Case Report

Median cleft of mandible and lower lip with ankyloglossia and ectopic minor salivary gland on tongue

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ABSTRACT

Median cleft of lower lip and mandible is a rare anomaly. This Cleft has also been described as Cleft No. 30 of Tessier's classification. In minor forms only lower lip is cleft. Frequently, the cleft extends into the mandibular symphysis and the tongue is attached to the cleft alveolar margin. At times the tongue may be bifid or absent, hyoid absent, thyroid cartilage underdeveloped, strap muscles atrophic, manubrium sterni absent, clavicles widely spaced etc. The earliest report of this anomaly was by Couronne' in 1819. Since then very few cases have been reported in literature with variations. We describe a male child who presented at the age of 6 months with an ectopic salivary gland on the dorsum of the tongue in addition to median cleft of lower lip, ankyloglossia and notching of the mandible. Excision of mass on dorsum of tongue, release of ankyloglossia and lip from the alveolus followed by repair was done. No bony work was done since the mandible was only notched. On post-operative follow-up at 18 months, dentition was delayed in both maxillary as well as mandibular teeth and there was a gap between the lower central incisors. At the age of 2 years 4 months, the dentition is still not complete and the gap between the lower central incisors is very apparent. There is a supernumerary upper central incisor on right side. There is no mobility between the two segments of mandible. Speech is normal. A regular follow-up will be done to study the eruption of permanent central incisors at the age of 7 years and till eruption of all permanent teeth to assess the occlusion and to decide whether any bony work is needed or not.

KEY WORDS

Median cleft mandible, Median cleft lower lip, Tessier cleft 30

INTRODUCTION

Median cleft of lower lip and mandible is a rare anomaly. It has also been described as Cleft No. 30 of Tessier's classification. Couronne' reported the first account of this anomaly in 1819[1]. Since then very few cases have been reported in literature with different variations. We came across a patient with associated ectopic salivary gland on the dorsum of tongue.

CASE HISTORY

A six-month-old, first born, male child presented to us

with median cleft of lower lip (Figure 1). The tongue was attached to the mandible and cleft margins of lower lip (Figure 2). There was no difficulty in feeding. There was no significant antenatal history except that the mother had taken anti-tubercular drugs during pregnancy. The child was delivered normally at full term. Intra-oral examination revealed a soft tissue mass of 2 x 1 cms in size, about two cms from the tip in midline on the dorsum of tongue (Figure 3). Chromosomal studies showed 46XY pattern. CT scan showed notching of the upper and lower borders of mandible in midline (Figure 4). No other anomaly was present.

Under general anaesthesia, the soft tissue mass over dorsum of the tongue was excised and the defect sutured. On histopathology, it was reported to be a minor salivary gland. The tongue and the lip were released from the alveolus. The tip of the tongue was sutured in layers. Vestibular flaps were raised on either side and sutured to cover the bare alveolar ridge. Paragingival incisions in the mandibular labio-buccal sulci were taken to mobilise the mucosa of the lip and the lip was sutured in layers (Figure 5). No bony work was done since the mandible was only notched. Post-operative course was uneventful with good healing. The deciduous dentition began late in both maxillary as well as mandibular teeth. The upper incisors erupted at the age of 11 months, the lower central incisors at 12 months and lateral incisors at 13 months. Photograph at 18 months shows the gap between the lower central incisors (Figure 6). Speech started at 11 months and is normal. At 2 years 4 months age the dentition is still incomplete. A supernumerary upper

