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CASE REPORT

Atypical Craniofacial Tessier 3-4 Cleft: A Very Rare Congenital Complex Facial Cleft Type

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ABSTRACT

Oblique and transverse craniofacial cleft are very infrequently encountered congenital anomaly. This complex craniofacial entity is mostly linked with developmental anomalies of the first and second branchial arches. The pathophysiology for this special cleft is still not well understood as opposed to the cleft lip and palate that is attributed to the failure of fusion of mesoderm. They are usually sporadic with no syndromic association or gender predilection. We intend to present one of such rare case of Tessier's cleft type 3- 4.

Keywords: facial cleft, malformations, Z-plasty

INTRODUCTION

Facial clefts that occur obliquely and/or laterally are very rare craniofacial congenital entity believed to be in the range of 0.24% of all facial clefts [1]. The overall incidence of facial cleft ranges between 1.43 and 4.85 per 100,000 births [2]. The pathophysiology of the cleft palate and lips can be explained through a failure in the union of the mesoderm during the embryonic process. However, it is difficult to explain the oblique or lateral complex type of craniofacial clefts such as the lateral oro-ocular, nasoocular, and medial oro-ocular clefts with this concept. Some researchers are of the view that that the presence of amniotic band plays a part in the congenital development of these very rare types of craniofacial clefts.[3] Some other investigators have also suggested that the etiopathogenesis of such complex clefts can be attributed to primary arrest of development, a neurovascular inadequacy or tears in the evolving maxillary process. [4] Literature also advocates that these anomalies could be caused by an interplay of directly tethered tissue movement and increased local pressure that results to cellular vascular insufficiency.[5] It should be noted that, most of the known cases of complex craniofacial clefts are sporadic with no syndromic link or gender preference.[3] This group of facial deformities has been best classified by Tessier 6 who has been described as the father in the field of craniofacial surgery. He uses the orbit as the center of the defect from which the clefts radiate like the spokes of a wheel as demonstrated in fig. 1.

Dr. Paul Tessier also developed a classification system for craniofacial clefts that is known to be the most extensively applied. The Tessier numbering system is based on precise anatomic disorders that fall along embryonic lines of fusion within the face. Tessier's classification system is numbered from 0 to 14 with clefts 0 to 7 describing facial clefts and that of 8 to 14 describing cranial vault clefts. Each cleft has distinct soft tissue and bone lines of splitting.

CASE REPORT

A 10-year-old female child born as the second child to unrelated healthy parents via spontaneous vaginal delivery. Height and weight at birth were 48cm and 2.9kg respectively. There was no history suggestive of exposure to teratogenic substances or any information pointing towards a similar genetic disorder in the families of either parents. As part of the investigations, facial and orbital CT scan with contrast was carried out and the results shows a right lower lip cleft, right paramedian facial cleft (Tessier 3) and left orbital cleft (Tessier 4). Both globes and lens, optic nerve and extra ocular muscles as well as the brain parenchyma were grossly unremarkable as shown in fig. 2 to 7.

