CASE REPORT

Tessier 30 Facial Cleft: A Rare Craniofacial Anomaly

Lakshmi Palukuri[1], Sharada Reddy J[2], Vaishali Naidu D[3]

Abstract

Aim: Surgical correction of median mandibular cleft with ankylossia.

Background: Orofacial developmental abnormalities that involve the upper lip and face are the most common variety. The midline cleft of the mandible is one of the rarest categorized as Tessier #30, which extends along the midline of the mandible, along with ankyloglossia or aglossia. The morphogenesis of craniofacial clefts could be due to the lack of fusion or normal development of the facial processes in the first branchial arch or failure of the mesodermal penetration into the midline.

Case description: This article presents a case of a 3-year-old female child with complete median cleft of the mandible and tongue with ankyloglossia. Single-stage mandibular cleft union with the concept of osteosynthesis and surgical correction of ankyloglossia was performed to restore function and esthetics at the earliest.

Clinical significance: This case signifies the rarity, and as very few cases have been reported worldwide, it is mandatory and worthwhile bringing to light whenever it occurs.

Keywords: Ankyloglossia, Mandibular cleft, Osteosynthesis, Tessier 30.

International Journal of Clinical Pediatric Dentistry (2023): 10.5005/jp-journals-10005-2506

Introduction

Craniofacial congenital clefts are malformations of the cranium and face with defects along the anatomic lines of fusion. The estimated incidence of these clefts is about 1.4–4.9 in 1,00,000 live births. Midline mandibular cleft is one of the rarest among orofacial anomalies.[1] Although the first case was notified by Couronne in 1819,[2] only 75–80 cases have been reported worldwide to date. Unveiling such defects is essential and highly significant whenever it occurs. Tessier[1] classified the craniofacial clefts as cleft #0–14, which radiate around the orbital rims, nose, and upper lip. Tessier #30 is the caudal extension of cleft #0–14, which extends along the midline of the mandible, along with ankyloglossia or aglossia.[1] The degree of skeletal and soft tissue involvement of Tessier #30 cleft varied widely. It might range from an indentation in the vermilion border of the lip to a complete cleft lip, extending along the tongue and the mandible through the supporting structures of the neck along the midline till the manubrium sterni.[2]

This article presents a peculiar case of the complete cleft of the mandible and the tongue along the midline with ankyloglossia, which was surgically intervened at the earliest possible.

Case Description

A female child of 3 years reported to the department with complete cleft of the mandible and the tongue along the midline with ankyloglossia. The child was delivered at term, and the pregnancy was uneventful. There was neither history of consanguineous marriage nor any record of congenital deformity in the family. On examination, the normal contour of the upper face and head with intact upper lip and nose was observed. The lower lip showed a healed scar extending from the vermilion border until the base of the chin, depicting surgical repair of the complete lower cleft lip, done at 6 months of age. A complete median cleft mandible and tongue with ankyloglossia were observed. There was fibrous adhesion of the tongue tip to the mobile mandibular segments at the cleft margins. At the point of juncture of the tongue, lower lip, and mandible, there was a large, firm reddish mass. The dentition showed a wide split in the midline between 71 and 81 and a fusion of 72 and 73 (Fig. 1). No other associated facial anomalies were observed. Clinically, the patient was unable to protrude her tongue and occlude the teeth with functional impairment of speech and mastication. The diagnosis of Tessier #30 was confirmed by the computed tomography (CT) image (Fig. 2) and diagnostic upper and lower alginate impressions.

Surgical/Operative Procedure

Department of Craniofacial Plastic Surgery and Reconstruction had planned a stepwise single-stage treatment:

- To relieve the ankyloglossia and excise the fibrous mass.
- Open reduction and internal fixation of the mandible.
- Surgical correction of the split tongue and the soft tissues.
- Stabilization of the upper border of the mandible.

