Isolated congenital fusion of the gums

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ABSTRACT

Isolated congenital fusion of the gums is a rare anomaly. Early surgical treatment is indicated, as longstanding cases will impair normal feeding leading to nutritional and growth problems. We report the first case in the Yaoundé Gynaeco-Obstetric and Paediatric Hospital, which was successfully managed surgically.

Key words: Congenital fusion, gums, surgical treatment, Yaounde, Cameroon

INTRODUCTION

Congenital fusion of the gums is a very rare malformation. The true incidence is unknown. Fusion may be partial or complete, and may be associated with other facial dysmorphologic malformations such as facial hemiatrophy or temporo-mandibular ankylosis.[1] Complete fusion may interfere seriously with feeding leading to severe nutritional problems. Major difficulties may also arise during intubation for surgery. We report a case referred to us in the Yaoundé Gynaeco-Obstetric and Paediatric Hospital from a rural hospital. The rarity of this entity merits its report.

CASE REPORT

A 14-day-old female was referred from a rural hospital for fusion of the gums noticed at birth. The mother is G3P3003, a 22-year-old housewife. The father, 32 years old, is a driver. The pregnancy was regularly followed up but no obstetric ultrasounds were done and delivery was at 40 weeks gestational age. The Apgar score was not specified in the referral letter and the birth weight was 3.4 kg.

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On physical examination she was febrile with a temperature of 38.5°C, weight 2.85 kg, with complete anterior and bilateral fusion of the gums [Figure 1]. Neurologically she was slightly hypotonic. The work-up included a complete blood count, C-reactive protein, blood, urine and cerebrospinal fluid cultures. All were normal except for the blood cultures which revealed *Enterobacter cloacae* sensitive to cefotaxime. Antibiotherapy was instituted and feeding was by gavage through a nasogastric tube. Chest x-rays and abdominal ultrasound were normal.

By the 4th day of admission, she was afebrile and taken to the theatre. Local anaesthesia with low dose lidocaine was instilled into the fibrous tissue joining the gums. The anterior and lateral parts of the gums were separated by carefully sectioning the fused tissues. A Guedel's cannula was inserted into the mouth and fixed. Examination of the oral cavity was normal.

The next day, incomplete opening of the mouth and difficulty when crying were noted. X-rays of both temporo-mandibular joints (TMJ) and the entire body were normal. She was taken to the theatre four days later and on examination residual fibrous cartilaginous tissue was found bilaterally at the temporo-mandibular joints. Adhesiolysis was done with Heggar's boogies until adequate opening of the mouth was obtained [Figure 2].

The child was able to suck and breastfeed adequately. There was minimal bleeding in both operations. She was discharged after 18 days of admission with a weight of 2.98 kg.

She was re-evaluated two weeks later, and examination and feeding were normal.

DISCUSSION

Congenital maxillomandibular fusion is a rare group of anomalies varying in severity from simple mucosal adhesions (synechiae) to extensive bony fusion (syngnathia).[2] In the presence of fusion with intra-alveolar synechiae, syndromic associations should



Figure 1: The gums of the patient before surgery with a nasogastric tube in place



Figure 2: The mouth of the same patient after surgery

be looked for, although isolated instances do occur. The most frequent syndrome setting is the Van der Woude syndrome.[3] This syndrome is a hereditary developmental malformation transmitted by autosomal dominant mode. Lower lip pits, cleft lip with or without cleft palate and isolated cleft palate are its cardinal signs.[4] Our patient had complete fibrous synechiae on the gums extending to the temporo-mandibular joints, with no other oral or morphologic anomalies. It may also occur in the context of a mandibulofacial dysostosis caused by a genetic delay in the development of the visceral arch.[5]

The exact aetiology is not known. Several hypotheses have been put forward.[6-11] It is hypothesised that during the 7th to 8th week of embryological alveolar ridge development when the tongue and palatal shelves are in close contact, the ensuing palatal closure depends on downward and forward contraction of the tongue. When the tongue protrudes from the mouth as a result of medial movements of the oral cavity walls, it prevents the alveolar ridges from fusing. Genetic, teratogenic or mechanical insults during this critical period may lead to prolonged close quiescent contact between oral structures, thereby predisposing to abnormal fusion. Other possible aetiologic factors put forward are: trauma late in pregnancy, abnormality of the stapedial artery, teratogenic agents, hereditary factors and remnants of the buccopharyngeal membrane.

Congenital fusion of the gums may present many problems to the newborn. Fusion prevents mouth opening, feeding, swallowing and respiration resulting in nutritional disorders and failure to thrive. So early treatment is warranted, not only to correct these problems, but also because in the early stages, this condition can be treated easily.[7] In long-standing cases, temporo-mandibular joint ankylosis often occurs because of immobility and lack of function, necessitating more complicated surgical treatment.[7,8,12] TMJ ankylosis can lead to lack of mandibular growth and facial and deformities.[8]

It should be noted that although surgical treatment is short and straightforward, general anaesthesia presents a challenge because of impossibility of oro-tracheal intubation.[7] Nasotracheal intubation whether done by so-called blind intubation or by indirect laryngoscopy using fiberoptic visualization is the method of choice.[13] The possibility of failure in this procedure, even in experienced hands, demands equipment and personnel for emergency tracheostomy. Blind nasal intubation may also fail due to technical difficulties or be complicated by laryngospasm. However, the use of fibre optic visualisation presents suction and ventilation problems.[14] Other surgeons have used elective tracheostomy successfully.[6, 9]

Some prefer local anaesthesia.[12] We used local anaesthesia in our patient because of unavailability of equipment for fiberoptic visualization or nasotracheal intubation.

Concerning outcome, normal mouth opening is expected several weeks after successful surgery.[7] Our patient achieved adequate mouth opening and was able to breastfeed, and cry normally 3 days after the second operation.

CONCLUSION

Isolated fusion of the gums is a very rare congenital

anomaly. Early surgical treatment is necessary to avert feeding problems. However anaesthesia for this procedure is challenging.

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