Case Report

Two-stage Corrections of Rare Facial Tessier's Cleft - 3,4,5,6,7

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Abstract

Congenital transverse and oblique facial clefts are rare congenital anomalies, and cooccurence of these is still more a rare anomaly. The condition most commonly is often associated with anomalies of the first and second branchial arches. The manuscript intends to present one such rare case of Tessier's cleft type 3–7 and present its early management as well as long-term treatment plan.

Keywords: Congenital defects, facial cleft, India, maxillary dystopia, oblique facial cleft, reconstruction, Tessier's clefts, transverse cleft

Introduction

Facial clefts are the common craniofacial malformations, often involving the palate and lips.[1] Less often, they occur obliquely and are estimated to be the rarest form of congenital deformities. Although the reported incidence is not estimated, it is believed to be in the range of 0.24% of all facial clefts.[1,2]

It was Tessier who postulated the anatomy-based classification system for craniofacial clefts. He assigned a number to each craniofacial cleft on the basis of its position relative to the sagittal midline and the orbit. The Tessier type 3 cleft extends from the philtrum of the upper lip, through the wing of the nostril, and reaches the medial canthus of the eye. Tessier numbers 3, 4, and 5 are positioned through the maxilla and the orbital floor, while the Tessier number 6 runs from the orbit to the cheek bone. Tessier number 7 is positioned on the line between the corner of the mouth and the ear.[3] The combinations of Tessier's lateral and transverse clefts are extremely rare, and there are very few reports of such instances. The etiology of such complex craniofacial deformities can be best explained by the developmental field reassignment theory.[4] The loss of some crucial elements in the formation of craniofacial bone, by virtue of its downcascading nature, causes failure of formation and fusion of bones, leaving a huge defect.[4] The aim of this manuscript is to report a complex Tessier's cleft and to describe the staged management of the case.

Case Report

A 2-year-old male child of Iraqi parents was referred to our hospital for evaluation and correction of his facial deformity. The baby was the first child of healthy, nonconsanguineous parents (mother was 19 years old and father was 24 years old) delivered at 40 weeks' gestation by cesarean section, weighing 3775 g and 50 cm long. His 1- and 5-Apgar scores were 10. There was no history of any relevant abnormalities in the family and no teratogenic factors identifiable clinically through history. Prenatal ultrasonography reports were absent and hence deemed noncontributory. Since birth, from about the 2nd week of life, the child wore feeder plates and not much corrective surgery was attempted. This was the first surgical rehabilitation attempt.

An examination of the left side of the face revealed a wide unilateral cleft lip with soft-tissue deficiency involving the entire cheek region, left external ear, and macrostomia. The cleft passed through the lateral part, the alar, and vestibular areas of the nose and bone was almost absent or markedly hypoplastic and deficient. Three-dimensional (3D) computed tomography revealed the absence of left side of maxilla, palate, zygoma, orbital floor, and medial third of the orbital rim, left side mandibular ramus, condyle, and coronoid process (Pruzansky type III). An ophthalmic examination revealed the presence of

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a functional eyeball with prolapse of orbital contents due to the absence of an orbital floor. The presence of lateral and transverse facial clefts suggestive of Tessier's hard and soft-tissue cleft type 3 through 7 was identifiable [Figure 1a-d]. Owing to the presence of the defect, loss of left half maxilla, hypoplasia of the zygoma, sectional hypoplasia of temporal bone and mandible, and general hypoplasia of the structures innervated by the fifth and seventh cranial nerves were observed. Owing to the age of the child, hypoplasia of the salivary glands, particularly parotid, could not be assessed. Furthermore, the defective eyesight was attributed to the abnormal anatomy rather than the ocular defects [Figure 1c and d].

A treatment plan of the following order was devised. The first step was to close the soft-tissue defect by reconstructing the lateral part of the nose, ala, and upper lip. This would ensure the proper feeding of the child, help minimize social stigmatization, develop speech, and minimize recurrent infections. The second-stage surgery would be following the primary stage closely. It would include reconstructing the orbital floor and rim with bone grafts to correct ocular dystopia. The medial canthal ligament is to be repositioned using a Y plate canthopexy approach. The mandibular ramus and condyle were also to be reconstructed using a rib graft at the age of 4 followed by distraction, if required at a later stage once growth is completed. Ear reconstruction using rib graft from the 6th, 7th, and 8th rib with excision of skin tag would be performed once the growth is completed. If necessary, fat graft may be considered for creating a symmetrical effect.

Surgical procedure

After ensuring adequate fitness for surgery, the child was operated under standard general anesthesia.

