CASE REPORT

CONGENITAL ISOLATED BILATERAL SOFT TISSUE SYNGNATHIA IN A 4-DAY OLD BABY BOY

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Congenital maxillomandibular syngnathia is characterized by fusion of jaws. Depending on the severity, it has a wide range of clinical presentations. It can be complete /incomplete and may be unilateral or bilateral. Primary concern in such patients is maintenance of airway and feeding difficulties. Therefore, early recognition and management is important to reduce nutritional, feeding, airway difficulties and growth-related problems in such new-borns. This case report presents a case of syngnathia in a 4-day infant with bilateral fusion of maxilla and mandible, leaving a small anterior portion. Early intervention was planned and the fusion was released to facilitate feeding. Good mouth opening was seen on 1week follow-up.

Keywords: Congenital; Maxillomandibular fusion; Syngnathia; Synechiae

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INTRODUCTION

Syngnathia is one of the rare congenital deformities of head and neck region involving the fusion of maxilla and mandible resulting in inability to open mouth adequately. This results in difficulty in feeding, swallowing, breathing which leads to ankylosis of temporomandibular joint if timely treatment is not provided.[1,2]

Depending on the character of connecting tissue it can either be fibrous or bony fusion. When fusion involves only soft tissue fibrous bands it is termed as Synechiae.

Bony adhesion of both jaws or extensive bony fusion of mandible with zygoma, hard palate and temporal bone is termed as Synostosis which has less occurrence rate than synechiae.[2]

First case of syngnathia was reported by Burket in 1936.[3] Exact cause and incidence of the condition is still unknown.[2] It can present as an isolated problem but is most often seen in association with other syndromes, i.e., with Horner syndrome, Popliteal syndrome, Van der Woude syndrome and Aglossia adactyllia syndrome.[2] Many cases with combination of cleft palate, cleft lip, hemifacial microsomia, bifid tongue, aglossia have also been reported.[1]

CASE REPORT

Four days old, full term, male presented to the Paediatric dentistry department, Children hospital, Pakistan Institute of Medical Sciences, Islamabad. The chief complaint of parents was that the baby had inability to open the mouth and difficulty in feeding (Figure-1). Baby was born via normal vaginal delivery at home. His weight at birth was 2.8 kg, body length was 51 cm which is 50th centile, with positive parental consanguinity.

Intraoral examination revealed presence of four anterior and posterior bands bilaterally. Bands were approximately 1×1 cm^2 in dimension and soft in texture with no bony involvement (Figure-2). Thorough intraoral examination could not be done due to negligible mouth opening. Paediatric consultation was done to rule out other associated abnormalities. Patient was considered otherwise normal with no chest, CVS, CNS and gastrointestinal abnormalities.

Surgical excision of synechia was planned under general anaesthesia. Since it was confirmed by thorough clinical examination and tissue palpation that no bony fusion is involved, radiographic investigations were considered unnecessary. Patient was admitted in the hospital for treatment and all preliminary investigations including Complete blood picture, PT/apTT, INR, serum electrolytes RFTS, echo and chest x-ray were done. To minimize haemorrhage during the procedure, intravenous injection of 10 mg/kg tranexamic acid (Vit K) was given. General anaesthesia was attained through nasotracheal intubation with the help of laryngoscope, head was gently stabilized, and local infiltration in the areas of fibrous bands were given with 1.8 ml cartridge of 2% lidocaine with epinephrine (1:100,000) and complete surgical excision of all fibrous bands were done using bard parker #15 blade.

High vacuum suction, catheter size 8 (Foyomed^R) were used to prevent aspiration. Mouth opening improved immediately after removal of bands and patient started oral feeding

on the day of surgery without difficulty (Figure-3). Postoperative examination revealed normal palate, tongue, and para-oral structures. Patient was discharged 24 hours after surgery. At one-week follow-up appointment, mouth opening was adequate and recorded 20 mm with no complaint of feeding and breathing difficulties. Patient was recalled after four weeks and then eight weeks, healing occurred unremarkably, and no post-operative complications were observed. Baby had gained weight appropriately with normal sucking reflex and parents were counselled that recurrence of the lesions is nearly impossible because of only fibrous tissue involvement and instructions were given for 3–6 monthly clinical follow ups, for one year.

