Eastern Journal of Medicine 16 (2011)90-93

Case Report

Tessier no. 7 unilateral cleft: two case reports

Daghan Isık[a,*], Mustafa Tercan[b], Mehmet Bekerecioglu[c]

[a] *Department of Plastic, Reconstructive and Aesthetic Surgery, Medical School of Yuzuncu Yil University, Van, Turkey*
[b] *Department of Plastic Reconstructive and Aesthetic Surgery, Haydarpaçsa Numune Teaching and Research Hospital, Usküdar, Istanbul, Turkey*
[c] *Department of Plastic, Reconstructive and Aesthetic Surgery, Medical School of Gaziantep University, Gaziantep, Turkey*

Abstract. Congenital macrostomia (transverse facial cleft) is a rare congenital anomaly. It is most commonly associated with anomalies of the first and second branchial arches. We present two patients with Tessier number 7 unilateral cleft and their treatment.

Key words: Tessier's cleft number 7 cleft, craniofacial cleft, treatment

1. Introduction

Macrostomia, also called a transverse facial cleft, is an uncommon congenital deformity when compared to the cleft lip and the palate. The exact incidence of Tessier no 7 cleft is unknown (1, 2). The etiology of facial cleft is multifactorial (1, 2). Poswillo postulated that this cleft occurs due to injury or disruption of the stapedial artery early in embryogenesis. (3) The etiopathogenesis behind the formation of atypical facial clefts remains unknown. It is seen more commonly in men than in women. Clinically, it can be seen in a variety of presentations, ranging from a small excess of skin in front of the ear to serious deformities including the soft tissue and the skeletal framework (4). Unilateral facial cleft cases have been encountered 6 times as frequent as bilateral facial clefts. Bilateral facial clefts generally present as symmetrical anomalies, but they can rarely be seen asymmetrically (5). Tessier no 7 cleft does not affect the patient, but nevertheless causes an abnormal facial appearance and disordered daily life functions. In this report, we present two patients with Tessier no 7 cleft, aged 7 months and 9 years old.

[*]Correspondence: Dr. Daghan Isık, MD.
Yuzuncu Yil Universitesi Tip Fakultesi,
Plastik Cer. A.D. 65100 Van, Turkiye
Tel: +90 432 215 04 71 (6535)
Fax: + 90 432 214 05 01
E mail: daghanisikmd@yahoo.com
Received: 23.01.2010
Accepted: 18.07.2010

2. Case presentations

2. 1. Case 1

A 7-month-old baby had macrostomia on the right side of the face and skin tags in front of the right ear, adjacent to the right oral commissure (Figure 1). The distance from the left philtrum to the left oral commissure was 25 mm, and that from the right philtrum to the right oral commissure was 45 mm. Hypoplasia of the right ear was not prominent, but there was cartilaginous excess in the skin tag at the preauricular area. The systemic examination of the patient revealed no other pathology. There was no congenital malformation in family history. The baby had been delivered via caesarean section at term. Blood and urine analyses were all within normal limits.

2. 2. Case 2

An 8 –year-old girl was admitted with skin excess in front of her left ear and a wide corner of the mouth on the left (Figure 2). Her systemic examination revealed no pathology. The history obtained from the patient and her parents was unremarkable. Local examination revealed an excess skin in front of the ear which was solid and palpable. The distance from the left philtrum to the left oral commissure was 50 mm, and that from the right philtrum to the right oral commissure was 27 mm.

