Case Report

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An isolated case of maxillomandibular syngnathia in a new born

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Abstract:

Congenital maxillomandibular syngnathia is a rare disorder. It is usually seen in association with various syndromes such as Van der Woude, popliteal pterygium syndrome, or in association with other defects. Isolated case reports of bony fusion of jaws are sparse. We report an isolated case of congenital fusion of the mandible and maxilla in a newborn baby and describe the clinical and radiological features of this anomaly. A brief review of literature is also presented.

Keywords:

Congenital maxillomandibular fusion, new born congenital diseases, syngnathia

Introduction

Congenital maxillomandibular fusion is a rare disorder which was first reported by Barker in 1936. Thirty-four cases have been reported in the literature since then, the last case was reported by Hegab *et al*. Congenital fusion of jaws presents as the fusion of soft tissues (synechiae) or bone (synostosis).[1-3] The former is more common than the later.[2] Syngnathia is a fusion between the maxilla and the mandible, which may be in the midline or laterally placed and may be unilateral or bilateral. Anomalies associated with syngnathia are 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, and oblique facial clefts.[1-3]

It is usually diagnosed immediately after birth because of the baby's inability to open the mouth or feed normally.[2,4] Associated problems include difficulty in maintaining airway patency, anesthesia management, and retardation of facial growth and development.[1] The management of children with this condition is not clear. No proven methodology is described in the literature because of variable presentation and also because some patients have survived without treatment.[3]

We present a case report of bony fusion of maxillary and mandibular alveolar processes in a newborn baby.

Case Report

A newborn baby with an inability to open the mouth was referred. The baby was the eighth child of parents. None of the previous children survived. There was no history of consanguinity between the parents. The mother gave a history of drug ingestion of unknown composition during pregnancy. On clinical examination, the head circumference and shape of the eyes, ears, and nose were normal. The lips were protuberant. The maxilla and mandible appeared to be relatively small. Complete fusion of the upper and lower alveoli was present with the absence of oral aperture. Due to the lack of mouth opening, examination of the tongue and palate was not possible. The rest of the baby's body was normal [Figures 1 and 2]. The computed

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tomography examination revealed relatively small maxilla and mandible. The ascending and horizontal rami of the mandible were fused to maxillary tuberosity on either side. The zygoma appears normal. The tongue was seen at the posterior aspect of the oral cavity due to fused bones. There were no other calvarial, facial, and spinal abnormalities [Figures 3-5]. All these findings were suggestive of congenital maxillomandibular syngnathia.

The parents were informed about the baby's condition, but they were unwilling to let the baby undergo treatment. A nasogastric tube was advanced for the purpose of feeding. Unfortunately, the baby died on the 3rd day of birth. The exact cause of death was unknown. Severe respiratory distress could be a possible cause.

Discussion

Congenital bony fusion of the maxilla and mandible is a rare condition. This condition is hardly ever seen in isolation and is usually associated with syndromes such as Van der Woude, popliteal pterygium syndrome, or in association with other defects or anomalies.[1,2] The exact etiology is unknown. Many theories have been proposed to explain congenital fusion of jaws such as abnormality of the stapedial artery, the early loss of neural crest cells, teratogenic agents, trauma, persistence of buccopharyngeal membrane, and amniotic constriction bands in the developing regions of the first branchial arch.[1-4]

Burket *et al*. 1936 first reported a case of true congenital bony temporomandibular joint (TMJ) ankylosis with the fusion of gums and associated facial hemiatrophy as well as Horner's syndrome. In a literature review, Laster *et al*. reported 24 cases since 1936 and Daniels in 2004 reported a case; thus, there are a total of 26 cases in the literature.[4]

Isolated cases of congenital maxillomandibular syngnathia have been reviewed from 2009 to 2019 [Table 1].

