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Case Report

Congenital Unilateral Maxillo-Mandibular Syngnathia: A Brief Case Report and Review of Literature

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Received: January 28, 2019 **Published:** February 25, 2019

Abstract

Introduction: Congenital maxillomandibular fusion is a very rare condition with few cases reported. Cases of combinations of bony or soft tissue adhesions between the mandible and maxilla with cleft of the lip or palate, aglossia, popliteal pterygium and van der Woude syndrome have been reported.

Case Report: The aim of this paper is to report a case of 3 days old baby with congenital unilateral maxillo-mandibular syngnathia, which was referred to the Paediatric surgery department of Sher-e-Bangla Medical College and Hospital, Barisal. Separation of the fusion was done under nitrous oxide/oxygen sedation.

Conclusion: Maxillomandibular syngnathia is considered rare. This report adds more information to the literature. Prompt, definitive diagnosis and management of this type patient may helps in early breast feeding and prevent facial deformity.

Keywords: *Congenital Maxillomandibular Fusion; Mandibular Syngnathia*

Introduction

Congenital maxilla-mandibular fusion is a rare condition which was first reported by Barker (1936). The last case was reported by A Hegab., *et al.* [1]; thus, there are about 34 cases have been reported in the literature since then. Congenital craniofacial disorders represent approximately 20% of all birth defects [2].

Congenital maxillomandibular fusion (syngnathia) is a rare deformity seen in infants that often appears as part of a syndrome or other anomalies (Table 1) and can vary in severity from single mucosal bands (synechiae) to complete bony fusion(synostosis), which is less common than synchiae. It may affect soft and/or hard tissue and can restrict maximum mouth opening causing problems with feeding , respiration and general anaesthesia [3,4]. In case of 'Syngnathia there is fusion between the maxilla and the mandible, which may be in the midline or laterally placed and may be unilateral or bilateral.

Popliteal pterygium and van der Woude syndrome
Cleft palate
Mandibular hypoplasia
Absent or abnormal tongue morphology
Hemifacial microsomia
Congenital amputation of arms and legs
Glossopalatine ankylosis
Horner s syndrome
Coloboma
Scoliosis
Oblique facial clefts

Table 1*: Other anomalies associated with syngnathia.*

Regarding management protocol, no proven methodology was described in the literature because of variable presentation. Treatment may be problematic in complex cases [5]. The author reports a new case of unilateral congenital syngnathia, who achieved successful release after simple incision in the fusion site.

Case Report

A 3 days old female baby presented with inability to open the mouth since birth was brought to the paediatric department of Sher-e-Bangla Medical College and Hospital, Barisal on 21 July 2017. The baby was associated with respiratory distress and difficulty in feeding problems. She was transferred to pediatric ICU immediately to overcome respiratory distress. Her nutrition was maintained by naso gastric tube feeding. The child care unit refers the baby to our department of Oral and Maxillofacial surgery for better opinion. According to her antenatal and obstetric history her mother gives birth to a twin baby, interestingly no congenital anomalies were detected in the next baby. No other records were available about the mother's history of past illness. On physical examination the syngnathia baby was associated with restricted mouth opening. After retracting the lip saliva was discharged in the right side. We used small plastic spatula for checking the cause and extension of fusion in between the arches. There was separation of arches in the right side and complete unilateral fusion of maxilla and mandible on the left side. The posterior portion of the maxilla and mandible, palate and tongue could not be assessed preoperatively due to restricted mouth opening. No other anomalies were detected on general examination. The baby seems to attempt mouth opening and crying, but, she failed.

