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Case report

Median cleft of the upper lip associated with a mass: A rare case

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ABSTRACT

Median cleft lip is a midline vertical cleft through the upper lip. This is a very rare anomaly described in the literature. Median cleft lip is caused by the failure of fusion of the medial nasal prominences. In this case report, a 4-month-old boy with a median cleft associated with a mass of the upper lip is presented. The patient has no other anomalies of the nose or alveolus. The patient has normotelorism. A Z-plasty technique was used on the skin of the base of the columella. A vertical excision of the cleft with muscle approximation was performed on the white roll and the wet-dry border of either side of the defect of the upper lip. Postoperatively, the patient had a satisfactory result. The incisive scars were not visible. Cupid's bow was appropriately aligned, and the height of the upper lip was equal on both sides.

1. Introduction

Median cleft lip is a midline vertical cleft through the upper lip. Within cranio-maxillofacial anomalies (Hou et al., 2011; Laure et al., 2010; Mohamed et al., 2011), this is a very rare anomaly described in the literature (Ghildiyal et al., 2003; Kawamoto, 1990). In 1935, Davis (Davis, 1935) found five oblique clefts of the face and four median clefts of the lip in a group of 945 patients. Fogh-Andersen's (Fogh-Andersen, 1965) study included 3988 cleft patients seen over 30 years of which 15 had a median cleft of the upper lip. The exact incidence is not known, but estimates range from 1.4 to 4.9 per 100,000 live births (Kawamoto, 1990). At the 1976 Interdisciplinary Workshop Conference in Chicago, Tessier (Tessier, 1976) presented a comprehensive classification of craniofacial clefts based on his personal experience with 336 patients. In the Tessier 0 through 14 cleft, the location of the median or midline cleft of the upper lip has a spectrum of dysmorphic gradation. This varied from a simple central vermilion notch to a wide cleft accompanied by a bifid nose and hypertelorbitism. Median cleft lip is caused by the failure of fusion of the medial nasal prominences (Mazzola, 1976). In this case report, a 4-month-old boy with a median cleft associated with a mass of the upper lip is presented.

2. Case report

A 4-month-old boy presented to the Xiangya Hospital of the Central South University with median cleft of the upper lip associated with a mass. The pregnancy was full-term with no complications. The mother did not have any diseases during the pregnancy. His mother did not smoke or drink alcohol, nor was she exposed to any environmental risk factors during the pregnancy. The patient was the first-born male to healthy parents, delivered at 39 weeks gestation via spontaneous vaginal delivery with outlet forceps. The patient's birth weight was normal and presented with no other anomalies. There was no family history of clefts. On examination, there was a median cleft of the upper lip. There was a mass in median cleft between the base of the columella and the vermilion of the upper lip, this small mass was actually a 'blob-like' piece of tissue between the cleft hemilabellum of the upper lip (Fig. 1A and Fig. 2A). The vermilion was separated in the midline, but the total length of the vermilion and the vertical height of the upper lip were normal. There were no bony abnormalities, and the alveolus was normal. The nose and the intercanthal distance were normal (Fig. 1A). Hematological and biochemical investigations revealed normal values, and hormonal studies were also within normal limits. Magnetic resonance imaging indicated normal position of the carotid and vertebral vessels.

The median cleft lip was repaired using the following steps. First, on the skin of the base of two nostril, the point 4 and the point 5 were marked, on the skin of the dry border of either side of the defect of the upper lip, the point 1 and point 6 were marked; then, a vertical line linked the point 4 with point 1, another vertical line linked the point 5 with point 6. The distance of the point 4 to the point 1 equaled to the distance of the point 5 to the point 6. The distance of the point 4 to the point 1 or the distance of the point 5 to the point 6 is actually the vertical height of two philtral columella. On the skin of the base of the columella, a point 3 was marked, on the dry vermilion of either side of the defect of the upper lip, the

