A chronicle of Tessier no. 0 and 1 facial cleft and its surgical management

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Abstract Description and successful management of a patient with Tessier no. 0 and 1 facial cleft is being presented. This bizarre coexistence of two of the rare facial clefts in a single patient deserves a place in the cleft database. Appropriate evaluation was done to rule out the presence of median cleft face syndrome. Lip cleft was repaired by straight line technique with staggering at the vermilion border. Nasal cleft was reconstructed by replacing the missing lower lateral cartilage with conchal cartilage via endonasal approach. Satisfactory outcome was achieved for this singular deformity by conforming to the basic tenets of plastic surgery.

Keywords Facial cleft · Tessier no.0 · Tessier no.1

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Introduction

Cleft surgery is the perfect blend of art and science; designing a beautiful cupid's bow requires an artist's imagination while science lies in understanding the dysfunctional cleft anatomy and restoring it. Tessier P [19] classified the rare facial clefts, a colossal effort, which simplifies the management of these patients. Still rarities keep appearing which cross the defined boundaries and cast a doubt on the completeness of these classifications. Despite this, an undeniable superior contemplation of all aspects of the dysmorphogenesis is achieved when one follows the footsteps of the legendary author. It is immensely difficult to perceive the morphogenesis of a rare cleft which spans a zigzag path through the embryological processes of the foetal head enlarge. This patient presented to us with a bizarre facial cleft which was classified as a combination of Tessier no. 0 and 1 cleft. Our perplexities, difficulties and finally successful management of this singular case are being described in the following lines.

Case description

A 17-year-old girl from poor socioeconomic status presented to our clinic for esthetic repair of her deformed

upper lip and nose. On eliciting, it was found that she was born from a healthy mother after full term pregnancy via spontaneous vaginal delivery. She had no siblings with clefts. Her infancy and childhood were uneventful except for the ridicule and pathos consequent to the facial stigmata. She had achieved expected intellectual and physical milestones commensurate with her age. She had normal forehead with no ocular abnormality but for the squint in her left eye. The dome of left ala was collapsed with a shortened nostril. There was a groove between the dorsum of the nose and the deformed ala. The nasolabial area was apparently normal as were the lower eyelid and infraorbital regions. The columella was normal dimensionally but was slightly deviated to the right side. The upper lip vermilion was parted in the centre with widened philtrum. The distance between the two philtral columns was 23mm at the columellar end and 28mm in between the peak of cupid's bow. Intraorally the maxillary central incisors were carious and rotated. Slight depression was palpable in the intact alveolus in the midline. Rest of the oral structures was normal. Computed tomography of the craniomaxillofacial skeleton was performed to rule out median cleft face syndrome and holoprosencephaly. All the midline structures except the upper lip and alveolus were inviolate. The left alar

dome was collapsed with missing lower lateral cartilage (Fig. 1a, b, c, d). Based upon these findings, a diagnosis of Tessier no. 0 and 1 facial cleft was assigned to this patient.

Morphopathogenesis

It is very difficult to predict the exact sequence of events that must have occurred in this particular patient during the fetal period. At best we can speculate the dysmorphogenetic process based upon the current knowledge of normal embryogenesis. Sulik [18] explains atypical clefts by the variability in the location of ossification centres. He states that more familiar Tessier clefts are formed at the seam of two minor ossification centres. Urata and Kawamoto [21] have reviewed the median clefts of upper lip and described them to various factors like infectious diseases, drugs, vascular compromise, radiation, and external mechanical factors. Losee et al. [11] have reviewed the embryologic aspects of nose.

To avoid parroting, we summarize that the middle part of the upper lip is derived from the frontonasal process, specifically the ectodermal nasal placodes and the elevated mesodermal ridge surrounding it. The lateral nasal process forms the ala of the nose, lateral crus of lower lateral

cartilage, upper lateral cartilage, nasal bones and external wall of nose. The median nasal process ultimately gives rise to one half of the nasal septum and medial crus of lower lateral cartilage (Skevas [17]). The caudal portion of the two median nasal processes i.e. the globular process gives rise to the primary palate, nasal tip, columella, prolabium and frenulum (Apesos [2], Litersarson [10]).

It will not be too impudent to theorize that the primary insult in our case must have been to the lateral nasal process resulting in

1. Flawed morphogenesis of the destined structures (missing lateral crus)
2. Impeded inferior and medial migration of the nasal placode to its final position.