- Stage 1: Reconstruction of the ala and lateral margin of the nose: An incision was placed along the lateral margin of the nose on the cleft side to rotate the lateral nasal wall and build the lateral nasal wall, nostril, and alar margins [Figure 2a-d]. On the upper lip, a rotation incision was placed along the philtral column. A subperiosteal flap was elevated on both sides of the cleft. A large Abbe-Estlander flap was raised and rotated from the lower lip preserving the blood supply (inferior labial artery) at one end to reconstruct the soft-tissue defect between the medial canthus and the lateral nasal wall and to create an upper lip. The Estlander flap was sutured to the defect using 4-0 vicryl and 5-0 and 6-0 ethilon sutures. A Z-plasty medial to the medial palpebral fissure may be required to reposition the eye in a more superior position
- Stage 2: Reconstruction of mandibular ramus and zygomatic region.

Under general anesthesia and orotracheal intubation, the right 5th and 6th rib were harvested through the anterior chest skin incision. Muscles retracted to expose the rib, periosteum carefully dissected by down and curved elevators, and

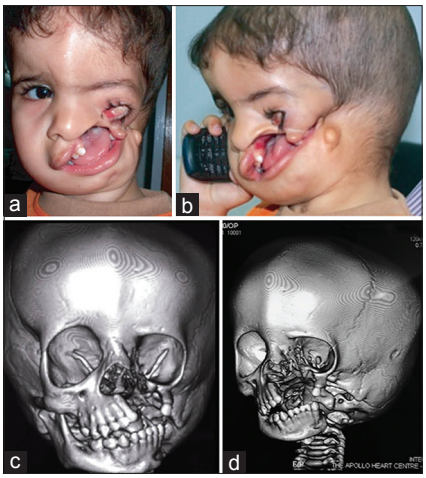

Figure 1: (a and b) Preoperative view showing Tessier's cleft numbers 3–7 and lateral view showing the cleft from commissures to preauricular region. (c) Three-dimensional image showing frontozygomatic deficiency. (d) Computed tomography showing complex distortion of middle-third of the face and loss of midfacial tissues. Note the soft tissue abnormalities

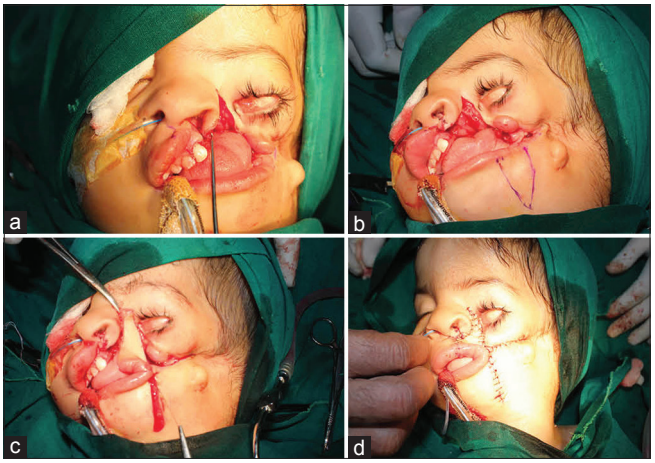

Figure 2: (a) Nasal soft tissue dissected to rotate nasal ala. (b) Abbe's flap. (c) Abbe's flap rotational advancement. (d) Immediate postoperative view of Stage 1 surgery

