Case Report

Congenital Syngnathia in Bangladesh: A Very Rare Case Report

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Abstract

Syngnathia is a rare anomaly involving fusion between the maxilla and mandible. The fusion may be due to soft tissue adhesion between the two or true bony fusion between maxilla and mandible. This condition ranges from only soft tissue fusion between gums known as synechia to bony union known as synostosis with different grades of fusion. It was first reported in 1936, with only 41 cases reported till 2009.

In this case a 11 years old girl reported to the maxillofacial department of Dhaka Dental College Hospital, with the complaint of inability to open the mouth and as a diagnosed case of alveolar bone fusion of mandible with palatal alveolar bone in the left side & bone fusion of alveolar process of mandibular angle region with maxilla in the right side. She was also bone fusion of the coronoid process with zygoma in the right side. Immediate after birth she had a history of synechia (Bilateral) & it was correct by making stoma by maxillofacial surgeon. The patient was subjected to a surgical procedure, after successful operation she enabled to open her mouth up to 35 mm.

Key Words: Synechia, Bony Maxillomandibular Fusion, Syngnathia

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Introduction

Congenital maxillomandibular fusion is a rare disorder. Maxillomandibular fusion is usually discovered immediately after birth, because the baby is unable to open his/her mouth or feed normally[1]. The fusion defects can present with a wide range of severities, ranging from a single mucosal band (synechiae) to complete bony fusion (syngnathia). It was first reported in 1936, with only 41 cases reported till 2009[2].

Cases were represented by either unilateral fusion between the mandible and maxilla[3-4],which is usually associated with other congenital anomalies such as those involving the central nervous system, which lead to mental retardation[4]; the cardiac system, which manifest as patent ductus arteriosus[4] and tetralogy of Fallot[5]; or the skeletal system, which include

hemivertebrae and fused ribs feet syndactyly, and hand brachydactyly. Cases may also have associated anomalies affecting the endocrine systems, such as type I diabetes mellitus and hypothyroidism, and the reproductive systems, which manifest as micropenis or hypospadias.

In cases involving bilateral fusion between both jaws with varying degrees of severity, anomalies in structures other than the craniofacial structures are rarely observed.

In 1997, Dawson et al[4]. classified syngnathia into the following types: type 1 (simple syngnathia), bony fusion between mandible and maxilla or the zygoma in the absence of other congenital anomalies in the head and neck; and type 2 (complex syngnathia), syngnathia occurring with other congenital anomalies in the head and neck. Type 2 syngnathia was further classified as type 2a syngnathia (syngnathia with aglossia) and type 2b syngnathia (syngnathia with agenesis or hypoplasia of the proximal mandible).

This report presents a case of bilateral maxillomandibular fusion in a 11 years old girl showing no other associated abnormalities. The patient was subjected to a surgical procedure, after successful operation she enabled to open her mouth up to 35 mm.

Case report

An 11 years girl was admitted at the Maxillofacial Surgery Department of Dhaka Dental College Hospital, Dhaka, being diagnosed with bilateral jaw fusion at 23rd September 2013. This patient has a history of synechia immediate after birth. And the problem was corrected immediate after birth by making stoma at Dhaka Dental College Hospital in 2003. Now she taking liquid diet for last 11 years. The maximum mouth opening was 0 mm in the symphysis region and any mandibular movement in all direction was impossible. The opening of mouth gradually decreased and become restricted for last two months. With this complaint she was admitted in Dhaka Dental College Hospital for better management.

The patient was born after a full-term pregnancy from related parents who were cousins. The patient family history did not reveal any similar condition, but one of her younger sister has the same problem. The patient had fused posterior upper and lower gums, small oral aperture, prominent nose, marked micrognathia.

Initial clinical examination revealed bony fusion of the right and left mandible, maxilla, and gums, with no interincisal opening. Additional abnormalities included micrognathia, retrognathia, apparently small head, and long neck (Figure 1).

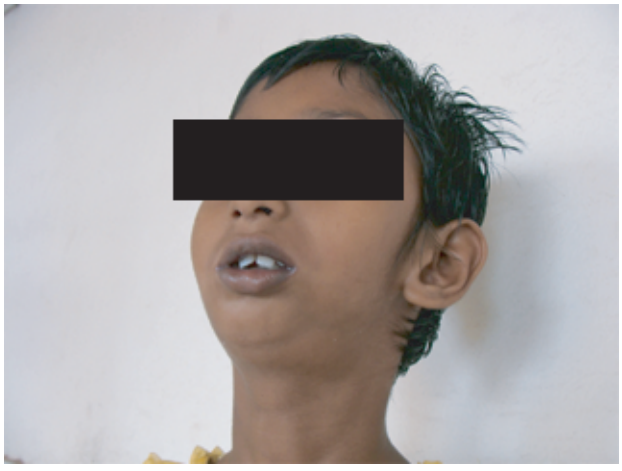

Figure 1: Preoperative: Showing restricted mouth opening with no interincisal opening.

