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Case Report

Atypical Mouth/Lip Asymmetry Affecting a Newborn with Bilateral Cleft Lip and Palate: A Case Report

PMY ***Tang***, EKW ***Chan***, NSY ***Chao***, KH ***Lee***

Abstract ***Introduction:*** Congenital hypoplasia of depressor angularis oris muscle (CHDAOM) is a rare cause of asymmetric crying facies in newborn. The association of CHDAOM with bilateral cleft lip and palate has not been reported. ***Aims:*** We report a case of a neonate with bilateral cleft lip and palate and CHDAOM. ***Methods:*** A full term baby girl with bilateral cleft lip and palate was transferred to our hospital for centralised care of cleft anomalies. She was born by spontaneous vaginal delivery with a birth weight of 2.4 kg and no history of birth trauma. Clinical examination showed complete bilateral cleft lip and palate. Upon further evaluation, while her facial features remained symmetrical at rest, there was drooping (downward and lateral movement) of the left sided lower corner of the mouth on crying, with preserved upper face symmetry. Magnetic resonance imaging brain confirmed normal brainstem structure. Genetic test did not reveal any microdeletion of the chromosome 22q11. Echocardiogram showed no significant cardiac structural anomalies. As the baby was unable to tolerate oral feeding with significant choking and recurrent aspiration, endoscopy was performed and it showed right partial vocal cord paralysis. Ultrasound of the lips showed thinner and smaller right depressor angularis oris muscle, confirming the diagnosis of CHDAOM on the right. ***Results:*** As the baby remained tube - feeding dependent, elective laparoscopic gastrostomy and surgical repair of the bilateral cleft lip were performed at 4 months of age. Post-op recovery was uneventful. ***Conclusions:*** While surgeons might be well familiar with the pre-operative management of baby with cleft lip anomalies, it is important to observe the baby for other unusual facial anomalies which in this case only become apparent at crying while manifesting a contralateral pathology. Timely documentation, appropriate investigations and anomalies screening, counselling on expectations on surgical outcomes are important when managing patient with cleft lip/palate anomalies and CHDAOM.

Key words *Cleft lip; Depressor angularis oris*

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Introduction

Bilateral cleft lip anomalies are often diagnosed antenatally nowadays, with the increasing use of routine antenatal screening ultrasound, giving parents the opportunities to learn more about the condition. However, a dynamic facial disorder, such as the asymmetric crying facies would be almost impossible to be detected antenatally. There is currently scanty literature describing patients suffering from these two conditions

simultaneously. We report a full term newborn girl who is suffering from bilateral cleft lip and palate and congenital hypoplasia of the depressor angularis oris muscle (CHDAOM).

Case Report

A female neonate was born by spontaneous vaginal delivery at term. There was known antenatal diagnosis of bilateral cleft lip and unremarkable family history. The delivery was smooth with no apparent birth trauma. The birth weight was 2.4 kg. The baby was then transferred to our hospital for the centralised care of babies with cleft anomalies. At physical examination, apart from bilateral cleft lip and palate, there was no other obvious facial dysmorphism. Echo-cardiogram did not reveal any significant structural anomaly. Pre-op lip taping was applied and oral motor training was commenced, as per our hospital protocol for all patients with cleft anomalies. However, upon swallowing assessment by the speech therapist, it was found that the baby had very poor swallowing effort. Desaturation was often observed during oral feeding and there were also multiple episodes of aspiration pneumonia. In view of suspected anatomical anomalies that might be impairing her swallowing, examination under anaesthesia was performed by the Ear, Nose & Throat surgeon, showing partial paralysis of the right vocal cord. Clinical examination showed no cranial nerve palsy. With time, we started to notice an increasing obvious asymmetry of the lower face when the baby was crying, with the left sided corner of the mouth moving downwards and laterally when the right sided corner of the mouth remained unmoved (Figure 1). The upper face musculature including the frontalis, orbicularis, oculi and the zygomaticus remained symmetrical at rest and when crying. Clinical evaluation of the symmetry of the nasaolabial fold was difficult given the baby's bilateral cleft lip. Magnetic resonance imaging brain was performed to exclude neurological cause for the asymmetric facies and it confirmed no structural anomaly in the brain and along the course of the facial nerve. Ultrasound of the lips showed thinner and smaller right depressor angularis oris muscle, confirming the clinical diagnosis of CHDAOM on the right (Figure 2). The diagnosis of congenital hypoplasia of depressor angularis oris muscle was made, the natural course of the disease and management options were explained to the parents. With the mother's consent, genetic testing was performed and it did not reveal any micro-deletion of the chromosome 22q11. Elective laparoscopic gastrostomy and repair of the bilateral cleft lip was performed at 4 months of age and the post op recovery was uneventful. The drooping of the left sided corner of the mouth when crying remained similar after the cleft lip repair (Figure 3).

