

Congenital Fusion of the Mandible and Maxilla- A Case Report from Ajman, United Arab Emirates

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Abstract Congenital fusion of the mandible to maxilla is a rare phenomenon. The fusion can be a bony fusion in which case it's referred to as Syngnathia or soft tissue fusion when it's addressed as Synechia. [1] Soft tissues fusion is involved most often in the oral cavity but it may involve both hard and soft tissues of the child. This results in inability of child to thrive as it doesn't allow mouth opening and feeding. This is a case report of a 3-day-old baby boy with partial maxilla-mandibular fusion from right canine to posterior jaw who was brought to Oral and Maxillofacial department of the dental centre in GMC Hospital, Ajman, UAE for treatment because of inability to thrive.

Keywords: congenital fusion, syngnathia

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1. Introduction

Congenital fusion of the mandible to maxilla is a rare phenomenon. The fusion can be a bony fusion in which case it's referred to as Syngnathia or soft tissue fusion when it's addressed as Synechia. [1] Soft tissues fusion is involved most often in the oral cavity but it may involve both hard and soft tissues of the child. This results in inability of child to thrive as it doesn't allow mouth opening and feeding. Treatment should be provided as early as possible as it could eventually result in the ankylosis of the temporomandibular joint. [2] The bony fusion can be further classified as partial or complete and then as syndromic and nonsyndromic. The etiology remains unknown although various theories have been postulated. Treatment has to be case to case basis [1].

2. Case Report

A 3-day-old baby boy with partial maxilla-mandibular fusion from right canine to posterior jaw was brought to Oral and Maxillofacial department of the dental centre in GMC Hospital, Ajman for treatment because of inability to thrive. The patient's jaws were found to be fused in the posterior regions on the right side. In the anterior region and left side of the jaw there was no fusion. Except for the fact that the parents were consanguineous, the medical history of both of them was insignificant. The mother gave a history of healthy pregnancy and denied any illness, trauma, or drug use. The baby was their 1st child born by

normal vaginal delivery. Both temporomandibular joints seem to function normally.

3. Discussion

Syngnathia is a quite a rare disorder and is often seen with syngnathia. [3] This pathology is associated with various syndromes such as Van der Woude, cleft palate alveolar syngnathia, and oromandibular limb hypogenesis syndrome. Since the first case reported in 1936 by Burket, only 27 cases of syngnathia have been reported. [4] Laster et al [5] reported 24 cases. Daniels reported a case of bony fusion of the mandible to the maxilla and to the zygomatic complex. Parkins and Boamah [7] reported a case of fusion of the jaws, synechia of the buccal mucosa and the gingiva, with cleft of the hard palate.

Gartlan et al. (1993) [8] reviewed synechia and were relegated as buccopharyngeal membrane remnants or as ectopic membranes on the substratum of their surmised inchoation. Particularly its isolated occurrence, bony fusion (syngnathia), is profoundly infrequent. A very few cases reported in the literature are in description, inconsistently erratic and perplexing in nomenclature and with circumscribed subsidiary conventional imaging [8].

The etiology is unclear. Neural crest cell derived osteogenic patterning is affected in the absence of Foxc1, as osteoblasts develop ectopically in the maxillary prominence and fuse with the dentary bone. Kimberley Inman has provided evidence that Foxc1 and Fgf8 genetically interact and it is Fgf8 dosage responsible for variation in the syngnathic phenotype [9].

Because the neonate is not able to open the mouth or feed normally, Maxillomandibular fusion is usually discovered immediately after birth [10].

The reclassification for syngnathia regarding other anomalies was proposed by Dawson et al [11] as follows: type 1: simple syngnathia, with no other anomalies in the head and neck; type 2: intricate syngnathia, with subgroups, type 2a: syngnathia coexistent with aglossia and type 2b: syngnathia coexistent with agenesis or hypoplasia of proximal mandible.

After their experience with a case of fusion of the mandible to the zygomatic involute and maxillary tuberosity, however, Laster et al [5] proposed a modified reclassification for bony syngnathia: type 1a: simple anterior syngnathia, characterized without other congenital deformity in the head and neck and by bony fusion of the alveolar ridge only and; type 1b: complex anterior syngnathia, characterized by associated with other congenital deformity in the head and neck and by bony fusion of the alveolar ridges only and; type 2a: simple zygomaticomandibular syngnathia characterized by bony fusion of the mandible to the zygomatic complex causing only mandibular micrognathia; type 2b: complex zygomaticomandibular syngnathia, characterized by bony fusion of the mandible to the zygomatic complex and associated with clefts or temporomandibular joint ankylosis.

The rigor of this disorder has no fine-tuned pattern. The range of jaw fusion elongates from consummate fusion, unilateral fusion, or bilateral fusion with a diminutive anterior opening. [10] Our case belonged to the unilateral fusion type.

Interfered with feeding, breathing, general health of the patient, this condition is problematic and (aspiration pneumonitis), growth and development, induction of anesthesia (intubation) etc. The infrequency of this condition imposes some inhibitions on standardization of the treatment [8].

It is more problematic in involute cases in terms of jaw function and its outcome. Dawson, et al [11] proposed a system of reclassification and elaborated on treatment recommendations focusing on functional outcome. In the management of any newborn with trismus, the airway is the first priority to be secured. Afterwards, feeding problems should be overcome by placing a nasogastric or gastrostomy tubes.

There is high rate of association between bony syngnathia and systemic malformations and the patient

should be under the supervision of a team of clinicians to aid proper diagnosis and treatment [12].

Under general anesthesia (blind intubation or via tracheostomy), surgical division of the bony fusion, is the optimal treatment for the simple syngnathia in isolated occurrence or cases with the availability of other anatomic abnormalities. The infant should be encouraged to feed normally as soon as possible [8] and proper physical therapy should be started immediately,

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