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Case report

Two rare cases of simultaneous Tessier number 3 cleft, contralateral cleft lip, and signs of amniotic band syndrome

Yi Xu, Yue Mu, Renji Chen*, Zongmei Zheng, Wenjing Zhang

Department of Plastic and Traumatic Surgery, School of Stomatology, Capital Medical University, No.4, Tiantan West, East District, Beijing, 100050, China

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ABSTRACT

The Tessier number 3 cleft is rare. In this paper, we report two extremely rare cases of simultaneous Tessier number 3 cleft, contralateral cleft lip, and signs of amniotic band syndrome. In the two cases, we confirmed that amniotic bands were the probable cause of the Tessier number 3 cleft, where swallowed fibrous strands of amniotic bands entangle a typical cleft lip and cause the more severe Tessier number 3 cleft. In this study, Z-plasty was performed for one case, and a straight-line method was used for the other. Postoperatively, the appearance of both patients was satisfactory, as expected. Consequently, treatment for the Tessier number 3 cleft should be designed individually based on the severity of deformity.

1. Introduction

Differing from typical cleft lip, craniofacial clefts are rare congenital anomalies. Their incidence has been estimated to range from 1.43 to 4.85 per 100,000 live births (Kawamoto, 1976). In 1976, Paul Tessier greatly contributed to the knowledge of these anomalies, presenting an anatomical classification for craniofacial clefts based on their positions relative to the sagittal midline and the orbit (Tessier, 1976). His work facilitated the classification and discussion of the wide spectrum of craniofacial clefts.

Among the craniofacial clefts, the Tessier number 3 cleft is extremely rare. According to Tessier (1976), it is a medial orbitomaxillary paranasal cleft that extends through the lip in the region of the typical cleft lip and ascends obliquely to involve the inner canthus and lower eyelid medial to the inferior lacrimal punctum. In 1989, David et al. used computed tomography (CT) and revisited Tessier's classification specifically for osseous deformities (David et al., 1989). They found that the maxilla on the side of the cleft is hypoplastic, with markedly reduced pneumatization; there is no separation between the maxilla and the nasal cavity on the side of the cleft; and there is distortion of the frontal process of the maxilla.

So far, the etiology of the Tessier number 3 cleft is still unclear, but previously reported cases can give hints. The previous reports have shown that craniofacial clefts can occur alone or accompanied by Amniotic band syndrome or/and typical cleft lip (Chattopadhyay et al., 2013; Eichhorn et al., 2015; Gawrych et al., 2010; Mishima et al., 1996; Robin et al., 2005). Amniotic band syndrome is a complex collection of asymmetric congenital anomalies. Early amniotic rupture leads to the formation of mesodermal fibrous strands that entangle fetal tissues, especially the limbs and appendages; consequently, distal ring constrictions, intrauterine amputations, and acrosyndactyly are the most common findings (Walter et al., 1998). From their case reports, these authors suspected that the etiology of craniofacial clefts was likely not only failed fusion of the facial processes but also the presence of amniotic bands during gestation.

Amniotic bands should be considered a major cause of craniofacial clefts, but this theory cannot be confirmed with certainty due to the rare incidence of craniofacial clefts. Here, we present two rare cases with simultaneous Tessier number 3 cleft, contralateral cleft lip, and signs of amniotic band syndrome, to provide clinical evidence for the possible etiology of the Tessier number 3 cleft.

2. Case reports

2.1. Case 1

The patient was a male infant of healthy non-consanguineous parents (25-year-old father and 23-year-old mother) with full-term delivery (38 weeks). There was no history of any relevant anomalies in his family and no identifiable teratogenic factors. The

* Corresponding author.
E-mail address: chenrenjicmu@sina.com (R. Chen).

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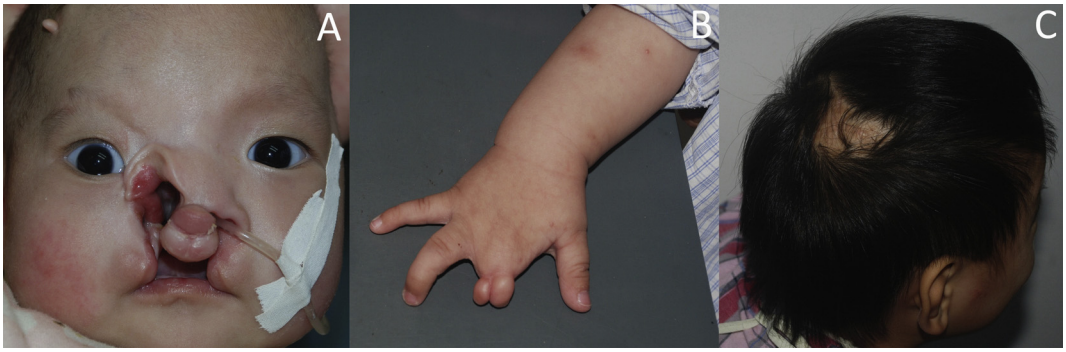

Fig. 1. Craniofacial deformities and signs of amniotic band syndrome in Case 1 patient.

mother did not undergo ultrasonic examination throughout her pregnancy, as they lived in a poor, remote mountain region in China.