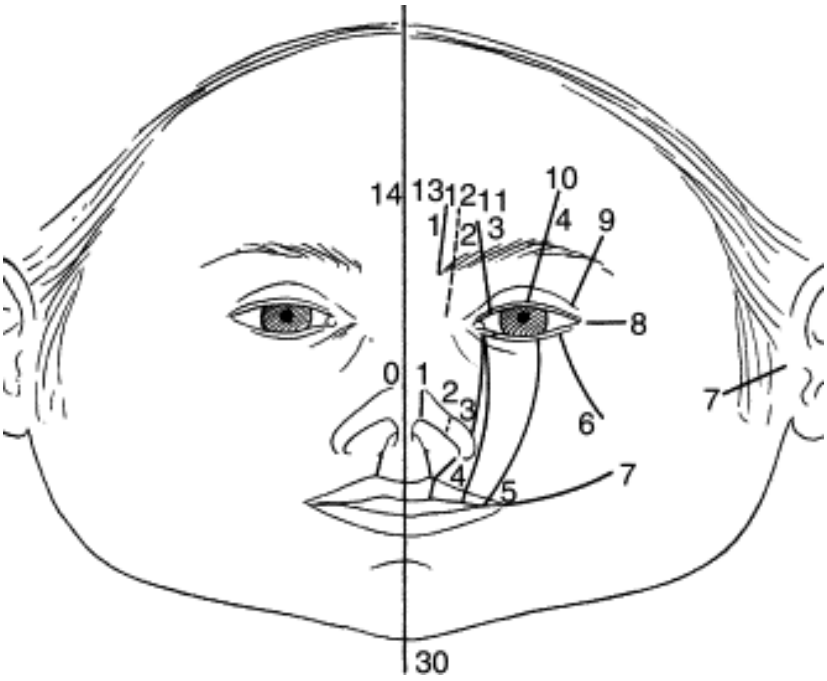
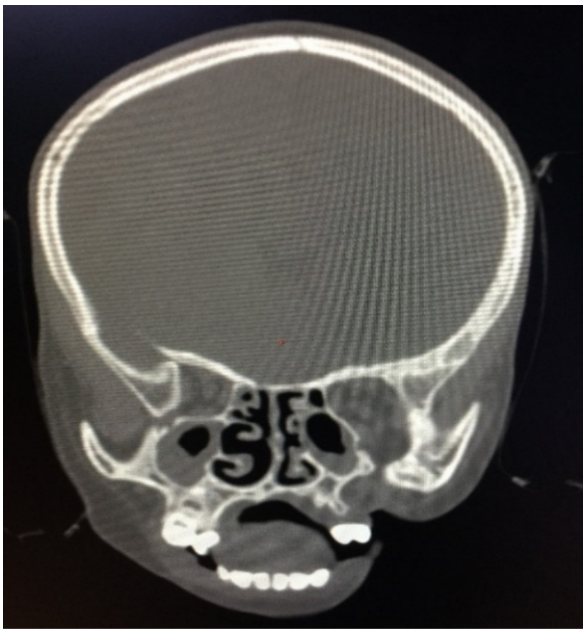
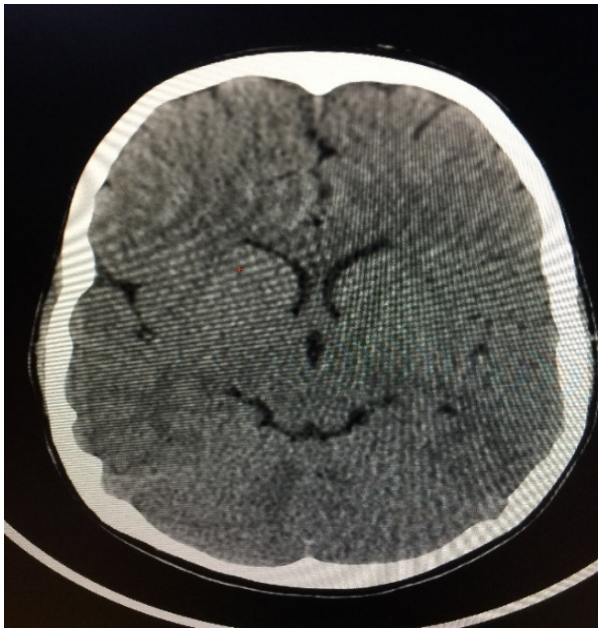