[1]Department of Plastic surgery, Osmania Medical College, Hyderabad, Telangana, India

[2,3]Department of Pedodontia, Government Dental College and Hospital, Hyderabad, Telangana, India

Corresponding Author: Sharada Reddy J, Department of Pedodontia, Government Dental College and Hospital, Hyderabad, Telangana, India, Phone: +91 9290970400, e-mail: sharadagdchyd@gmail.com

How to cite this article: Palukuri L, JSR, DVN Tessier 30 Facial Cleft: A Rare Craniofacial Anomaly. Int J Clin Pediatr Dent 2023;16(1):177–179.

Source of support: Nil

Conflict of interest: None

Patient consent statement: The author(s) have obtained written informed consent from the patient's parents/legal guardians for publication of the case report details and related images.

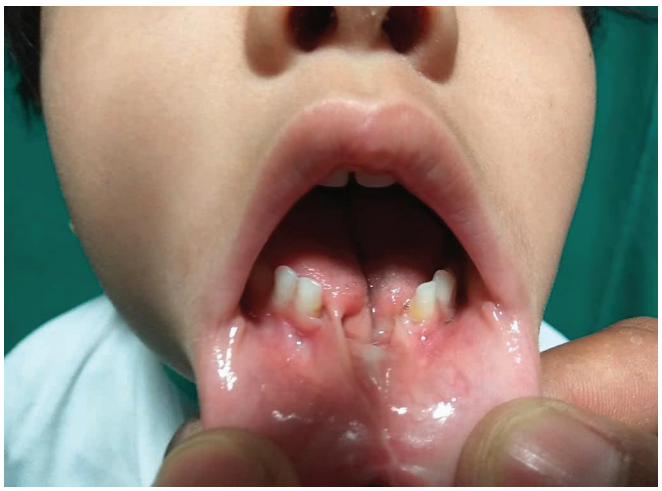

Fig. 1: Preoperative clinical image

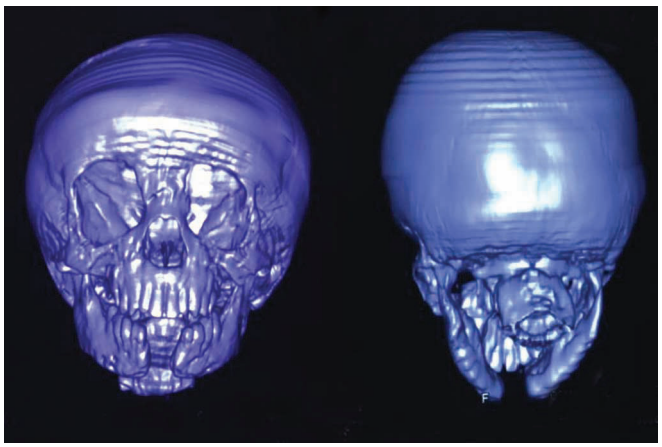

Fig. 2: Preoperative CT image

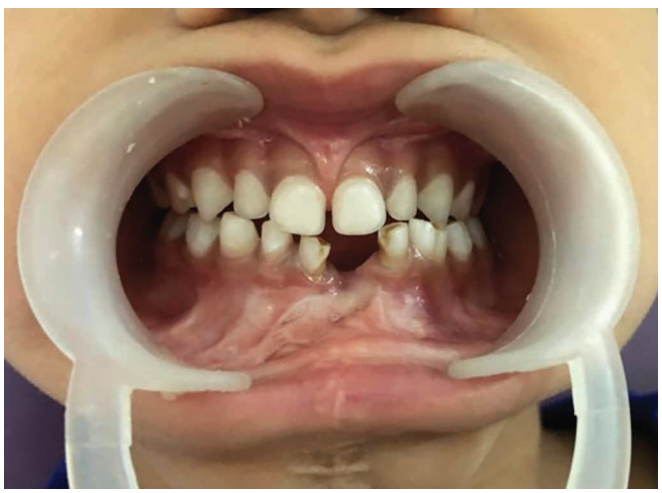

Fig. 3: Postoperative maxillo-mandibular occlusion

Under general anesthesia, the two halves of the tongue were relieved for proper access to the mandible, followed by the excision of the fibrous mass. The cleft edges of the mandible were exposed with a subperiosteal incision and freshened with the micromotor burs, and the segments were secured together with a two hole titanium miniplate and 6 mm screws at the inferior border of the mandible. Surgical correction of the split tongue, along with