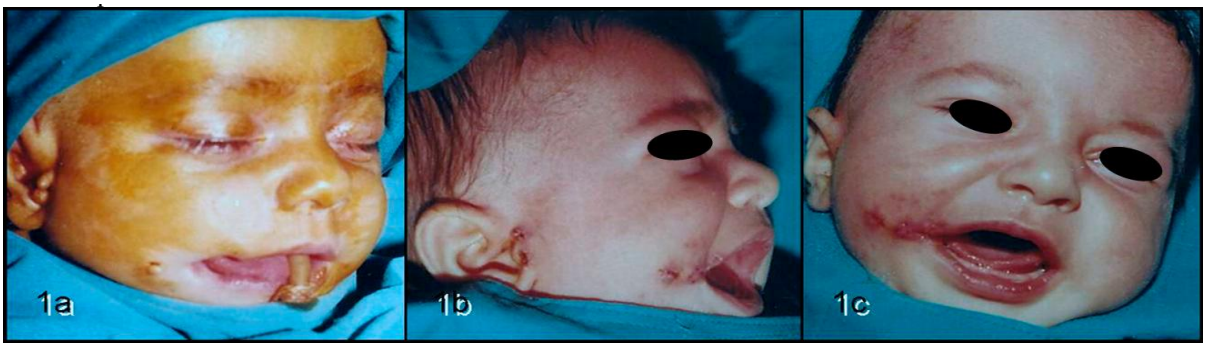

Fig. 1. (Case 1) (a) Preoperative display of a tessier number 7 cleft, (b) Early postoperative lateral photography, (c) Postoperative anterior view.

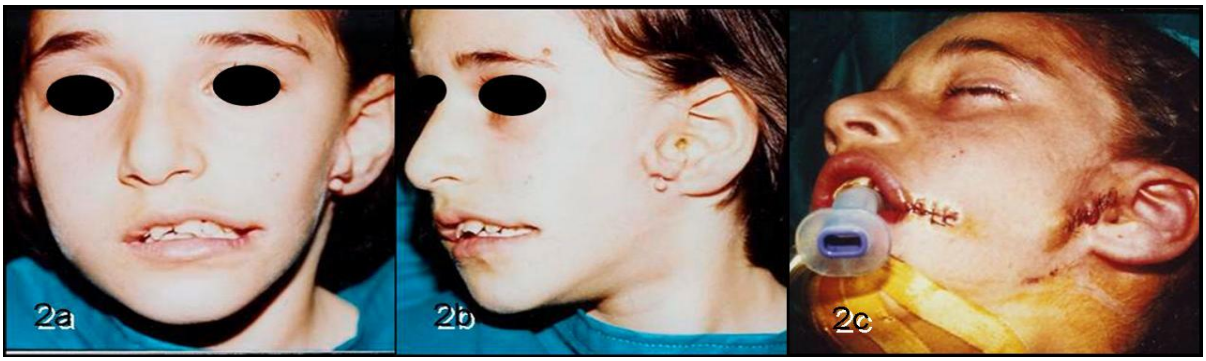

Fig. 2. (Case 2) (a) Preoperative anterior appearance showing the macrostomia corresponding to Tessier number cleft 7, (b) Preoperative lateral view, (c) Early postoperative appearance.

Dental occlusion was normal, but articulation was not normal.

Hypoplasia was evident in the left parotido-masseteric area causing facial asymmetry. There was a prominent deformity of the left ear without hypoplasia. The bony framework of the Maxilla and the zygomatic bone was normal. Mild hypoplasia at the left side of the mandible was observed in inspection.

2. 3. Treatment

Cases 1 and 2 underwent operative intervention according to the protocol of New York University (6). According to this protocol, excision of excessive soft tissue and repair of oral commissure was planned first. Both cases were operated under general anaesthesia at the supine position. This procedure was performed in the following order.

The philtral dimple was marked using a surgical pen. The distance between the philtrum and the oral commissure was measured on the normal side and the same distance was measured and marked on the cleft side. Thereby the point of the commissure was determined on the upper lip of the cleft side. Opposite point of this commissure point was marked on the lower lip. A horizontal incision on the border of the skin and the vermillion was made between the commissure point and the end of the cleft. The vermilions on the upper and the lower lip were dissected from the orbicularis oris muscles and sutured each other for creating the inside mucosa of the commissure. Congenitally interrupted orbicularis oris muscles were exposed.

