Journal of Cranio-Maxillo-Facial Surgery 41 (2013) 598–602

Contents lists available at SciVerse ScienceDirect

Journal of Cranio-Maxillo-Facial Surgery

journal homepage: www.jcmfs.com

Severe bilateral Tessier 3 clefts in a Uighur girl: The significance and surgical repair

Dandan Wu [a,b], Guomin Wang [a,b,*], Yusheng Yang [a,b], Yang Chen [a,b], Teng Wan [a,b]

[a] Center for Cleft Lip and Palate, Department of Oral & Cranio-Maxillofacial Science, Ninth People's Hospital, Shanghai Jiao Tong University, School of Medicine, PR China
[b] Shanghai Key Laboratory of Stomatology, Shanghai 200011, PR China

ARTICLE INFO

Article history:
Paper received 24 May 2012
Accepted 20 November 2012

Keywords:
Bilateral Tessier 3 clefts
Severity
Surgical repair
Further treatment

ABSTRACT

The Tessier 3 cleft is one of the rarest congenital craniofacial clefts, which often extends through the upper lip, the alar groove and the medial canthus. Only a few cases have been reported. There is no standardized method for the surgical treatment for this condition in the literature, and to obtain an acceptable outcome is difficult. A Uighur girl with severe bilateral Tessier 3 clefts and associated orofacial deformities is described here, and a novel protocol for clefts of this severity and rarity is presented. This study focuses particularly on describing the surgical procedures and techniques. Further treatments required for the cleft-associated deformities during later growth and developmental stages are also discussed in detail.

1. Introduction

Oblique facial clefts, which were classified into types 3, 4, and 5 by Tessier (1976), are an extremely rare and disfiguring congenital anomaly of the face. The Tessier 3 cleft is the most medial type of oblique facial cleft, and is known as a naso-ocular cleft by the American Association for Cleft Palate Rehabilitation (Natsume et al., 1999). The cleft often extends from the philtrum of the upper lip, through the wing of the nostril and reaches the medial canthus of the eye, with associated deformities such as cleft palate, nasolacrimal abnormality, disruption of the medial wall of the antrum, coloboma of the lower eyelid, anophthalmia, microphthalmia and telorbitism (Gawrych et al., 2010).

The aetiology of the cleft is not very clear, but may relate to drug ingestion, nutritional insufficiency or disease during pregnancy. It is thought to occur sporadically without any familial or gender association, and no abnormal karyotype has been detected by chromosomal investigation in existing patients (Wenbin et al., 2007). Its exact incidence is unknown, but the overall incidence of oblique facial clefts ranges from 0.75 to 5.4 per 100 common clefts (Mavili et al., 1992). Due to its rarity, only a few cases of Tessier 3 cleft have been published, especially in the bilateral form (Table 1). Each case is unique and surgical management of such cases is still a challenge.

Here, we present a Uighur girl with severe bilateral Tessier 3 clefts and other associated orofacial deformities. We also provide a detailed description of surgical procedures and techniques, which may be used for the treatment of these severe and rare types of clefts. The aim of this study is to provide guidance for surgical repairs in treating this rare craniofacial deformity.

2. Patient and methods

The study was carried out according to the principles of the Declaration of Helsinki; informed consent was obtained and Shanghai Ninth People's Hospital Ethics Committee approved the study. Permission to use these images herein has been obtained from the parents of the girl who participated in this study.

A Uighur female infant was first referred for repair of bilateral Tessier 3 clefts to the Centre for Cleft Lip and Palate at 20 months of age. The patient was the second child of a healthy 33-year-old father and a non-consanguineous 30-year-old mother, with a full-term normal vaginal delivery. The birth weight was 2250 g. There were no identifiable teratogenic factors other than dystrophia in the mother during her pregnancy, no family history of clefts or other diseases, and the elder sister is healthy.

On examination, the facial deformities revealed symmetrical bilateral oblique clefts extending from the vermilion of the upper lips, through the bilateral alar grooves, up to the lower eyelids adjacent to both medial canthi. Additional deformities included

* Corresponding author. Center for Cleft Lip and Palate, Department of Oral & Cranio-Maxillofacial Science, Ninth People's Hospital, Shanghai Jiao Tong University, School of Medicine, No. 639 Zhi Zao Ju Road, Shanghai 200011, PR China. Tel.: +86 (0) 21 23271699x5149; fax: +86 (0) 21 53072423.
E-mail address: guomin@sh163.net (G. Wang).

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http://dx.doi.org/10.1016/j.jcms.2012.11.040

Table 1
Reported cases of bilateral Tessier 3 clefts.

Case	Clinical features	Authors and year
1	Bilateral Tessier 3 clefts with severe microphthalmia and a protruding premaxilla	Schlenker et al., 1979
2	Symmetrical bilateral Tessier 3 clefts	Komatsu et al., 1999
3	Bilateral Tessier 3 clefts, complete on the right and incomplete on the left side in combination with a no. 0 cleft	Sieg et al., 2004
4	Bilateral Tessier 1–13 and 3–11 clefts with bilateral anophthalmia, severe orbital hypertelorism, and a premaxilla located at the level of the orbits	(Monasterio and Taylor, 2008)
5	Bilateral Tessier 3 clefts with no clefting of the secondary palate	Mishra and Purwar, 2009
6	Milder bilateral Tessier 3 clefts with normal premaxilla and rudimentary prolabium	Mishra and Purwar, 2009
7	Bilateral Tessier 3 clefts with left anophthalmia	da Silva Freitas et al., 2010
8	A right number 3, 11 cleft and a left number 3, 10 cleft with an expanding fronto-orbital encephalocoele	Wan et al., 2011
9	Bilateral Tessier 3 clefts with both anophthalmia	Sesenna et al., 2012
10	Symmetrical bilateral Tessier 3 clefts with rudimentary prolabium	Chen et al., 2012

