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CASE REPORTS

CONGENITAL ASYMMETRIC CRYING FACE SYNDROME: A CASE OF TWO NEWBORNS

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ABSTRACT

In the head examination of the newborn babies, palpebral fissures, symmetry of the movements of the eyelids, nose, mouth and auricles should be evaluated. Asymmetric weeping face (ACF) is a congenital disorder resulting from unilateral hypoplasia or agenesis of the depressor anguli oris muscle or, rarely, the depressor of the rim and depressor of the labii inferioris. Although isolated asymmetric weeping face (ACF) is considered a cosmetic defect, it can also accompany a wide range of developmental disorders, including congenital facial nerve palsy. Each case with asymmetric weeping face should be evaluated morphologically and anatomically in detail and molecular examination should be performed if necessary.

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KEYWORDS

Newborn, facial asymmetry, depressor anguli oris muscle.

Introduction

Congenital asymmetric crying face syndrome (ACF) is characterised by facial asymmetry in the corner of the mouth when the baby cries. Asymmetric crying face usually occurs as a result of unilateral agenesis or hypoplasia of the congenital depressor anguli oris muscle. Symptoms are present from birth. It may be an isolated anomaly and sometimes associated with other congenital anomalies. In the literature, association with cardiovascular anomalies and chromosomal disorders has been reported most frequently. It has also been reported that this condition is associated with neuroblastoma, mediastinal teratoma and neurofibromatosis type 1.[1,2] In this case report, two newborns with congenital facial asymmetry due to hypoplasia of the musculus depressor labii inferioris are presented because of its rarity.

Case Report

Case Report 1

There was no history of traumatic birth in the history of a female baby who was born from the fifth pregnancy of a 34-year-old mother with a weight of 3240 grams at 39 weeks of gestation by normal spontaneous vaginal delivery. In the family history, it was learned that there was no consanguinity between the parents and there was no other individual with congenital anomalies in the family. On physical examination, the patient, who appeared normal in neutral condition and had a prominent bilateral nasolabial sulcus, developed asymmetry in the left lower lip, especially during crying. However, it was determined that the forehead wrinkling, eye closure and nasal wing movement functions were normal (Figure 1). With these findings, isolated hypoplasia of one of the subgroup mimic muscles, possibly the musculus depressor labii inferioris muscle, was considered. Karyotype analysis from peripheral blood was found to be compatible with 46,XX. Echocardiography was normal in terms of additional anomalies that may accompany and no pathology was detected in cranial and abdominal ultrasonography.

Figure 1. The patient does not appear to have left commissure depression during crying.

Case Report 2

A 26-year-old patient who was born 2950 g by caesarean section from a 26-year-old mother was admitted to our unit on the second postnatal day with the complaint of maternal jaundice. It was learnt that there was no problem during delivery and the delivery was easy. Prenatal follow-up was

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normal. On physical examination, the corner of the mouth was drawn to the right side when she cried (Figure 2). Cranial and abdominal ultrasonography and echocardiography were planned for additional anomalies and reported as normal. The karyotype analysis sent from the patient was reported as 46,XY. Unilateral hypoplasia of the depressor anguli oris muscle was considered with physical examination findings.

Figure 2. Asymmetric facial appearance of the patient.

Discussion

Congenital asymmetric crying face is a minor congenital anomaly seen in 0.5-1% of newborns. When the child cries or laughs, asymmetry is noticed in the corner of the mouth on one side of the face. The left side is affected in 80% of cases. The appearance of the face is symmetrical at rest and one side of the mouth is pulled down when the child cries. It is 2 times more common in boys than in girls. It occurs as a result of agenesis or hypoplasia of the depressor anguli oris muscle.[3] The left side was affected in both of our cases. Almost all system findings such as central nervous system, cardiac system, gastrointestinal and genitourinary systems, musculoskeletal system, cervicofacial region, respiratory system, skin and soft tissues can be seen in ACF. Cervicofacial region and cardiovascular system anomalies are frequently detected. Mitral stenosis, aortic regurgitation, pulmonary stenosis, persistent left superior vena cava, tricuspid regurgitation, total anomalous pulmonary venous return, transposition of great arteries, hypoplastic left heart syndrome, tricuspid atresia, right aortic arrhythmia, cardiomyopathy are the most common cardiac pathologies. In addition, ACF may be observed alone or as a part of another syndrome such as VACTERL syndrome and Digeorge syndrome.[4,5] Both of our two patients had no additional anomalies both on physical examination findings and echocardiography on cardiac screening. The disease presents in the early neonatal period and is most commonly confused with peripheral facial nerve palsy due to birth trauma. The mandibular branch of the facial nerve is a superficial nerve that can be easily damaged by pressure on the face before or during labour. The most important risk factor for this type of injury is prolonged, difficult labour and use of forceps. Other risk factors include fetal macrosomia, multiple pregnancy, first pregnancy, head-pelvis incompatibility and uterine anomalies. In traumatic nerve damage, all lower lip muscles are paralysed. However, since only the depressor anguli oris muscle is involved in congenital ACF, other facial mimic muscles remain intact. Therefore, while the symmetrical structure of the face is intact at rest, the downward and outward movement of the corner of the mouth is restricted during crying. Unlike facial paralysis, movements such as frowning, eye closure, forehead wrinkling are normal and nasolabial sulcus depths are normal in both directions. There is no problem with tearing and sucking. Saliva flow is not observed from the affected side.[6] The clinical findings of our case were compatible with ACF and facial paralysis was ruled out. The facial muscles can be detected by both computed tomography (CT) and magnetic resonance imaging, but these imaging modalities can be difficult to perform. CT also carries an additional radiation risk for neonates. Therefore, Roedel et al. recommended ultrasonography to confirm the absence of facial muscles.[7] However, the differential diagnosis is still based on anamnesis and physical examination. In our case, a diagnosis of congenital asymmetric crying face was considered based on clinical findings. Muscle ultrasonography and electromyelography may be used to support the diagnosis. In this report, we present two isolated cases with congenital asymmetric crying face without a history of trauma at birth and no accompanying anomaly was found on investigations. Although congenital asymmetric crying face is a minor anomaly, it should be kept in mind that it may be associated with major congenital anomalies and other anomalies should be investigated in case of this finding. It should be known that a multidisciplinary approach is required in terms of complications and appropriate treatment follow-up

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