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CASE STUDY

A RARE CONGENITAL ANOMALY: PARTIAL MAXILLO-MANDIBULAR FUSION

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ABSTRACT

Congenital maxillo-mandibular fusion is a rare anomaly involving simple mucosal fusion or complete bony fusion that may lead to nutritional or respiratory problems in early and after birth due to restricted mouth opening. Early intervention is crucial to prevent ankylosis of the temporomandibular joint and to achieve complete jaw development. Herein, we present a rare case of maxilla-mandibular fusion in the context of associated therapeutic challenges.

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INTRODUCTION

Congenital soft tissue (synechia) or bony (syngnathia) fusion of maxilla and the mandible is a rare anomaly (Daniels, 2004) that was first described by Burket in a patient with ankylosis of the temporomandibular joint, gingival fusion, and facial asymmetry (Parkins and Boamah, 2009). This anomaly, whether bilateral or unilateral, may lead to problems in the respiration, feeding, and growth of the baby due to restrictedmouth opening (Uluçay *et al*., 2010), necessitating early intervention. Delayed intervention may result in the occurrence of more complicated problems such as the ankylosis of temporomandibular joint (Bali *et al*., 2010).

Case presentation

No mouthopeningwasobservedduringthepostnatalexamination of a female infant born at termwith a birth weight of 3500 g from a 26 year-old mother. Reportedly, the mother had an uncomplicatedp regnancy and the baby was the fourth off spring of the parents. No familial history of congenital a nomalies was present, although there was a 1st degree consanguinity marriage between the parents. Blood tests and other systemic examinations were normal. The alveo larfusion involved the gingiva only, commencing at the right canine teeth and extending upto the left canine. Also, associated bonyfusion was observed extending from the left canine teeth upto the left retro-molararea (Figure 1A). A maxillofacial tomography showed bonyfusion between the left mandibular corpus-ramus and maxilla, with a secondary displacement of the mandible superiorly from the symphysis (Figure 1B). The written informed consent was obtained from the infant's parents. The infant's parents have also provided consent to have the case details, and three dimensional maxillofacial CT and pre- and post operative photos published. Surgical treatment under general anesthesia was planned for the 7 day-old baby following the workups. A consultation was requested from the department of otorhinolaryngology which pointed out to a potentially increased risk of tracheostomy-associated complications owing to immature development of the trachea in the newborn period. Tracheostomy was not performed, also considering the requirement for general anesthesia and reliable airway. Under sevoflurane anesthesia with maintenance of the spontaneous respiration, blind nasal intubation was attempted with no success. Then, a decision was made to start the surgical intervention to create oral opening under dissociative anesthesia, followed by endotracheal intubation. The anterior gingival fusion could be easily and rapidly released with a mosquito clamp, while a periosteal elevator and scissors were used to open the bony fusion extending from the left canine teeth to the retromolar space.

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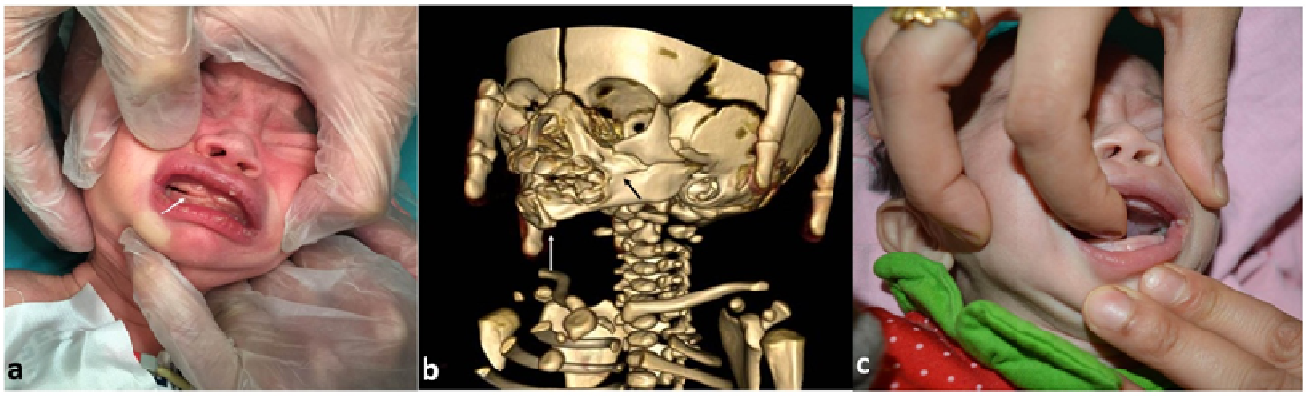

Fig. 1. a) Fusion starting at the level of the canine tooth (white arrow) extending into the left retromolar area. b) Three dimensional maxillofacial CT showing the fusion between maxilla and mandible (black arrow), displacing the mandible superiorly at the level of symphysis (white arrow). c) Adequate mouth opening attained after second surgery

Minimal bleeding occurring at the site of surgery was aspirated. Endotracheal intubation was performed after adequate oral opening was attained and fusion was completely opened. Postoperative breastfeeding was allowed and jaw physiotherapy was commenced. Due to recurrence of the bony and mucosal fusion between the left canine and left retro-molar area, the patient was taken into surgery. Anterior mouth opening allowed endotracheal intubation through video laryngoscopy. After opening the mucosal and bony fusion, silicon material was placed on the non-epithelized gingival area to prevent the relapses that was removed after completion of epithelization on the 7th day. At 3rd postoperative month, a good mouth opening was found (Figure 1C).

DISCUSSION

Congenital maxilla-mandibular fusion is a rare anomaly of unknown etiology that can occur at different anatomical sites within the oral cavity. Medial movement of the oral cavity walls and the outward movement of the tongue through the oral opening prevent the fusion of alveolar processes. Abnormal fusion occurring at the 7th and 8th weeks of embryonic development is thought to result from genetic, teratogenic or mechanical factors (Haydar *et al*., 2003). The fusion may involve epithelial membranes or bands of tissue as well as fibrous tissue, bone or muscle in varying degrees. Isolated fusion between soft tissues istermed as synechia, while those between bony structures are referred toas syngnathia (Gupta *et al*., 2008). Our patient had synechia between the mucosal membranes anteriorly, while syngnathia was present in the lateral part of the left canine tooth level. This anomalymay occur alone or in association with a number of other conditions including cleft palate, microglossia, micrognathia, etc. (Tanrikulu *et al*., 2005) Maxillo-mandibular fusion is generally identified soon after birth by the restrictionor total absence of the mouth opening (Daniels, 2004). The timing of the surgery depends on the general condition of the patient, although total parenteral or nasogastric tube feeding may be required until the time of surgery to prevent malnutrition. Early surgery is recommended to prevent the risk of aspiration and nutritional problems, and also for attaining normal mandibular function and facial development as well as for the prevention of TME ankylosis (Hegab *et al*., 2012). Nasal blind intubation or intubation through fiber-optic indirect laryngoscopy are generally preferred under general anesthesia, while tracheostomy represents a last resort (Bozdag *et al*., 2011). After release of fused tissues, mandible should be mobilized during the surgery for achieving mouth opening. However, forceful maneuvers may result in jaw fractures, due to soft alveoli and fragile mandible in newborns. A variety of applications such as silicone plaques may be utilized to prevent re-fusion of bone and mucosa. Early jaw mobilization and physiotherapy also reduce the risk of recurrence (Hegab *et al*., 2012). The patient should be closely followed and the family should be well informed about the condition to help prevent recurrences.

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