The lateral view of skull with mandible revealed unilateral alveolar bony fusion of maxilla and mandible in left side and under developed mandible in other site (Figure). After 3 days of clinical examination the baby was developed with neonatal jaundice. Her bilirubin level was 18.5 mg/dl; her body wt was 2.5 kg, other blood picture was within normal limit. She was then treated by photo therapy for 2 days to correct bilirubin level. Early surgery was decided to aid the baby for breast feeding as well as to overcome respiratory difficulties. After normalization of the baby's physical condition 50 ml whole blood transfusion was done to improve immunity. After three days the baby was undergone surgery done under nitrous oxide/oxygen sedation. The fusion site was released by simple incision to create a gap of 10 mm. Dressing gauze soaked in neobacrin ointment was interposed with sialastic sheet at the gap for 3 hours. Then the baby was sent to her mother for breast feeding. After 5 days of post-operative period she was released with good physical condition. The parent was advised for long follow-up with the baby. But, the patient did not return for further reporting within last 10 month.

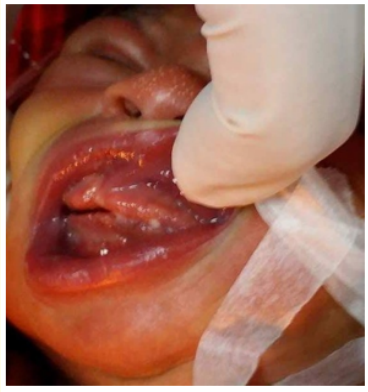

Figure 1: *Pre-operative view.*

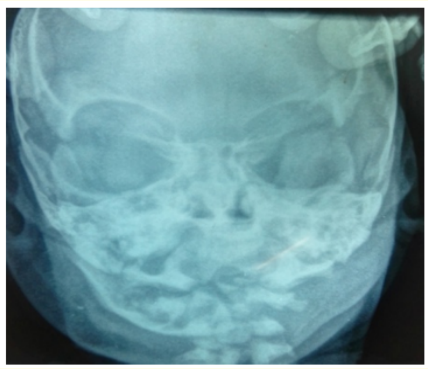

Figure 2a: *Plain X-ray Skull with mandible [P/A View].*

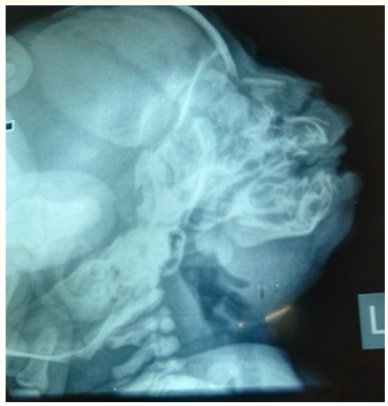

Figure 2b: *X-ray lateral view of skull.*

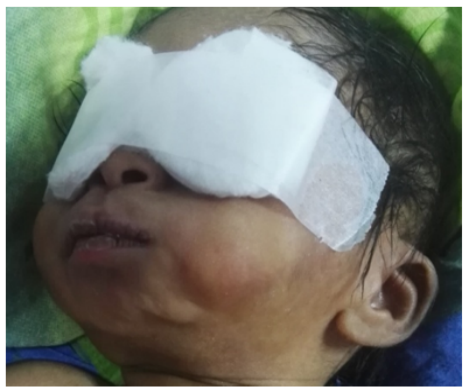

Figure 3a: *Post-operative view.*

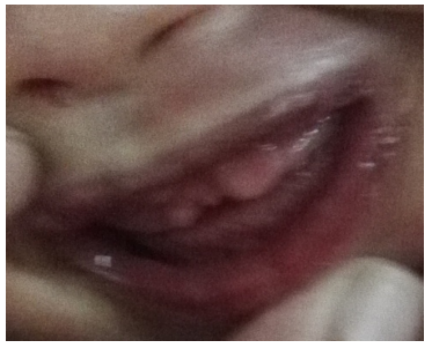

Figure 3b: *Post-operative view.*

Treatment protocol

The need for surgery and the timing of surgical treatment in this patient depends on their general condition at the time of presentation. Due to inability to mouth opening these patient are often being given total per enteral nutrition or fed by naso gastric tube. Study have shown that children less than 6 month old are more prone to critical events during anesthesia, such as cardiac arrest and air way problems [6]. A literature review shows that only a few patient have undergone surgery on an emergency basis [7].

