Surgical correction of Tessier number 10 cleft
Correção cirúrgica de fenda número 10 de Tessier

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RESUMO


SUMMARY

Purpose: To review different surgical techniques used to manage patients presenting Tessier cleft #10 and associated deformities. Method: A retrospective study including Tessier #10 cleft patients treated between 1996 and 2009, in Hospital de Clínicas of Federal University of Paraná and Assistance Center for Cleft Lip and Palate (CAIF) – Curitiba, PR. Results: There were ten patients with Tessier #10 cleft, five presented as isolated deformity, three had association between cleft #10 and 7, and two cases of Tessier #10 and 2 cleft. The main craniofacial deformities diagnosed were: upper eyelid coloboma (7), frontal encephalocele (4), amblyopia (4), macroblefaron (3), hipertelorism and orbital dystopia (3), and frontal hairline alteration (4). Six cases of upper eyelid coloboma were treated with primary closure, and one case with Cutler-Beard flap. Patients with frontal encephalocele were submitted to the resection of the herniated cerebral tissue and calvarium reconstruction. Conclusions: The Tessier # 10 cleft has two presentation forms (eyelid coloboma and frontal encephalocele). It is the first exclusive study on this rare malformation.

INTRODUCTION

Craniofacial clefts are rare deformities of unknown cause. The incidence range from 1.4 to 4.9 per 100,000 live births. The exact cause of craniofacial clefts is unknown and those theories of cleft formation that apply to typical cases of cleft lip and palate may also be applicable to the craniofacial clefts. It is probably caused by bone incomplete fusion or primary ossification centers failure during the embryonic development. Tessier classification system is based on anatomical parameters and is universally accepted to describe rare craniofacial clefts.

The bony defect of Tessier #10 cleft patients is located on the upper central orbit, lateral to supraorbital nerve, with extension through orbital roof and frontal bone, it may be associated to frontal encephalocele (Figure 1). The upper eyelid coloboma is located on medial third, and it may be the form of commitment of soft tissues. The eyebrow may be divided into two regions: lateral with vertical positioning, sometimes together with the hairline and the medial region which is atrophic and occasionally absent. Tessier described that cleft #10 represents the northbound of cleft #4.

Few studies on rare craniofacial clefts were published. Specifically on cleft #10, we did not find in the English literature studies or series on this subject. Tessier et al., in their book, presented methods of repair of some of these deformities. Our study consists in a revision of different surgical techniques used to manage patients presenting cleft #10 and associated deformities.

METHODS

This is a retrospective study including Tessier #10 cleft patients treated between 1996 and 2009, in Hospital de Clínicas of Federal University of Paraná and Assistance Center for Cleft Lip and Palate (CAIF) – Curitiba, PR.

Data were collected including soft tissues and osseous commitment.

All the tomographic studies and surgical procedures were re-evaluated.

RESULTS

There were ten patients with Tessier #10 cleft, five presented as isolated deformity, three had association between cleft #10 and 7, and two cases of Tessier #10 and 2 cleft (Table 1). The craniofacial deformities diagnosed were: upper eyelid coloboma (7), frontal encephalocele (4), amblyopia (4), macroblefaron (3), hyperelorism and orbital dystopia (3), and frontal hairline alteration (4). There was also association with other malformation such as: preauricular appendix (3), microtia (1), cleft lip and palate (2), macrostomia (3), mandibular branch agenesis (1), external ear auditory conduct atresia (1), lacrimal duct bifid (1), hemifacial microsomia (1).

Three patients were only evaluated in our service, but they were not operated. There were seven patients with upper eyelid coloboma (Figure 2), six were treated with primary closure, and one case was treated in another service with Cutler-Beard flap. There were 2 cases of leukoma, prior to our intervention. The displaced eyebrow was treated with the remaining eyebrow flap rotation, hair transplantation and tattooing. The loss of frontal hairline was corrected with the resection of the committed area. The number of surgical procedures per patient ranged from two to ten, according to the deformity degree.

Patients with frontal encephalocele needed the resection of the herniated cerebral tissue, associated to calvarium reconstruction (Figure 3). Two patients were grafted with inner layer of parietal bone, after splitting. One patient, who received previously split costal graft in another hospital, was submitted to alloplastic implant of porous polyethylene, to solve the contour irregularity. The forth case of encephalocele was only evaluated in our service, and was not submitted to surgical procedure.

DISCUSSION

Basically, Tessier #10 cleft has two presentation forms: upper eyelid coloboma and/or frontal encephalocele (Figure 1).
Corneal protection is the primary goal in the medical treatment of eyelid colobomas. The eyelid deformity may be most commonly triangular in shape, with the base at the eyelid margin. It is usually located on the medial half of the upper eyelid and may vary in size from a small indentation of the eyelid margin to near absence of the entire eyelid (ablepharia). The treatment depends directly on the defect’s size. The rule of 25% for eyelid reconstruction is imperative to choose the method of repair. Defects until 25% of eyelid extension may be closed primarily; between 25-50%, a lateral canthotomia may be performed avoiding skin retraction.

Tessier described that the #10 cleft represents the continuance of cleft #4. In our personal experience of 310 cases of rare craniofacial clefts, we did not observe the association between these two clefts. Cleft #10 was associated to clefts #2 and 7 in this series. Tessier #4 clefts did not present any cranial commitment.

The most challenging deformity of Tessier #10 cleft is the macroblefaron. The eyelid lengthening is elongated, with medial and lateral canthus laterally displaced. So it is very difficult to be completely corrected. In our three cases, after the orbital dystopia correction, medial canthopexia was associated with bony fixation. The lateral canthus was medially positioned to reduce the width of the eyelid. We could not achieve the perfect correction of the macroblefaron in our cases. We did not find either a surgical technique description to correct this malformation.

Ablepharia of the upper eyelid is defined where the remnants represent less than a third of the eyelid. Tessier referred that a large dermatocele may be covered by cutaneous remnants represent less than a third of the eyelid. Tessier #10 cleft represented 2% of the malformations. In our personal experience of 310 cases of rare craniofacial clefts, we did not observe the association between these two clefts. Cleft #10 was associated to clefts #2 and 7 in this series. Tessier #4 clefts did not present any cranial commitment.

### CONCLUSION

Children born with facial cleft represents a surgical challenge and their treatment requires both aesthetic sense and technical skills to repair form and function. The treatment of rare craniofacial cleft patients represents a great challenge to the craniofacial surgeon, it should be managed according to the complexity, localization, variability and extension of the
deformities. The Tessier #10 cleft has two presentation forms (eyelid coloboma and frontal encephalocele). It is the first exclusive study on this rare malformation.

REFERENCES