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Case report

Lateral facial cleft associated with accessory mandible having teeth, absent parotid gland and peripheral facial weakness

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ABSTRACT

Transverse facial cleft is a very rare malformation. The Tessier no. 7 cleft is a lateral facial cleft which emanates from oral cavity and extends towards the tragus, involving both soft tissue and skeletal components. Here, we present a case having transverse facial cleft, accessory mandible having teeth, absent parotid gland and ipsilateral peripheral facial nerve weakness. After surgical repair of the cleft in 2-month of age, improvement of the facial nerve function was detected in 3-year of age. Resection of the accessory mandible was planned in 5–6 years of age.

1. Introduction

Transverse facial cleft is a very rare malformation which results from failure of mesodermal migration. It can be associated with the first and second branchial arches syndromes (Franco et al., 2007; Rogers and Mulliken, 2007). Transverse facial cleft is seen in every 100–300 facial clefts (Franco et al., 2007; Chen et al., 2009; Kajikawa et al., 2010; Bütow and Botha, 2010). They constitute 1.5% of all clefts (Chen et al., 2009). Lateral or transverse clefts of the lip have generally been documented to have an incidence of 0.02% of live births (Kuriyama et al., 2008).

The Tessier no. 7 cleft (Tessier, 1976) is a lateral facial cleft which originates from the oral cavity and extends towards the tragus, involving both soft tissue and skeletal components (Borzabadi-Farahani et al., 2013). The deformity generally affects both the commissure and the musculature. Sometimes, deep facial structures may be involved, and complete division of the face may be seen (Chen et al., 2009). The number 7 cleft usually presents with both a soft tissue (cleft lip and preauricular tags) and a bony component (temporo-zygomatic cleft). Van der Meulen et al. (1983) classified this cleft as a maxillo-mandibular dysplasia.

The ipsilateral tongue, soft palate, and muscles of mastication may be underdeveloped. The parotid gland and parotid duct may be absent. Facial nerve paralysis may be seen. External ear deformities ranging from preauricular skin tags to complete absence can be seen.

Temporomandibular joint and zygomatic arc deformities, defects of eyelid, accessory maxilla (Cheung et al., 1993), accessory mandible, polydactyly and other congenital anomalies can be seen (*e.g.* Bradley and Kawamoto, 2007). Bony involvement in the pterygomaxillary junction, or hypoplasia of the alveolar process (molar region) or involvement of the maxilla, zygomatic body, mandibular condyle, coronoid process and the sphenoid bone, especially the pterygoid plates, can also be seen (Bütow and Botha, 2010). In this article, we present a case having transverse facial cleft, accessory mandible with teeth, absent parotid gland, the distorted auricle and ipsilateral peripheral facial nerve weakness (Fig. 1).

2. Case report

A 50-day-old girl presented with a congenital transverse cleft on the right side of the face. The patient was born by caesarean section, on term weighing 2750 g. Her parents were healthy with no hereditary disease history and not consanguineously married. The mother had taken no drugs, had no viral infection, radiation exposure or trauma during the pregnancy (Fig. 1). Physical examination revealed a complete full thickness cleft running from the right oral commissure up to the tragus of the ear. The mandible and its alveolar bone were hypoplastic.

An accessory mandible was present at the same side (Fig. 1). Ipsilateral peripheral facial weakness was accompanied with the cleft. The right eyelid could not be closed during crying and sleeping. Right nasolabial fold and right frontal creases were absent

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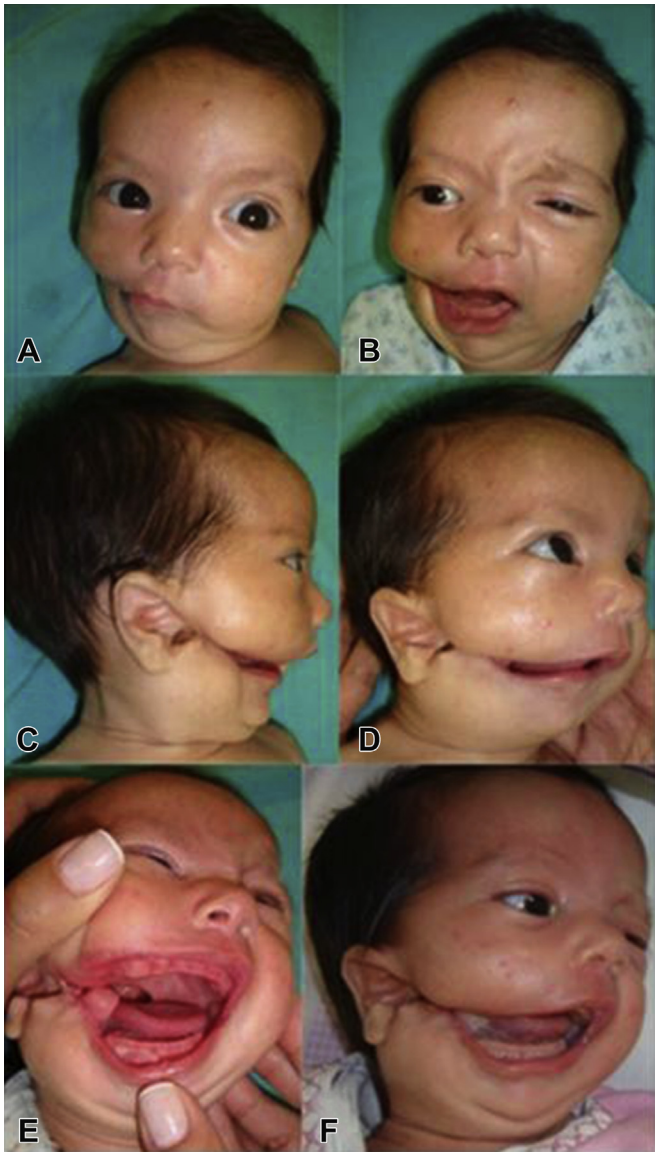

