Accessory Maxilla in a Tessier 07 Cleft - A Case Report

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Abstract

The Rationale: Accessory maxilla is a rare condition often associated with Tessier type-7 clefts with fewer than 25 cases recorded in the literature. This manuscript reports a unilateral accessory maxilla with six supernumerary teeth. **Patient Concerns:** A 5-1/2-year-old boy, a treated macrostomia case, on follow-up visit showed evidence of accessory maxilla with teeth on radiological examination. The structure was interfering with growth, and hence, surgical removal was planned. **Diagnosis:** Based on clinical history, diagnosis and imaging, accessory maxilla with supernumerary teeth was diagnosed. **Treatment and Outcomes:** The accessory structures and teeth were removed surgically via an intraoral approach. Healing was uneventful. The growth deviation was arrested. **Take-Away Lessons:** Intraoral approach is a good option to remove an accessory maxilla. Tessier type-7 cleft may be accompanied by type-5 clefts and such accessory structures when impinging on vital structures such as temporomandibular joint or facial nerve should be immediately removed to facilitate proper form and function.

Keywords: Accessory maxilla, macrostomia, maxillary duplication, supernumerary teeth, tessier cleft type-7

Background

Partial duplication of the maxilla is an uncommon condition with fewer than 25 cases reported in literature and associated with Tessier-7 type of clefts. This condition is referred to as 'accessory maxilla' or 'maxillary duplication' and is used to describe a clinical entity that is characterized by the presence of extra bone, lying posterior to the maxillary tuberosity. Often the condition is associated with abnormal zygomatic arch and facial clefts, notably Tessier no. 7 cleft.[1,2] Oblique facial clefts are a rare craniofacial deformity that can manifest in a variety of patterns and severity. Macrostomia is a unique type of oblique facial cleft that results in a large mouth by involving commissures of the involved side. Conventionally, Tessier-5 and Tessier-7 (soft tissue) types are associated with macrostomia.[3,4] In such cases, the occurrence of supernumerary teeth and odontomes are not uncommon.[5] The aim of this manuscript is to present a maxillary duplication with several supernumerary teeth occurring in a Tessier-5,7 case.

Case Report

A 5-1/2-year-old male patient, known case of Tessier-7 (soft tissue) facial cleft with evidence of mandibular hypoplasia managed surgically in infancy presented for regular post-operative check-up. At 6 months of age, patient was operated by the author for the closure of macrostomia, which was successfully done [Figure 1]. No other abnormalities were noted. The next stage of correction was to be done later at an appropriate age. Patient was lost to follow-up subsequently.

After the subsiding of the third wave of COVID-19 pandemic, patient presented again. In the intervening time, the active growth ensured and entire dento-alveolar apparatus structures were disrupted. There were many misplaced teeth and supernumerary teeth observed. The patient had clinically and radiologically age appropriate full complement of left maxillary quadrant in the bony segment mesial to a bony cleft. Radiologically, in the left maxilla, there was a bony cleft. Radiologically, right maxillary quadrant was age appropriate. The mandibular right quadrant set of teeth corresponded numerically while the position of teeth varied caused by the tilting of the anterior teeth while the entire left quadrant teeth was pushed distally. To compensate, the left mandibular growth was also distorted [Figure 2].

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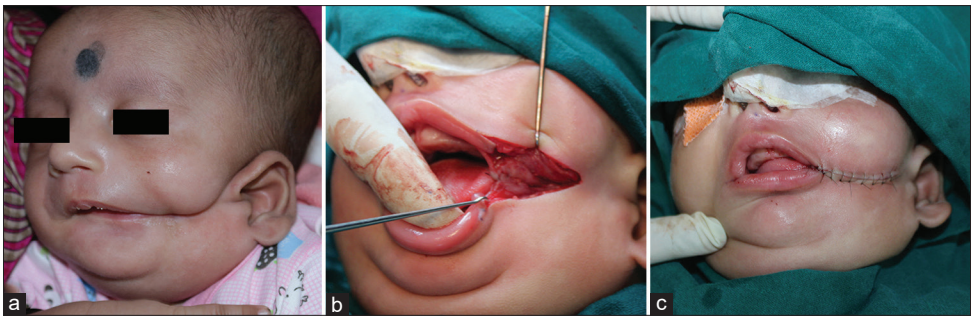