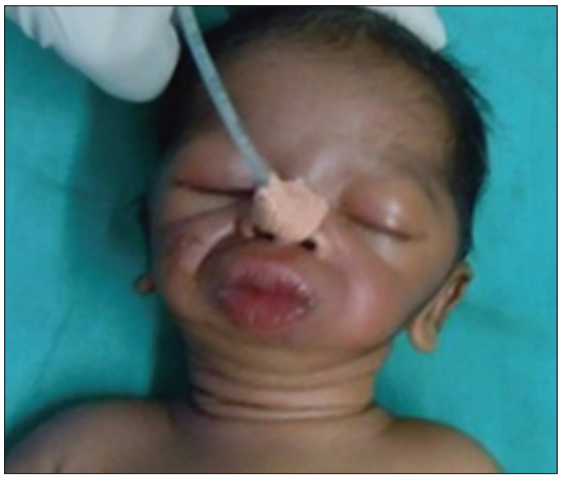

Figure 1: Frontal view of the baby with a nasogastric tube *in situ*

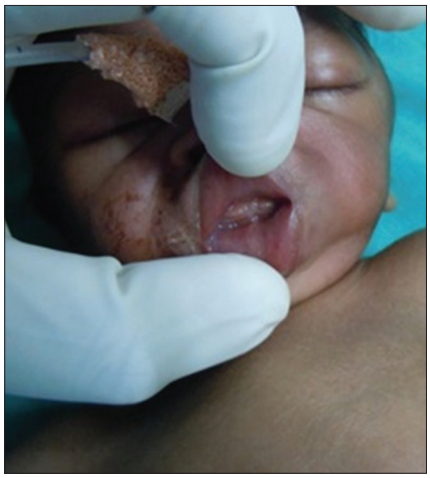

Figure 2: Intraoral view showing the fusion of the upper and lower alveoli

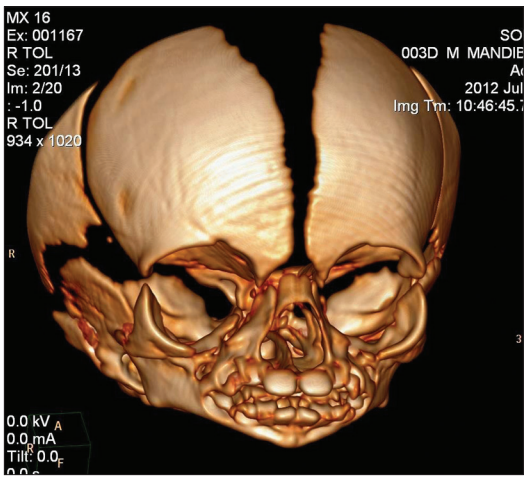

Figure 3: Three-dimensional-computed tomography image showing bilateral maxillomandibular fusion

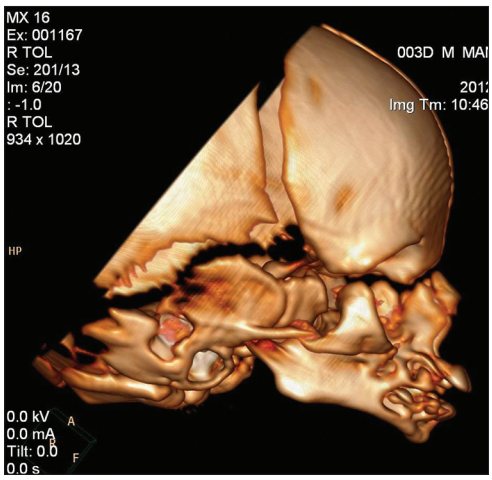

Figure 4: Three-dimensional-computed tomography image showing the fusion of the posterior aspects of the maxilla and mandible on the right side