Cranial, axial, and coronal computed tomography (CT), as well as a 3-dimensional CT scan, was performed, revealing bilateral fusion between the mandible and maxilla (Figure 2).

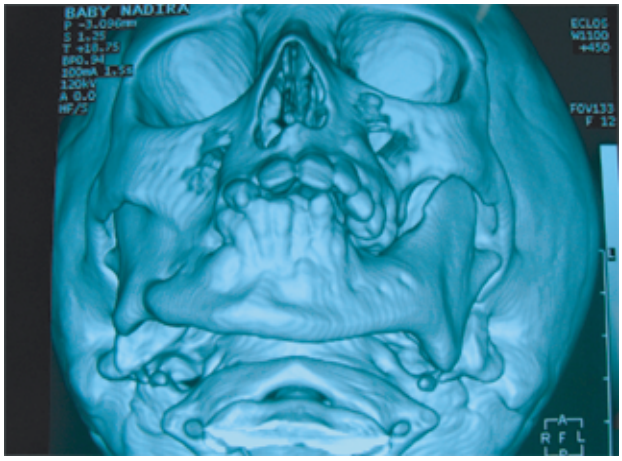

Figure 2: Bilateral Fusion of Maxilla & Mandible

CT scan revealed the exact fusion between the maxilla and ramus of the mandible, with the developing teeth crypts extending in the fusion area, also showing the normal condyles free from any fusion or ankylosis with the surrounding structures. The mandibular rami were completely fused to the maxillary tuberosity & zygoma in the right side.

Surgical Procedure

The patient was brought to the operation theater and seated in supine position, no attempted of blind awake nasal intubation was taken. Tracheostomy was performed and anaesthesia was maintained with nitrous oxide & oxygen. After intubation, the patient was prepped and draped in the usual sterile manner. Local anaesthesia lidocaine with epinephrine was injected for infiltration in the mandible and in the field area.

Through a left and right submandibular incision the mandible were exposed and the coronoid process revealed the mandibular rami were fused to the maxillary tuberosity & zygomatic area in (Rt) side & alveolar bone of mandible with palatal alveolar bone of maxilla on left side, without TMJ ankylosis. Using electric hand piece & TC bar all bony fusion are separated. The mouth opening was restricted, and then bilateral condylectomy & coronoidectomy was done and mouth was opened. The soft tissue fusion in the alveolar area was separated using diathermy.

A 3.5-cm mouth opening was achieved. The previously fused area was checked, and soft tissue resistance was observed posteriorly in the retromolar area and released by multiple mouth openings.

Suturing was completed using 3-0 vicryl sutures and 4-0 proline in a continuous and interrupted manner. The throat pack was removed; the patient recovered successfully from anesthesia and was brought to the recovery room in a satisfactory condition.

The surgery was performed on the 30th November 2013 and the patient was discharged on the 5th postoperative day in a good condition with improved mouth opening.

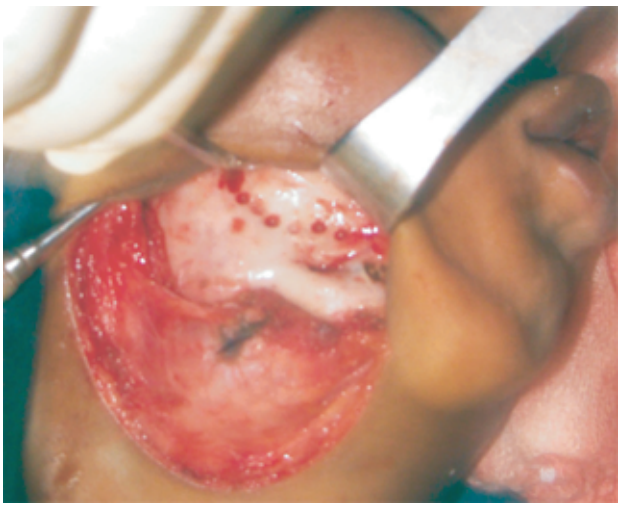

Figure 3: Land mark for separation of fusion between mandibular rami & zygoma.