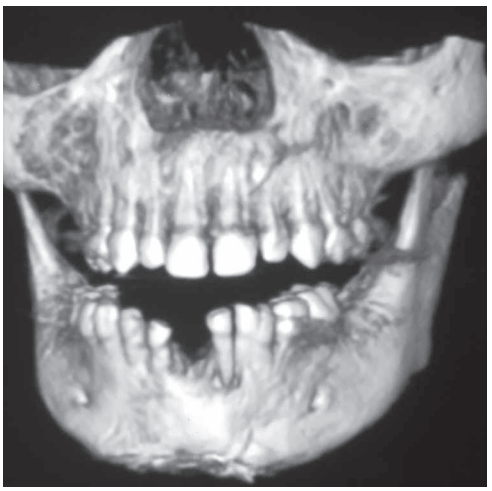

Fig. 4: CBCT after 1 year

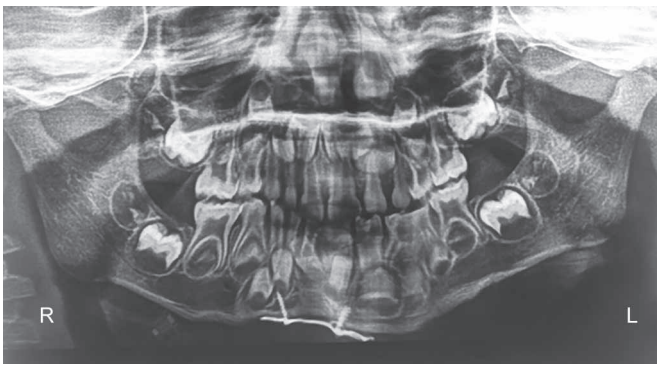

Fig. 5: OPG after 1 year

reconstruction of the soft tissues, was done. To stabilize the splaying of the upper border of the mandible, circummandibular wiring of the acrylic cap splint was done. The cap split was removed after 4 weeks, and follow-up was done regularly.

Postoperative

After 1 year, the growth of the mandible was in accordance with the chronological age of the child. There was appreciable maxilla–mandibular development with a stable mandible and an ideal occlusion (Fig. 3). Satisfactory healing of the mouth with improved tongue protrusion was observed. Postoperative cone-beam CT (CBCT) (Fig. 4) and orthopantomogram (OPG) (Fig. 5) showed a complete bony reunion of the mandibular segments.

Discussion

The fetal mandible undergoes a considerable transformation during its growth and development. The morphogenesis of craniofacial clefts is thought to be largely due to a lack of fusion or deficient mesodermal penetration along the midline of the first branchial arch. The severity of the cleft increases if a defect occurs in the early embryonic period, that is, interference in the development of neural crest cells along the fusion planes during the early formative stages of the facial skeleton.[3]

Eventual optimization of the orofacial growth and development is the ultimate goal of the surgical management of the cleft mandible and the anomalies. Due to the significant variations in its severity and rarity of Tessier #30, the treatment schedule has not yet reached a consensus. The timing to correct the defects of the mandible varied from 10 months to 15 years of age.[4] Millard

et al.[4] reported initial rectification of the midline lower cleft lip at 6 months, followed by the mandibular cleft closure at 8 years of age, as early mandibular defect correction could damage the tooth buds. On the contrary, Sherman and Goulian[5] successfully managed the median mandibular defects at 20 months of age. Armstrong and Waterhouse[6] rectified lower cleft lip deformity at the age of 5 months, while the mandibular defect was corrected with a bone graft at 4 years of age. Oostrom et al.,[7] Seyhan and Kylynr[8] and da Silva Freitas et al.[9] stated that osteosynthesis along the inferior border of the mandible is foresighted as it does not harm the tooth buds and aids in better occlusion. But, Ishii et al.[10] stated that some amount of the damage would be inevitable due to the lower position of developing tooth buds in younger age-groups.