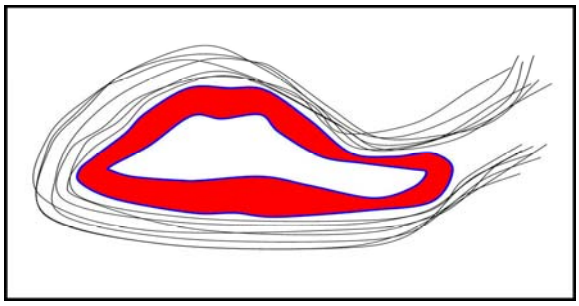

Fig. 3. Schematic view of the orbicularis oris muscle in cases of Tessier number 7.

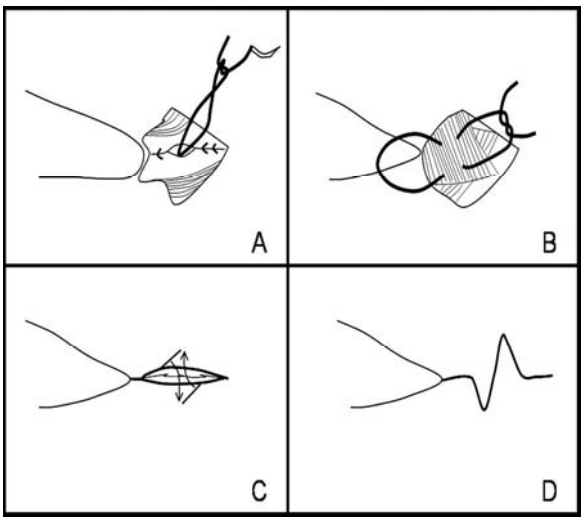

Fig. 4. Operative procedures: (a) Oral mucosa correction, (b) Formation of oral sphincter continuity, (c) Z-plasty and skin closure planning, (d) Appearance of the final scar.

One side of the orbicularis oris muscle was placed over the other side and sutured for oral sphincter continuity. A minor Z-plasty was performed on the skin of the oral commissure (Figures 3, 4). Then, the skin tags and their cartilaginous excess at the cheek and the preauricular area were excised and the defect was sutured primarily. In order to restrict the opening of the mouth, fluid food was only given for the first 5 postoperative days.

On the 6th postoperative day, the skin sutures were removed. No complications were observed in the post-operative period. Case 2 and her parents did not accept any intervention for the hypoplastic bone.

3. Discussion

Tessier no 7 clefts are more rarely encountered than cleft lip and palate deformities. Physical restricting forces such as amniotic bands my lead to the formation of these unusual clefts (7, 8). This hypothesis is supported by an experimental study by Stelnicki et al. (7, 8). There are several classification systems, such as that of the American Association of Cleft Palate Rehabilitation, Karfik, Boo-Chia, Demeyer, and Van der Meulen (1, 2, 4, 9). However, the best-known classification system of facial clefts is that devised by Paul Tessier (4). The clefts are numbered 0 to 14 with cleft number 8 forming the equator. Hence, clefts numbered 0 to 7 of the lower hemisphere represent the facial clefts and the clefts of the upper hemisphere numbered 9 to 14 are their cranial prolongation. According to Tessier, cleft number 7 is a lateral cleft. The side of involvement is not significant. The incidence of Tessier number 7 cleft has been determined to be 1/3000 to 1/5642 (2). The clinical expression of number 7 cleft is highly variable. In addition to the facial findings, hypoplasia of the zygoma, temporal bone, maxilla and mandible, parotid gland and parotid duct, and hypoplasia of the innervation area of the fifth and seventh cranial nerves, palate and tongue have been seen (10). The severity of the deformity, the pathophysiology of the disorder with regard to growth potential and the psychological aspect of the deformity all contribute to the decision regarding the surgical intervention. Treatment involves the reconstruction of soft tissue followed by reconstruction of the bony frame. Soft tissue reconstruction should be performed in the pre-school period. In this period, excision of skin tags, and the correction of macrostomia and prominent ear are appropriate (10). When unnecessary, the bone frame is not reconstructed in the early ages. Bone grafting and augmentation must be applied at older ages. At 5 years of age, cranio-orbito-zygomatic development is at 85% of adults. For this reason, skin and bone reconstruction of the middle face have to be done after age 6, and that of maxilla and mandibular interventions must be performed in the adolescent period. Since case 2 did not agree with correction of prominent ear and minimal mandibular hypoplasia, we only performed macrostomia repair. Osteotomy of the maxilla, mandible and zygoma, distraction osteogenesis constitute the alternatives in treatment. Soft tissue augmentation must be planned after the bone and the face development.