The hypogenetic boy was brought to the Department of Plastic and Traumatic Surgery, School of Stomatology, Capital Medical University, when he was 6 months old. He had a stomach tube due to food intake difficulty. The infant had an oblique facial cleft on the right side, corresponding to the Tessier number 3 cleft, which ran from the upper lip at the philtrum column, similar to the typical cleft lip, to the lower eyelid, and he had complete cleft lip on the left side (Fig. 1A). His oral cavity contained a bilateral complete cleft palate and a bilateral complete cleft alveolar ridge. We also observed signs of amniotic band syndrome, such as a skin tube pedicle on the right side of the forehead (Fig. 1A), syndactyly of the right index and middle fingers, ring constriction and intrauterine amputation of these two fingers (Fig. 1B), and congenital alopecia on the top of his scalp (Fig. 1C).

Three-dimensional CT (3D CT) examination of the craniofacial skeleton when he was 2 years 11 months old revealed deformities of the nasal, maxillary, and palatal bones, markedly reduced right maxillary sinus pneumatization, and bilateral alveolar cleft medial to the canine (Fig. 2).

So far, the patient has been managed with the following four separate surgical steps, but more operations will be needed in the future:

(1) At 6 months: Primary closure of the complete cleft lip with a straight-line method; this was not routine unilateral cheiloplasty, but more precisely, semi-bilateral cheiloplasty (Fig. 3C);
(2) At 12 months: Primary closure of the Tessier number 3 cleft with Z-plasty for the mid-face defect and straight-line method for the upper lip. Based on the methods of Tessier et al. (1977) and Resnick and Kawamoto (1990)) for repairing oblique facial clefts, the Z-plasty that we used can elevate the caudally displaced medial canthal area and rotate the nasal ala downward. The straight-line cheiloplasty performed here was the same as the semi-bilateral cheiloplasty mentioned above (Fig. 3D);
(3) At 2 years 6 months: The soft and hard palates were closed with the two-flap technique (Bardach, 1995), and the skin tube pedicle was excised simultaneously (Fig. 3E);
(4) At 2 years 11 months: A second Z-plasty was performed on the right nasal facial groove to descend the nasal ala, and lip revision was performed to narrow the philtrum and repair the Cupid's bow (Fig. 3F).

2.2. Case 2

The patient was a male infant of healthy non-consanguineous parents (30-year-old father and 30-year-old mother) with full-term delivery (39 weeks). There was no history of any relevant anomalies in his family and no identifiable teratogenic factors.

The infant had an oblique facial cleft on the right side, corresponding to the Tessier number 3 cleft, which began on the right side of the Cupid's bow, undermined the base of the nasal

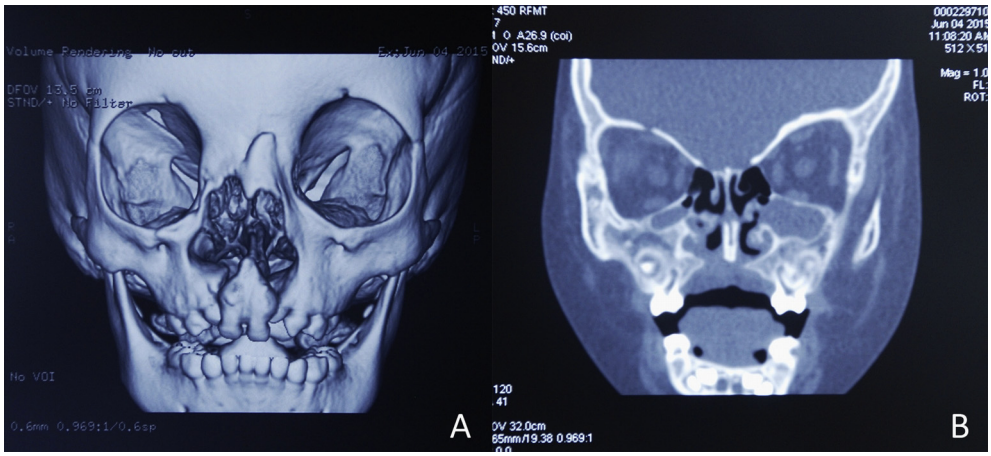

Fig. 2. 3D CT scan of the skull of Case 1 patient at 2 years 11 months of age. (A) 3D view shows nasal bone and maxillary bone deformities (especially on the right side). (B) 2D view shows marked reduction in right maxillary sinus pneumatization.