Correction of Bilateral Tessier No. 2, 3, and 12 Facial Cleft with Anopthalmia

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Abstract

Oblique facial cleft is a rare congenital deformity. Its incidence has been reported as 0.24% of all reported cases of facial cleft. We report on a patient who had a left-sided oblique facial cleft with anopthalmia, including lip and palate, nose alar base, and medial canthus. The patient also had a right-sided oblique facial cleft, which included lip and palate, nose alar base, medial canthus, and upper eye brow. Primary closure of the facial cleft was performed using multiple Z-plasty after excision of scar tissue.

Key words: Oblique facial cleft, Anopthalmia

Introduction

Craniofacial cleft is an extremely rare disease, with a prevalence rate of approximately 0.24% in all reported cases of facial cleft[1]. The incidence of facial cleft was reported as 1.43 to 4.85 out of 100,000 newborn infants[2]. Tessier’s classification, reported in 1976, has been the representative system for classification of facial clefts. Its classification is based on anatomical position. Tessier’s classification of facial cleft types ranges from 0 to 14, based on the central line, which connects the sagittal plane and both orbital fossa[3]. Fearon suggested another classification system for use in deciding on the treatment method; using this system, facial clefts are classified according to four types, depending on anatomical position[4].

In the embryo, facial development occurs over a period of 4 to 8 weeks. Craniofacial cleft is created by failure of fusion of mesoderm of the facial area during the state of embryonic development. An oblique facial cleft is formed by extraordinary fusion of the lateral nasal process and maxillary process. Oblique facial cleft may appear without relationship to other syndromes, and there is no sexual difference[1].

We experienced a case of bilateral Tessier No. 2, 3, and 12 facial cleft with anopthalmia. This patient was treated with primary closure of the facial cleft, performed using multiple Z-plasty after excision of scar tissue.
Case Report

The patient, a three-year-old female without specific systemic disease, was found by a medical volunteer who was visiting the Philippines. The cleft on the right side of her face was classified as Tessier type 2, defined as a facial cleft running from the upper lip to the ala of the nose and from the eyebrow of the upper eyelid to the forehead, and Tessier type 12, defined as an independent cranial cleft. On the left side, her eyeball could not be seen and the lower eyelid was dislocated toward the latero-down direction. The nasolacrimal duct was obstructed and the oblique facial cleft extended from the medial canthus through the ala of the nose to the cleft lip and palate.

In addition, a severe Tessier type 3 facial cleft with anophthalmia was observed on the left side (Fig. 1). According to Fearon’s classification, a paramedian cleft (type II) was observed on both the left and right sides.

Defect of palate, alveolar bone, medial and anterior wall of the left maxilla, and medial and inferior wall of orbit were observed on computerized tomography images (Fig. 2). Surgery performed on this patient focused on removal of all of visible scars and primary closure of the facial clefts using multiple Z-plasty. Through dissection of the inferior of left orbit and cheek, sound tissue was drawn to the ala of the nose and then sutured in layers. For correction of the right ala of the nose, Z-plasty was performed at the medial canthus and lower eyelid. For correction of the left nasal ala and lower eyelid, Z-plasty was also performed at the medial canthus and lower eyelid. Straight line closure was performed at the left nasal ala and upper lip, where a broad defect was observed (Fig. 3). No procedure for correction of bony defects was performed. There was no postoperative complication. One year after the operation, the patient’s facial appearance was acceptable (Fig. 4).

Fig. 1. Preoperative photograph. Right facial cleft is classified as Tessier No. 2 and 12, left facial cleft is classified as Tessier No. 3.

Fig. 2. (A) Axial computed tomography (CT) images. (B) Three-dimensional reconstruction from CT images. Bone defect was observed on alveolus, palate, left medial, and anterior wall of the maxilla, and orbital rim.