Dawson *et al*. classified syngnathia as follows: Type 1: simple syngnathia with no other congenital anomalies in the head and neck; Type 2: complex syngnathia with subgroups, Type 2a: syngnathia co-existent with aglossia, and Type 2b: syngnathia coexistent with the agenesis or hypoplasia of the proximal mandible. Later, Laster *et al*. modified classification as Type 1a: simple anterior syngnathia characterized by the bony fusion of the alveolar ridges only and without other congenital deformities in the head and neck; Type 1b: complex anterior syngnathia characterized by the bony fusion of the alveolar ridges only and associated with other congenital deformity in the head and neck; Type 2a: simple zygomatico-mandibular syngnathia characterized by the bony fusion of the mandible to the zygomatic complex causing only mandibular micrognathia; and Type 2b: complex zygomatico-mandibular syngnathia characterized by the bony fusion of the mandible to the zygomatic complex and associated with clefts or TMJ ankylosis.[2,4]

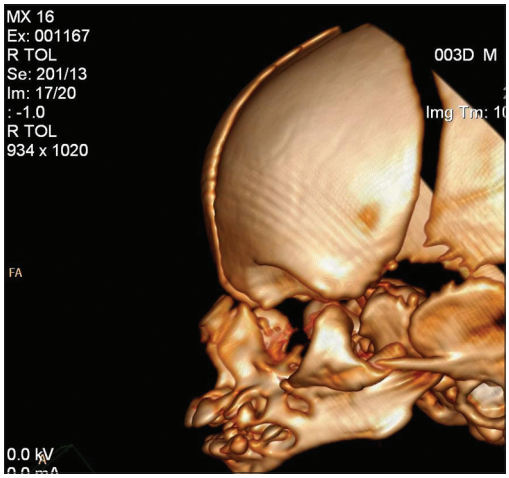

Figure 5: Three-dimensional-computed tomography image showing the fusion of the posterior aspects of the maxilla and mandible on the left sidethe posterior aspects of the maxilla and mandible on the left side

The patient presented here had a bony fusion of alveolar ridges only without other congenital deformities. Therefore, it belongs to the Type 1a category of Laster classification.

This condition is usually diagnosed at the time of birth. This unanticipated diagnosis can potentially result in major complications in airway management and feeding

Table 1: Patients reported with isolated congenital maxillomandibular syngnathia in the literature

Authors	Clinical features	Associated deformities	Age and gender
Alam[5]	Unilateral fusion of the maxilla and mandible on the left side		3 days/girl
Nagaprasad *et al*.[6]	Unilateral fusion of the maxilla and mandible on the left side		4 months/girl
Khasigwala *et al*.[7]	Bilateral fusion of the maxilla and mandible	Tracheoesophageal fistula	2 days/boy
Raban *et al*.[8]	Bilateral fusion of the maxilla and mandible	Microcephaly (<3rd centile), dense bilateral cataracts, low anterior hairline, low set and posteriorly rotated ears, short palpebral fissures, bilateral syndactyly of toes 2-4, and a hypoplastic prepuce of the genitalia	Premature baby/ boy
Shao and Yang[9]	Unilateral bony fusion between maxillary tuberosity and ascending rami of the mandible	Polydactyly of the left hand	42 days/girl
Rattan[10]	Bilateral bony fusion of the maxilla to the mandible in its posterior aspect		8 years/boy
Hegab *et al*.[1]	1. Bilateral bony fusion of maxilla, zygomatic bone to horizontal and ascending rami of the mandible 2. Right side fusion of the maxilla and mandible 3. Right side fusion of maxilla, zygomatic bone to mandible	Retrognathia	4 months/girl 3 months/boy 3 years/boy
Fallahi *et al*.[2]	Unilateral right ascending rami and alveolar arch of the mandible fused to the upper alveolar arch and zygoma	Ear lobe deformity, small skin pits anterior to tragus, abnormal lateral canthal fissure	6 months/girl
Subramanian *et al*.[3]	1. Bilateral fusion of the maxilla and mandible on the posterior aspect 2. Fusion of the maxilla and mandible on the right side 3. Lateral side of the maxilla and mandible on both sides were in close contact with each other 4. Bony fusion at the midline between the maxilla and mandible	Bilateral TMJ fusion Underdeveloped mandible, 6 digits in the forearm Reductive limb deformity	9 months/girl 3 months/boy 2 years/boy 2 years/boy
Halli *et al*.[11]	Unilateral fusion of the maxilla and mandible on the left side	1. Hemifacial hypoplasia of the left side 2. Anti-mongoloid slant of the medial palpebral fissure of the left eye with a classical bird face deformity	8 years/boy

