Single Stage Correction of Bilateral Tessier 4 Cleft

Abstract

Tessier facial clefts are among the rarest facial clefts reported in literature and many contradicting issues have always been rising over the management and surgical approaches involved during the craniofacial cleft repair. Among the craniofacial clefts Tessier no. 4 is an extremely rare facial anomaly, and there are very few evidence which clearly describe the surgical approaches and techniques. Often these type of craniofacial clefts yield very poor surgical results, and they require multidisciplinary sequential corrective surgeries. This article presents a rare case of an 18-month-old baby with bilateral Tessier no. 4 clefts and its successful rehabilitation.

Keywords: Bilateral cleft, Facial clefts, Tessier 4

Introduction

Craniofacial clefts are the result of the disruption of the normal developmental program during embryogenesis affecting the face, the cranium, or both. Most of these clefts cause significant psychosocial ramifications and disfiguring of the face and cranial structures with either deficit or excessive tissues that cleave through the anatomic planes in a linear fashion. Very frequently, most cleft lips involve lip with or without nose and alveolar ridge but, facial clefts not only can involve these structures but also can split the bones and skin or make some anomalous facial features. They exist in varying degrees of severity, and they occur either as complete or incomplete forms fruste-type (characterized by minimal soft and bony tissue deformities) clefts and can appear alone or in association with other facial clefts. In addition to this, they can also appear as two different types of Tessier clefts on either side of the face. The incidence of rare craniofacial clefts compared with common cleft lip and palate malformations may range from 1.43 to 4.85 per 100,000 births.

A wide range of classification systems is proposed for cleft lip and palate. The complex clefts falling under the category of “Rare Craniofacial Clefts” are also classified by many authors differently. In 1976, Tessier presented a unique classification of craniofacial clefts which is generally the most accepted one for the description of all the complex rare craniofacial clefts. It is the most complete and has withstood the test of time which represents an orderly anatomic classification system in which all the clefts, major and minor and despite its position are considered and numbered from 0 to 14 with a number 30 for a medial symphysis in the mandible. This system significantly simplifies the nomenclature of clefts. It is a purely descriptive system not related to embryological or pathological factors.

Tessier no. 4 is often referred to as an oro-ocular cleft (American association of cleft palate rehabilitation by Harkins et al. in 1962) or medial maxillary dysplasia (Boo-Chai 1970) that begins lateral to the Cupid’s bow, between the tubercle and the oral commissure, lateral to the nasal ala skirting through the nose and passes onto the cheek extending into the lower eyelid lateral to the inferior punctum. Bony involvement begins between the lateral incisors and the canine tooth extending lateral to the pyriform aperture involving the maxillary sinus which ends at the infraorbital rim (lateral to the lacrimal groove and medial to the infraorbital foramen), all of which result in maxillary sinus exostrophy, inferior and medial globe prolapse and meloschisis.

Craniofacial clefts constitute the most challenging malformations as they are
never the same. Due to their complexity, the individual degree of cleft formation and the different structures and organs involved, these clefts should be managed appropriately to restore to near normalcy and to avoid complications, unnecessary anxiety, and exhaustion of the patient and family members. Successful reconstruction and rehabilitation need much more complex diagnostic and treatment modalities. The surgeon must be extremely skillful in craniofacial surgery, maxillofacial techniques, soft tissues procedures for soft tissue reconstruction, and no less important, have a solid background in the management of craniofacial anomalies.

Besides careful examination, imaging techniques are necessary to assess the individual degree of skeletal involvement in the cleft formation. For correct diagnosis, modern imaging techniques such as computed tomography (CT), magnetic resonance imaging, and three-dimensional CT allow better preoperative understanding of the problem and planning of the surgical procedures.

Considering these requirements I would like to present my experience in the surgical management of 18-month-old patient with rare facial cleft Tessier no. 4.

Case Report

An 18-month-old toddler with bilateral Tessier no. 4 cleft was referred to our hospital for rehabilitation.