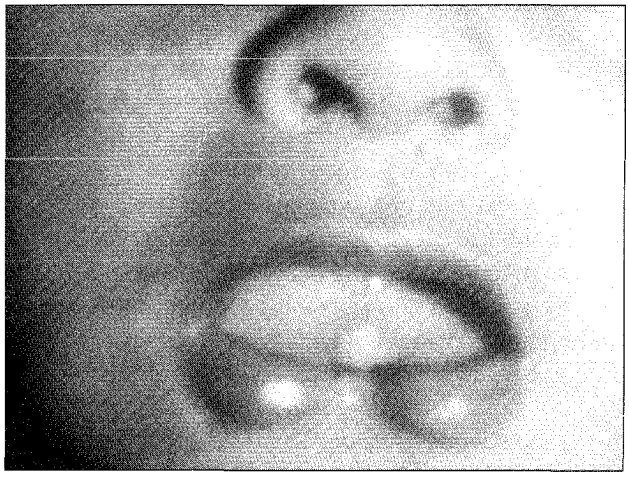

Figure 1: Shows median cleft of lower lip

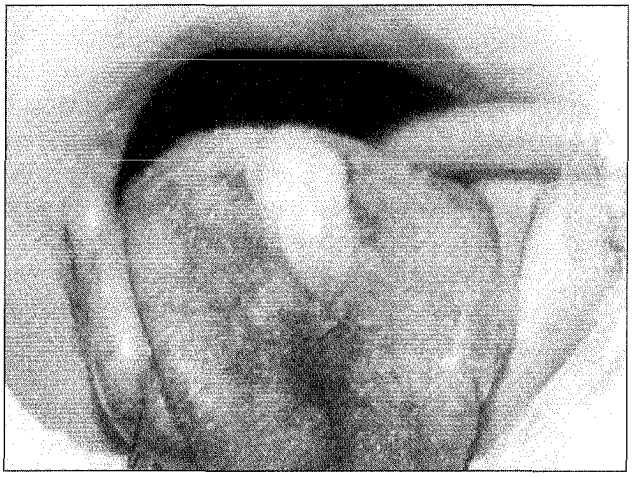

Figure 3: Shows ectopic minor salivary gland on dorsum of tongue

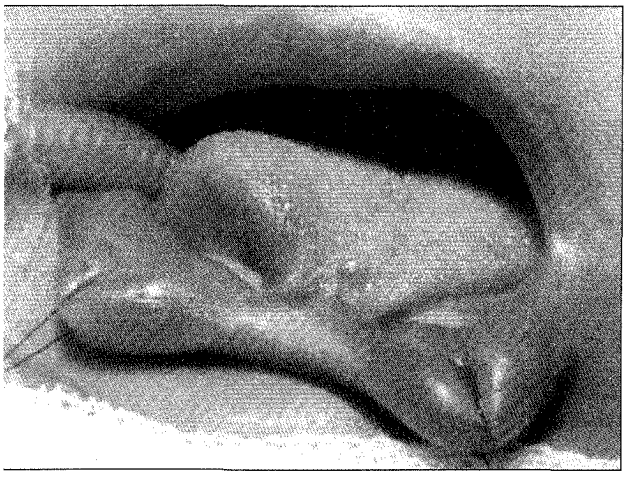

Figure 2: Shows median cleft of lower lip and ankyloglossia

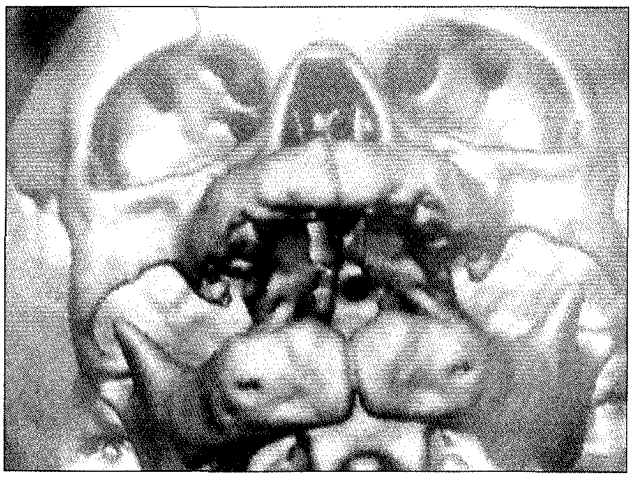

Figure 4: CT scan showing notching of mandible

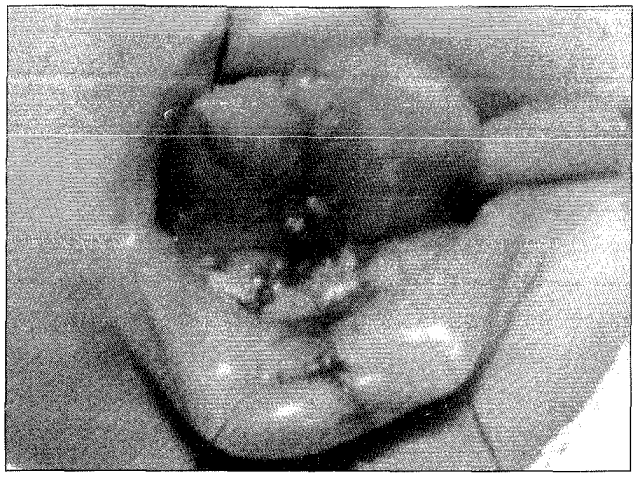

Figure 5: Immediate post-operative result

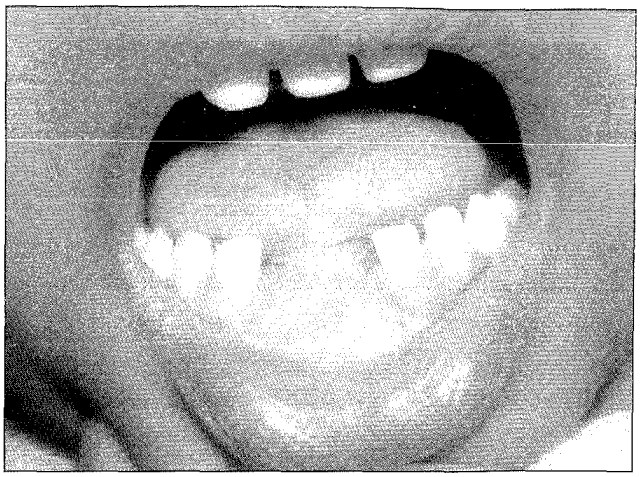

Figure 6: Shows appearance at the age of 18 months

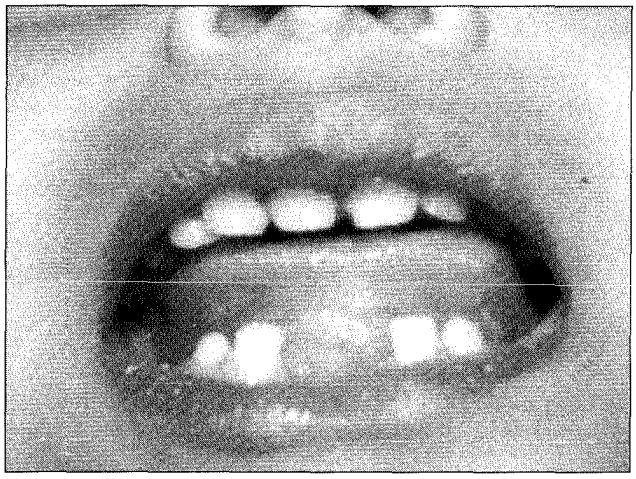

Figure 7: Shows appearance at 2 years 4 months

central incisor on right side is present. The last molars have still not erupted (Figure 7).

DISCUSSION

Cleft of lower lip and mandible is a developmental anomaly of the first branchial arches. Several hypotheses concerning its pathogenesis have been proposed in literature. Most authors consider it to be due to a failure of fusion of the first pair of branchial arches or a failure of mesodermal penetration into the midline of mandibular part of the first branchial arch. Morton and Jordan[2] feel that the second theory may explain the absence of hyoid, thyroid cartilage, strap muscles and manubrium in severe varieties. In 1996, Oostrom et al[3] proposed that, there is only one branchial arch during early embryonic period (7th week) into which two mandibular processes grow with a groove in midline. Hypoplasia of the mandibular processes during early embryonic period will lead to severest cleft of the mandible extending into the neck. During the late embryonic period less severe median clefts will develop.

Thus the deformity can range from minor to severe variety and in various combinations. In literature many variations have been described. In minor forms only the lower lip is cleft. Frequently, the cleft extends into the mandibular symphysis. The tongue may be bifid and attached to the cleft alveolar margin[4]. At times, the tongue may be absent. Armstrong[5] reported bifid uvula and ankyloglossia. In severe forms, epiglottis or hyoid may be absent.[2] Thyroid cartilage may be underdeveloped[2]. There may be a midline cervical cord. Strap muscles may be atrophic and represented by fibrous tissue causing bulging of neck during straining.[2] Monroe[1,2] reported flexion contracture of the neck. There may be a midline dermoid in the neck.[2,6] The clavicles may be widely spaced and manubrium sterni may be bifid[2,7] or absent. There may be presternal tags.[7] Seyhan[7] reported a case with associated bifid sternum with a subcutaneous mass and ventriculo-septal defect.

