Congenital Fusion of Maxilla and Mandible: A Case Report

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Abstract

Objective: Congenital fusion of jaws is a rare disorder which is observed in infants and cab ne syndromic and non-syndromic. Limited mouth opening in patients can affect the child growth and cause difficulties in feeding, swallowing and breathing.
Case: A In this study, a female infant from Afghanistan, settled in Qom referred to AlZahra maternity with upper and lower fusion jaws is reported. According to clinical and laboratory examinations, infant had no other disorders except the above said disease, and her jaw fusion was non-syndromic.
Conclusion: Treatment of this disease in early stages not only is easier, but also can be effective in terms of child's growth and feeding; because in most cases TMJ ankylosis occurs due to the lack of mobility and loss of function which leads to difficulties in oral operations.
Key words: Ankylosis, Blind Nasal intubation method, Congenital, Jaw fusion.
Please cite this article as:
Mehdizade M, Vadiati Saberi B, Noori Bayat SH, Pishvaie MH. Congenital Fusion of Maxilla and Mandible: A Case Report. J Dent Sch 2015; 33(1): 118-122.
Received: 09.02.2014 Final Revision: 16.03.2014 Accepted: 19.04.2014

Introduction:

Congenital craniofacial includes about 20% of birth defects which one of them is fusion (joining) jaws (1). Congenital fusion of jaws is a rare (2-5) which was observed in infants and reported in 1936 by Burket for the first time (3). This fusion can be unilateral or bilateral and includes synechiae or syngnathia. Although synechiae had been reported several times, but syngnathia is rare which is less common than other types (5)and have been reported only 24 cases from 1936 to 2004 (2). Jaw fusion has two types:syndromic and non-syndromic that the latter one is very rare (2). Syndromic type has been observed in syndromes like cleft palatal late alveolar synechiae, vanderwoude. Limitation in mouth opening in these patients can affect child's growth and cause difficulties in feeding, swallowing and breathing.

Case:

Baby girl from Afghanistan and settled in Qom, with approximate weight of 3.5 kg from AlZahra maternity was referred to maxillofacial surgeon for examination and treatment. Initial examination shows upper and lower fusion (figure 1) and impossibility of mouth opening for infant. Due to the lack of equipments, maxillofacial surgeon avoided to perform the surgery at first. So the patient was referred to hospitals in Tehran; but after three weeks, while no action has been taken except gastrostomy to help infant's feeding by pediatrician, patient referred to the MaxiofacialSurgen. Due to the

insistence of infant's family, patient underwent oral and maxillofacial surgery at Imam Reza Hospital of Qom. Initial consultations with pediatricians and anesthetist were carried out and usual examinations were taken before the operation. General health of infant and non-syndromic malformation was confirmed by pediatrician. Also, no abnormality was reported in her family history. According to consultation, initial plan for Blind Nasal Intubation was considered. Although, crycothyrotomy is notusually prescribed for infants, but tracheostomy equipments was considered before intubation and injection of anesthetizing drugs for infant. Fortunately, after two attempts, Blind Nasal intubation was carried out successfully. It should be mentioned that the infant was 29 days old on operation day and feeding was done only with the tube implanted by gastrostomy (figure 2).

