Congenital bilateral syngnathia and tracheoesophageal fistula: A rare presentation

Ankit Khasgiwala, Sagar Jangam[1], Shashi Sharma[2], Vilas Newaskar[3]

Abstract

Congenital syngnathia (CS), first reported by Burket in 1936, is a rare condition, with <50 reported cases and is associated with other conditions and syndromes. CS restricts mouth opening, causing difficulty in feeding, swallowing, and respiration. This report puts forth the clinical findings and management of this challenging condition in association with tracheoesophageal fistula in a neonate.

Keywords: Aspiration pneumonitis, congenital, syngnathia, tracheoesophageal fistula

Introduction

Congenital syngnathia (CS) is an unusual condition characterized by fusion of the jaw with inability to open the mouth at birth. Fusion can be unilateral or bilateral or complete and range from simple mucosal bands (synechiae) to complete bony fusion (synostosis).[1] We present a case of a 2-day-old neonate with fusion of the maxillary and mandibular alveolar complex (Laster's Type 1b congenital maxillomandibular syngnathia) along with tracheoesophageal fistula (TEF) probably the first of its kind.

Case Report

A 2-day-old male neonate, preterm (36 weeks), normal delivery, weighing 2.0 kg, presented with a history of severely restricted mouth opening and frothing from mouth and nose and abdominal distention. The mother was healthy without any history of illness, trauma, or drug exposure. There was no history of consanguinity. The baby was the first child, and there was no incidence of similar kind of anomalies in close family members. Routine blood investigations were within normal limits.

The child had a slit-like space of 5–6 mm between the upper and lower arches in the region of the central incisors with rest of the alveolar areas showing fusion preventing detailed intraoral examination [Figure 1]. Radiological examination showed fusion of the upper and lower alveolar arches [Figure 2].

The patient was being given parenteral nutrition in view of reduced mouth opening and inability to pass infant feeding tube. The child had signs of aspiration pneumonitis (rattling respiration, cough, and cyanosis) and was rushed to emergency or to release the syngnathia under general anesthesia. A fiberoptic intubation through nasal route was performed using 2.2 mm ultrathin fiberoptic bronchoscope permitting passage of a 3.0 mm endotracheal tube (ETT). Bronchoscopy revealed a TEF Type C (Gross).[2] The position of the ETT was verified and fixed subsequently. This finding correlated with clinical observation of frothing from mouth and nose at birth.

Syngnathia was released transorally; fusion on the left side was bony which was released by osteotome and mallet whereas scalpel blade was used to release the soft-tissue

Department of Plastic and Reconstructive Surgery, Amandeep Hospital, Amritsar, Punjab, [1]Department of Oral and Maxillofacial Surgery, Government Dental College, Aurangabad, Maharashtra, [2]Department of Surgery, MGM Medical College and M Y Hospital, [3]Department of Oral and Maxillofacial Surgery, Government College of Dentistry, Indore, Madhya Pradesh, India

Correspondence: Dr. Ankit Khasgiwala,
Department of Plastic and Reconstructive Surgery,
Amandeep Hospital, Amritsar, Punjab, India.
E-mail: ankitkhasgiwala@yahoo.in

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fusion on the right side. Mouth opening achieved after the release was 25 mm [Figure 3]. Hemostasis was attained, and a collagen membrane dressing was given to cover the raw area. Tongue, palate, and floor of the mouth were found to be anatomically normal after the release. No attempt at repair of TEF repair was done in this surgical appointment which was subsequently repaired after 48 h under general anesthesia. Unfortunately, the child developed septicemia likely due to previous aspiration pneumonitis and succumbed after 7 days.

