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The Tessier number 3 cleft: A report of 10 cases and review of literature

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Summary The Tessier number 3 cleft is one of the most intricate and destructive of all facial clefts, presenting surgeons with a difficult task for reconstruction. We present a series of 10 patients with this rare cleft all treated by a single surgeon over 30 years. All patients with Tessier number 3 clefts treated between 1978 and 2008 by the senior surgeon were reviewed. Demographic data and all associated clinical findings including cranial and extracranial anomalies were recorded. Methods used to reconstruct each patient were also noted. Seven males and three females were identified and age at initial treatment ranged from 12 months to 12 years. Mean follow-up was 6.3 years. Multiple craniofacial anomalies were appreciated including other rare facial clefts, hypertelorbitism, lacrimal obstruction, anophthalmia, choanal atresia, and hemifacial microsomia. Amniotic banding was the most prominent extracranial finding noted in these patients. Tessier number 3 clefts can be associated with multiple other craniofacial anomalies making reconstruction challenging. Soft tissue and bony reconstruction must be considered separately, and a variety of tools may be employed to accomplish each goal. As the presentation can be highly variable, an individualized treatment plan must be made to meet each patient's specific needs.

Introduction

Historically, the first case of an oblique facial cleft was recorded in Latin by von Kulmus in 1732.[1] In 1823, Laroche made the distinction between what he called cheek clefts and ordinary clefts of the lip.[2,3] Subsequently, Walter Dick of Glasgow described the first case of an oblique facial cleft

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in the English medical literature. Pelvet later separated oblique facial clefts involving the nose from other cheek clefts.[1] In addition, in 1887, Morian provided a classification for oblique facial clefts, dividing them into naso-ocluar clefts which extended from the nostril to the lower eyelid, and oro-ocluar clefts which extended from the eye to the lip.[1,2] This classification was later adopted in 1962 by the Nomenclature Committee of the American Association for Cleft Palate Rehabilitation.

Paul Tessier provided one of the most elaborate and comprehensive numerical classification systems for rare facial clefts in 1976.[4] In his scheme, Tessier assigned numbers to each craniofacial cleft on the basis of its position relative to the sagittal midline and the orbit. Based on this classification, the number 3 cleft extends from the philtrum of the lip to the medial canthus of the eye, with foreshortening of this distance.[5] The bony cleft occurs at the lateral incisor/canine area of the alveolus, extending through the frontal process of the maxilla to the lacrimal groove of the medial orbit.[6] Soft tissue defects, including colobomas of the nasal ala and lower eyelid, and an inferiorly displaced medial canthus and globe, are characteristic.[6] Concurrent absence or dysfunction of the nasolacrimal system is predictably high.[7]

The etiology of rare facial clefts remains unknown. Lack of fusion, insufficient mesodermal penetration, or failure of the naso-optic groove to invaginate and form the tubular nasolacrimal system, however, have all been suggested.[8,9] The exact incidence of Tessier number 3 clefts is unknown, but cases are typically sporadic with no syndromic association or sex predilection.[7] Complete, incomplete, unilateral, and bilateral forms have all been described in the literature.[1,9] Furthermore, Tessier clefts number 7, 9, 10, or 11 have been associated with number 3 clefts as the cranial extension.[1,9] Frequently, patients with these clefts have also been found to have epibulbar cysts. Given these findings, the number 3 cleft is perhaps the most intricate and destructive of all the facial clefts.[4]

As Tessier number 3 clefts rank near the top in challenge and degree of difficulty presented to the reconstructive surgeon, treatment is often individualized to the patient. This article aims to describe different clinical features and associated malformations in 10 patients with the rare number 3 craniofacial cleft, as well as the obstacles faced in functional and aesthetic restoration. These patients span 30 years of experience by the senior author (HKK) and highlight the evolution in surgical treatment.

Patients and methods

We reviewed all Tessier number 3 craniofacial cleft cases seen and treated by the senior author (HKK) during the past 30 years (1978—2008). The assessment included retrospective evaluation of demographic data (age of initial treatment and sex distribution), prenatal history (infection, exposure to radiation, chemicals, or medications), and family history of craniofacial anomalies. All clinical findings including cranial and extracranial anomalies were recorded. Different surgical procedures employed to correct deformities were recorded. The surgical treatment varied according to each case, time of presentation, severity of the clefting condition, and associated cranial and extracranial anomalies. Preoperative and postoperative photographs were taken of patients for diagnostic purposes and for evaluation of different surgical treatments.

Results

The study included 10 patients, seven males and three females; their age at initial treatment ranged from 6 weeks to 20 years. Mean follow-up was 6.3 years, ranging from 12 months to 12 years. Three patients were previously operated on by other surgeons before initial evaluation. In all patients, antenatal history was negative for serious infections, exposure to radiation, chemicals, or illicit drugs. All patients had no family history of craniofacial malformations. One patient was noted to have associated craniosynostosis which was released during early childhood.

On physical examination, five cases were affected on the right side of the face, three on the left, and two bilaterally. The cleft was complete in six patients and incomplete in four patients. Of the four incomplete clefts, one patient had medial canthus dystopia, one was noted to have a coloboma of the nasal ala, and two were found have both alar and medial lid colobomas with foreshortened naso-ocluar distances. The lacrimal system was obstructed in four patients. Associated cranial and extracranial anomalies are summarized in Table 1.

Other rare facial clefts were present in six patients. Of the two patients with bilateral involvement, one was noted to also have bilateral Tessier number 10 clefts as the cranial extension. Among unilateral cases, cranial extension of the number 3 cleft was present in three patients, assuming the form of number 10, 11, or 13 clefts. A contralateral Tessier number 4 cleft was present in one case with a number 10 northbound extension. Number 7 facial clefts with other features of hemifacial microsomia were found in three patients. Interestingly, the number 7 clefts were noted to be ipsilateral to the number 3 cleft in two patients and contralateral to the number 3 cleft in one. An associated midline 0 cleft with a bifid nose was found in one patient. Bilateral cleft lip and palate was noted in five patients, two of whom had bilateral number 3 clefts while the other three patients had only unilateral involvement (Table 1).

Hypertelorbitism was present in three patients. Unilateral anophthalmia was present in one case who also had right ear grade III microtia, a right transverse number 7 facial cleft, an absent right mandibular ramus, hydrocephalus, a low-set left ear with preauricular tags, and a large ventricular septal defect (VSD). This patient only underwent placement of a ventriculoperitoneal shunt and died before any further craniofacial surgery was performed (Figure 1).

Bilateral microphthalmos was present in one patient who also had left choanal atresia, left mandibular hypoplasia, left mala hypoplasia, left orbital dystopia, a protruding left ear, and a plagiocephalic skull with synostosis release. Left torticollis with kyphosis and scoliosis were also noted. Finally, this patient was found to have blindness in the right eye with limited vision in the left.

With respect to limb malformations, three patients had amniotic band syndrome. Auto-amputation of multiple