Figure 1: (a) Patient at six months of age showing macrostomia along left commissure of the mouth, (b) Intraoperative view of closure of macrostomia, (c) Immediate post-operative

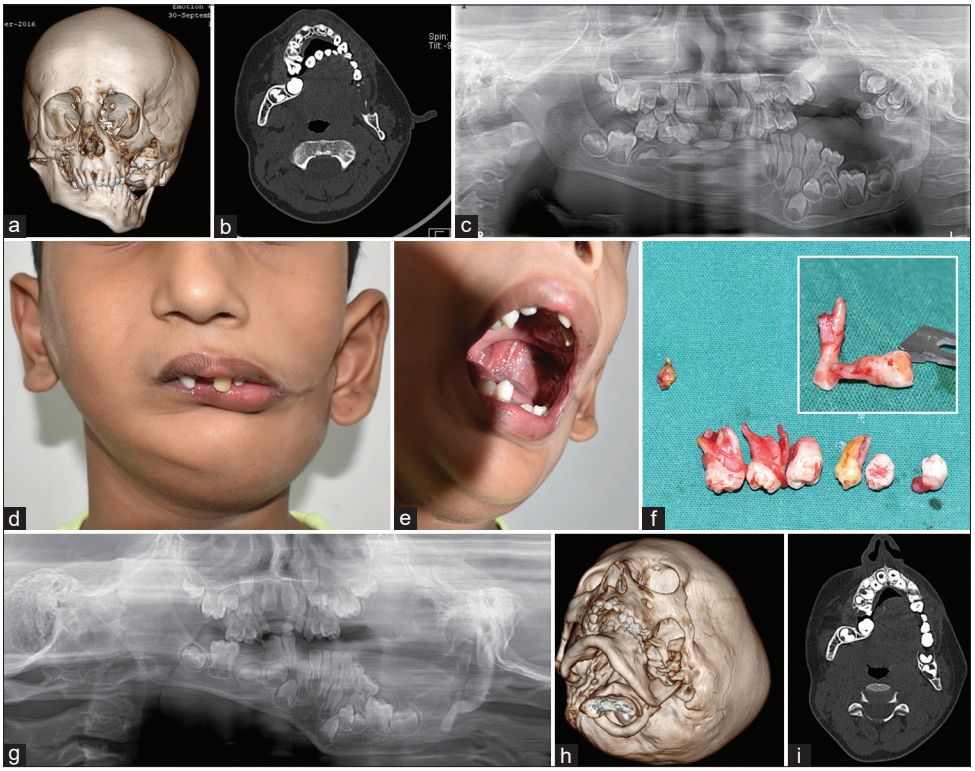

Figure 2: (a-c) Pre-operative 3D CT scan and OPG at 5-1/2 years showing the cleft, accessory maxilla posterior to tuberosity with supernumerary teeth, Note the corresponding changes in the mandibular arch, (d) Frontal profile of the patient. Note the difference between right and left malar arch, (e) Intraoral appearance of the cleft, (f) Resected hard tissue from the accessory maxilla, (g) Immediate post-operative – OPG X-ray, (h and i) Post-operative 3D CT scan. OPG = Orthopantomogram; 3D = Three-dimensional; CT = Computed tomography

Posterior to the tuberosity, a separate bony structure resembling accessory maxilla was observed. This entity had as much as six supernumerary teeth all clumped together, suggestive of a possible developing odontome distal to the cleft with the structures being pushed into the coronoid area with the teeth lying in proximity to the root of the zymgomatic process of temporal bone. The outgrowth appears to extend diffusely from the inferior border of the right zygomatic bone to the maxillary tuberosity with a clear gap between the normal maxilla and the outgrowth of bone on the left maxilla. Based on the patient's history, clinical and imaging findings, a diagnosis of accessory maxilla with left side Tessier no. 7 cleft with a possible Tessier type-5 cleft was arrived at. The developing accessory maxilla caused wide spread pressure effect and distortion to the base of the skull and adjoining structures.

The surgical goal was to remove the accessory bony and teeth elements that were impeding regular function and growth of the vital native tissues including maxilla, mandible and temporomandibular joint (TMJ) area.