Treacher Collins syndrome and hemifacial microsomia exist in the differential diagnosis. Former, Treacher Collins syndrome characterized by hypoplasia / aplasia of the body and arch of the zygoma, a significantly increased facial convexity, mandibular hypoplasia, a retrusive chin with increased vertical height, and external and middle-ear anomalies. An important distinguish feature of this anomaly is that it is bilateral and symmetrical (11). Latter, hemifacial microsomia (HFM) is a variable and asymmetric malformation involving first and second pharyngeal arch derivatives. HFM primarily affects the orbit, maxilla, mandible, ear, cranial nerves, and facial soft tissues (12).

As a conclusion, neither Treacher Collins syndrome nor hemifacial microsomia should be

eliminated in the differential diagnosis for an uncomplicated surgical procedure and improvement of the patients' daily life functions at the result of the surgery. On the other hand, applications of the some surgical steps such as overlapping of the orbicularis oris muscle and z-plasty of the skin to break the straight scar are important manoeuvres for improving of the surgical result.

References

1. Kawamoto HK. Rare craniofacial clefts. In: McCarthy JG (ed.). Plastic Surgery (1st ed). Philadelphia: WB Saunders Company, 1990, pp 2922-2973.
2. Kawamoto HK. Craniofacial clefts. In: Aston SJ, Beasley RW, Thorne HC (eds). Grabb and Smith 's Surgery (1st ed). New York: Lippincott-Raven, 1977, pp 349-363.
3. Poswillo D. The pathogenesis of the first and second branchial arch syndrome. Oral Surg Oral Med Oral Pathol 1973; 35: 302-328.
4. Tessier P. Anatomical classification of facial, craniofacial and latero-facial clefts. J Maxillofac Surg 1976; 4: 69-92.
5. Uzunismail A, Gencosmanoğlu R, Yuksel F, Özdemir A. Tessier's cleft no.7 with asymmetrical involment. Plast Reconstr Surg 1995; 96: 224-225.
6. McCarthy JG, Grayson BH, Coccaro PJ, Wood-Smith D. Craniofacial microsomia. In: McCarthy JG (ed.). Plastic Surgery (1st ed). Philadelphia: WB Saunders Company, 1990, pp 3054-3100.
7. Stelnicki EJ, Hoffman WY, Vanderwall K, et al. A new in utero model for lateral facial clefts. J Craniofac Surgery 1997; 8: 460-465.
8. Stelnicki EJ, Hoffman W, Foster R, Lopoo J, Longaker M. The in utero repair of tessier number 7 lateral facial clefts created by amniotic band-like compression. J Craniofac Surg 1998; 9: 557-562.
9. Van der Meulen JC, Mazzola R, Vermey-Keers C, Stricker M, Raphael B. A morphogenetic classification of craniofacial malformations. Plast Reconstr Surg 1983; 71: 560-572.
10. Stradoudakis AC. Craniofacial anomalies and principles of their correction. In: Georgiade GS, Riefkohl R, Levin LS (eds). Plastic, Maxillofacial and Reconstructive Surgery (3rd ed). Baltimore: Williams & Wilkins, 1997, pp 273-296.
11. Havlik RJ. Miscellaneous craniofacial conditions. In: Beasley RW, Aston SJ, Barlett SP, Gurtner GC, Spear SL (eds). Grabb & Smith's Plastic Surgery (6th ed). Philadelphia: Lippincott Williams & Wilkins, 2007, pp 281-296.
12. Fan WS, Mulliken JB, Padwa BL. An association between hemifacial microsomia and facial clefting. J Oral Maxillofac Surg 2005; 63: 330-334.