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Case report

Congenital bilateral zygomatico-maxillo-mandibular fusion associated with gumfusion: Case report

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ABSTRACT

Introduction: Maxillomandibular sygnathia is a rare and severe craniofacial deformity defined by gingival mucosal fusion (synechia) or bony fusion (synostosis).
Case report: We will present a case of complete closure of the mouth since birth from eastern Morocco treated in our department of maxillofacial surgery at the University Hospital of Casablanca. The patient was referred after two months by the pediatrician of the provincial hospital and following the placement of a nasogastric tube. On admission, she presented with facial dysmorphosis, signs of malnutrition with closure of the mouth and fusion of both gums with pro-alveoli and retromandibulia. CT scan with 3D reconstruction confirmed maxillomandibular synostosis. 5 days later, she underwent a fixed mucosal incision with osteotomy at the maxillomandibular joints with early and prolonged active physical therapy. The evolution was marked by the recurrence of mouth closure, the little girl was operated a second time and then she died by a mucous plug at the level of her tracheostomy cannula during resuscitation.
Discussion: Maxillomandibular sygnathia is a very rare pathology whose origin remains unknown. Very few cases published in the literature.
Conclusion: The therapeutic difficulties encountered outside the ideal age of the operation and the management of recurrences were also linked to socio-economic factors making it difficult to ensure adequate postoperative follow-up.

1. Introduction

Maxillomandibular sygnathia is a rare and severe craniofacial deformity defined by a fusion of the gum mucosa (synechia) or bone fusion (synostosis). It represents several types such as mandibulo-maxillary fusion, alveolar fusion and gum fusion [1]. Sixty-two cases of congenital bone syngnosis have been reported in the literature up to 2019 [2]. Sygnathia manifests itself from birth by a complete closure of the mouth leading to difficulties in feeding and swallowing. It can be complicated by poor facial development and/or joint ankylosis (TMJ).

Currently there is no precise protocol for proper management. We will present the case of maxillomandibular sygnathia followed in our unit to illustrate the different therapeutic difficulties that can be encountered.

There are several risk factors such as the consumption of high doses of vitamin A, a teratogenic agent, and the notion of consanguinity.

The diagnosis is obvious clinically at birth, however, indirect signs can be detected by prenatal ultrasound.

The scanner with 3D reconstruction allows us to detect with precision the type of synostosis and the bones of the face involved in the malformation.

Several classifications have been proposed, notably that of Dawson et al., and that of LASTER et al. Fig. 1.

The treatment is surgical with immediate, early and prolonged post-op rehabilitation.

2. Case report

14-month-old child, second of a two-year-old sibling, with limited mouth opening since birth. Resulting from a full-term pregnancy, uneventful vaginal delivery, with the presence of a similar case in the family as a family history, presence of the notion of consanguinity. The

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Type 1a	Simple anterior syngnathia	Bony fusion of the alveolar ridge without other congenital deformity in the head and neck
Type 1b	Complex anterior syngnathia	Bony fusion of the alveolar ridges associated with other congenital deformity in the head and neck
Type 2a	Simple zygomatico-mandibular syngnathia	Bony fusion of the mandible to the zygomatic complex causing only mandibular micrognathia
Type 2b	Complex zygomatico-mandibular syngnathia	Bony fusion of te mandible to te zygomatic complex and associated with clefts or TMJ ankylosis

Fig. 1. Laster classification.

