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Case Report

Median Cleft of Lower Lip with Central Cleft Palate and Ankyloglossia: A Case Report

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Abstract ***Purpose:*** Median cleft of the lower lip is an uncommon condition. We report a case of a non-syndromic Chinese girl with a central cleft palate, median lower cleft lip and anklylossia. ***Methods:*** A full term 6 momths old female child presented to our clinic for central cleft palate. Her palate was repaired uneventfully at 12 months of age. During the regular post palatoplasty follow up when she was 5-year-old, we noticed a subtle notch in the midline lower lip and ankyloglossia. ***Findings:*** As subsequent speech assessment showed hyponasal speech, velo-pharyngeal insufficiency corrective surgery together with surgical tongue tie release and surgical repair of the median cleft lower lip were offered. The parents needed more time to think about it. ***Conclusions:*** This case report highlighted the importance of longitudinal clinical follow up of patient with cleft anomalies.

Key words *Ankyloglossia; Median cleft lower lip*

Introduction

Median cleft of the lower lip is an uncommon condition. Despite a cleft lip is often considered as a congenital anatomical anomaly, sometimes, the clinical presentation can be so subtle that it can be missed by multiple healthcare professionals, especially when the lesion is asymptomatic. The incidence of median cleft lower lip in the Chinese population has not been reported. Hereby we report a case of a non-syndromic Chinese girl with central cleft palate, and a delayed presentation of median lower cleft lip and anklyloglossia.

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Case Report

A full term 6 months old female child was brought to our clinic for incidental finding of central cleft palate. She was born in China with unremarkable antenatal history. Her parents were of non-consanguineous marriage and there was no known maternal exposure of drugs or radiation. She had no family history of cleft lip or palate. On clinical examination at 6 months old, there was no obvious cleft lip and a wide central cleft palate involving the soft and hard palate. There was no cleft alveolus. She also had a 4 cm non-complicated haemangioma at the buttock, otherwise she has no other dermatological or skeletal anomalies. Audiological and clinical assessment by the Ear, Nose and Throat surgeon revealed bilateral otitis media effusion with moderate hearing loss. Elective cleft palate repair and bilateral grommet insertion was performed at one year of age as per our centre's protocol. Pre-op routine ultrasound examination and echocardiogram confirmed no structural cardiac, urinary or central nervous system anomalies. Von Langenbeck's palatoplasty with intravelo-veloplasty was performed under general anaesthesia uneventfully. Her post op recovery was uncomplicated and she continued to receive regular

clinical follow up with interval assessment of her speech and skeletal development.

Continual regular speech assessment revealed mild audible nasal airflow for high vowels. Perceptive speech evaluation showed mild hyponasality, age appropriate speech intelligibility and mild disarticulation. Clinical examination at 5-year-old showed healed palate with no palatal fistula and palatal movement was observed during phonation. However, there was also a subtle but noticeable notch in the midline lower lip mucosa (Figure 1). There were no associated lip pits at both the upper and lower lip. Upon further questioning, the mother explained while she had noticed the child's lower midline lower lip mucosal notch at birth, and it had gradually become more apparent clinically with age. She even volunteered a positive family history with the child's paternal grandmother having a similar median lower lip submucosal notch, with no surgical intervention was required. In addition, we also noticed tongue tie with restricted tongue movement, potentially impacting on her current articulation. The clinical diagnosis of a median lower lip microform cleft with ankyloglossia and hyponasal cleft speech was made. The girl was then scheduled for standard velo-pharyngeal insufficiency assessment including naso-endoscopy and multi-view fluoroscopic studies. And she was advised to continue with the intensive speech therapy. If significant velo-pharyngeal insufficiency is to be confirmed upon further investigations, we planned to offer velo-pharyngeal insufficiency corrective surgery together with tongue tie release. Moreover, we also offered surgical repair of the median cleft lower lip. Upon detailed discussion with the mother, she decided to complete all relevant investigations first before considering the lower lip corrective surgery, as it was aesthetically acceptable to the patient and was not causing any symptom.

Discussion

Median cleft lower lip is an uncommon condition. It is formally classified as the Tessier no. 30 facial cleft.[1] It was first described by Couronne in 1819[2] and till now, only about 75 cases have been reported in the literature worldwide.[3] According to the theory proposed by Oostrom et al,[4] lower lip median cleft is caused by the failure of fusion of the first branchial arch during the early embryonic period at 7th week, and possible associated anomalies included cleft palate[5] and ankyloglossia, which were present in our case. Another theory proposed by Morton and Jordan[6] suggested that the median lower cleft lip is caused by the failure of mesodermal penetration into the midline of mandibular part of the first branchial arch, which could also explain the absence of hyoid, thyroid cartilage, strap muscles and manubrium in the severe varieties.