Conventional radiograph helps to determining the site and types of fusion in limited cases, but, CT scan gives a clear picture of fusion site and aids in planning surgery.

Surgical exposure depends on the site of fusion. Intra oral or extra oral incision has been used. In unilateral fusion without TMJ involvement intra oral incision is followed. Extra oral (pre auricular) incision preferred for fusion related to TMJ region. Some author prefers sub mandibular incision in delayed surgical cases. Late surgical treatment provides better result, because patient can be easily manipulated but, it can result in TMJ ankylosis, deficiency in jaw growth and facial deformities [8]. Literature review shows that the frequency of recurrence is higher in syndromal rather than non syndromal cases. The goal of surgery is to relieve fusion and prevent recurrence. So, a careful surgical technique and subsequent meticulous attention to long term physiotherapy are considered to achieving a satisfactory result. In addition, a long follow-up should be considered to prevent recurrence of bony fusion and to allow further evaluation of facial growth patterns [1].

Discussion

Syngnathia with resultant inability to open the mouth is extremely uncommon. This condition occurs in association with other Oral and Maxillofacial abnormalities including various syndromes (Table 1). According to literature review congenital syngnathia can be unilateral, bilateral with an anterior slit or complete. A modified classification for bony syngnathia was Suggested by Laster., *et al.* [2] as follows:

- Type 1a, simple anterior syngnathia- characterized by bony fusion of the alveolar ridge only and without other congenital deformity in the head and neck.
- Type 1b, complex anterior syngnathia- characterized by bony fusion of the alveolar ridge only and associated with other congenital deformity in the head and neck.
- Type 2a, simple zygomatico-mandibular syngnathia-characterized by bony fusion of the mandible to the zygomatic complex causing only mandibular micrognathia.
- Type 2b, complex zygomatico-mandibular syngnathia-characterized by bony fusion of the mandible to the zygomatic complex and associated with cleft or TMJ ankylosis.

The case presented in this report is classified as type 1a according to Laster., *et al.* [2] as there was bony fusion of alveolar ridge without any other anomalies. There was alveolar fusion only in left side which restrict mouth opening, difficulty in feeding, swallowing and respiration. The aetiology of Syngnathia is unknown. Goodacre and Wallace (1990) proposed persistence of the buccopharyngeal membrane or amniotic constriction bands in the region of the developing branchial arches as the cause. Dawson., *et al.* [9] reported there was no evidence of familial influence, environmental insult or consanguinity to support this condition. The case reported here is not associated with any other abnormalities and the mother had no history of drugs, toxin or trauma during pregnancy. Both condyles of the case were normal without ankylosis in the OPG X-ray. It is challenging to keeping such an infant stable for CT scan, but the diagnosis can be confirmed by CT scan (if possible), because the later yields more information about the site and type of fusion condition of the TMJ, as well as any dysmorphia of the facial skeleton [9]. The problems associated with syngnathia include issues with maintenance and protection of the air way, feeding difficulties and problems with anaesthesia management [1]. The management of congenital fusion when associated with multiple malformations needs a team approach. In late cases the baby may need tracheostomy to manage respiratory obstruction. Early surgical division of the occlusion is recommended for normal feeding and avoidance of upper airway obstruction and to allow for normal mandibular function and growth. Early intervention also reducing the possibilities of TMJ ankylosis.

Conclusion

Unilateral maxillo-mandibular fusion is a rare disorder. This report adds more information to the literature. Prompt, definitive diagnosis and management of this type patient may help in early breast feeding and prevent permanent deformity. The parent should be aware of the problems and motivated for physiotherapy and follow-up appointment at regular interval.

Conflict of Interest

None declared.

Ethical Approval

Not mentioned.

Acknowledgment

I like to appreciate my senior colleagues and teachers of the department of paediatrics and paediatric surgery of sher-e-bangla medical college and hospital for their nice cooperation regarding preparation of this case report.

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Volume 18 Issue 3 March 2019