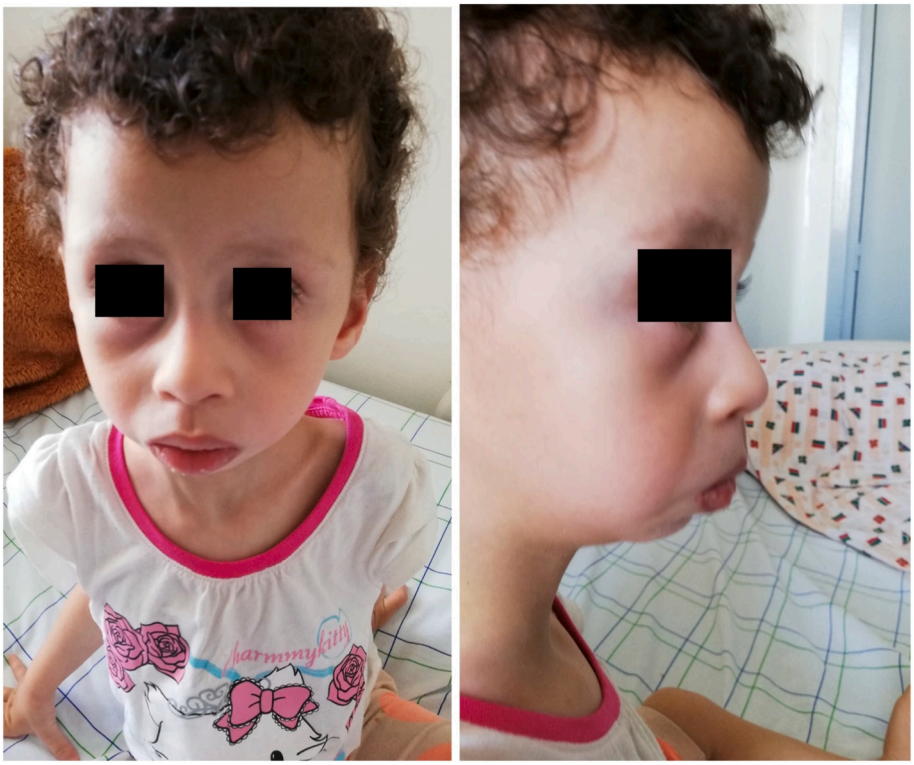

Fig. 2. Preoperative photograph of the patient

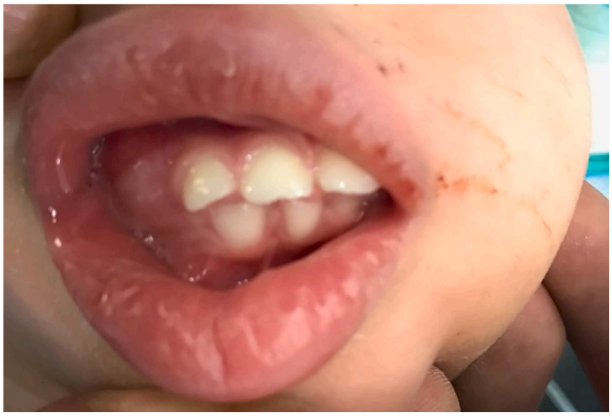

Fig. 3. Oral view showing the fusion of the upper and lower gums.

newborn was seen at the provincial hospital and his pediatrician put him under a nasogastric tube and sent him after two months to our department.

The clinical examination on admission showed a malnourished child with a nasogastric tube in place, The examination of the oral cavity on admission showed a complete closure of the mouth with fusion of the two gums associated with retromandibulia and upper proalveolus, the exobuccal examination showed a facial dysmorphosis with low set ears, round eyes. The patient generally showed signs of malnutrition Figs. 2, 3.

CT scan with three-dimensional reconstruction showed bilateral maxillomandibular synostosis Figs. 4–5, our patient was classified as type 2a according to Laster et al. In view of this symptomatology, surgical management was necessary.

A multidisciplinary staff meeting was held in collaboration with the resuscitators, pediatricians and nutritionists, and the staff's decision was surgical management.

Nasotracheal intubation was difficult using fine fiberoptic laryngoscopes. Five days later, the patient was operated on by the department head (who had 16 years of experience) and underwent a vestibular intraoral incision along the fused occlusal surfaces of the upper and