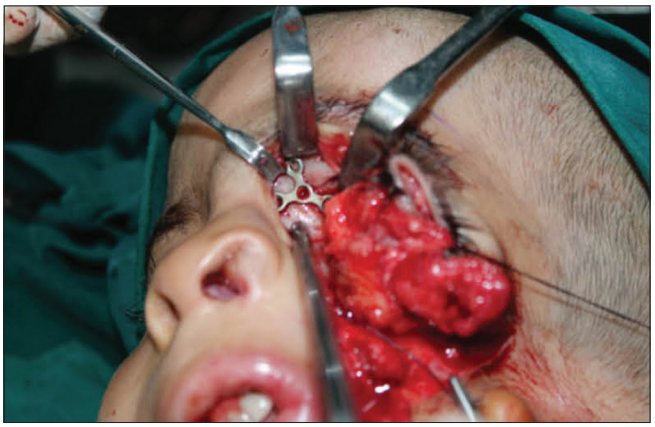

Figure 3: Placement and securing Y-plate in the medial canthal region

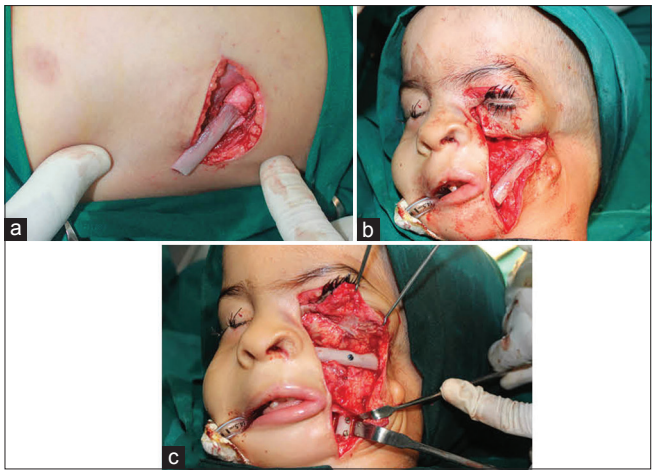

Figure 4: Stage 2 surgery, intraoperative pictures. (a) Rib graft harvested. (b) Ramus reconstruction with rib graft. (c) Reconstruction of zygoma and ramus

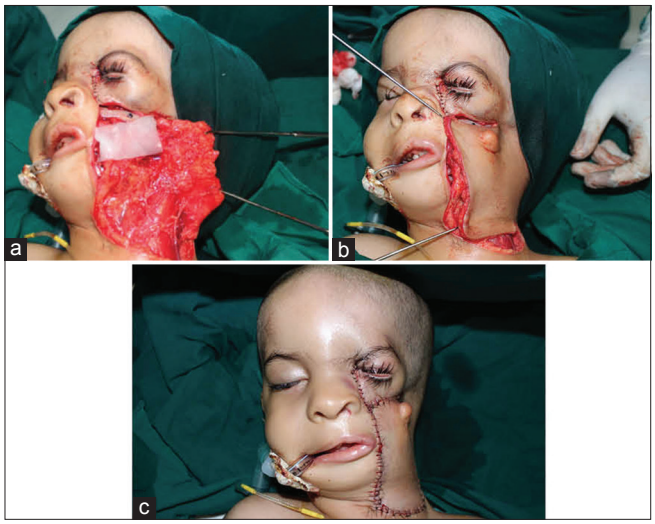

Figure 5: Stage 2 surgery, intraoperative pictures. (a) rhBMP2 to stimulate bone growth. (b) Approximation of the flap after graft placement. (c) Immediate postoperative view

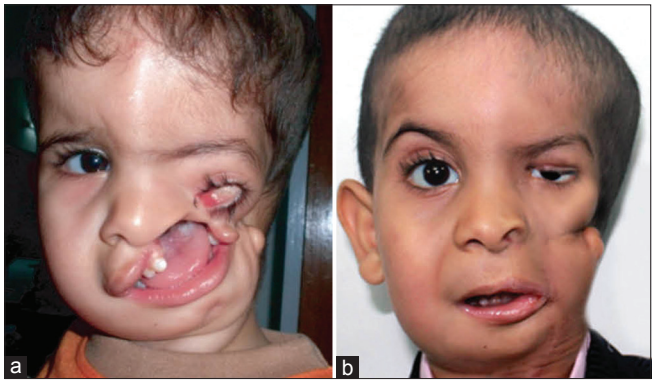

Figure 6: (a and b) Pre- and post-operative view

continuous palpation of the underlying rib are essential to avoid pleural injury. The 2 pieces of bone grafts were stored was temporarily in saline. Donor site was closed in layers. A Y-shaped incision was placed along the left lateral cheek with modified Z-plasty, involving the medial canthal region and a Y plate canthopexy done [Figure 3].[5] The lower arm of the Y-incision was further extended for adequate exposure for graft placement. In the mandibular ramus region, an incision placed 2 cm below the mandible, to preserve marginal mandibular branch of facial nerve. Grafts are used in the reconstruction of ramus of the mandible and zygomatic regions [Figures 4 and 5]. Later closure was done in layers. Appropriate antibiotics and NSAIDS cover were provided.

The patient recovered without any complication [Figure 6]. The left eye regained sight. The patient must now wait for up to 6 years and when the first molar erupts, checking of persistence of hypoplasia of jaws, zygoma, and temporal bone needs to be done and appropriate treatment needs to be instituted.

Discussion

The primary goal of the rehabilitation of the complex facial (transverse and oblique clefts) based on complex facial clefts as mentioned in literature is "(1) the initial priority is to protect the vision/corneas; (2) the soft-tissue component may be treated in the 1st month of life, to restore a relatively normal appearance to grotesquely deformed patients; (3) each component (lip, nasal, alveolar, ocular, and orbital deformities) should be evaluated and treated; and (4) because many of these patients do not have normal facial growth potential, it is unnecessary to wait for full facial development before starting surgical repair."[4,6]

As in the present case, severe cleft face malformation, besides causing difficulties in food intake and repeated, recurrent infections, also leads to psychological problems for both the parents due to the child's appearance and the child. The problem is further accentuated in marginalized societies as well as those suffering from civil unrest societies. Iraq region, for unknown reasons, has several incidences of complex craniofacial clefts.[7-9] Probably, a genetic approach will reveal peculiar mutations or traits in the population.

