Bilateral Tessier's 7 Cleft with Maxillary Duplication

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Abstract Tessier's 7 cleft or lateral facial cleft are unusual lesions that result from failure of the embryonic mandibular and maxillary process of the first branchial arch to fuse properly and form the corners of the mouth. It may be seen alone or in combination with other anomalies, accompanied by varying degrees of severity. We report an extremely rare case of bilateral Tessier's 7 cleft along with maxillary duplication, macrostomia, bilateral posterior maxillary cleft, and mandibular retrusion in an 18-year-old male patient.

Keywords Tessier's 7 cleft · Macrostomia · Maxillary cleft

Introduction

Craniofacial clefts are very rare congenital deformities. It is found in one out of 80,000 births and occurs more frequently in males. Most cases are unilateral, but bilateral involvement can be seen in 10–20% of cases [1]. Although, the transverse or lateral cleft is the most common type of craniofacial cleft excluding cleft lip and palate, the associated maxillofacial skeletal deformities and dental anomalies have rarely been reported in the literature [1]. Earlier documentations of the posterior maxillary clefts have used the term “maxillary duplication”. Duplication of jaws carrying teeth may occur in maxilla or mandible, either as an accessory process or more rarely, a complete jaw [1, 2].

The first and second branchial arch involvement during early embryonic development results in a wide spectrum of anomalies that may include macrostomia and lateral facial clefts [2]. The lateral facial cleft, which results due to incomplete development of the perioral muscles of the face, gives an appearance of macrostomia giving rise to potential psychological and esthetic problems. We report an extremely rare case of bilateral Tessier's 7 cleft along with maxillofacial skeletal deformities and dental anomalies in an 18-year-old male patient. The surgical rehabilitation of the case is also discussed.

Case Report

An 18-year-old male patient reported to the department of oral and maxillofacial surgery with the chief complaint of deformed face, wide mouth, and multiple teeth in the upper jaw since birth. The deformities had worsened with age. Both the patient and his parents had been aware of the deformity but delayed evaluation because of economic circumstances.

Extraoral examination revealed macrostomia with zygomatico-maxillary deformities, dental anomalies, and mandibular retrusion. The frontal view showed that both oral fissures extended horizontally into the cheek, nearly 1.5 cm from the normal lip point, and the two lips were not at the same level, with the right side a little higher with a skin tag on the left side of the cheek (Figs. 1, 2, and 3). At rest the lips were incompetent with significant upper and lower tooth exposure. The lateral view presented with macrostomia and the maxilla was severely protrusive with associated protrusion of the upper teeth. The width of the open mouth was 3.5 cm. Intraoral examination revealed polydontia with a total of 32 teeth present in the maxilla

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resulting in severe crowding of the arch thus impairing speech, mastication, and occlusion (Fig. 4). Orthopantomograph revealed polydontia and a bilateral posterior cleft of the maxilla (Fig. 5). CT scan showed bilateral posterior cleft in the maxilla with polydontia (Figs. 6, 7). There was hypoplasia of both zygomas, especially the zygomatic arch. A diagnosis of bilateral Tessier's 7 cleft with duplication of maxilla was made based on the clinical and radiological features. Management of this deformity was planned in three stages, first correction of macrostomia, then maxillary arch correction by orthodontics and mandibular advancement at a later stage. Patient was posted for surgery under general anesthesia, Pfeiffer wavy incision technique was used for bilateral Tessier's 7 cleft closure (Fig. 8). After marking the proposed oral commissure, the vermillion turnover flap was raised and the upper and lower muscle bundles were skeletonized and divided. The upper bundle was sutured over lower one in a crisscross manner to create the modiolus and a straight-line three-layer skin closure was obtained. Teeth which were interfering with occlusion and out of the arch were extracted at the same time. Sutures were removed after a week and healing was satisfactory (Fig. 9).

Fig. 1 Frontal view showing macrostomia with bilateral Tessier's 7 cleft with skin tag on the left side of the cheek

Fig. 2 Right lateral view showing extension of the Tessier's 7 cleft

Fig. 3 Left lateral view showing extension of the Tessier's 7 cleft along with skin tag

Discussion

The number 7 cleft presents as macrostomia and pre-auricular tags. It also presents with a temporo-zygomatic cleft with the absence of a zygomatic arch, deformities of the mandibular ramus, condyle, and coronoid process. The number 7 cleft is the commonest atypical craniofacial cleft, with the incidence of the macrostomia malformation reported to be between 1:3,000 and 1:5,642 births [2]. The possible etiology is insufficient merging of the maxillary process with

Fig. 4 Intraoral view showing polydontia along with bilateral posterior maxillary cleft

the mandibular process of first branchial arch. In addition, these processes are hypoplastic. Therefore, the cranial clefts are often associated with hypoplasia of the jaw bone and the surrounding soft tissue. The severity of the cleft can vary, and thus, additional symptoms may or may not be present. Examples include pre-auricular tags; microtia; zygomatic arch deficiency; deformities of the mandibular ramus, condyle, and coronoid process; and other craniofacial clefts. The cleft can also be part of an underlying syndrome and present with an absent orbital rim and eyelash hypoplasia in Treacher Collins syndrome, facial asymmetry in hemifacial microsomia, and vertebral anomalies in Goldenhar syndrome.

The pathogenesis of lateral cleft as well as the posterior maxillary cleft is not firmly established. Most commonly cleft occurs due to non-union between different processes forming the face (the maxillary and mandibular and the maxillary and the zygomatic process). This was challenged by the proponents of the mesenchymal penetration theory,

Fig. 5 Orthopantomogram (OPG) showing polydontia along with bilateral posterior maxillary cleft

Fig. 6 CT axial view showing polydontia with accessory maxilla posteriorly

which holds that there is deficiency in mesenchymal penetration of this embryonic region resulting in epithelial breakdown and subsequent facial cleft formation. The etiological causes of the lateral facial cleft appears to be heterogeneous: (1) vascular cause, attributable to embryonic hematoma formation from disruption of the stapedia artery stem, the severity of the condition depending on the size of the hematoma and its time of resolution as brachial arch development occurs, which is similar to hemifacial microsomia; and (2) environmental factors, including an increase in number of infectious diseases, drugs or chemical agents capable of inducing embryonic dysmorphogenesis [3].

Fig. 7 CT axial view showing bilateral posterior maxillary cleft with hypoplastic zygomatic arches

Fig. 9 Postoperative view showing closure of bilateral Tessier 7 cleft

Fig. 8 Frontal view showing markings of Pfeiffer wavy incision technique bilaterally

Cheung et al. [4] postulated that in the deep tissue between embryonic maxilla and mandible, a late compensatory growth from the embryonic temporal bone could have penetrated into this region before the complete breakdown of the overlying ectoderm. The ectodermal cover of this mesodermal budding will form the tooth germs in the future accessory maxilla [4, 5].

Management of this condition varies with age and systemic associations and is mainly cosmetic in uncomplicated cases [6]. In patients with mandibular hypoplasia advancement can be done by using either orthognathic surgery or distraction osteogenesis [7]. Maxillary protrusion can be corrected by orthognathic surgery. In the present case we initially operated for macrostomia and later planned for orthodontic treatment followed by orthognathic surgery.

Conclusion

Macrostomia not only jeopardizes the normal facial anatomy and structure, causing saliva or food flowing out of the mouth, but may also impair the occlusal relationship and the development of the mandible [8]. Therefore, the treatment, especially the surgery for the facial cleft, should be performed as early as possible during infancy. In our

case, however, it was reported in an adult patient we had decided to carry out surgical treatment in three different stages with regular follow up.

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Conflict of interest None declared.

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