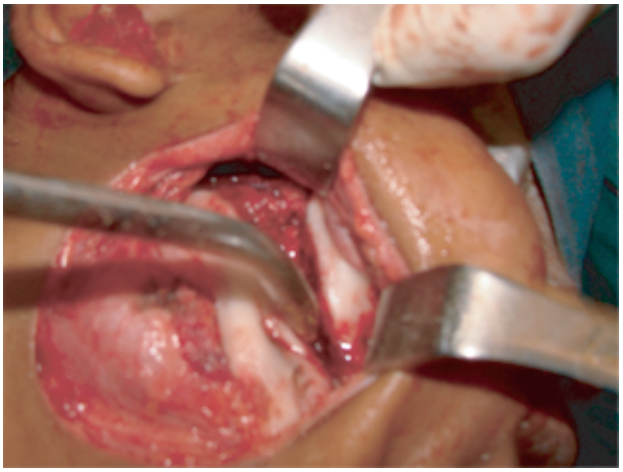

Figure 4: Separation of the fusion.

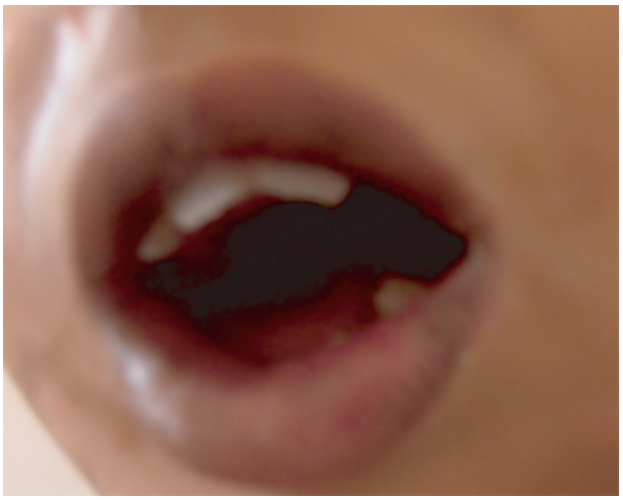

Figure 5: Postoperative View.

Discussion

According to previous studies, the pathogenesis of congenital maxillomandibular fusion remains unclear, although it can be related to a developmental defect in the region of the first branchial arch, where separation of the maxilla and mandible fails to occur during the 7th to 8th week of embryonic development[6]. Moreover, genetic, teratogenic, and mechanical insults contribute to this condition[7]. Trauma late in pregnancy, the presence of an abnormal stapedial artery, and teratogenic agents have been suggested to be causative factors[8]. Persistence of the buccopharyngeal membrane, amniotic constriction bands in the region of development of branchial arches, environmental insults with drugs like meclizine, and large doses of vitamin A have been postulated as other possible reasons[4].

A case of a human fetus with sygnathia without any clinical history and also a case of vitamin A induced bony syngnathia with a cleft of the secondary plate in rats, treated with vitamin A has also been reported. The condition is problematic and interface with feeding, breathing, general health of the patient (Aspiration, Pneumonitis), growth and development induction of anaesthesia (Intubation) etc.

The cause of this condition was not apparent from the parent's history, and consanguinity was the only apparent risk factor for development of this condition. However, neither the case reported by Daniels nor the 5 cases reported by Dawson et al.[4] mentioned any consanguinity of parents. According to the system proposed by Dawson et al.[4] our case could be classified as a case of type 1 syngnathia.

To treat a patient with congenital syngnathia effectively, it is important to secure the airway of the patient, ensure proper nutrition, and secure enough mouth opening with no recurrence pattern. Therefore, it is recommended to prevent any recurrence after surgery by using the proper operation, including maintaining the mouth opening to prevent ankylosis[2].

References

1. Daniels J. Congenital maxillomandibular fusion: a case report and review of the literature. J Cranio-Maxillofacial Surg 2004;32:135-139.
2. Kim CH, Kim MY. Congenital Syngnathia: A case report. J Korean Assoc Oral maxillofac Surg. 2012;38:171-6.
3. Traahold LM. Cleft lip and palate associated with alveolar synechia and filiform palpebral synechia. Plas Reconstr Surg. 1990;86:337-339.
4. Dawson KH, Gruss JS, Myall RW. Congenital bony syngnathia: a proposed classification. Cleft palate craniofac J. 1997;34:141-146.
5. Brown DM, Marsh JL. Agnathia and associated malformations: a case report. Cleft palate J. 1990;27:415-418.
6. Tanrikulu R, Erol B, Gorgun B, Ilhon O. Congenital alveolar synechiae as an isolated anomaly: a case report. J Clin Pediatr Dent. 2003;28:81-83.
7. Haydar SG, Tercon A, Uckan S, Gurakan B. Congenital gum synechiae as an isolated anomaly: a case report. J Clin Pediatr Dent. 2003;28:81-83.
8. Laster Z, Temkin D, Zarfin Y, Kushnir A. Complete bony fusion of the mandible to the zygomatic complex and maxillary tuberosity: case report and review. Int J oral Maxillofac Surg. 2001;30:75-79.