Congenital Fibrous Maxillomandibular Fusion

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Abstract Congenital fibrous fusion of the jaws (synechiae) without any concomitant oral anomalies is a very rare malformation. Less than 40 cases have been documented in the literature. Early division of fibrous bands not only allows effective feeding but also prevents development of facial deformity. Here we present a case of congenital maxillomandibular fusion in a neonate.

Keywords Congenital · Maxillomandibular · Fusion · Syngnathia

Case Report

A 20 days old male neonate was referred from the department of pediatric dentistry to our department with complaint of inability to feed and restricted mouth opening following full term unremarkable pregnancy and vaginal delivery. Apgar score was 8 and 9 at 1 and 5 min after birth. Weight at birth was 2500 g. He was being fed orally with fluids, although with great difficulty. There was no

history of systemic abnormality in the family. However, the parents did have a consanguineous marriage. Intraorally, there was soft tissue fusion of maxillary and mandibular gum pads with fibrous bands present bilaterally in the posterior region on the buccal aspects; the thickness of the bands was approximately between 3 and 4 mm. Anteriorly the mouth opening was 4 mm (Fig. 1). Detailed intraoral examination could not be carried out due to restricted mouth opening. However on limited examination all other oral structures seemed to be normal. On palpation bilateral temporomandibular joint movements were within normal limits. Patient was referred to the department of pediatrics to rule out any intracranial abnormality. Investigations were done including complete blood count, C-reactive protein, and urine analysis. All were in the normal range. CT scan of the head and ECG were also normal. Lateral cephalogram was done to rule out any bony fusion. Bilateral excision of soft tissue bands was planned under local anesthesia and sedation. Antibiotics were instituted parenterally under the supervision of a pediatrician.

There are difficulties in intubating patients with limited mouth opening for general anaesthesia; since fibreoptic intubation was not available at our centre and parents did not give consent for tracheostomy the procedure was planned under conscious sedation. Patient was sedated by an anesthetist using Midazolam [0.1 mg/kg intravenously]. After stabilizing the infant's head gently with hand, local anesthesia with adrenaline was infiltrated in the area of fibrous bands and all the fibrous bands were cut using bard parker knife (no. 15). High pressure suction was attached to prevent blood aspiration (Fig. 2). After incision of the fibrous bands bilaterally, index fingers of both hands were inserted in between the alveolar ridges and mouth opening of 20 mm was achieved. A thorough intraoral examination revealed all structures including uvula, palate and tongue to

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Fig. 1 Restricted mouth opening with fibrous maxillomandibular synechiae on right side

Fig. 3 'U' shaped Iodoform pack secured in between the ridges to prevent reattachment of raw surfaces

Fig. 2 Excision of bands under high power suction under LA with sedation

Fig. 4 Postoperative mouth opening after 1 month

be normal. Complete hemostasis was obtained by applying finger pressure for 5 min.

To ensure hemostasis in the immediate post operative period and to prevent reattachment of the raw surfaces, a 'U' shaped Iodoform gauze piece measuring 5 × 2 cm was placed in between the alveolar ridges and secured in place with the help of a 4-0 vicryl suture bilaterally. (Fig. 3).

The gauge piece was removed after 48 h and the patient was discharged with the advice to use a pacifier for 10 min every hour for next 4 days to avoid reattachment of the cut surfaces till complete epithelization could take place. The child had a normal sucking reflex so he could be fed using both bottle and breast milk. Uneventful healing occurred in one week. The mouth opening as recorded after 1 week was 20 mm and after two months it was 26 mm. (Fig. 4). There has been no recurrence till the last follow up six months post operatively.

Discussion

Congenital maxillomandibular fusion is a rare anomaly involving fusion between the maxilla and the mandible. It

varies in severity from simple mucosal adhesions (synechiae) to extensive bony fusion (syngnathia), soft tissue fusion being more common. It may be partial or complete and may occur unilaterally or bilaterally [1]. Bilateral fusion in the posterior region is more common presentation. No sex predilection has been reported [7]. Less than 40 cases have been reported in literature; most commonly in association with popliteal pterygium syndrome [2]. Maxillomandibular fusion is generally discovered immediately after birth since the baby is not able to open the mouth or feed normally. Congenital maxillomandibular fusion may be associated with other anomalies like Van der Woude syndrome [3], popliteal pterygium syndrome [4] or Aglossia-adactylia syndrome [5]. Gartlan et al. [2] reviewed soft tissue fusion and classified synechiae as buccopharyngeal membrane remnants or ectopic membranes depending upon their presumed origin.

The exact etiology of syngnathia is not known but some of the reported causes including Goodacre and Wallace's [6] experimental studies include persistence of buccopharyngeal membrane, amniotic constriction bands in the region of the developing first branchial arch, environmental insults, drugs such as meclozine and large doses of vitamin A.

Dawson et al. [7] in a review classified syngnathia as:

Type 1	Simple syngnathia—with no other anomalies in the head and neck
Type 2	Complex syngnathia—with subgroups
*Type 2a	Syngnathia co-existent with aglossia
*Type 2b	Syngnathia co-existent with agenesis or hypoplasia of proximal mandible

Their study did not provide any evidence of familial tendency, history of drug exposure or consanguinity. However, there was a clear history of consanguinity in our case. Ahmad Mir et al. [1] also reported a case of synechia, in which the parents were first cousins. This suggests the possibility of autosomal recessive inheritance.

Once recognized bony fusion can be ruled out by conventional radiography or high resolution spiral CT scan. Surgical excision should be done as soon as possible so that nutritional deficiencies can be avoided in developing neonate. Also it is important to provide early mobilization of TMJ for unhindered growth of maxillofacial complex. Anesthesia is challenging in such patients. Awake blind nasal intubation is the ideal way of introducing anesthesia. In case of failed awake blind nasal intubation, fiberoptic intubation or tracheostomy may be required. However when there is no evidence of body fusion, soft tissue maxillomandibular union can be excised under local anesthesia with sedation under expert anaesthetic care. In this case we excised the fibrous bands under local anes-

thesia under sedation with high pressure suction to prevent blood aspiration.

The raw surfaces have to be kept separated from each other till their epithelialization takes place. We achieved this by a combination of Iodoform gauze interpositioning in the early post-operative, followed by active use of pacifier thereafter. Physiotherapy should also be advised to parents by inserting little fingers in between the gums and rotating the fingers so that the ridges can be separated.

References

1. Ahmad Mir M, Iqbal S, Hafeez Adil (2007) Syngnathia without any other associated anomaly: a very rare case report. Internet J Plast Surg 4(1). Available on : URL http://www.ispub.com/journal/the_internet_journal_of_plastic_surgery/volume_4_number_1_44/article/syngnathia_without_any_other_associated_anomaly_a_very_rare_case_report.html
2. Gartlan MG, Davies J, Smith RJ (1993) Congenital oral synechia. Ann Otol Rhinol Laryngol 102:186–197
3. Puvabanditsin S, Garrow E, Sitburana O (2003) Syngnathia and Van der Woude syndrome: A case Report and Literature Review. Cleft Palate Craniofac J 40(1):104–106
4. Hamamoto J, Matsumoto T (1984) A case of facio-genito-popliteal syndrome. Ann Plast Surg 13:224–229
5. Johnsson GF, Robinow M (1978) Aglossia–adactylia. Radiology 128:127–132
6. Goodacre TE, Wallace AF (1990) Congenital alveolar fusion. Br J Plast Surg 43:203–209
7. Dawson KH, Gruss JS, Myall RW (1997) Congenital bony syngnathia. A proposed classification. Cleft Palate Craniofac J 34(2):141–146