic or syndromic cause is extremely unlikely.

In conclusion, maxillomandibular syngnathia is a rare finding with a wide variety of presentations. Given the effects this can have on the infant's development, treatment should not be delayed. In the majority of cases, imaging will be necessary to determine the extent of the fusion and will assist in guiding the management. The presented case is an extremely unusual presentation in that the extent of the fusion was able to be determined on physical examination. For these unique cases, simple division of the fibrous bands can be curative.

REFERENCES