Figure 1: Tessier classification for orthodontist. http://www.slideshare.net





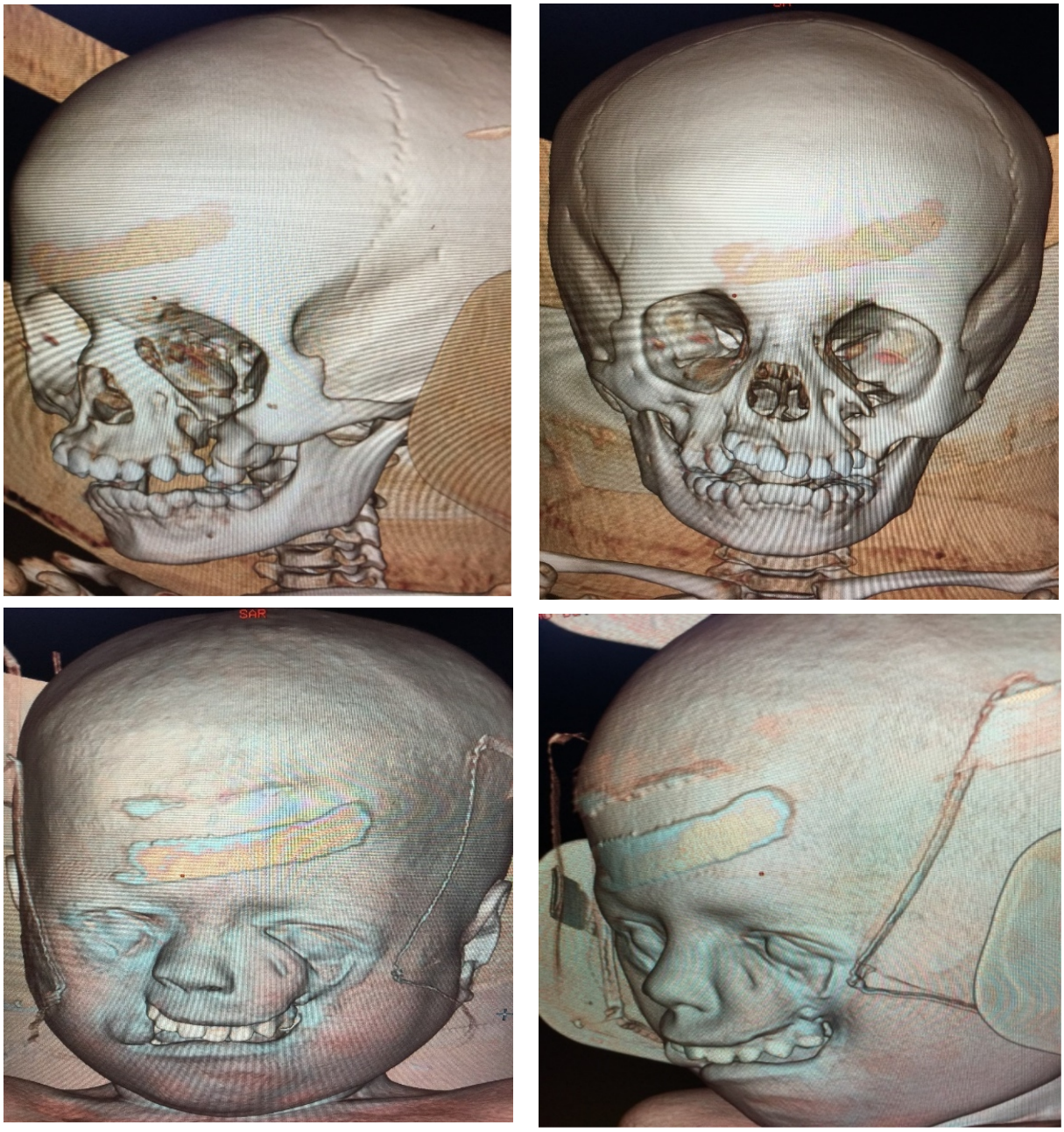


Figure 2a; CT showing the bony defects, Fig. 2e&2f; CT 3D volume reconstruction showing soft tissue defects.

DISCUSSION

The mouth, nose and orbit are important landmarks through which Tessier craniofacial follow constant axes. Craniofacial clefts encompass a defect of the underlying cranial and/or facial osseous structures. In this present case, soft tissue and the underlying facial skeleton were involved. The right facial cleft involves the upper lip and hard palate traversing the maxillary bone between the canine and first premolar teeth, runs along the anteromedial wall of the right maxillary antrum and terminates just below the floor of the orbit. For the left side, it extends from the upper lip, hard and soft palates and traverses the maxillary bone between the first and second premolar teeth, passing along the anteromedial wall of the left maxillary antrum laterally to the left infraorbital foramen and ends to the floor of the left orbit. A cleft in the right lower lip is also present with no involvement of the maxillary bone. Soft tissue abnormalities are noted with downward retraction of the bilateral lower lids as shown in figures 6 and 7. Tessier proposed an anatomy-based classification system for craniofacial clefts in 1976, assigning a

number to each cleft based on its placement in relation to the mid sagittal orbit. The Tessier type-3 craniofacial developmental anomaly extends from the philtrum of the upper lip to the medial canthus of the orbit, passing through the wing of the nose. Tessier numbers 3, 4, and 5 are found in the maxilla and orbital floor, respectively, while Tessier number 6 extends from the orbit to the maxillary bone. Tessier number 7 is located on the line between the mouth angle and the ear.[6]

It has been stated that each of these complex craniofacial clefts has its own uniqueness and poses a different challenge in management.[6] Where both soft and bony tissues are involved, as in this case, a detailed radiological reconstruction and volume rendering of the maxillofacial region is needed to evaluate the morphology of the cleft in other to plan for treatment. Surgical intervention for these group of patients is aimed at restoration of function and to improve facial cosmetic. However, when there is tissue hypoplasia of the specialized facial structures as in the nose, eyelids or lips, it creates the greatest impediment in restoring normal appearance and function. This is even more compounded when there is involvement of many structures such as nose, orbit, sinus, palate, alveolus, and soft-tissue. Correction of these congenital anomaly based on the complexity of the defect as mentioned in the literature ranging from Z-plasty7 local flaps,[8] cheek rotation flap including the lower eyelid, [9] rotation and advancement flap of the cheek,[10] and the tissue expansion methods[11]. Depending on the complexity of the anomaly, in some instances, the aftermaths of surgical interventions are not really superlative.

CONCLUSION

Tessier's oblique and transverse clefts in combination are extremely uncommon, with only a few cases reported. We are delighted to describe a very unusual case with lateral and oblique facial clefts.

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