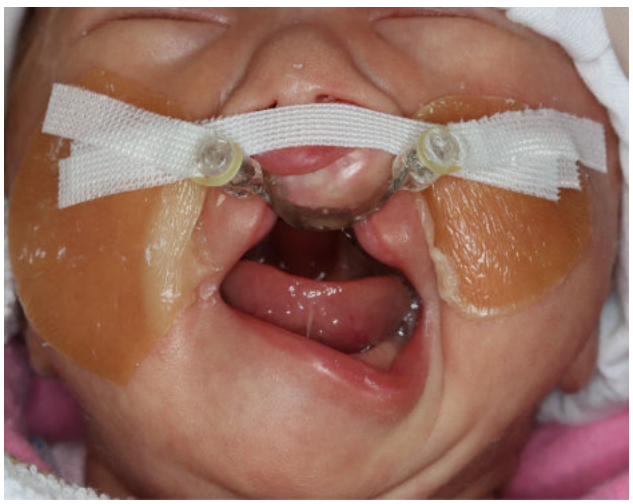

Figure 1 Pre-operative photo showing bilateral cleft lip and left sided dropping of the mouth when crying, with the pre-op lip taping and oral gastric feeding tube.

Discussion

CHDAOM had been reported to have an incidence of 6 per 1000 live birth and it was often confused with facial nerve palsy.[1] It attaches to the skin and the mucous membrane of the lower lip, drawing the lower corner of mouth downward and everts the lower lip.[2] Hence, when the baby is crying, the angle of mouth would be pulled down on normal side due to unopposed action of depressor angularis oris muscle while no movement on the hypoplastic side. The lower lip on the affected side would usually appear thinner and smaller due to lack of eversion and muscle agenesis. According to the literature, although the cause of hypoplastic development of the depressor angularis oris muscles remained idiopathic, intra-uterine subclinical viral infection and familial heredity have been speculated to have some causative influence.[3] CHDAOM is often characterised by the isolated involvement of the corner of the mouth, sparing the upper face, resulting in symmetrical forehead wrinkling, eyes closure and nasolabial fold depth. Although CHDAOM was usually a clinical diagnosis, such diagnosis in a patient with underlying bilateral cleft anomaly would be difficult, especially with early pre-op surgical adjuncts such as lip

