CASE REPORTS

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Congenital Maxillomandibular Fusion: A Rare Case of Isolated True Bony Syngnathia

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ABSTRACT

Objective: To present a rare case of congenital maxillomandibular fusion or syngnathia.

Methods:

Study Design: Case Report
Setting: Tertiary Public Teaching Hospital
Participant: One patient

Results: A 3-year-old girl with oral adhesion (syngnathia) caused by a mandibular to maxillary fibrous band with bony fusion underwent successful surgical division and release. Subsequent monitoring and serial oral dilations were performed post operatively, resulting in mouth opening of 24mm over a period of three months. Currently, the patient is able to tolerate a general liquid diet.

Conclusion: Congenital maxillomandibular fusion is a very rare condition with few cases reported. We hope this report contributes to its diagnosis and management in other children.

Keywords: *congenital maxillomandibular fusion, syngnathia*

From birth, the upper aerodigestive tract of neonates plays an important role in simultaneous nose-breathing, suckling and swallowing. It is crucial for this mechanism to function properly to ensure survival. Problems that interfere with this design, such as bilateral choanal atresia, may result in death, unless attended to emergently. This report a case that could have had similar consequences over a slightly longer course.

CASE REPORT

A newborn girl from Nueva Ecija, Philippines was noted to have maxillo-mandibular fusion after full-term spontaneous home delivery to a 31-year-old G3P3 (3003) mother attended by a midwife. The mother was a non-smoker and non-alcoholic beverage drinker with irregular prenatal check-ups but had multivitamins and ferrous sulfate during pregnancy. She took unrecalled medications for an upper respiratory tract infection at 1 to 2 months of pregnancy and denied exposure to viral exanthems, chemicals, radiation or teratogenic drugs.

On examination, there was complete bilateral fusion of the bony upper and lower jaws. A nasogastric tube was inserted through the right nasal cavity. The rest of the physical examination findings were normal. Complete blood count revealed a white cell count of 18.45 with segmenter predominance. She was started on Ampicillin 60 mg/IV every 8 hours, Gentamicin 14 mg/IV

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OD, Vitamin K 0.5 mg/IM and Oxytetracycline eye ointment, and she eventually improved. The assessment was congenital maxillomandibular fusion or syngnathia. *(Figure 1)*

CT scan of the facial bones with 3D reconstruction showed complete bony fusion of the maxilla and the mandible with hypoplasia of the temporomandibular joints. *(Figure 2)* Radiographs showed no definite lung infiltrates, cardiothymic shadow within normal limits, intact diaphragm and costophrenic angles, gastrointestinal tract patterns within normal, no abnormal mass or calcifications, no pneumoperitoneum or organomegaly and no osseous, joint space or soft tissue abnormalities appreciated.

At two months of age, elective tracheostomy under local anesthesia was performed due to dyspneic episodes associated with desaturations exacerbated by recurrent upper respiratory tract infections. Anesthesia was maintained through the tracheostomy tube while a gastrostomy was carried out. She tolerated the procedure well and was eventually discharged, breathing spontaneously and well oxygenated. Several follow-up visits included a repeat CT scan of the facial bones with 3D reconstruction (including reference markers) at 1 year and 5 months of age, but she was not seen again until 3 years of age.

At 3 years and 6 months of age, she underwent surgical division of the bony fusion. An extended right pre-auricular incision and dissection of the superficial temporal fascia over root of zygoma were carried to the fascia and periosteum which were incised over the zygomatic root. The skin flap and parotid were retracted anteriorly and the coronoid process and zygomatico-maxillo-mandibular area were exposed. With maximum retraction to facilitate optimal exposure of a long tunnel medial to the masseter muscle, the mandibular fusion was released and the mucosal incision was gradually extended to the midline. The bony fusion was exposed by dissecting the mucosa away from the bone using a periosteal and freer elevator. Osteotomy of zygomatico-maxillo-mandibular fusion was achieved by tunneling from the zygomatic area to the midline by alternately using an osteotome, oscillating saw and high-speed surgical drill with different-sized cutting bur tips. The same procedure was replicated on the left side up to the midline until the division from the right side was reached. The bony division was then pried open until all fibrous bands were detached. Sharp edges were smoothened using a surgical drill with diamond bur tips. Avulsed central and lateral incisors were removed. The oral cavity was established and packed with medicated gauze impregnated with antibiotic ointment. The patient underwent serial oral dilation on succeeding follow-ups, eventually achieving mouth opening of 24mm within 3 months. *(Figures 3, 4)* Currently, the patient is able to tolerate a general liquid diet.

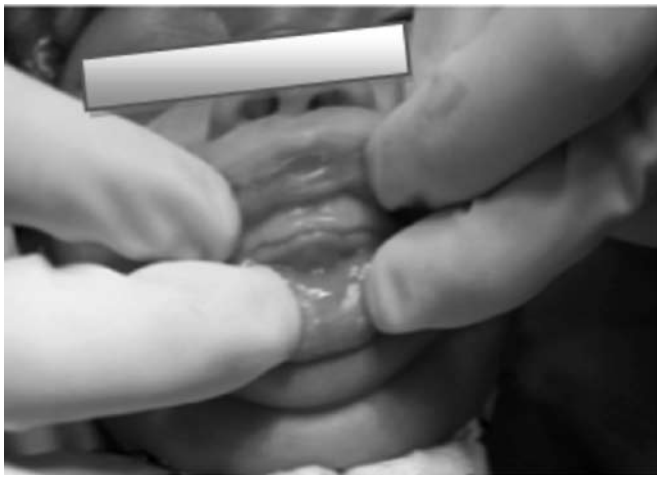

Figure 1. Complete bilateral fusion of the bony and soft-tissue components of the upper and lower jaws.