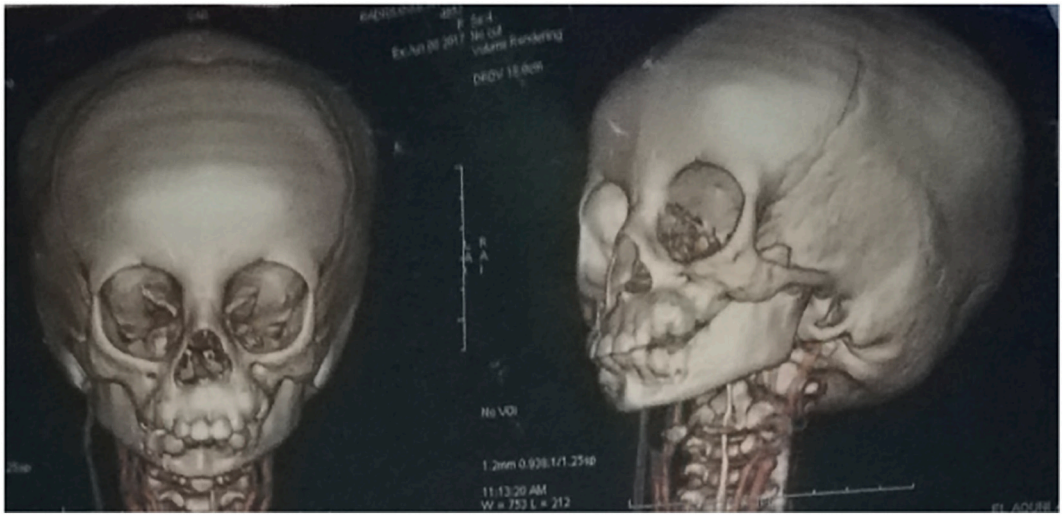

Fig. 4. Three-dimensional CT scan showing bilateral fusion of the maxillary and mandibular ridges.

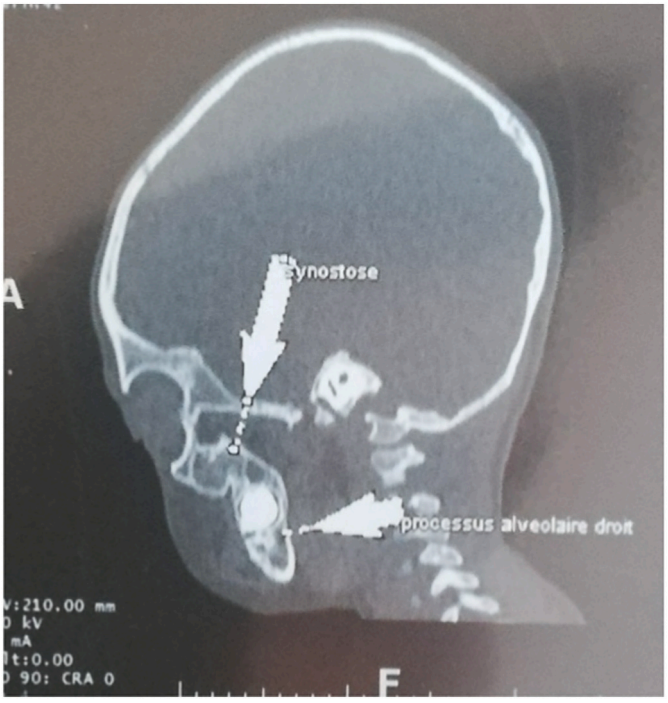

Fig. 5. Sagittal view of the facial scanner showing the synostosis block.

lower gingiva. The mucosal surfaces were separated anteriorly. Osteotomes were used posteriorly (bilaterally) to separate the bony fusions. At the end of the operation, the mouth opening was much improved.

The immediate postoperative course was favorable (mouth opening 35 mm), and the evolution was marked by the appearance of a scarred neck. Active physiotherapy was prescribed.

The patient was lost to follow-up for 10 months and then returned to the department with complete closure of the mouth. The CT scan showed the presence of synostosis between the two coronoid processes and the posterior aspect of the maxilla with TMJ ankylosis.

A second operation was performed after 4 days, intubation was difficult and required a tracheotomy. The infant benefited from a bilateral osteotomy with placement of a silicon plate. The child died after 24 h due to a mucous plug in the tracheostomy tube in the intensive care unit Fig. 6.

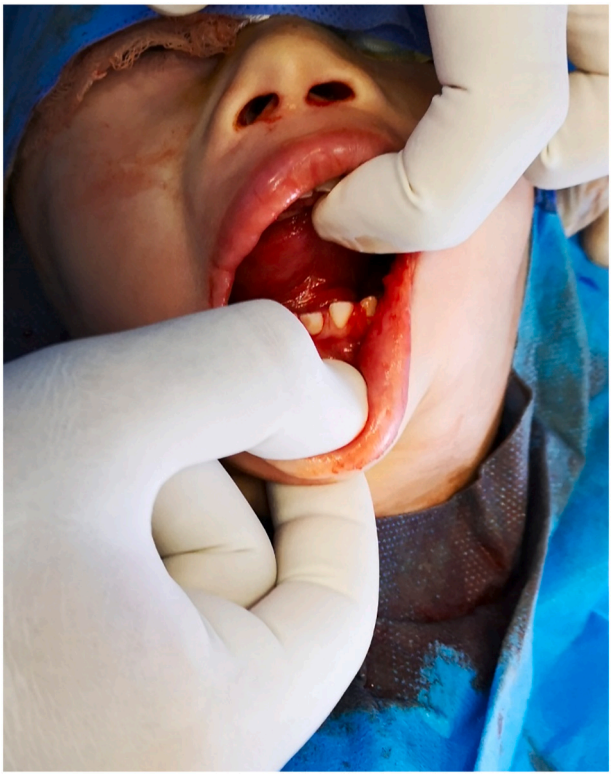

Fig. 6. Immediate post-op image showing the mouth opening obtained.

