and multiple cysts with well demarcated margins on T2-weighted images and low signal intensity on T1-weighted images. (1) Although the T1-weighted images usually demonstrate low signal intensity, the cyst may demonstrate mixed or even hyperintense signal depending on the age of the hemorrhage. (4) In this case, the MRI did reveal a heterogeneously hyperintense mass on T2-weighted images and a significant fluid-fluid level contained within the cyst with relatively more hypointense fluid layering posteriorly in the supine patient. On post-gadolinium T1-weighted imaging, cystic lymphangiomas typically show peripheral rim enhancement as well as enhancement of internal septations. The mass in this case was determined to be a large lymphatic malformation with associated internal hemorrhage.

As discussed, these lesions typically present prior to age 2 years. This child's presentation at age 13-years-old was atypical, but enlargement after injury in older patients has been reported. (6) Fung, et. al. described in their study that MRI produced highly detailed images that proved to be diagnostic and predictive of intraoperative findings, as was evident in this case. (1) Hemorrhage into the lymphatic malformation and the distinct appearance on MRI led to high confidence in the diagnosis and strategy of resection.

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CASE REPORT

Congenital Fusion Of Maxilla And Mandible (Bony Syngnathia)

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Abstract

Congenital fusion of maxilla and mandible as an isolated occurrence is very rare. Syngnathia is often associated with other intra oral and maxillofacial anomalies. We present a rare condition of isolated complete bony fusion of maxilla and mandible in a neonate without any other associated anomaly.

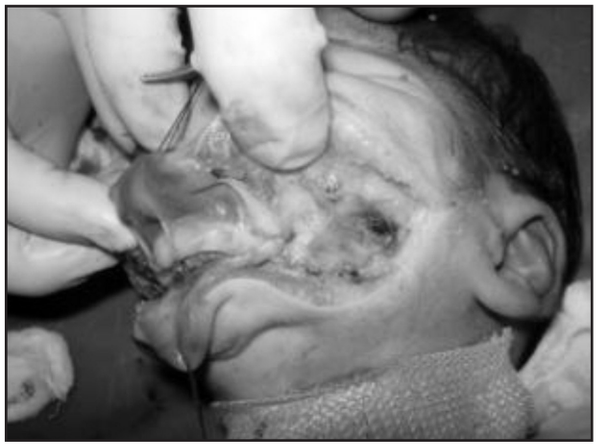
Introduction

Congenital craniofacial defect comprise 20% of congenital birth defects. (1) The fusion defects can present in a wide range of severity from single mucosal band (synechiae) to complete bony fusion (Syngnathia). Only 25 cases of syngnathia have been reported in literature. (2) We present a rare condition of isolated complete bony fusion of maxilla and mandible in a neonate without any other associated anomaly.

Case Report

A full term neonate 16 hours of age, weighing 3770 grams was referred to our medical newborn unit, for management of birth asphyxia. This was the first-born baby of consanguineous parents. Mother had a healthy gestation period and no occurrence of any illness, trauma or drug use. Baby was delivered by caesarean section and had not cried since birth. On admission baby was lethargic, tachypneic with no cyanosis or grunting. Physical examination revealed hypoplasia of mandible. There was severe trismus and mouth could not be opened even to pass an oropharyngeal tube. There were no other obvious external anomalies. Cardiovascular and respiratory systems were normal. Blood sugar, calcium and renal parameters were normal. Chest X ray was normal. On the second day of admission baby developed seizures which was controlled with Inj. Phenobarbitone. Baby's respiratory distress worsened and at 56 hours of life oxygen saturation could not be maintained with oxygen hood. Baby was started on nasopharyngeal CPAP with 5 cm H2O pressure and there was a brief improvement in oxygen saturation.

Radiography of the skull and facial bones revealed bilateral bony fusion of maxilla and mandible from anterior to posterior ends. In view poor respiratory efforts and declining saturation blind nasal endotracheal intubation was done to maintain oxygen saturation. ENT surgeon secured the airway with an emergency tracheostomy and baby was mechanically ventilated. Saturation was maintained between 85-87% for 24 hours. Repeat X-ray chest revealed bilateral infiltrates on the third day of hospitalization. Baby developed progressive desaturation and succumbed at 78 hours of hospitalization. Postmortem examination revealed



a complete bony fusion of maxilla and mandible from anterior to posterior ends on either side (Fig 1). Oral cavity was otherwise normal. Airway examination revealed normal vocal cords. There was no other anomaly.

Discussion

Synechiae are adhesions between anatomic structures. Syngnathia has been reported in association with cleft lip, cleft palate, aglossia, oral soft tissue synechiae, hypoplasia of proximal mandible. (3) Pathogenesis of this condition remains unclear though it can be related to developmental defect in the region of the 1st branchial arch, where separation of maxilla and mandible during the 7-8 week of embryonic development has failed to occur. (4) The cause of congenital bony fusion is not certain. Genetic, teratogenic, mechanical insults do contribute. (5) Trauma late in pregnancy, abnormal stapedial artery and teratogenic agents have been suggested. (1) Persistence of buccopharyngeal membrane, amniotic constriction bands in the region of development of branchial arches, environmental insults with drugs like meclozine and large doses of vitamin A have been postulated. The proposed classification for syngnathia is as follows (6). 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, Type 2 Complex syngnathia with other congenital anomalies in the head and neck, Type 2a syngnathia with aglossia, Type 2b syngnathia with agenesis or hypoplasia of the proximal mandible.

Congenital bony fusion can be clinically recognized and diagnosed. Conventional radiography and high resolution or spiral CT can support the diagnosis. Syngnathia interferes with feeding, breathing, growth, development and induction of anesthesia. However based on the scarce literature on isolated syngnathia functional results are likely to be good. Airway is the first priority to be secured in the management, thereafter feeds should be cared for by nasogastric or orogastric tubes. Fiberoptic nasotracheal intubation under spontaneous ventilation using low doses of ketamine offers a safe and non-invasive technique compared with tracheostomy or blind nasotracheal intubation. (7) Surgical division of bony fusion under general anesthesia is the optimal treatment in all types of syngnathia and is to be followed by physical therapy. Complete mouth opening may not be possible immediately after the surgical procedure and it may take 1- 2 weeks post operatively. When treatment for this fusion is not undertaken early or in long standing cases growth anomalies from temporo-mandibular joint ankylosis may occur (8).

Contributors

All 3 authors were involved in the management, design, drafting the article and approval of the final version. VP will act as the guarantor of the paper.

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