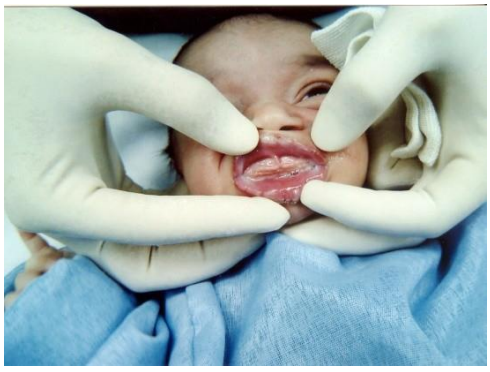

Figure 1- Upper and lower infusion

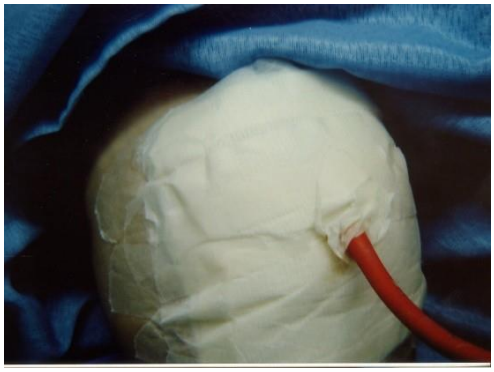

Figure 2- Implanted tube by gastrostomy

Operation Method

On operation day, infant was lied down in a supine position and doctors tried to keep general anesthesia as follows: First, cardiac monitoring and pulse oximetry were connected and after getting appropriate IV line, 0.05mg Atropine and 5mg ketamine were injected to infant, and adequate pre-oxygenation was done. Also, surgical premed injected 100,000 unit of penicillin G and 0.5mg of dexamethasone through the same IV line. Then, by breathing sound and sense of touch, Blind Nasal Incubation was done successfully using tube cuff 3 after two trying. Low cough reflex was observed; after that, 5mg Fentanil, 10mg Thiopentaland 1mg Atracurium were injected in intravenously; after approving the intubation by bilateral lung auscultation, infant was connected to ventilator of anesthesia machine and halothane was prescribed with $\frac{3}{10}$ % concentration. Then, anesthesia tube was fixed around the nose by leucoplast tape. What is common in oral and maxillofacial surgeries is putting a pharyngeal that was impossible in this special case. Since anesthesia tube was without cuff, during surgery operation team was careful for hemostasis and preventing the blood and saliva leakage into pharyngeal space. Then, separation from anterior to posterior was done by scalpel no.15. Jaw fusion was fibrotic in anterior and fibro osseous in the posterior region, so the team was careful not to damage tooth buds. Posterior jaw separation was performed using a scalpel and surgical osteotome. It should be mentioned that, there was not so much bleeding in the surgical area. After separation, suturing was done by chromic 4.0. The whole time of surgery, from induction of anesthesia until the end of the surgical procedure, lasted about two hours and forty minutes. After completion of surgery and extubation, breathing and hemodynamic status of patient was normal and transferred to the recovery with good general health; oral mucosal bleeding was under control (figure 3,4). After consciousness with good general health, the

infant was transferred into the ward transcribing penicillin V, 1000mg every 6hours, ibuprofen syrup 50 mg, every 6 hours; on the next day visit, there was no problem and patient referred to pediatricians to remove gastrostomy tube. One month after surgery, the patient was in good health. For the first time, mouth feeding was started from the first days after surgery and there was no problem in clinical examinations.

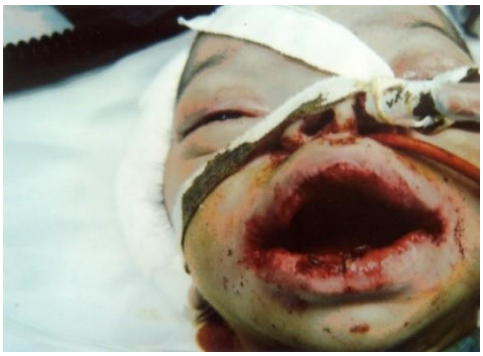

Figure 3- Patient after surgery

Discussion:

Maxilla and mandible fusion are very rare. Bruket reported jaw fusion with ankylosis in TMJ and attachment to the gums for the first timeand patient was suffered from unilateral atrophy like Horner syndrome (3). Since that, sporadic reports have been seen about this abnormality. Defects in jaw fusions are usually diagnosed after birth because the child can not open the mouth. The extension of this disease in variable and includes different degrees such as full attachment to gums (6), unilateral fusion with an incision on the front of the mouth (3), or bilateral fusion with a small incision in the anterior (2). Fusion of hard tissues of alveolar ridge in maxilla and mandible occurs after development; therefore, it should not be considered as a congenital craniofacial disorder but it is very similar to congenital disorders (1). Dawson, *et al*. (1997) (7) classified jaw bone fusions (7); this classification was revised by Laster, *et al.* (2001) (1):