Consequently the philtral columns and adjacent vermillion were not hypoplastic but the philtrum was widened with cleft in the vermillion.

However, exactly how and why this occurred and what must have been the primary insult is still left to conjecture and speculation as no positive history of any known etiologic agent was elicited.

Management

Appropriate management of any facial cleft hinges upon recognition of the deficient

Fig. 1a Preoperative front view

Fig. 1b Preoperative basal view

Fig. 1c CT scan axial section

Fig. 1d CT scan coronal section

part of the framework. When considering the frequently seen clefts of lip and palate our mind automatically analyzes the hypoplastic area and visualizes the

reconstruction utilizing the familiar repair techniques. This mechanized mental exercise dawns a more conscious robe when we encounter its rarer counterpart. Initial effort is directed upon determining the flawed parts and then attention is drawn to plan the repair technique. The missing lower lateral cartilage was restored by a free graft from the auricle. We used the anterior auricular approach for harvesting the graft and endonasal for placing it. Philtrum was refurbished to its normal dimension by excising the excess tissue. A full vermillion was created by designing vermillion flaps which were joined by a staggered line. There was no indication for opening the alveolar cleft. The entire procedure was carried out under LA and sedation. The steps of the surgery are depicted in the photo sequence (Figs. 2a, 2b, 2c, 2d). The patient tolerated the procedure well and good postoperative results were achieved (Figs. 3a and 3b). Subsequently the patient was referred to ophthalmic surgeon for squint correction.

Fig. 2a Incision markings

Fig. 2b Auricular cartilage harvest

Fig. 2c Cartilage graft

Fig. 2d Immediate postop

Discussion

A rare case is akin to an unsolved riddle and never fails to fascinate the acumen of a witty surgeon. It stimulates us to leap out of the slumber of commonplace, but in no

Fig. 3a Postoperative front view

Fig. 3b Postoperative basal view

way inferior surgical problems. When managed successfully such patients become beautiful feathers in the cap of our surgical careers.

Initial attempts to describe the rare facial clefts were done by Davis [4], Blackfield and Wilde [3], Fogh-Anderson [5], Popescu [15], Pitanguy [14] and more recently by Shewmake and Kawamoto [16]. Numerous attempts have been made to classify the facial clefts. In 1962, Harkins and associates [6] proposed a classification system endorsed by American association of cleft palate rehabilitation (AACPR). In 1966, Karik [8] proposed a detailed classification of rare craniofacial clefts based on embryologic and morphologic criteria.

The most accepted and purely utilitarian categorization of facial clefts was given by Paul Tessier [19] and Kawamoto [9]. The clefts are numbered from 0 to 14 starting in the midline and using the orbit as the reference point. According to this scheme no.0 and 14 constitute median craniofacial dysrhabia. The cleft goes through the frontal bone with duplication of the crista galli, the midline of nose with nasal septal duplication, the columella, midline of upper lip and maxilla. No.1 is a paramedian craniofacial cleft. It comes through the dome of alar cartilage and frontal bone, olfactory groove of cribriform plate, between the nasal bone and frontal process of maxilla. All the patients with midline dysmorphogenesis must be evaluated for the presence of holoprosencephaly and median cleft face syndrome.

Thus, juxtaposing the feature in our case it will be apt to designate it as 'forme fruste' of Tessier no.0 and 1 facial cleft.

The rarity and diverse presentation of the median cleft bear the burden and blame for the limited volume of literature dedicated to the surgical management of this entity (Urata [21] and Kawamoto [9]). The correction of a midline cleft of upper

lip has been described by various authors (Millard [12], Iregbulen [7], Nakamura et al. [13], Apesos [2], Akoz [1], Turkaslan et al [20]). We adopted the technique described by Apesos [2].

Shewmake [16] and Kawamoto [9] have nicely elucidated the principles of management of cleft nose. They have concluded that each nasal cleft is unique and should be completely evaluated to determine the specific degree of anatomic pathology. We decided to use the conchal cartilage as a substitute for the alar cartilage and restore the symmetry of alar dome.

Thus, the correction of this rare surgical case derives its design and emphasizes upon the golden rules of diagnosing before treating and replacing like with like after restoring the displaced to its normal position. These axioms of plastic repair always come handy while managing a stereotype case.

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