

Tessier 30 Facial Cleft- Median Cleft of Upper and Lower Lip, Lower Alveolus and Mandible, Ankyloglossia and Cervical Teratoma

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Abstract

Tessier cleft no. 30 is a rare congenital anomaly. It may range in severity from median cleft of the upper and lower lip to cleft of the manubrium sterni involving the mandible, floor of the mouth, tongue, hyoid bone, thyroid cartilage and strap muscles. Ankyloglossia and median web in the neck extending from chin resulting in neck contracture are frequently found. This is a case report of median cleft of upper and lower lip, lower alveolus, mandible with severe ankyloglossia, a fibrotic band extending from the chin to the suprasternal notch and cervical teratoma; in a new born female. The deformity was corrected as staged procedure i.e. removal of cervical teratoma, release of the tongue from floor of the mouth and lower alveolus, repair of the cleft lower lip, excision of the fibrotic band and correction of neck contracture with multiple Z-plasties. Fixation of the mandibular cleft, orthodontic treatment of upper teeth, rhinoplasty and otoplasty will be done by surgeries in future.

Keywords: Tessier cleft 30; Median cleft; Cervical teratoma; Ankyloglossia

abdomen and echocardiography, further confirmed the finding and ruled out any other anomaly.

Introduction

Cleft lip and palate are common, whereas median mandibular clefts are extremely rare anomaly found in cleft no. 30 in Tessier's classification. This anomaly was first time reported by Couronne in 1819 [1]. So far upto 70 cases have been described in history. Associated anomalies are lower lip hemangioma, facial anomalies, ankyloglossia, cleft palate, cervical dermoid cyst, hyoid bone agenesis, laryngeal cartilage malformation, epiglottis aplasia, cardiac malformation and chromosomal abnormalities, which further worsen the prognosis. Failure of union of the first branchial arch in midline results in lower midline facial cleft defect whereas failure of fusion of the lower branchial arch elements are thought to lead to associated deformities of the neck [2].

Case History

A two hour born female was brought with chief complaints of deformity of lips, swelling neck and difficulty in breathing (Figures 1 and 2). There was neither history of consanguineous marriage nor any record of congenital deformity in the family. Pregnancy was uneventful, with no medication, or known exposure of X-rays. The patient was delivered at term after an uncomplicated labour, but developed respiratory distress soon after birth. There was cleft of upper and lower lip, alveolus, mandible was split in the midline, ankyloglossia and swelling 10 × 10 cm on left side of the neck crossing midline and encroaching upon right side. The rest of the physical examination was within normal limits. X-ray mandible, ultrasound



Figure 1: A two hour born female with chief complaint of swelling neck and difficulty in breathing.



Figure 2: A two hour born female with chief complaint of deformity of lips.



Figure 4: Patient is on regular follow up and is stable.

Total excision of the cervical teratoma was done at the age of 48 hours. Post-operative period was uneventful and histopathological examination came out to be teratoma. At the age of 3 months, repair of upper lip by 'Z' plasty, lower lip by 'V' excision and repair and chin repair was done. After removal of teratoma, extending from the chin down to the manubrium sternum was a firm band over which the skin was freely movable. Third surgery to release cervical contracture and full thickness skin grafting was done. Fourth surgery for complete excision of scar and contracture with multiple unequal 'Z' plasty as performed (Figure 3). The post-operative period was uneventful and weight of the patient gradually increased. Patient is on regular follow up and is stable (Figure 4). Further surgeries required in future are orthodontic repair of teeth and fixation of mandible.



Figure 3: Fourth surgery for complete excision of scar and contracture with multiple unequal 'Z' plasty.

Discussion

Couronne in 1869 was the first to describe the median cleft of the lower jaw with very few case reports in literature afterwards [3,4]. Developmental anomaly of the first branchial arches may result in cleft of lower lip and mandible. Several hypotheses concerning its pathogenesis have been proposed in literature. Regarding pathogenesis several hypothesis have been proposed in literature which include neural crest cell development disruption along normal fusion planes in the developing facial skeleton, 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 feel that the second theory may explain the absence of hyoid, thyroid cartilage, manubrium and strap muscles in severe varieties [3,4]. In 1996, Oostrom et al proposed that only one branchial arch during early embryonic period (7th week) into which two mandibular processes grow with a groove in midline. Defect in early embryonic period will lead to severe cleft of the mandible extending into the neck whereas in late embryonic period will lead to median clefts with less severity [5]. Deformity can range from minor to severe variety and in various combinations. Thus, there may be a midline notch in lower lip to complete midline cleft of the lower lip; associated mandible notching, complete cleft of symphysis of mandible, bifid or absence of tongue, hyoid bone agenesis, thyroid cartilage hypoplasia, strap muscles hypoplasia, bifid or absent manubrium sterni and bifid sternum and ventriculoseptal defect [6]. There may be other associated facial anomalies like cleft of upper lip, cleft palate, Pierre-Robin anomaly, hemifacial microsomia, mucus pits lower lip and dermoid cyst of nose, eye, chin, and ear deformities [7-9].

In our patient, median cleft of both lips, alveolus with severe ankyloglossia, notching of mandible associated with cervical teratoma is a rare presentation of the median cleft of lower lip and mandible. The rarity and variation of severity of the condition are responsible for the lack of a concordance regarding nature and timing of the corrective surgery. The majority of the authors advice corrective surgery of the soft tissue structures at earliest, to avoid feeding and speech abnormality [10,11]. Armstrong and Waterhouse have suggested that reconstruction should be done after the age of 10 years to avoid damaging developing tooth buds years unless there is an emergency like breathing difficulty or feeding problems [6,10]. Mandible surgeries

include fixation of both mobile bone segments with stainless steel wire and titanium plate with or without bone graft [11].

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