Macrostomia in association with pre-auricular tags: a case report

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Abstract:
Macrostomia is a rare congenital anomaly with an incompletely described pathogenesis and several distinct non-syndromic phenotypes are associated. Macrostomia and pre-auricular tags usually can be correlated with syndromes, but in isolated cases are less common. Case report: In this case show a girl presenting macrostomia and pre-auricular tags isolated without association syndromic or other apparent causes. In the anamnesis, parents reported having a pregnancy without complications. In ectoscopic tests, ultrasound, and intra-oral physical examination were not significant alterations found beyond macrostomia and pre-auricular tags. It performed surgery to correct macrostomia and withdrawal of pre-auricular tags. Discussion: Macrostomia is a rare malformation that can only be treated surgically. Various surgical techniques have been described in the literature for the correction of these defects. In this case, the surgical option was the Z-plasty technique, however others techniques can be chosen according to the position of facial cleft. Conclusion: Although rare in occurrence, requires further studies to a better knowledge leading to a more effective diagnosis and treatment of the patient.

Keywords: Macrostomia; Craniofacial Abnormalities

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INTRODUCTION

Macrostomia is a rare congenital anomaly whose main result of expansion of the oral cavity resulting in the mouth deformation. There is a prevalence of 1: 60,000-300,000 newborn and males are affected more often than females. Macrostomia is correlated with syndromes such as Apert, Crouzon, Goldenhar, Treacher Collins, Simpson-Golabi-Behmel, Fryns and Barber-Say, but it can also present in isolated cases without syndromic association, as for example hemifacial microsomia.

The Tessier classification ranks deformations and orofacial clefts according to their position in the face, ranging from 1-14. The macrostomia is classified as defect Tessier at position seven, which is located in the temporo-zygomatic area, just above the mandibular branch, behind the zygomatic arch, together the coronoid process of the mandible. This macrostomia is due to failure of fusion of the maxillary and mandibular processes of the first and second branchial arch or due to disruption in the processes after fusion. It is commonly associated with others defects of the first branchial arch, as cleft and appendices (tags) pre-auricular. This cleft can present as slight widening of the mouth to a cleft extending up to the ear. It can be bilateral, but most of the reported cases are unilateral and do not extend beyond the anterior border of the masseter. Problems associated with macrostomia include aesthetic disharmony and functional problems like feeding difficulties, drooling, speech incoherence, and difficulty in blowing. The goal of surgical correction of these clefts includes good aesthetics and better function of orbicularis oris muscle. The aesthetic outcome of these surgeries depends not only on the placement of scars along the natural skin creases but also on their show up during facial expressions. No guidelines exist for preoperative workup of patients with macrostomia. However, the surgical repair as early as possible is correlated with more normal speech development.

The purpose of this study was to relate a clinical case of unilateral Tessier’s 7 cleft along with macrostomia and pre-auricular tags in female patient and your treatment.

CASE REPORT

A 2 years-old female was attended in Oral Pathology Clinic of State University of Montes Claros – Minas Gerais state, Brazil, accompanied by her responsible, forwarded by the of pediatric neurology. In anamnesis, the mother reported to have had a pregnancy and quiet and uneventful birth, as the child’s cognitive development has also been normal. The mother also reported that already has two daughters and clinical parameters of the same would be normal. Upon ectoscopic examination it was found that the patient would have a facial asymmetry (Figure 1) located in the right commissure lip region with extension to angles of the mouth to the masseter area (Figure 2). Pre-auricular tags were also found on the same side of facial. In intra oral clinical examination, no changes were found or noteworthy (Figure 3).

In tests ultrasound showed typical homogeneous echogenicity in the anterior region of the left side, and labial one hypoechoic image, the longitudinal right to the orbicularis muscle of the lip disagreeing with very image of a normal muscle of the mother (Figure 4). Further physical examination did not show any other
abnormalities. The patient after the clinical diagnosis of Macrostomia was referred to plastic surgery service where surgery to reposition and junction of the labial tissue was performed by Z plasty technique, and removed the pre-auricular tags (Figure 5). The patient remains in follow-up with speech therapy and dentist.

**DISCUSSION**

The incidence of macrostomia is present more in males. The presence of unilateral deformity is more prevalent that in the bilateral and 60% to 70% of the unilateral cases were associated with ear deformities and pre-auricular tags. Their presence of macrostomia and pre-auricular tags usually is associated with other syndromes, such as Treacher Collins, Fryns, Barber-Say and Goldenhar, however with these all syndromes is present systemic alterations as renal and cardiac anomalies, polydactyly, mandibular asymmetry, and other systemic alterations, but this in case presented, we see that involvement was a girl and without systemic changes beyond the macrostomia and pre-auricular tags, which are embryonic problems.

The etiopathogenesis of macrostomia remains unknown, many theories have been proposed. The mandibular dysplasia and transverse soft tissue deficiency could be explained by a lack of growth of the mandibular process. Others have suggested a vascular etiology, explained by the presence of a hematoma in the territory of the stapedial artery preventing fusion of the maxillary and mandibular processes. In the case of bilateral macrostomia, however, both stapedial arteries need to be pathological, making this explanation less plausible. Buonocore et al. believes that both uterine trauma as amniotic bands syndrome resulting in the failure to fusion of the lower jaw and secondary processes would be capable of producing such an anomaly. Furthermore, the diastasis of facial muscles can result in delayed and subsequent fusing mesodermal penetration, resulting in the failure of fusion of the mandibular and maxillary process. What could be a possible cause of bad congenital formations, causing macrostomia and pre-auricular tags.

The recent genetic study in a large family of Chinese Han origin identified that a germline PTCH2 mutation may be associated with macrostomia, suggesting the importance of considering genetic in characterization of etiologic risk factor of macrostomia. In this case, there was no story of medication or illness during pregnancy. In individual cases, it is often impossible identify a specific etiology factor. Therefore, this case reports macrostomia non-syndromic with pre-auricular tags.

Aesthetics as well as function, especially in subjects without other anomalies, is a great concern. Surgery should be done in young age to avoid unwanted anxiety and psychological impacts on both child and family.
as well as correcting sialorrhea, speech problems and compromised chewing ability. The objective of surgical treatment includes to restore of symmetric lip length and height and contour, create of mobile and symmetric commissures, myoplasty to restore labial function for speech, eating, and blowing, skin closure with minimal visible scarring and mucosal repair with adequate vestibular space. In the present case was an unilateral macrostomia. In contrast to bilateral involvement, the measurements of the unilateral cases are taken from the normal side and are transposed to the abnormal side. In addition, in repair process reconstruction of commissural part needs precise technique. It is already described in the literature that muscle suture should obey an order of application of the upper orbicularis muscle before the lower portion, but without real function commitment. In relation to skin repair, technical Z-plasty and W-plasty are widely used and with good aesthetic and functional results. Scars fragmented by the techniques previously mentioned often avoid retractions and give harmony to the lip. The nomenclature of these surgical techniques is due to the scar format after the surgery. In this case, the surgical option was based on the reconstruction of the labial and retail site rotation with Z-plasty technique with good anatomical positioning and aesthetic result.

CONCLUSION

In summary, although the unilateral macrostomia associated with pre-auricular tags is rare in occurrence, requires further studies to a better knowledge leading to a more effective diagnosis. Various surgical techniques have been proposed for the correction of defects. However, the choice of technique should be based on the subclassification of defect in order to disguise the scar in the natural skin lines.

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