Type 1 → simple fusion without other anomalies in head and neck (like this case).
Type 2 →complex fusion with subclasses: a) fusion along with aglossia, b) fusion along with agenesis or hypoplasia of the mandible.
Laster *et al*. had another classification of hard bone fusion in maxillofacial area:
Type 1a → simple bone fusion to the alveolar ridge anterior without any other congenital anomalies in head and neck (like the patient reported in this article).
Type 1b →complex fusion in alveolar ridge along with another congenital anomalies in head and neck.
Type 2a → simple fusion ofzygoma and mandible which makes unusual small mandible.
Type 2b→osseous compound fusion of zygoma and mandible with cleft or TMJankylosis(1)
The main cause of fusion between upper and lower jaws seems to be unknown during the seventh and eighth weeks of development of congenital alveolar ridge, when tongue and palatal arches are in contact and closure of palate depends on moving towards anterior and end of the tongue,fracture in this movement leads to the jaws fusion. Teratogenic factors may be involved in prolonged contact of oral structures and abnormal fusion (7). However, there are other theories for its ethology: Sijman and Prinsloo (1966) stated that the reason for this fusion depends on immaturity of ectoderm or probably abnormal processes during embryonic development (2). Hegtvedt (1993) proposed a theory about the lack of neural crest cells (8); Laster, *et al.* (2001) considered trauma or teratogenic materials as the main cause of this anomaly (1); although remaining of buccopharyngeal membrane (2), abnormal developmental processes (8), abnormal stapedial (9), environmental factors and using some drugs like Meclizine or high levels of Vit A (10) also can be the reasons for this abnormal fusion. Stenderg, *et al*. (1983) also said aboutprobable stages of such anomalies: first fusion of the

gums of upper and lower jaws, then palatal posterior cleft due to the lack of exposure of tongue in mandible and eventually TMJ fibrosis and ankylosis due to the immobility of mandible. Maxilla and mandible fusion can limit the growth of mandible and also cause severe malnutrition due to patient's inability to eat the food; so, children with this anomaly are seemed to be younger than their age (6). In this regard, the relevant anomalies are reported such as aglossia (11, 12), absence or hypoplasia of proximal mandible (13), TMJ ankylosis and cleft palate (14, 15), vertibal anomalies (16), ocular anomalies, coloboma(17, 18) and mental retardation (16). Anesthesia methods for these patients are different including: tracheostomy (19), fibroptic intubation (20), and Blind nasal intubation (21). Beside the most common diseases that make oral or nasal intubation difficult such as TMJ ankylosis, also head and neck infections, ankylosing spondylitis, obesity etc., congenital jaw fusion should be considered too. However, with advancements of biomedical engineering, now in most cases it is possible to perform intubation successfully for such patients using fiberoptic systems. Although, it is impossible to adjust the anesthesia tube no. 3 with the diameter of lumen of fiberoptic devices due to the diameter of anesthesia tube and lumen of fiberoptic devices. In addition, because of the high price and maintenance costs, not all the therapy centers can afford to buy these devices, especially the center in which this operation has done. In this particular case, due to the infant's age, it was impossible tocrycothyrotomyand there was no fiberoptic device for intubation in operation room; before the induction of anesthesia, certain conditions were considered to prepare tracheostomy equipments; but, fortunately after two attempts, the Blind Nasal intubation method was performed successfully and after lung auscultation, bilateral ventilation was heard. In most of the reported cases, the best time for this type of surgery is exactly before infancy ends up. During this time, feeding is done by NG (22,23); in this particular case, feeding was done by inserted tube through gastrostomy which was performed by pediatricians.

Conclusion:

Not only treatment in early stages can be easier, but also it can help the growth and nutrition of the child; because by passing time, TMJ ankylosis happens in these people due to the immobility and lack of function which leads to difficulties in surgery (4). Fortunately, intubation was performed through Blind nasal for this special patient, but if it didn't work, that was inevitable to perform tracheostomy. It seems that this intubation technique was used in patient's previous surgery.

Conflict of Interest: "None Declared"

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