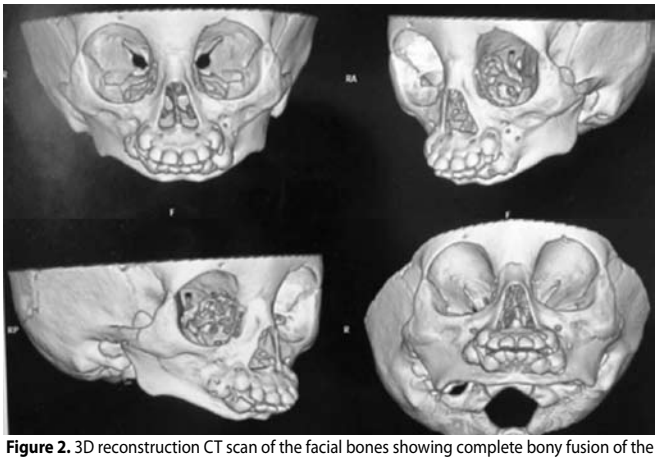

Figure 2. 3D reconstruction CT scan of the facial bones showing complete bony fusion of the maxilla and the mandible with hypoplasia of the temporomandibular joints.

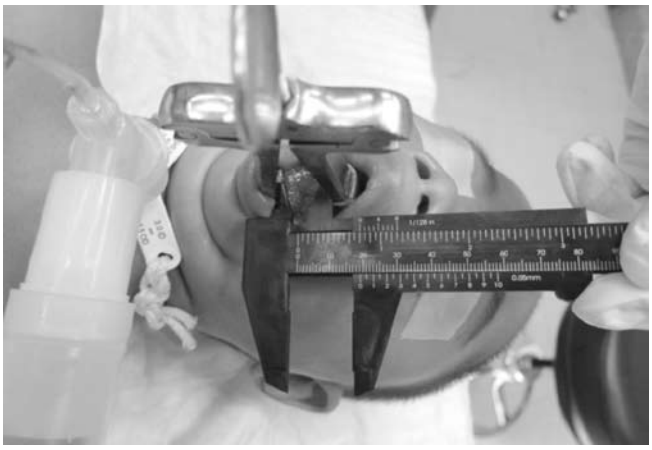

Figure 3. Oral dilation at 15mm (4 weeks postoperative)

Figure 4. Mouth opening of 22 mm (8 weeks postoperative)

DISCUSSION

Congenital causes of limited mouth opening involving fusion of the maxilla and mandible (syngnathia) are a rare group of anomalies. Cases are classified into those involving bony tissue and those involving soft tissue alone. Occurrence may be unilateral or bilateral, partial or complete.[1,2] Among the abovementioned classifications, cases involving solely soft tissue and bilateral fusion in the posterior region are more common.[2]

Isolated occurrence of bony syngnathia is a very rare condition with few reported cases. In the literature, the congenital defect is associated with other anomalies like Van der Woude syndrome, popliteal pterygium syndrome[2,3] and aglossia–adactylia syndrome. Our patient has none of these syndromes or any other intraoral or maxillofacial abnormalities. No sex predilection has been reported. [2,4] However, it is interesting to note that eight out of 11 cases reviewed for this study have male subjects.

The exact pathogenesis of congenital bony fusion is unknown. Some of the etiologic hypotheses proposed by Goodacre and Wallace[5] include "persistence of buccopharyngeal membrane, amniotic constriction bands in the region of the developing first branchial arch, environmental insults, drugs such as meclozine and large doses of Vitamin A."[1,2] According to a review by Dawson *et al.*, there is no familial tendency, history of drug or toxin exposure and consanguinity.[1,2,4] However, there is a possibility of autosomal recessive inheritance. Mir *et al.* and Poovazhagi *et al.* reported cases which revealed a history of consanguinity.[1,6]

Our patient presented with maxillomandibular fusion discovered immediately after birth. Physically, oral cavity deformity was evident and the baby was not able to open her mouth or feed normally. Once recognized, the diagnosis was confirmed by CT scan. These events are congruent with the observation that congenital bony fusion is clinically diagnosed soon or after birth as the neonate presents with airway and feeding difficulties.[1] The modality of choice is high resolution or spiral CT scan which has the advantage of revealing the condition of the temporomandibular joints or any hypoplasia of the other facial bones.[1]

The usual accompanying problems in such patients include airway and respiratory impairment, feeding difficulties, speech limitation, poor oral hygiene, interference with salivation and mastication, and induction of anesthesia.[1] In spite of these complications, studies show that functional results especially in isolated occurrence are likely to be good.[1] For our patient, feeding problems were addressed initially by insertion of nasogastric tube and subsequently, by gastrostomy tube.

Treatment consists of surgical division of the maxillomandibular fusion under anesthesia.[1,6] Awake blind nasal intubation is the ideal manner of inducing anesthesia, although a tracheostomy may be required if blind intubation fails.[1,2] A less invasive technique using fiberoptic nasotracheal intubation may also be administered.[3,6] Our patient underwent tracheostomy insertion for maintenance of airway and anesthesia. Following surgery, active physical therapy is recommended and the infant should be encouraged to feed normally as soon as possible[1,3,4] to achieve adequate mandibular range or motion and function. Acceptable mouth opening may be appreciated 1-2 weeks post operatively. [6] The mother of our patient was advised to perform such manual therapy techniques as carefully opening the baby's mouth with a tongue depressor and rotating fingers to press the gums.

Indeed, congenital maxillomandibular fusion is a very rare condition with few cases reported. We hope this report contributes to its diagnosis and management in other children.

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