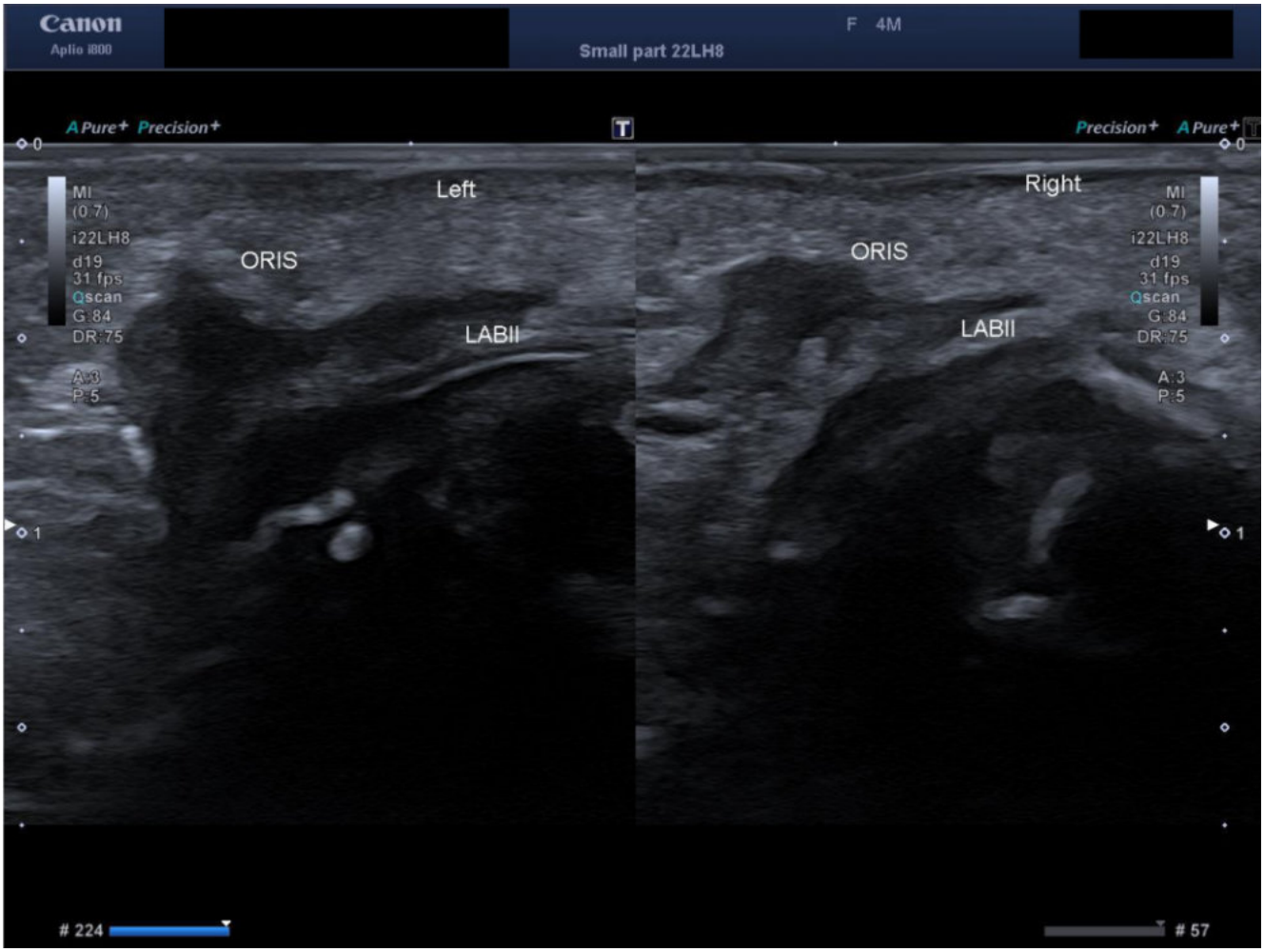

Figure 2 Ultrasound finding of thinner and smaller right depressor angularis oris muscle (2.4 mm) compared to the normal left depressor angularis oris muscle (3.2 mm).

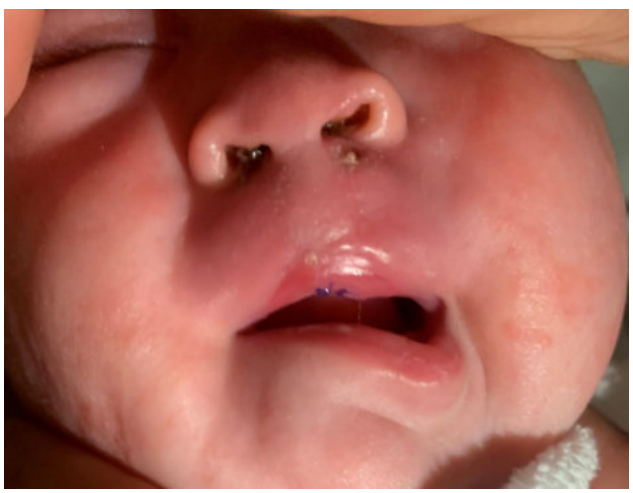

Figure 3 Post-operative photo showing persistent left sided drooping of the mouth when crying.

taping or naso-alveolar molding, which might physically interfere with the interpretation of facial dynamic movement. In addition, in our case, as the patient was dependent on oral-gastric tube feeding, the presence of the feeding tube would further complicate the clinical picture. The partial unilateral vocal cord palsy could potentially explain the choking and aspiration, and since the baby has improved oral feeding clinically, no follow up endoscopic assessment was arranged. While facial nerve pathologies such as nerve compression in utero, trauma to the facial nerve during delivery, or congenital facial nerve palsy may give rise to similar clinical presentation, the facial asymmetry in these cases would usually be present both at rest and while crying. Electro-diagnostic testing of the facial nerve has been advocated in some centers to rule out facial nerve pathology,[4] however, it was not done in our

case due to limited resource. Instead, ultrasound was utilised to demonstrate the thinning of the muscle at the affected side, confirming the diagnosis of CHDAOM.[2] Fortunately, we were able to make the diagnosis of CHDAOM prior to the surgical repair of the bilateral cleft lip, and timely appropriate counseling could be given. CHDAOM is a benign condition and it does not affect speaking or eating in the long term follow up, while botulinium-A toxin injection to the unaffected side of the depressor angularis oris muscles to obtain temporary facial symmetry had been described in the literature, the parents opted for conservative management for the time being. The prognosis of the return of muscle function has not been well documented, some mentioned that the dynamic facial asymmetry would become less noticeable after the child becomes older as the adjacent muscles may develop some compensatory movement. Intervention is usually considered when there is presence of social inhibition or psychological difficulties. While CHDAOM usually occurs as an isolated clinical finding, 45%-70% of cases could occur with other congenital malformations. Thorough physical examination and comprehensive newborn screening should be performed to rule out potential associated syndromes and to facilitate prompt treatment of the more serious co-existing abnormalities.[3]

Declaration of Interest

There are no conflicts of interests.

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Nil.

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