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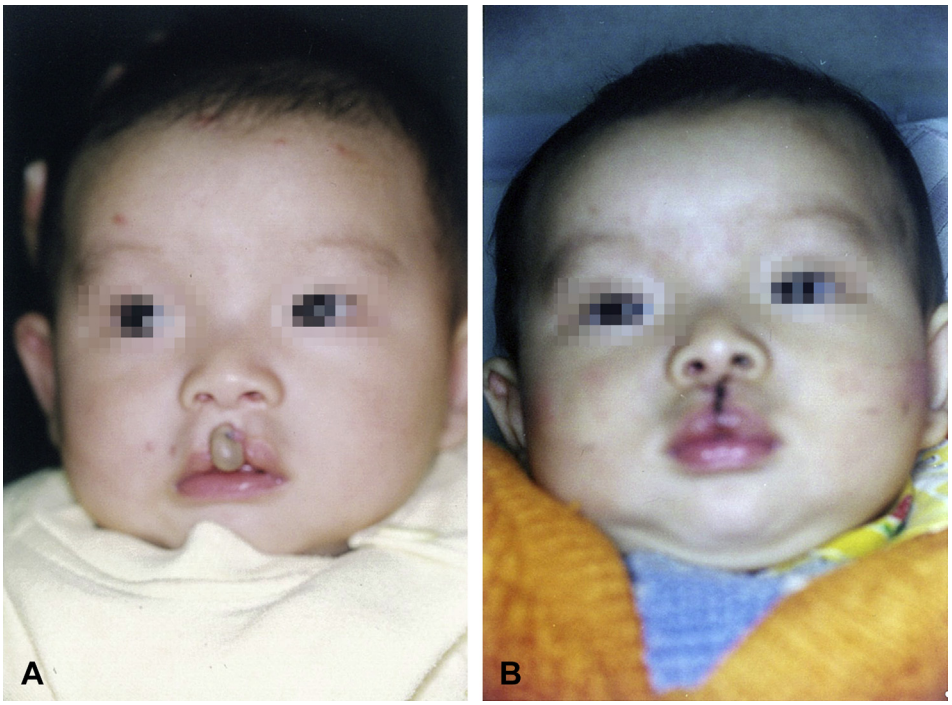

Fig. 1. A 4-month-old boy with a median cleft lip with a mass in the center of the lip separating the vermilion. A. Preoperative frontal view; B. Postoperative frontal view.

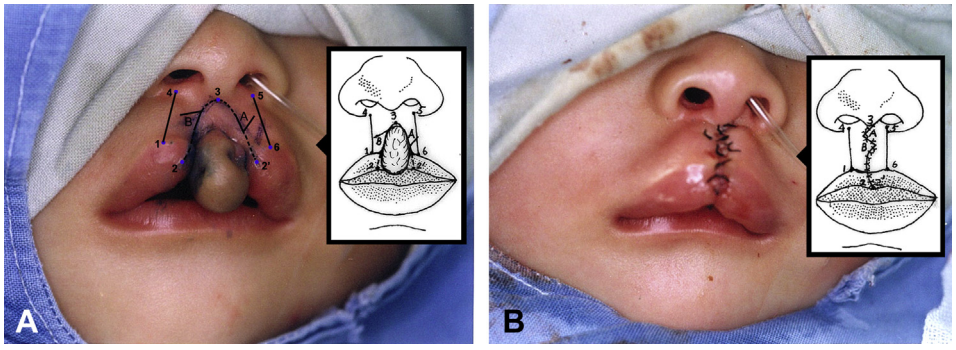

Fig. 2. Surgical plan. A. Preoperative design; B. Postoperative condition.

point 2 and the point 2′ were marked, length of the point 3 to point 2 equaled to the length of point 3 to point 2′. The two side limbs were marked on the two side of the point 3 to point 2 and point 3 to point 2′, the three lines of the Z must be equal in length. The angles of Z two flaps were 60° (Fig. 2A). These incisive lines were marked with methylene blue. Next, a Z-plasty incision was made, the tissue in the midline and mass were excised via a vertical incision. The skin of the upper lip was dissected from the orbicularis oris muscle on both sides of the defect via the same incision. Next, a 5-0 Vicryl was used to close the muscle layer. The positions of the Z-plasty flaps of the skin of the base of the columella of the upper lip were exchanged and were first closed by reapproximation, then the white roll and the wet–dry border were sutured (Fig. 2B and Fig. 1B). The surgical pathology report was “a teratoid polyp with skin appendages, fibrous connective tissue, skeletal muscle, bone”. No evidence of malignancy noted.

Postoperatively, the patient had a satisfactory result. The incisive scars were not visible. Cupid’s bow was appropriately aligned, and the height of the upper lip was equal on both sides (Fig. 3).

3. Discussion

A median cleft of the lip is defined as a vertical cleft through the centre of the upper lip. This is a rare anomaly, which have been divided into two categories: true and false. Millard and Williams (1968) stressed that any congenital vertical cleft through the centre of the upper lip, no matter to what extent, in the absence of a prolabial remnant should be classified as a median cleft of the upper lip. These clefts are best explained by the failed mesodermal migration into fused frontonasal and maxillary process (Stark, 1954). It is a rather rare condition among craniofacial anomalies.

At present, the exact incidence is not known. There have been a few reports in the literature describing different variations of the median cleft lip (Table 1). Veau (Veau, 1937) classified three varieties of median clefts: notch of the lip, median cleft extending to the columella, and a defect due to atrophy of midline facial structures. Our case may be a secondary type of Veau classification, namely median cleft extending to the columella. DeMyer (DeMyer, 1967) described two groups of syndromes associated with the