ABSTRACT

Cleft lip and palate (CLP) are commonly occurring birth defects which occur due to failure of fusion of various developmental processes of the face, leading to a visible defect in the lip and palate. In severe cases, this defect may extend over the face, thus causing facial clefts. The present case of a 7-year-old girl is unique in the sense that she has right and left side facial clefts of different severity with bilateral accessory maxilla, extra set of dentition, unilateral CLP of the right side, and a double soft palate. Other less associated findings in the case are severe maxillary protrusion, complex open bite, downward and backward rotation of mandible, and incompetent lips with apparently no limb deformities. With some relevant data in hand, we present this case for various suggestions and best possible treatment plan.

Keywords: Accessory maxilla, open bite, orofacial cleft, unilateral cleft lip and palate

INTRODUCTION

During embryogenesis of face, failure of proper fusion of developing frontonasal prominence, paired maxillary processes, and paired mandibular processes lead to malunion or nonunion of lip and palate.[1] When these clefts extend over the face and cheekbone, facial clefts occur. Tessier classified facial clefts in numbers ranging from 1 to 30 depending on the location and extent of clefting.[2] Another condition which may occur separately or with facial clefts is a double maxilla or accessory maxilla. Only a few cases of maxillary jaw duplication are reported.[3,4]

The present case seems to be the 27th reported case of Tessier’s cleft no. 5 on the right side of the face. Moreover, along with the Tessier’s cleft no. 6 on the left side of the face, whose prevalence data could not be found after an extensive research, the patient also has bilateral accessory maxilla with extra set of dentition, unilateral cleft lip and palate (CLP) of the right side, and double soft palate. With these conditions occurring in a single patient and after an extensive literature search, we conclude this case report to be the first of its kind.

CASE REPORT

A 7-year-old girl reported with the chief complaint of grooves and scar marks over the cheek since birth and forwardly placed upper front teeth. Clinical and radiographic findings revealed Tessier’s no. 5 (mild form) on the right side and no. 6 on the left side [Figure 1] along with cleft lip and alveolus of the right side and incomplete hard palate cleft and complete soft palate cleft [Figure 2]. She also has bilateral accessory posterior maxilla with an extra dentition and a double soft palate. Due to transverse constriction of main maxilla posterior occlusion is possible only with the molars of accessory maxilla [Figure 2]. The patient is a known case of the repaired cleft lip at 6 months of age (Millard’s technique)

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and cleft palate at 1.5 years of age. Soft-tissue sclerotic furrow on the left side cheek was attempted for repair at 3.5 years of age, but it reoccurred and is still present. Simultaneously, lip revision was also done. All the above operations were done elsewhere.

Facial cleft on the left side of the face is on the maxilla and the zygomatic bone, opening the infraorbital fissure. This feature differentiates it from the right side no. 5 where only orbital and maxillary bones are affected [Figure 3]. The left side of the cheek has marked vertical sclerodermic furrow with a coloboma on the lower eyelid, in the lateral third, corresponding to the orbital cleft present [Figure 1]. The mandible is micrognathic and retruded, whereas the maxilla is severely prognathic. Cephalometric readings revealed her to be skeletally Class II with hyperdivergent growth pattern, increased overjet (about 15 mm), and compound open bite as well as incompetent lips [Figure 4 and Table 1]. Hands and feet are totally normal [Figure 5].

The right side cleft alveolus, mesial to deciduous canine, is a differentiating feature of no. 5, as it is not present in no. 6. She has bilateral accessory posterior maxilla with an extra set of teeth [Figures 6 and 7]. Hard palate is shallow and narrow anteriorly [Figure 2].

Orthopantomogram and cone-beam computed tomography evaluation showed that the main maxillary arch has 53, 54, 55, 11, 22, 63, 64, and 26 erupted into occlusion, whereas 15, 14, 13, 21, 23, 24, 25, and 27 (palatally placed) are unerupted. Similarly, the accessory posterior maxillary arch has 16, 5E, 6E, 26, and 27 erupted (doubtful, may be an extra premolar), whereas 17 (tooth bud), 14, 15, 25 (successor of E), 24, and 25 are unerupted. In the right posterior alveolar cleft region, one extra premolar remains unerupted. There is agenesis of 16 in the main maxilla, whereas it is present in the accessory maxilla. Mandibular dentition is following a normal eruption pattern. Presently it is in mixed dentition stage [Figures 6 and 7]. The patient is an obligatory mouth breather and hence

Figure 1: Pretreatment extraoral photographs

Figure 2: Pretreatment intraoral photographs

Figure 3: Cone-beam computed tomography – three-dimensional reconstruction of the skull

Figure 4: Lateral cephalogram showing Class II pattern with open bite and protrusive maxilla
has a generalized gingival inflammation. No chromosomal aberrations were reported through genetic mapping.

**Comprehensive treatment plan (surgico-orthodontic management)**

Surgical management of Tessier’s no. 5 and 6 consists of soft tissue and bony component. Soft-tissue repair warranted only if the defect is wide and will be proceeded by bony correction wherever possible. Surgical intervention shall be delayed till the patient has not crossed the circumpubertal period and the second permanent molars are not erupted. Meanwhile, an oral screen (to increase lip competency) has been given. This also acted as a psychological support to the patient and the parents. Transverse expansion of the maxilla will be done at appropriate time. After that, either orthodontic alignment of the accessory maxilla with the main maxilla will be done or surgical repositioning of the accessory maxilla will be done, depending on the progress of the case. Final orthognathic surgery, postsurgical orthodontics, and/or plastic revision surgery will be planned postadolescence.