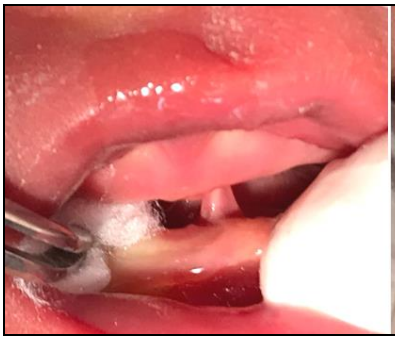

Figure-1: Restricted mouth opening and approximation of upper and lower jaws

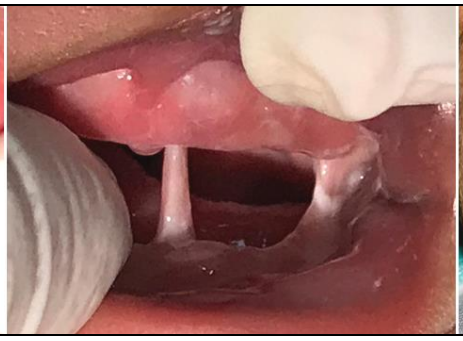

Figure-2: fibrous synechie anteriorly and posteriorly on buccal side

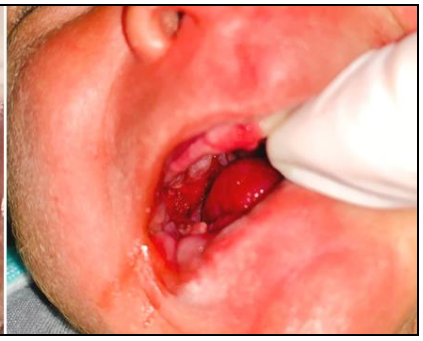

Figure-3: Improved mouth opening after fibrous bands removal

DISCUSSION

Congenital maxillomandibular fusion is a rare anomaly involving the adhesion of maxilla and mandible. These may consist of membranes or bands of epithelium (synechiae) supported by various amounts of connective tissue, and possibly even muscle or bone (synostosis).[4] They may be unilateral, bilateral, leaving a slit-like opening or complete.[5] Isolated cases of synechie are very rare.[1] About less than 40 cases have been documented in the literature.[6]

Oral synechia can be adhering at various locations and of various degrees. These can be classified into five types: Synechia by cord-like adhesion of the alveolar mucosa on one or both sides of the upper and lower jaws (alveolar synechia); synechia by a membranous fusion on the hard palate and floor of the mouth, excluding the rear of the tongue (lateral synechia); synechia involving the hard palate and tongue partially; synechia involving the soft palate and tongue widely, such that continuity is interrupted between the oral cavity and the pharynx; and synechia by a membranous adhesion between the hard palate and lower lip.[7] In our case alveolar synechiae was observed.

Bilateral fusion is more commonly observed in the posterior region.[6]Although in our case bands were present both anteriorly and posteriorly. No gender predilection has been reported.[8] Though the exact aetiology is not known, many theories have been proposed to explain the aetiology of intraoral synechiae such as teratogenic, genetic, or mechanical insult during 7th and 8th stages of embryological development of tongue, alveolar ridges, and palatal shelves, abnormality of the stapedial artery, presence of amniotic constriction bands in the region of the developing branchial arches, hypervitaminosis A, drugs like meclozine[6] and trauma late in pregnancy. Mathis postulated that such adhesions were remnants of the buccopharyngeal membrane.[9]

Dawson *et al*. has described syngnathia but has given no evidence about family history, consanguinity or drugs whereas in our case consanguinity was present.[8] Ahmed Mir *et al.* and Rishi Bali *et al*. on the other hand, have reported parental consanguinity in their cases of oral synechiae which is in consistent with our report.[10,6]

It is commonly observed with other craniofacial abnormalities, including Horner syndrome, orofacial-digital syndrome, popliteal syndrome, Van der Woude syndrome and Aglossia–adactylia syndrome.[11]Many cases with combination of cleft palate, cleft lip, hemifacial microsomia, bifid tongue, aglossia have also been reported.[12] But these were not observed in our case. And so, this rare case was diagnosed as isolated congenital synechiae.

Once bony fusion is ruled out by physical examination, conventional radiographs or high resolution spiral CT scans (for assessing the extent of adhesions), surgical excision should be done as early as possible for effective nutrition, respiration to provide early mobilization of temporomandibular joint (tmj) complex for unhindered growth and development of oral maxillofacial complex and prevention of tmj ankylosis.[6,13] Successful surgical corrections during late post-natal life also have been reported in the literature.

In the present case the patient, being 4 days old, without dysmorphic features, presented early, and surgery, being the mainstay of the treatment strategy, was timely planned. Airway management and maintenance of anaesthesia may present a problem to the surgical team. Different methods have been mentioned in the literature to excise these fibrous bands including surgical instruments, silk ligatures, electrocautery, scissors and scalpel.[14] Smriti Panda *et al*. used diode laser under endoscopic guidance for the release of fibrous bands. [15]In the present case, surgical excision with the use of scalpel was carried out under sedation along with constant anaesthetic monitoring.

No recurrence was observed on the follow up visits. Fibrous alveolar synechiae without bone or muscle abnormalities have good outcomes when properly excised. Complete mouth opening may not be possible immediately after the surgical procedure and it may take 1–2 weeks post operatively. In the present case, adequate mouth opening was observed after 1-week follow-up.

CONCLUSION

Though syngnathia is a rare case, still the operative management of such cases should be carried out as early as possible. If the patient is otherwise healthy, excision under general anaesthesia is the ultimate and successful treatment option. Regular physiotherapy and close follow-up will prevent reunion.

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