Under standard preparation, after nasotracheal intubation, through an incision in the left retromolar region mucoperisosteal flap was raised. The transalveolar extraction of teeth was carried out and the entire dento-alveolar structures removed. The area was closed in layers. Appropriate pain killers and antibiotic coverage were provided. Patient had no complication and healing was uneventful. Patient is being followed up for the later correction of jaws.

Discussion

Oblique facial clefting involving mouth is a rare genetic entity classified as Tessier-7 congenital cleft or macrostomia with an approximate incidence of 1-in-80,000 to 1-in-300,000 live births or 0.3%–1.0% of the cleft spectrum. It is believed to be a result of abnormal foetal development of the first and second brachial arches.[3] Isolated macrostomia with maxillary duplication is rare and the entity is more commonly associated with other forms of facial clefts.[1,2] Owing to this multitude of defects, patient was assessed and operated early in life to ensure the normal development of speech, eating, facial appearance and possible social interaction. Although some advocate delaying such correction and creation of new commissure when there is a need of mandibular and zygomatic repair, ensuring coordinated growth and proper feeding becomes paramount. Hence, in the present case, early correction of macrostomia was commenced and finished. There are several techniques and modification each having its own advantage and limitation. Author preferred the Kaplan's modification of four-layered technique to accommodate growth and meet the parent's expectation.[6] As observed, the goal of the surgery was achieved as a near normal left commissure could be seen in the patient even after five years of surgery.

Unilateral accessory maxilla is a rare condition often associated with Tessier Type-7 clefts with fewer than 25 cases reported worldwide. The Tessier Type-7 is a rare type of facial cleft that begins medial to oral commissure involving the maxillary bone and extending to various depths, even to the inferior orbital rim. Such types of clefts are rare. The bone involvement usually includes an alveolar cleft in the premolar region, extends across the maxilla lateral to the infraorbital nerve, up to the infraorbital rim and orbital floor.[1,2] Fortunately, in the present case, the infraorbital rim was spared.

As per the Neuromeric theory, the Type-7 clefts arise from the r2 mesenchyme and jugal developmental field with the zygomaticofacial neurovascular bundle being compromised. A classic type-7 cleft would cause malar flattening and soft tissue clefting from the inferior orbital fissure toward the commissure. Consequently, maxilla would be retruded leading to an Angle Class III occlusion. Incidentally, the zygomaticomaxillary buttress is spared. Rarely, the condition occurs in the isolated form and mostly partial. The present case is also a partial and has nearly all above reported conditions. In addition, these facial defects have been also attributed to hypoxia due to Stapedial artery disruption, amniotic membrane complex disruption and subtle genetic mutations.[7] The exact cause of maxillary duplication is still not clearly delineated.[1,2] A recent review by Sun *et al*. provides a pooled analysis of accessory maxilla and interestingly, most of the classic findings of the case are consistent with the results of the 24 cases of accessory maxilla.[2]

After the diagnosis, the most challenging part of the case was to choose the appropriate type of surgical approach to the accessory maxilla.[1,2] If to continue through an intraoral approach, it will be facial scar sparing but may result in the incomplete removal of the duplicated maxilla, limited surgical access and possible damage to already malformed adjacent bone and soft-tissue structures. While an external preauricular approach will damage the delicate root of zygomatic arch–temporal process and in the process may damage the facial nerve as well as TMJ tissues during tissue remodeling that often accompanies postsurgical removal of bone. The decision to pursue an intraoral approach was based on the intact preservation of TMJ apparatus and not damaging facial nerve during the external surgical approach. Although the access was limited, as age was in favour, removal of accessory tissues was feasible.

Conclusion

A rare case of unilateral accessory maxilla in a Tessier cleft-7 is reported along with successful management. The case highlights the need for proper, regular follow-up of cases that have complex facial clefting. Developing abnormal structures such as accessory maxilla and hyperdontia, inherently pose a challenge to space, speech, function, nutrition and may even hinder development of adjoining vital structures. By mechanical impingement, they could also disturb the neurovascular bundles. Incidentally, they also pose a threat to the developing occlusion warranting the removal of unnecessary tooth at the earliest possible time.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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