Distraction Osteogenesis in a Tessier 6 Cleft After Fronto-Orbito-Zygomatic Osteotomy - A Case Report

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Abstract

The Rationale: Craniofacial clefts are rare malformations. According to their localization, 15 types are classified. Patient Concerns: They referred to impaired eyelid function, lateral protection of the eye-bulb, and aesthetic appearance. Diagnosis: A three-dimensional-computerized tomography scan and a stereolithographic model led to the diagnosis of a rare intraorbital Tessier 6 cleft. Treatment: A newly designed lateral orbital rim osteotomy, presurgically simulated on a stereolithographic model, was performed to forward position the lateral orbital rims bilaterally by osteodistraction. Outcomes: Both lateral orbital rims were distracted anteriorly for around 12 mm over a period of 9 weeks, without any side effects to the patient. Take-away Lessons: In this intraorbital Tessier 6 cleft, an improved eyelid function with better lateral protection of the eye bulb and more favorable aesthetic appearance was achieved with this novel lateral orbital rim osteotomy.

Keywords: Classification, exophthalmia, facial cleft, nonsyndromic, orofacial cleft

INTRODUCTION

By definition, a Tessier 6 facial cleft involves the maxillary alveolar[1] process between the second and third molars, the lateral aspect of the zygoma, and the inferolateral orbital rim, with or without overlying cutaneous, subcutaneous, and/or muscle tissues.

Congenital nonsyndromic craniofacial clefts, as classified by Tessier[2-3] in 1976, can manifest themselves in very variable ways. In addition to cleft formation due to lacking development of nonexistent primordial tissue, cutaneous bridging at those sites may often be observed. Such variability in phenotype may render rather challenging both a precise diagnosis and a subsequent, appropriate treatment.[4-6] In most circumstances standardized surgical protocols prove futile, as due to their localization, such clefts present themselves uniquely related to their type of tissue defect and involved functional structures. Often, surgeons are left to rely on just their innovation, improvisation, and surgical skills during the surgery.

This report presents a very rare intraorbital Tessier 6 facial cleft, its diagnosis, presurgical haptic planning, a novel approach to surgical treatment, and its outcome and follow-up.

CASE REPORT

The patient’s chief concern was a bilateral incomplete eyelid closure, leading sometimes to dry eyes. Her parents were particularly concerned about her large eyes and the aesthetics associated with them.

The patient presented with substantially under-developed inferolateral and superolateral orbital rims on both sides, being 11–13 mm posterior to the standard and protrusive eye-balls with a slight hypertelorism. The eye mobility was unrestricted, neither strabismus nor visual impairment or keratitis were detected. She initially underwent the intrauterine diagnosis of a metopic synostosis and probably later due to exophthalmia and a slight hypertelorism the diagnosis of Crouzon syndrome [Figure 1a and b]. A three-dimensional computerized tomography and a

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subsequent stereolithographic skull model in this 10-year-old girl, however, proved the absence of bilateral coronal and sagittal synostoses with the diagnosis of an intraorbital Tessier 6 cleft, involving maxillary, sphenoid, zygomatic bones, and the orbital floor [Figure 2]. Further, a substantial bilateral setback of the lateral orbital rims, lateral frontal, and zygomatic bones, and the
frontozygomatic sutures [Figure 3] became visible. To correct the functional deficits, an anterior distraction of both lateral orbital rims became necessary.

During a haptic presurgical planning phase, a novel lateral orbital rim osteotomy was determined on the stereolithographic model together with the ideal positioning of the distractors and their footplates [Figure 4]. It was also decided to reinforce the atrophic fronto-zygomatic suture for distraction, using a 4-hole microplate 0.4 mm and four 1.5 mm miniscrews, Level 1 Micro-System (KLS-Martin, Tuttingen, Germany).

In supine position, under general anaesthesia with oral intubation, the surgical access was carried out via lateral eyebrow and latero-inferior orbital rim incisions, placed along the Langer relaxed skin tension lines [Figure 5]. To protect facial nerve branches at these sites, physiological saline solution 0.9% (Fresenius Kabi, Midrand, South Africa) was previously injected into the subcutaneous plane for expansion. The lateral canthal ligaments and their attachments at the Whitnall’s tubercles, as well as the peri-orbita, were protected throughout the procedure.

After placing the microplate onto the fronto-zygomatic suture, the osteotomy was performed according to the presurgical planning and the internal distractor (KLS-Martin – Micro Zurich II Distraction System, Tuttingen, Germany) was fixed bilaterally on the zygoma. The mobilized lateral orbital rims were anteriorly distracted according to the direction of the zygomatic arch. Wound closure was performed with 4.0 V-Loc 90 (Covidien, North Haven-Connecticut, USA) in single stitches.

The intraoperative and postoperative course was without complications. After a 4-day postsurgical latency period, distraction was performed over a period of 36 days, at a rate of 0.3 mm twice per day. The overall distraction length at the zygoma was 24 mm, the one at the fronto-zygomatic suture on average 12 mm, slightly varying between both sides [Figure 6].

After 4 weeks of retention with the orbital rim at its planned position, removal of the distractors followed at week 9 under general anaesthesia using the former incision lines [Figure 7]. The overall follow-up period ended with a final clinical control in week 20 after the surgery.

The wound care of the skin incisions was carried out with Dermastine + Vitamin A (Litha Pharma, Midrand, South Africa) throughout the entire treatment period.

**Take-away Lessons**

A database of 4868 cleft patients revealed 126 patients with oblique-facial and lateral-facial clefts (2.5%). However, 79 (1.62%) bilateral Tessier 0–3 syndromic midfacial clefts among 91 holoprosencephaly patients were excluded. Various records of Tessier 6 clefts (0.41%) were identified. Sixteen cases of Tessier 6-7-8 facial clefts combined with palatal clefts were found in Goldenhar and Treacher-Collins syndromes; 2 cases with Tessier 6 facial clefts combined with additional standard cleft lip alveolar palate or other lateral facial cleft variations; and 2 cases with a Tessier 6 cleft only (0.04%), including the hcre described rare intra-orbital Tessier 6 cleft (0.02%).

The diagnosis of a Tessier 6 cleft may be considered controversial, as its pattern and extension may vary considerably. By definition, it involves the maxillary alveolar process between the second and third molar, the lateral aspect of the zygoma, and the infraorbital orbital rim, with or without overlying cutaneous, subcutaneous and/or muscle tissues. However, it has been already described as a Tessier 5 cleft and it may even extend towards the cranium, like Tessier 8 or 9 clefts.

Due to the intrauterine diagnosis of a metopic synostosis, this patient underwent a surgical repair soon after her birth, using a coronal approach. As this previous reconstruction left behind bony defects in the frontal bone area, local skin incisions were chosen instead of a coronal approach, even though from an aesthetic point of view this surgical access might be considered controversial.

Tessier classified various osteotomy lines for facial and craniofacial corrections that, however, involved always the lateral orbital rim together with additional craniofacial osteotomies. An isolated osteotomy of the lateral orbital rim, a fronto-orbito-zygomatic osteotomy, has never been described so far.

To the authors’ knowledge, this is the first description of an intraorbital Tessier 6 craniofacial cleft, being treated with isolated lateral orbital rim osteotomies and distraction osteogenesis.

**Declaration of patient/parent consent**

The authors certify that they have obtained all appropriate patient consent forms. In there, both parents of the patient gave their consent that images and other clinical information can be used for scientific publication purposes. The parents understand that the names and initials will not be published and due efforts will be made to conceal their identity, but also acknowledge that anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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