The etiology of oblique and lateral facial clefts is poorly understood. It is usually cited to the unsuccessful fusion of the mesoderm during embryonic processes. However, in areas where such fusion does not occur, as in the present case, lateral oroocular, some types of nasoocular, and medial oroocular clefts cannot be explained by this theory. In such situations, etiopathogenesis cited includes a primary stop of development, a neurovascular insufficiency or necrosis, or tears in the developing maxillary process. Literature also suggests that these malformations could be caused by a combination of directly tethered tissue migration (such as amniotic bands) and increased local pressure that leads to cellular ischemia.[1,9] The amniotic bands as a cause of complex lateral clefts were reported by Eppley *et al*.[2] Such failure of fusions may be complete or incomplete, can involve the palate, and sometimes extend into the temporal region. As reported in literature, most of the known cases of oblique facial clefts are sporadic with no syndromic association or gender predilection.[1]

The severe facial deformity in this patient was not diagnosed prenatally owing to several factors. Even in normal clinical situations, missing a complex facial cleft is not uncommon, when employing the standard 2D approaches. 3D ultrasound would be valuable for diagnosing atypical facial clefts.[1]

Each rare craniofacial cleft is unique and poses a different challenge. The rarity of the condition and the diverse presentation render developing a common diagnostic and treatment algorithm a challenging one.[3,5] In the present case, there was involvement of both soft and bony tissue abnormalities, so clinical and a detailed radiological examination of the maxillofacial region is needed to assess the morphology of the cleft and plan treatment. Tissue hypoplasia of the specialized facial structures of the nose, eyelids, or lips constitutes the greatest obstacle in restoring normal appearance and function. The complexity of the cleft increases by the involvement of many areas – nasal, ocular, sinus, palate, alveolus, and soft-tissue components. Hence, depending on the age of presentation and maturation/growth pattern of these structures, often, many operations are necessary for the correction of these defects.[4,6] Furthermore, depending on the defect, in few cases, the outcomes are not ideal. The treatment goal of these patients is to restore function and improve facial harmony. Conventionally, the correction of these defects involved multiple local cheek and nasal flaps for Tessier numbers 3 and 4 clefts. It was Resnick and Kawamoto who suggested a closure of the soft-tissue cleft of the Tessier number 4 with cheek and nasal flaps.[10] In certain situations, the use of a nasal flap was impossible due to a significant defect of the lateral nose. A straight-line cleft closure may be substituted. However, chance of occurrence of fistula (due to tissue tension) and the cheek scar deformation are relatively high. It was Toth *et al*. who suggested the use of a forehead tissue expander to provide sufficient local facial skin to allow the elevation and rotation of a flap and to restore the nasal structures.[11,12]

However, for the present case, an extended Estlander flap was the choice as it provides more flexible tissue for closing of defect along with natural skin color and facial hair characteristics. If needed in future, *Z*-plasty medial to the medial palpebral fissure may be carried out to position the eye once the growth stabilizes.[8]

Early bone grafting for complex clefts is controversial. According to Resnick and Kawamoto, bone grafting is preferable before the age of 5 years. However, early grafting increases the chance of graft resorption, leading to secondary bone augmentation in later stages of life. It is also said that the positioned grafts may prevent natural remodeling of the facial skeleton by abnormal muscular forces. Bony defect repair was carried out early, with rib graft as bone block from the iliac crest, which could be resorbed, if introduced at an earlier stage of development.[1,10] In the present case, loss of vision owing to anatomical defects was a possibility. Hence, rehabilitative measures were undertaken without due consideration of the resorptive potential. Postoperatively, the vision was restored and functional capabilities of eye were regained.

At present, the child has a stabilized closure of the hard and soft tissue, and it is proposed to wait till the child reaches 6 years of age when an assessment is performed again. Based on the outcome, the treatment plan will be approached again.

Conclusion

A rare case of lateral and oblique facial cleft is presented, which is the most complex and difficult to repair because of the absence of frontomaxillary process, intersinusonasal septation, deficiency of the nose, and distortion of the ala. The treatment algorithm at the age of 2 years for rehabilitation is presented. Early therapy will instill confidence in patients as well as the caregivers. Speech also will be modified by this early approach. As there are only a handful of cases with this extent of hard- and soft-tissue defect, more such reports only would help formulate strategies to address the rehabilitation of these patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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