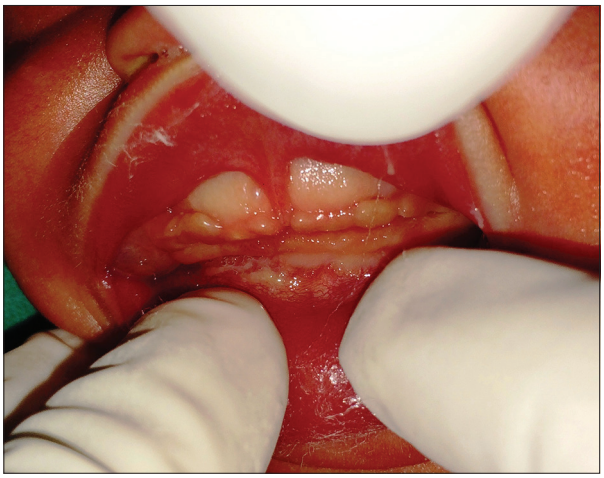

Figure 1: Preoperative frontal view

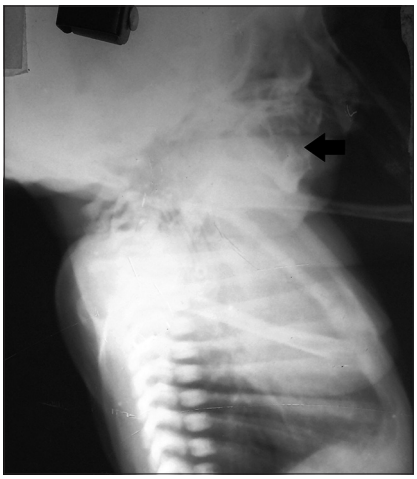

Figure 2: X-ray showing fusion of the maxillary and mandibular arches

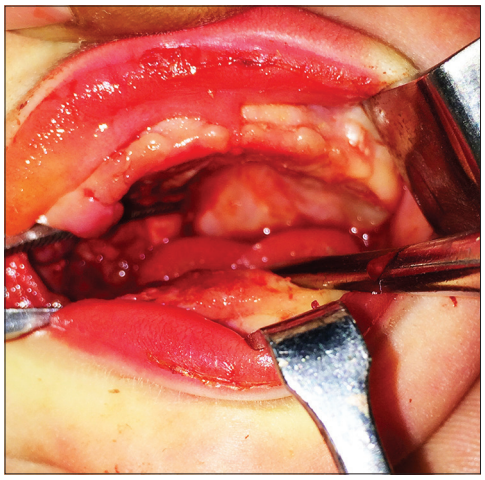

Figure 3: Mouth opening achieved postoperatively (2.5 cm)

Discussion

CS is a rare anomaly which presents as fusion of the jaws with inability to open the mouth at the time of birth. It can have varied presentations in maxillofacial complex ranging from simple fibrous union of alveolar ridges to bony fusion involving maxilla, mandible, temporomandibular joint, and even zygoma.

Laster *et al*. have classified CS based on site and extent of fusion and its association with other craniofacial anomalies [Table 1].[3,4] The case discussed previously had fusion of the upper and lower alveolus without any craniofacial anomaly, i.e., cleft, temporomandibular joint (TMJ) ankylosis, or other hence was classified under Laster's Type 1b congenital syngnathia.[3,4] However, he had neck abnormality, i.e., cTEF. In our literature search (PubMed Central® (PMC), and Google Scholar), we did not come across any case of CS associated with cTEF making this case a rare entity.

Various theories have been proposed to explain the etiology of this anomaly. These include:

- Persistence of buccopharyngeal membrane (Gorlin and Goldman)[5]
- Oropharyngeal atresia secondary to persistence of buccopharyngeal membrane (Longacre)[6]

Table 1: Laster's classification of congenital syngnathia

Type	Category	Clinical features
Type 1a	Simple anterior syngnathia	Bony fusion of alveolar ridge without other congenital deformity in the head and neck
Type 1b	Complex anterior syngnathia	Bony fusion of the alveolar ridges associated with other congenital deformity in the head and neck
Type 2a	Simple zygomatico-mandibular syngnathia	Bony fusion of the mandible to the zygomatic complex causing only mandibular micrognathia
Type 2b	Complex zygomatico-mandibular syngnathia	Bony fusion of the mandible to the zygomatic complex and associated with clefts or TMJ ankylosis

TMJ: Temporomandibular joint

- Amniotic constriction bands which lead to pressure in the region of the developing first branchial arch (Burket)[7]
- Increasing period of contact between the alveoli due to the small jaw and decreased swallowing reflex (Walker and Frase, Humphrey)[8,9]
- Antenatal exposure to drugs such as meclizine, large dose Vitamin A, which cause edema of the embryo and upsets skeletal growth (King, Kalter and Warkaney).[10-12]

The differential diagnosis includes congenital TMJ ankylosis and glossopalatine ankylosis. Therefore, a careful clinical and radiological examination forms an essential part in diagnosing CS.

Treatment of congenital intermaxillary syngnathia is surgical release under general or local anesthesia depending on the site and extent of fusion. Intubation is facilitated by pediatric fiberoptic bronchoscope or elective tracheostomy. Blind nasal intubation is not preferred because of the risk of laryngeal edema and its sequelae.[13] Alveolar fusion cases can be surgically approached through transoral route. However, fusion involving TMJ and zygomatic complex region requires extraoral approach. The use of interalveolar soft wedges has been recommended as a part of jaw physiotherapy to prevent the recurrence of the condition. Parental counseling and periodic recall to assess the craniofacial growth and possible recurrences form an essential part in the management of such anomaly.

Conclusion

A thorough literature search revealed that this is the first case of CS associated with TEF. There is no established treatment protocol for such anomaly considering the relative rarity of the condition. The management team should be aware of the wide range of severity of syngnathia along with other possible congenital deformities and adopt a treatment plan according to the situation and general condition of the patient. Furthermore, the parents must be counseled and informed about the surgical outcomes and associated comorbidities. Very little information is available on the long-term outcome of these patients.

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Conflicts of interest

There are no conflicts of interest.

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