Fig. 3. Correction of right ala of the nose. Z-plasty was performed at the medial canthus and lower eyelid. For correction of the left nasal ala and lower eyelid, Z-plasty was also performed at the medial canthus and lower eyelid. Straight line closure was performed at the left nasal ala and upper lip, where a broad defect was observed.
Craniofacial cleft is an extremely rare disease. Tessier’s classification, which has been widely used for treatment of facial clefts, is based on two lines connecting the mid-sagittal plane and a center-line of the orbit fossa according to anatomical position. The facial cleft (Tessier type 2) on the right side of this patient extended from the upper lip to the nasal ala, and the independent facial cleft (Tessier type 12) was located at the eyebrow. On the left side, a continuous facial cleft (Tessier type 2 and 3) extended from the upper lip to the nasal ala and the
reported three causes of anophthalmia. The first is the nose, lower eyelid, and inner canthus, and shows defects of soft and hard tissue at the same time. Stoll et al. reported that Tessier type 2 is a proxy for Tessier type 2. The inner margin of the eyebrow becomes deformed as a result of passing between the frontal process of the maxilla and the nasal bone.

Tessier type 3 facial cleft is accompanied by a cleft lip and palate. It extends from the upper lip to the ala of the nose, lower eyelid, and inner canthus, and shows defects of soft and hard tissue at the same time. Stoll et al. reported three causes of anophthalmia. The first is anophthalmia due to failure of optic outgrowth in the forebrain, the second is anophthalmia due to malformation in the anterior end of the forebrain, and the third is anophthalmia due to degeneration arising from the optic vesicle and tract. The third form usually appears closer to microphthalmia, rather than real anophthalmia. This case can be considered as the third form.

Due to the difficulty of antenatal screening, severe facial dysmorphia is generally diagnosed too late, or after delivery. The usefulness of three-dimensional ultrasonography, rather than two-dimensional, in diagnosis of an antenatal facial cleft has been reported. Radiological examination is also necessary for diagnosis and evaluation of a facial cleft. Axial and three-dimensional computed tomography can provide information about soft and hard tissue defects of facial cleft patients.

The ultimate goal of treatment of facial cleft is the functional correction of macrosomia, the soft tissue reconstruction of palpebra for protection of eyeballs, proper separation of oral, nasal, and orbital cavities, and esthetic correction of facial appearance. Tissue expander, advancement and rotation flap, autogenous bone graft, and multiple Z-plasty have generally been used for correction of facial clefts. The operation time for bone graft for treatment of bone defect has been controversial in facial cleft patients. Resnick and Kawamoto reported a preference for bone grafting before the age of five. However, premature bone graft could cause significant bone resorption and interruption of normal bone remodeling.

For correction of a Tessier type 2 facial cleft, the lateral side of the nose can be used as a donor site for the full thickness rotation flap. However, an asymmetrical nostril or a large defect after the primary repair may require secondary correction. Severe facial cleft including the entire nose can be corrected with a longer rotation flap or a forehead flap. Tessier type 3 defect can generally be corrected using a multiple cheek flap or a nasal flap. Nasal flap cannot be used for large defects on the lateral nasal surface, because tension of soft tissue is too great when correction is performed using a nasal flap. To overcome such tissue insufficiency, Toth et al. suggested using a tissue expander on the forehead. However, due to scar formation on the forehead, straight-line closure was used more often than a tissue expander. In our patient, multiple Z-plasty was performed at the medial canthus and lateral nasal surface for correction of the right nasal area. On the left side, straight-line closure was performed on the lateral surface of the nose, and multiple Z-plasty was performed for correction of the medial canthus and lower eyelid cleft.

Correction of both sides of the facial cleft is difficult. In the case of a midline facial cleft (Type I), correction of telorbitism is needed. In the case of a median facial cleft (Type II) and orbital cleft (Type III), the unilateral nose and lacrimal system should be restored and an eyelid flap should be used to cover the eyeball, respectively. Correction of a cleft lip and palate, and secondary bone graft are also needed in Type II and III cases. However, satisfactory performance of all of these procedures at one time is difficult, and repeated operations are needed.

The focus of treatment of our patient was scar excision and primary closure of each part of the bilateral facial cleft. The patient’s appearance has shown considerable improvement one year after the operation. However, the length of the follow-up period has been limited. In the future, scar revision, bone graft, and orthodontic treatment will be needed in this patient.
References