This case report has been reported in line with the SCARE Criteria [3].S.

3. Discussion

Congenital syngnathia is a very rare disease that presents several unique challenges. There is no consensus on the best operational approach to manage this condition [4]. It was first reported by Burket in 1936. It can occur with different degrees of severity, ranging from synechia (mucosal band fusion) to bony fusion (synostosis) [5].

Syngnathia is an uncommon disease with an uncertain etiology [6]. It may be related to a fetal stapedial artery anomaly, early loss of neural crest cells, teratogen(s), trauma, persistence of the buccopharyngeal

Table 1

Type 1a	Simple anterior syngnathia	Bony fusion of the alveolar ridge without other congenital deformity in the head and neck
Type 1b	Complex anterior syngnathia	Bony fusion of the alveolar ridges associated with other congenital deformity in the head and neck
Type 2a	Simple zygomatico-mandibular syngnathia	Bony fusion of the mandible to the zygomatic complex causing only mandibular micrognathia
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membrane, amniotic constriction bands in the region of the developing gill arch, environmental factors, drugs (such as high doses of vitamin A3), and consanguinity [7]. Cases of acquired sygnathia have been reported due to myositis ossificans and fibrotic pedicle flaps [6]. We note the notion of consanguinity and the presence of similar cases in the family in our case although there are isolated cases of fused jaws, syngnathia is often part of a syndromic group such as Van der Woude syndrome [8].

Dawson et al. divided syngnathia into several categories:

Type 1: Simple sygnathia characterized by the presence of bony fusion between the mandible and maxilla or zygoma in the absence of other congenital anomalies of the head and neck.

Type 2: Complex sygnathia characterized by the presence of bony fusion between the mandible and maxilla or zygoma in the presence of other congenital anomalies of the head and neck;

Type 2a: Sygnathia with aglossia.

Type 2b: Sygnathia with agenesis or hypoplasia of the proximal mandible.

Laster et al. proposed a modified classification for bony syngnathia of the maxillofacial region in 2001 (Table 1).

Prenatal 2D ultrasound in the sagittal plane allows for the objectification of signs in favor of sygnathia such as micrognathia and echogenicity of the zygomandibular area. This has led to a better understanding of the malformations by the parents and the resulting poor prognosis [9]. Because of the rarity of syngnathia, there are no treatment protocols [10]. As such, case reports can guide the maxillofacial surgeon toward optimal management that can minimize long-term complications.

Treatment should focus on securing the airway and placing a nasogastric tube to provide nutrition from birth. This is followed by early removal of the bony fusion with interposition of material to cover the bony ends according to DAWSON et al. and followed by active physical therapy. Early treatment has been recommended to avoid the problems that usually result from ankylosis, such as mandibular growth retardation and resulting facial deformities [11]. Our patient was treated surgically at an early age with rehabilitation to allow proper mouth opening and normal mandibular growth and to avoid complications, but the lack of parental cooperation in rehabilitation and the long distance between the hospital and the birthplace of both infants complicated their follow-up. Approaches used in surgery vary depending on the type and site of fusion, such as coronal, submandibular, preauricular, and intraoral approaches. Interposition of a flap or material, such as silicone sheets, gauze packs, and has been described to cover the gross area of bone [5].

4. Conclusion

This is one of the first cases of maxillomandibular sygnathia reported and operated in our country.

The therapeutic difficulties encountered outside the ideal age of the operation and the management of recurrences were also linked to socio-economic factors making it difficult to ensure adequate postoperative follow-up.

To improve the prognosis and quality of life of the patients, it is necessary to involve all the specialists (maxillofacial surgeons, anesthesiologists and pediatricians) in the management and it is necessary to meet for each treated case in order to draw conclusions and useful recommendations.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Registration of research studies

None.

Guarantor

Raiteb Mohamed.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

CRediT authorship contribution statement

Raiteb Mohamed: Corresponding author writing the paper.
Elmrini Sanaa: writing the paper.
Fatemazahra Azami Hassani: writing the paper.
Faiçal Slimani: Correction of the paper.

Declaration of competing interest

Authors of this article have no conflict or competing interests. All of the authors approved the final version of the manuscript.

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