**DISCUSSION**

There is always a possibility of overlap of structures affected. Cleft of soft tissue and the underlying bone (if affected) may or may not coincide.\(^2\) Craniofacial clefts have a prevalence of 1.43–4.85/100,000 live births.\(^3\) Previous studies reported that about 50% or more cases with CLP or CPO had some associated anomalies.\(^6,7\) Anatomically, cleft no. 4, 5, 6, 7, and 8 seem to be close contenders.\(^2\) Macrostomia, tags, and hypoplasia of the zygoma are features of no. 7 cleft, which is also not present.\(^8\) Cleft no. 3 looks similar to 6, except that the latter comes through the infraorbital rim more laterally. When no. 6, 7, and 8 occur together, it is called “Treacher Collins–Franceschetti syndrome.” No. 7 is more specifically related to hemifacial microsomia and no. 8 to Goldenhar syndrome.\(^2,8,9\)

Multiple surgeries at specific age may be required. Menard *et al.*\(^10\) also did bone grafting of bilateral maxillae and alveolar cleft, bilateral cheek advancement flaps, rhinoplasty, and transcranial correction of orbital dystopia in their facial cleft cases. Tissue expanders for facial cleft reconstruction are safe and reliable.\(^11,12\) da Silva Freitas *et al.* recommended the use of autologous soft-tissue repair and bone grafting for such cases.\(^12\)

**Table 1: Cephalometric analysis of skeletal parameters**

| Parameters | Pre-treatment |
|------------|---------------|
| SN length (65 mm) | 57 mm |
| SNA | 90° |
| SNB | 67° |
| ANB (3.12°±1.8°) | 23° |
| Wits (-0.01 mm) | 14 mm |
| Maxillary length (44 mm) | 45 mm |
| Mandibular length (69 mm) | 50 mm |
| Effective Maxillary length (87 mm) | 78.5 mm |
| Effective Mandibular length (mm) | 80 mm |
| N ⊥ to A point (-4 mm) | 5 mm |
| N ⊥ to B point (-10 mm) | -21 mm |
| N ⊥ to Pog (-10 mm) | -23 mm |
| FMA (23.83±2°) | 30° |
| SN-MP (32-35°) | 44° |
| Basal plane angle (30°) | 33° |
| Y Axis (59.62°±3) | 61° |
| Bjork’s Sum (394°) | 405° |
| Jaraback Ratio (62-65%) | 55.32% |
| Gonial Angle (123±7°) | 135° |
| Upper anterior facial height (46%) | 39.60% |
| Lower anterior facial height (54%) | 60.40% |

![Figure 5: Normal hands and feet](image1)

![Figure 6: Axial view of cone-beam computed tomography section showing two sets of accessory posterior dentition on the accessory maxilla](image2)

![Figure 7: Panoramic radiograph](image3)
Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Sperber GH, Sperber SM. Embryogenetics of cleft lip and palate. In: Berkowitz S, editor. Cleft Lip Palate Diagnosis Manag. 3rd ed. New York: Springer Heidelberg; 2013.
2. Tessier P. Anatomical classification facial, cranio-facial and latero-facial clefts. J Maxillofac Surg 1976;4:69-92.
3. Borzabadi-Farahani A, Res M, Yen SL, Yamashita DD, Sanchez-Lara PA. Bilateral maxillary duplication: Case report and literature review. Oral Surg Oral Med Oral Pathol Oral Radiol 2012;113:e29-32.
4. Sjamsudin J, David D, Singh GD. An Indonesian child with orofacial duplication and neurocristopathy anomalies: Case report. J Craniomaxillofac Surg 2001;29:195-7.
5. Kawamoto HK Jr. The kaleidoscopic world of rare craniofacial clefts: Order out of chaos (Tessier classification). Clin Plast Surg 1976;3:529-72.
6. Shprintzen RJ, Siegel-Sadewitz VL, Amato J, Goldberg RB. Anomalies associated with cleft lip, cleft palate, or both. Am J Med Genet 1985;20:585-95.
7. Rollnick BR, Pruzansky S. Genetic services at a center for craniofacial anomalies. Cleft Palate J 1981;18:304-13.
8. Hou R, Feng X, Zhang J, Lu B, Liu G, Wang L. A rare bilateral tesser no 6 and 7 clefts. J Craniomaxillofac Surg 2011;39:93-5.
9. Harrison MS. The Treacher Collins-Franceschetti syndrome. J Laryngol Otol 1957;71:597-604.
10. Menard RM, Moore MH, David DJ. Tissue expansion in the reconstruction of tessier craniofacial clefts: A series of 17 patients. Plast Reconstr Surg 1999;103:779-86.
11. Galante G, Dado DV. The tesserer number 5 cleft: A report of two cases and a review of the literature. Plast Reconstr Surg 1991;88:131-5.
12. da Silva Freitas R, Alonso N, Shin JH, Busato L, Dall’Oglio Tolazzi AR, de Oliveira e Cruz GA. The tesserer number 5 facial cleft: Surgical strategies and outcomes in six patients. Cleft Palate Craniofac J 2009;46:179-86.