Accordingly, in the present case, the child underwent lip rectification at 6 months of age, followed by the single-stage mandibular cleft union with the concept of osteosynthesis and surgical correction of ankyloglossia at 3 years. This was planned because:

- The child had difficulty in mastication, speech, and tongue protrusion.
- Tooth buds wouldn't be damaged with careful placement and fixation of titanium miniplates.
- Functional movement of the mandible could be achieved at the earliest.
- Treatment might benefit the maxilla–mandibular growth and establishment of proper occlusion.

Conclusion

The present case report signifies the rarity and a unique single-stage treatment approach that succeeded conventional development of the mandible. The 1-year follow-up showed an improvement in facial profile, speech, and masticatory function. Therefore, it seems reasonable to normalize the function of the mandible at the very beginning of life.

A long-term investigation is planned to trace the growth of the mandible eruption of permanent teeth into an acceptable occlusion and esthetic development.

References

1. Sari E. Tessier number 30 facial cleft: a rare maxillofacial anomaly. Turk J Plast Surg 2018;26(1):12–19. DOI: 10.4103/tjps.tjps_7_18
2. Mahajan S, Patel PK, Duan Y, et al. The median mandibular symphysis Tessier 30 cleft:: 3D imaging analysis and review of the literature. Cleft Palate Craniofac J 2018;55(5):778–786. DOI: 10.1177/1055665618756073
3. Rattan KN, Singh B, Malik P. Tessier 30 facial cleft- median cleft of upper and lower lip, lower alveolus and mandible, ankyloglossia and cervical teratoma. Otolaryngol (Sunnyvale) 2016;6(4):247. DOI: 10.4172/2161-119X.1000247
4. Millard DR Jr, Lehman JA Jr, Deane M, et al. Median cleft of the lower lip and mandible: a case report. Br J Plast Surg 1971;24(4):391–395. DOI: 10.1016/s0007-1226(71)80096-6
5. Sherman JE, Goulian D. The successful one-stage surgical management of a midline cleft of the lower lip, mandible, and tongue. Plast Reconstr Surg 1980;66(5):756–759. DOI: 10.1097/00006534-198011000-00016
6. Armstrong AP, Waterhouse N. Tessier 30 median mandibular cleft: case report and literature review. Br J Plast Surg 1996;49(8):536–538. DOI: 10.1016/s0007-1226(96)90130-7
7. Oostrom CA, Vermeij-Keers C, Gilbert PM, et al. Median cleft of the lower lip and mandible: case reports, a new embryologic hypothesis, and subdivision. Plast Reconstr Surg 1996;97(2):313–320. DOI: 10.1097/00006534-199602000-00006
8. Seyhan T, Kýlýnr H. Median cleft of the lower lip: report of two new cases and review of the literature. Ann Otol Rhinol Laryngol 2002;111(3 Pt 1):217–221. DOI: 10.1177/000348940211100305
9. da Silva Freitas R, De Oliveira E Cruz GA, Alonso N, et al. Tessier number 30 clefts: surgical correction of a rare malformation. Rev Bras Cir Craniomaxilofac 2010;13(1):12–17.
10. Ishii M, Ishii Y, Moriyama T, et al. Seventeen-year follow-up of a patient with median cleft of the lower lip, mandible, and tongue with flexion contracture: a case report. Cleft Palate Craniofac J 2002;39(5):555–559. DOI: 10.1597/1545-1569_2002_039_0555_syfuoa_2.0.co_2