There may be other associated facial anomalies like cleft of upper lip, cleft palate[1], Pierre-Robin anomaly, mucus pits of lower lip, hemifacial microsomia, and dermoid cyst of nose or chin, eye and ear deformities.[2]

Congenital anomalies of heart[2,7] (single ventricle, transposition of great vessels[8]), hand anomalies[1,2] (syndactyly, brachydactyly) and anomalies of foot[2] (clubfoot, ectrodactyly) have also been reported. Although no genetic abnormality has been described, Subramani et al[9] have reported a mother having cleft tongue with ankyloglossia, linear band in the midline of palate and hyperpigmented patch over dorsum of tongue and her daughter with cleft tongue, ankyloglossia, cleft of soft palate, notched vermilion of upper lip and ectodermal dysplasia of face.

In our patient, median cleft of lower lip with ankyloglossia, notching of mandible associated with ectopic mass of salivary gland on dorsum of tongue is a rare presentation of the median cleft of lower lip and mandible. Lack of differentiation during late embryonic period may have led to these features also explaining the presence of ectopic salivary gland. Ingestion of antitubercular drugs by mother during antenatal period could be one of the contributing factors (antenatal exposure to radiation, infection, diabetes, and drugs like anticonvulsants, steroids, diazepam have been described as causative agents.)[2]

The primary treatment is soft tissue correction. When required, reconstruction of the mandible is done with rib graft[10] or iliac bone graft[11] along with vitallium[10] or titanium[11] reconstruction plate, or an acrylic splint. Seyhan[7] freshened the edges of mobile bone segments and fixed with stainless steel wires in a 10 months old child with good result. Sherman & Goulian[8] reported one-stage reconstruction using rib graft at 20 months age. Others[5,10,11] have described reconstruction 6 years onwards. Armstrong and Waterhouse[5] have suggested that reconstruction should be done after the age of 10 years to avoid damaging developing tooth buds. To simplify, if there is no gap between the two mandibular halves, the second surgery can be deferred till 10 years. While if the patient has feeding or breathing difficulty and the mandibular segments are mobile, an early attempt should be made to stabilise the mandible with bone graft or reconstruction plate.[7] As the mandible was only notched and there was no functional deficit in our patient, no bony work was done primarily. Since there is no mobility of mandibular segments, surgery, if required can be deferred till the age of 10 years.

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