bilateral defects of the nasal ala, inferior displacement of medial canthi, coloboma of the lower eyelids, and a conspicuously protruding prolabium. Intraoral examination showed bilateral wide alveolar and palatal clefts with absence of deciduous lateral incisors (Fig. 1). Ophthalmologic examination revealed blepharal dysraphism of both eyes and obstruction of the nasolacrimal ducts. There were no other congenital anomalies. Craniofacial three-dimensional CT demonstrated that the bony clefts extending through the alveolus of the lateral incisor areas, both nasomaxillary sutures, up to the bilateral inferomedial walls of the orbits, corresponded to the soft tissue defects, and also showed a severely protruding premaxilla (Fig. 2). Measurements of the width of the clefts between the prolabium and the lateral labium (30 mm), and between the premaxilla and the lateral maxilla (35 mm) were taken.

Because of the complexity of the deformities, the patient was assessed by a multidisciplinary team consisting of oral and maxillofacial surgeons, plastic surgeons, orthodontists, ophthalmologists, and otorhinolaryngologists. A protocol of sequential treatments was determined, and staged surgical procedures were chosen. The first operation for closing the bilateral cleft lips was performed under general anaesthesia. The lip landmarks and incision lines were marked as in the reconstruction of normal bilateral cleft lips. Due to the wider clefts, osteotomy of the anterior part of the vomer bone was performed, and a bone block of approximately 7 mm in length was removed to retrude the protruding premaxilla. The lateral lips were released supra-periosteally, and the orbicularis oris was dissected bilaterally adequately and reconstituted properly. Finally, the prolabium and the lateral labia were sutured together, and reconstruction of the vermilion tubercle was finished by resetting the muscles to build vermilion in the midline with an everted profile. The labial arch was used to reduce tension after surgery. The wound healed well, and the patient was discharged with sutures removed 1 week postoperatively.

1.5 years later, the child with a good configuration of the upper lip was re-admitted for the second phase of the surgery (Fig. 3A). The maximal width of the clefts had shortened to 25 mm. With the patient safely under general anaesthesia, the incision lines, which consisted of nine flaps as key elements (a superiorly-based nasofrontal flap, two inferiorly-based nasal flaps, two medially-based cheek flaps, two laterally-based cheek flaps, and two upper eyelid flaps) were marked as shown in Fig. 3B. The superiorly-based nasofrontal flap was raised off the underlying nasal bone and the maxillary process of the frontal bone to allow downward advancement of the shortened nasal tissue. The bilateral small inferiorly-based nasal flaps were then elevated from the nasal cartilages and prepared for lateral and inferior rotation to correct the nasal ala. Following this the cheek flap on each side was separated into two flaps along the incision lines. The laterally-based cheek flap with a horizontal incision parallelled to the lower lid margin was released from the anterior wall of the maxilla on the periosteum to allow its medial advancement without creating undue tension, and the displaced facial musculature was dissected and freed from abnormal attachments. Care was taken to identify and preserve the inferior orbital neurovascular bundle. The medially-based cheek flap was then turned over to be nasal lining used for filling the gap. The nasalis muscles were re-orientated and sutured to the facial muscle groups to restore a natural facial expression. Next, medial canthoplasty was performed by fixation of the remnant medial canthal ligament to the paranasal periosteum, repositioning of the orbicularis oculi, and superior rotation of the laterally-based cheek flap to combine with the upper eyelid flap. Finally, the laterally-based cheek flap was advanced medially to suture with the superiorly-based nasofrontal flap along the junctions of facial units and the inferiorly-based nasal flap which had been rotated to correct the nostril shape. The patient tolerated the procedure well, and her recovery was uneventful with no complications (Fig. 4A). At 6 years of age, the patient underwent palatoplasty.

3. Results

The wide facial and palatal clefts were completely closured and the defective nasal ala and the dislocated medial canthi effectively reconstructed. The patient had an acceptable facial appearance with inconspicuous scars and natural facial expression (Fig. 4B). The outcomes of these operations were functionally and aesthetically satisfactory.

4. Discussion

Tessier (1976) proposed a classification of facial, cranio-facial and latero-facial clefts by numbering from 0 to 14 around the orbit, counterclockwise, to indicate the location of cleft. According to the classification, the Tessier 3 cleft is characterized by cleft lip and palate, superior displacement of the alar base, inferior displacement of the medial canthus, coloboma of the lower eyelid, nasolacrimal abnormality, disruption of the medial wall of the antrum, cleft of the inferomedial wall of the orbit, and telorbitism.

Embryologically, the commonly accepted primary mechanism of cleft formation is attributed to failed fusion of the mesoderm between the lateral nasal process which arises from the frontonasal process and the maxillary process, while the secondary mechanism relates to tissue destruction caused by amniotic bands (Gawrych et al., 2010). Proponents of the first theory believe that focal foetal dysplasia is the primary factor underlying the cleft and some manifestations would be difficult to explain by a band-related mechanism. The pattern of the cleft, particularly in bilateral symmetrical clefts, although subject to slight variation, is usually consistent, and could not be caused by randomly distributed bands. Several associated anomalies, such as cleft palate, anophthalmia, and microphthalmia, cannot be explained by amniotic bands. However, some other associated deformities, such as constriction rings, visceral and extremity defects, are considered to be the result of constriction by amniotic bands (Mavili et al., 1992).