On clinical examination, he had bilateral facial clefts extending laterally from the cupids bow, running lateral to the nasolacrimal duct and terminating medial to the inferior punctum which leads to deformation of the eyes and eyelids. The left eye was rudimentary with the loss of vision. The right upper eyelid had excess tissue that made it difficult for the baby to close the eyes. There was a lack of soft tissue to provide proper closure of the eyelids, exposure keratitis over the right eye was evident. Due to the extensive involvement of the eyes in the cleft defect, the nasolacrimal ducts were damaged causing epiphora or continuous tear flow from the eyes. The vision of the right eye was reduced to about 40% since birth. He also had cleft palate. The nostrils and nasal septum were symmetrical and normal. Because of the cleft, the child could not be fed normally and had no chance of good speech formation. The central part of cleft was attached to the upper lip. CT scan showed facial cleft bilaterally from the distal surface of the lateral incisors extending up to the orbit, but the orbital floor and the infraorbital rim were adequately present to support the globe. Hematological and biochemistry investigations were within the normal limits.

The treatment plan was formulated to surgically correct the soft tissue cleft for the patient is as follows:
- **Bilateral cleft lip repair**
- **Using multiple Z-plasties:**
  a. Repair of the eyelid component and
  b. The cheek component.
- **Prosthetically rehabilitate the eyes**

Surgical procedure

Using standard surgical protocols, GA was induced and maintained. The first step of incision was initiated with the prolabium (modified Paul Black’s technique) where the prolabium flap along with the vermillion was raised and hinged superiorly with two lateral prolabial flaps in the form of a fork which was later used in the closure of the facial cleft followed by, lateral lip Z-plasty done bilaterally. On the lateral lips, the mucosa was separated from the muscle layer followed by a vestibular incision and subperiosteal dissection was done below the cheek areas up to the zygoma region to advance the lateral lips medially for the closure of the facial clefts. At this stage, the mucosal layer from both the lateral lips was advanced and sutured at the center of the premaxilla followed by suturing of the muscle layer below the prolabial flap. Now, the prolabial flap was turned downward and sutured along the skin of the lateral lips which completed the lip closure.

Now, the Z-plasty incisions were marked at the lateral canthus of both the eyes for the correction of the lateral dystopia with excision of all the excessive tissues from the eyelids which was then dissected for approximation before the suturing. Fornix deepening sutures were placed to increase the depth of the socket of the left eye for accommodating the prosthetic eye. Finally, the medial orbitomaxillary cleft that’s extending above the cupids bow to the medial canthus of the eye was closed bilaterally using the lateral prolabial flaps turned upward, and extreme caution was taken while suturing to avoid injury to the nasolacrimal duct and the lacrimal punctum. Postoperative clinical outcome demonstrated excellent healing and good esthetic results [Figures 1-3].

Discussion

Tessier no. 4 is a very rare craniofacial cleft anomaly that requires a multidisciplinary staged reconstruction of both the soft as well as hard tissues. Tessier proposed a classification system based on the clinical experiences and observation, in which he numbered the clefts from 0 to 14.
that followed a well-defined “time zones.” The primary axis of this system of classification was defined by the eyelids and the orbits, dividing the face into upper and lower hemispheres. Tessier used these landmarks as orbit belonged to both the cranium as well as the face which separated the clefts into northbound or southbound clefts. All of the craniofacial clefts are formed by the combination of northbound and southbound clefts. The incidence of rare facial clefts is between 1.43 and 4.85 per 100,000 births and 9.5 and 34 per 1000 clefts.\[11\]

Among atypical craniofacial clefts, the no. 4 cleft is very rare and very fewer cases have been reported in the literature. It is characterized by a cleft lip, cleft palate, decreased oro-ocular and oculo-alar distance, eyelid colobomas, orbital dystopia, and inferiorly displaced medial canthus.\[3\] It is this rarity that has left most of the craniofacial clefts without the establishment of a comprehensive treatment plan. Ortiz-Monasterio et al.\[12\] investigated a series of 345 atypical craniofacial clefts, among which only eight were Tessier no. 4 cleft. Since then, very few cases of Tessier cleft have been reported in literature so far.