Median cleft lower lip can present with variable clinical severity ranging from a mild submucosal notch in the lower lip to the most severe form involving the lower lip, tongue, alveolus, mandible, floor of the mouth and even midline bony structures in the neck. In our case, the median cleft lip was so subtle that we had missed it for almost five years with our regular multidisciplinary clinical follow up. In fact, the median lower lip microform cleft only became apparent when the girl opened her mouth to speak.

With the low prevalence and wide spectrum of severity of median lower cleft lip, there is currently no consensus on the mode of management and the timing of surgical intervention. In mild cases, the median cleft lip could be surgically repaired with a direct V excision of the cleft with a Z plasty closure.[7] In the mildest spectrum of median cleft lower lip, such as in our case, when the defect could only become clinically apparent when the patient opened her mouth, the need for surgical intervention would largely depend on the patient's expectation.

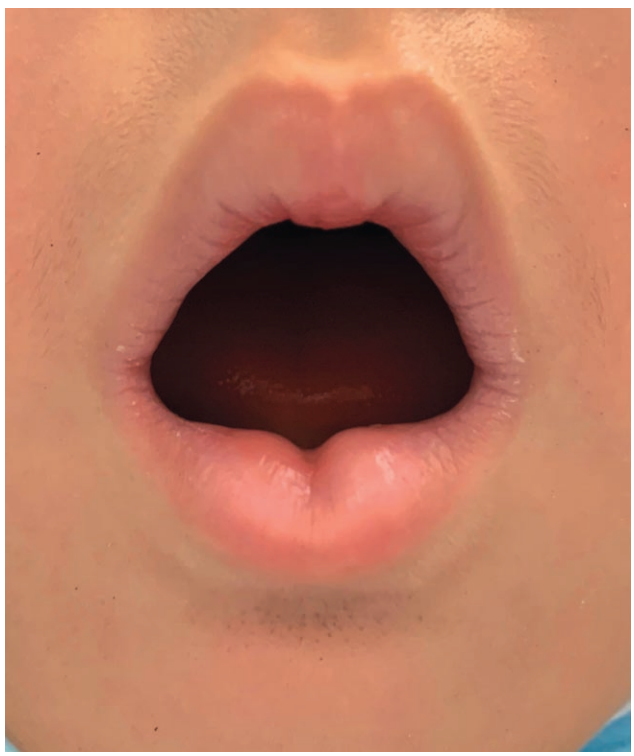

Figure 1 Median lower lip microform cleft lip.

And as per our centre's usual practice, release of tongue tie was usually only performed according to the National Institute for Health and Care Excellence guidelines[8] – when the tongue tie was impairing breastfeeding; therefore, we seldom perform "routine" tongue tie release in non-verbal patients as frequently, this anatomical anomaly could remain asymptomatic without surgical intervention. However, in this case, as the tongue tie would potentially be one of the contributing factors for her speech impairment, surgical release of tongue tie was offered after comprehensive speech assessment.

Interestingly, there seems to be degree of genetic inheritance in the median cleft lip in our patient as her paternal grandmother was said to have the similar clinical presentation. Unfortunately, we were unable to carry out genetic testing due to resource limitation. Apart from chromosomal aberrations,[9] other possible associated anomalies such as congenital heart deformities,[10] Pierre Robin anomaly,[11] hand anomalies[12] and foot deformities[13] have been reported in the literatures.

There was currently no literature describing any association between buttock haemangioma and median lower lip cleft. In our case, the buttock haemangioma spontaneously resolved with time without medical treatment. Given this natural disease course, it was likely to be the common infantile haemangioma and thus, it could very well just be an incidental finding of a common clinical condition (infantile haemangioma) in a patient with another uncommon condition (median lower cleft lip with cleft palate).

Conclusion

We described the clinical findings and management of a non-syndromic Chinese girl with central cleft palate, buttock haemangioma and a delayed presentation of median cleft lower lip and ankyloglossia. The importance of longitudinal clinical follow up of these patients was highlighted, and thorough physical examination and detailed clinical & family history should always be performed.

Conflict of Interest

The authors declare they have no competing interest.

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