TMJ=Temporomandibular joint

at birth. Thus, some studies have shown that prenatal diagnosis of this condition would allow appropriate prenatal planning to manage any complications at the time of delivery. Laster *et al*. first reported a prenatal sonographic image depicting the absence of mouth opening and contiguous appearance of the upper and lower jaws in case of maxillomandibular fusion. The general condition of the baby plays a major role in planning and management. The need for an emergency intervention occurs only when the babies show difficulty thriving. The syngnathia cases have been operated as early as the 2nd day of life to 4 years, and some patients have survived without surgery.[3] Early surgical intervention is required to prevent airway obstruction, for normal feeding, and to allow normal mandibular function and growth.[1,5] Although the surgery is simple, general anesthesia is often difficult. Arrangements to manage airway obstruction or respiratory distress should be made prior to the surgery such as open crib, equipment for an airway, tracheostomy, resuscitation, and mechanical ventilation. Fine fiber-optic laryngoscopes are required for naso-endotracheal intubation. Excessive force may easily lead to jaw fracture and should be avoided because TMJs are stiffened due to disuse, the alveolar ridges are soft, and infantile bone is fragile.

Unfortunately, the patient presented here did not undergo surgery due to the unwillingness of parents for surgical intervention even after proper counseling.

Conclusion

Congenital maxillomandibular syngnathia is a rare condition, and only 44 cases have been reported previously in the literature. We conclude that early diagnosis and prompt treatment would require to prevent fatality which otherwise results as in our case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Hegab A, ElMadawy A, Shawkat WM. Congenital maxillomandibular fusion: A report of three cases. Int J Oral Maxillofac Surg 2012;41:1248-52.
2. Fallahi HR, Naeini M, Mahmoudi M, Javaherforoosh F. Congenital zygomatico-maxillo-mandibular fusion: A brief case report and review of literature. Int J Oral Maxillofac Surg 2010;39:930-3.
3. Subramanian B, Agrawal K, Panda K. Congenital fusion of the jaws: A management protocol. Int J Oral Maxillofac Surg 2010;39:925-9.
4. Daniels JS. Congenital maxillomandibular fusion: A case report and review of the literature. J Craniomaxillofac Surg 2004;32:135-9.
5. Alam MD. Congenital unilateral maxillo-mandibular syngnathia: A brief case report and review of literature. EC Dental Science 18.3 (2019):479-84.
6. Nagaprasad N, Qureshi MR, Qureshi VF, Qureshi MF, Farhan H, Qureshi S. Rare occurrence of two cases of congenital syngnathia at Osmania general hospital, Hyderabad, India: A Case Report. Biomed J Sci Tech Res 2018;7:5773-5.
7. Khasgiwala A, Jangam S, Sharma S, Newaskar V. Congenital bilateral syngnathia and tracheoesophageal fistula: A rare presentation. Contemp Clin Dent 2016;7:566-8.
8. Raban MS, Muller SJ, Harrison MC. An unusual case of congenital syngnathia. J Neonatal Biol 2015;4:1-3.
9. Shao X, Yang G. Congenital unilateral maxillomandibular fusion. Br J Oral Maxillofac Surg 2014;52:e20.
10. Rattan V. Congenital maxillomandibular fusion: Report of three cases. Int J Oral Maxillofac Surg 2013;42:546-7.
11. Halli R, Kharkar V, Kini Y, Rudagi BM. Congenital unilateral maxillo-mandibulo-zygomatic fusion (syngnathia): A case report in an 8-year-old boy. Int J Oral Maxillofac Surg 2010;39:500-2.