In general, the primary surgical repair should be limited to the soft tissue reconstruction. The skeletal reconstruction such as bone grafting or osteotomy should be postponed until preschool ages, because early manipulation of the bony skeleton may disturb the normal midfacial development. Several procedures for the primary repair of Tessier no. 4 cleft have been reported. Multiple local flaps are frequently discussed, but the “patchwork” scarring created by the interdiggitating flaps resulted in a conspicuous appearance. In this case discussed above, I have used modified Z-plasties\[10\] on the lateral cheek to close the defects and modified Paul Black’s technique\[9\] was used to correct the bilateral lip; aligning the vermillion and restoring the muscle continuity was the prime concern which showed a good postoperative result as it was the prime concern of the child’s parents. In my opinion, most of these clefts can be treated using local flaps.

Similar procedures were described in a study by Van der Meulen\[13\] who first described cheek advancement flap technique. This technique has the advantage that the resulting scars are made along the esthetic facial units. Furthermore, even a wide cleft can be easily approximated when the flap is dissected as far as the lateral canthal area.

Figure 3: (a and b) Markings of the incision. (c and d) Cheek flaps raised and medially transposed to cover the defects bilaterally. (e) Defects closed, excess eyelid tissues removed and prosthesis placed. (f) Immediate postoperative view. (g) Facial appearance after suture removal

In 1990, Resnick and Kawamoto\[15\] described the surgical treatment of this congenital anomaly which included: (1) medial canthopexy and lower eyelid reconstruction to protect the eye, which may be an emergency procedure; (2) bone grafting for orbital bony continuity and maxillary

![Figure 2: Preoperative three-dimensional computed tomography. (a) Frontal view, (b and c) left and right lateral views](image-url)
bony deformities; (3) reconstruction of the soft tissue defects of the cheek (if local flaps do not suffice, tissue expansion may be a versatile technique for reconstruction); (4) cleft lip repair; (5) serial orbital conformers for orbital expansion of anophthalmic or microphthalmic orbits; and (6) if required, multiple surgical revisions may be performed for bone and soft tissue deformities as correction of medial and lateral canthal ligament malposition, treatment of lower eyelid defects, and augmentation of anterior maxillary region and the zygoma.

When coming to the eye defects in the above case, it was challenging as right eye had a blurred vision and left eye was a lazy eye. Coloboma was excised, and lid correction was done. Sulcus deepening was done in the left eye to provide a shelf for the artificial prosthesis. Cover shell of the prosthesis should have a proper fit. Ocular prosthesis provided aesthetic and a pleasing appearance. Similarly, Kawamoto[16] described that the eye is usually present and functional in Tessier no. 4 cleft, several cases accompanied with microphthalmia or anophthalmia have been reported in the literature.[16‑18] Most of these cases were unilateral or asymmetrical cleft, but bilateral no. 4 cleft associated with microphthalmia or anophthalmia is extremely rare, as was seen in our case. The soft tissue deficiency of the lower eyelid was not satisfactorily reconstructed. It is suggested that any single flap is not enough for the eyelid reconstruction in such a wide cleft as in our cases.

Correction with the other local flaps will be planned in the near future. Tissue expansion or free tissue transfer are good alternatives for soft tissue reconstruction. Tissue expansion also adds ideal similar skin color, texture, appearance, and aids in a perfect outcome of scar revisions.[19,20]

**Conclusion**

In the above case, surgical result in an 18 month old boy with tessiers no 4 was presented. There is no prescribed technique; every case is individually tailored. It is valuable to maximize the extent of diagnostic options before surgery to achieve complete correction of the malformations. Early repair using autogenous tissues and minimal discarding of healthy tissues as much as possible is recommended. Regular follow-up of these patients is also very important for the successful outcome of the surgical treatment.

**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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