Fig. 1. A) Preoperative frontal view of 50-day-old girl having right lateral facial cleft. Severe mandibular hypoplasia is seen. B) Preoperative frontal view. Presence of ipsilateral peripheral facial paralysis is observed. Nasolabial fold and frontal crease formation is absent on the cleft side during crying. Beside, right eyelid closure is absent. C and D) Preoperative lateral and oblique views. Complete full thickness cleft is running from the right oral commissure up to the tragus of the ear. E) Close-up intraoral view. Accessory mandible is seen. It is fixed to the underlying structure. F) Preoperative oblique mouth-open view. Note how shallow the lower gingivobuccal sulcus is.

(Fig. 1). During crying, the baby could depress her lower lip on the affected side (Fig. 1). Therefore, we thought that the facial nerve function was present at the inferior side of the cleft. The nose was normal in appearance. Because cleft extended to the tragal area of the ear; the helix and the tragus were divergent from each other. The crura of the anthelix were malformed. Internal ear structures were reported as normal by ENT department. All extremities were normal in appearance. CT scans of the brain revealed no further abnormalities. There was evidence of motor retardation.

2.1. Operative technique

We repaired the lateral cleft when she was 2-months-old.

Under general anaesthesia, we measured the distance between the midpoint of the upper and lower lips and the oral commissure

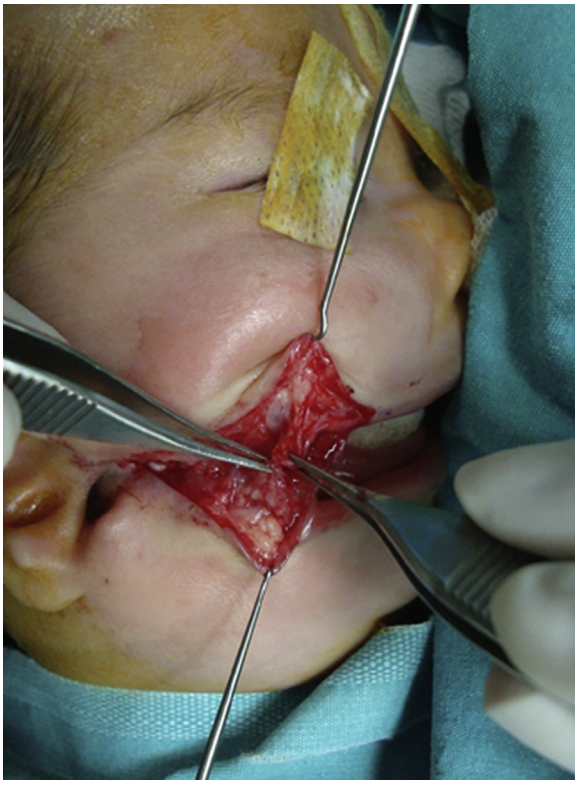

Fig. 2. Intraoperative view. The orbicularis oris muscle was reconstructed by cross-overlapping the muscular bundles at the angle of 90°. Skin, orbicularis oris and mucosa repair was achieved.

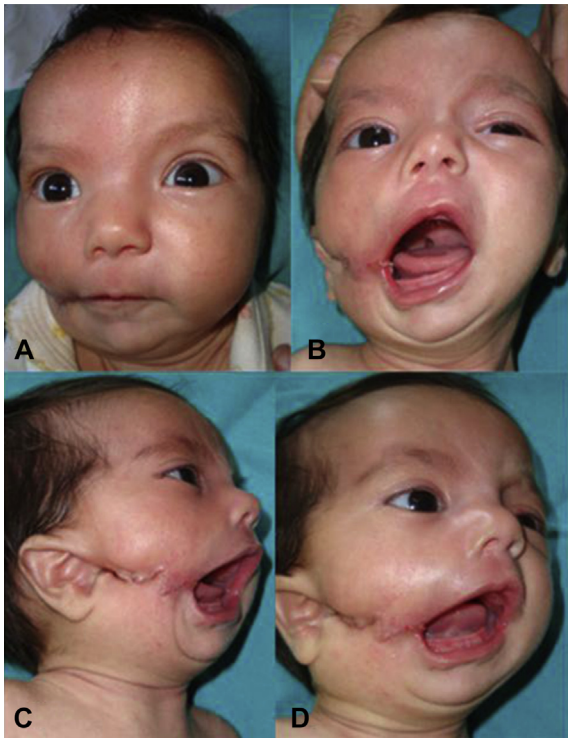

Fig. 3. A) Postoperative frontal view. When mouth is closed, appearance of mouth is good. B) Postoperative frontal view. Significant mandibular hypoplasia and underdeveloped soft tissue envelope at mandibular area prevents proper contour formation at the cheek area. C and D) Early postoperative lateral and oblique views. Z-plasty was performed on skin and mucosa closures.