Case Report

Congenital asymmetric crying facies: A case report with review of the literature

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ABSTRACT

Asymmetric crying facies (ACF) in a newborn is a condition which occurs in one in 160 live births, with an estimated prevalence in 0.2–0.6% of infants and the left-sided predominance was determined in 80% of neonatal ACF cases. When ACF is associated with other anomalies, it is known as ACF syndrome. Congenital ACF in a newborn is a rare condition caused by unilateral hypoplasia or agenesis of the depressor anguli oris muscle resulting in an ACF in the neonatal period. ACF is often confused with facial nerve compression of the fetus in the uterus or facial nerve palsy secondary to trauma. The present review is having a baseline single case study of ACF due to the left-sided congenital hypoplasia of depressor angularis oris muscle with no other anomalies. The differential diagnosis and complete physical examination are essential to rule out other associated anomalies in ACF cases.

Key words: Asymmetric crying facies, Depressor anguli oris, Neonatal asymmetric crying facies

Asymmetric crying facies (ACF) in a newborn is a condition which occurs in one in 160 live births, with an estimated prevalence in 0.2–0.6% of infants [1] and the left-sided predominance was determined in 80% of neonatal ACF cases [2,3]. This can be an isolated clinical finding or be coupled with other congenital malformations. ACF is associated with other birth defects in approximately 45–70% of cases [4], most commonly seen in the cardiovascular system. ACF having an association with a heart defect is known as Cayler syndrome. When ACF is associated with other anomalies, it is known as ACF syndrome. Congenital ACF in a newborn is a rare condition caused by unilateral hypoplasia or agenesis of the depressor anguli oris muscle resulting in an ACF in the neonatal period. Symmetric face at rest and asymmetric face observed during crying, drooping of one corner of the mouth during crying, while eye closure, nasolabial fold, and forehead wrinkling is symmetric.

CASE REPORT

A male neonate was born by lower abdominal cesarean section delivery at the hospital to a 20-year-old primigravida mother with full-term pregnancy, breech presentation, and uneventful antenatal and perinatal period. The birth weight was 2.4 kg. The Apgar scores taken at 1 and 5 min are 9 and 10, respectively.

On examination, the baby had asymmetric frowning, complete eye closure, and normal extraocular movements. The wrinkling, sucking reflex, and eye closure were normal and symmetrical at the time of physical evaluation. The nostrils position and dilation during breathing and cry are same. No facial asymmetry was observed when the baby was quiet and resting (Fig. 1a). However, on crying, the right corner of the mouth deviated downward and outward while the left corner did not show deviation (Fig. 1b).

The ultrasound (USG) and chest radiography reports of the newborn were normal. No obvious anomaly was noted. The newborn was diagnosed as ACF due to the left-sided congenital hypoplasia of depressor angularis oris muscle with no other anomalies. The diagnosis in this case is made on the clinical basis of a thorough physical examination and also looked for the symmetry of the jaw and asynclitism of the gums to rule out the nerve compression causing facial weakness.

Both the parents were absolutely normal and healthy carrying no viral/bacterial infections and no consanguinity. The mother was a registered patient of antenatal care and underwent all the necessary examinations during the antenatal care period which were normal. There was no family history of congenital defects in either of the sides. Since in this case, USG and other tests were done, parents were advised to undergo electrodiagnostic testing and a neurological opinion. Counseling of the parents was done regarding the congenital malformation of depressor anguli oris muscle and its causative factors. Parents were explained that a spontaneous recovery may possible as the child grow further.

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DISCUSSION

Congenital ACF which is also known as congenital unilateral lower lip palsy is an uncommon condition caused by congenital hypoplasia or agenesis of the depressor anguli oris muscle on one side of the mouth. The facial weakness in a newborn can be caused by two reasons as birth trauma and developmental problems during the gestational period. Birth trauma leads to compression of branches of the facial nerve. While in congenital ACF, the depressor anguli oris muscle which controls the lower lip movement may undergo incomplete development during pregnancy which is called as hypoplasia or the absence of muscle called as agenesis. Hence, a thorough physical evaluation of a child with proper birth and gestational history is very important in such cases.

Although the functional deficit is small, the anomaly may be associated with cardiovascular, head and neck, musculoskeletal, respiratory, gastrointestinal, central nervous system, or genitourinary anomalies in 45% of cases [5]. ACF in a newborn is characterized by facial asymmetry only when a newborn is crying wherein one angle of the mouth deviates to one side which is the unaffected side and downward while the other side does not move; however, the face is symmetrical at rest. The etiology of ACF is multifactorial and can be due to faulty muscle or nerve development [6]. Intrauterine viral infection, chromosomal aberrations, hereditary factors, or a defect located at the brain stem level can cause a defect in the depressor anguli oris muscle development [2], one of the muscles that control the movements of the lower lip on one side of the mouth. Depressor anguli oris muscle origins at the mandible and insertion at the angle of the mouth. Depressor anguli oris muscle depresses the corner of the mouth.

In ACF many times, the management depends on the cause and assessment of the case. If the cause is traumatic, then no further evaluation is needed, but in case of defective muscle development, further evaluation is needed to rule out the presence of cardiac abnormalities. Fluorescence *in situ* hybridization test can be done to look for chromosomal abnormalities like the deletion of a small piece of chromosome 22q11 which is reported in Cayler syndrome in which heart defects can be seen [7].

The asymmetry of the jaw and non-parallel gums may offer a clue for nerve compression causing the facial weakness where the chances of other anomalies are less. ACF is often confused with facial nerve compression of the fetus in the uterus or facial nerve palsy secondary to trauma. Congenital facial nerve palsy is manifested with facial asymmetry both at rest while crying without other malformations. Ultrasonography and electrodiagnostic testing could help to confirm the condition. Gupta and Prasad found that the use of USG to observe facial muscles could be helpful for differential diagnosis [8].The diagnostic criteria for ACF are shown in Table 1 [9]. A prospective study on congenital ACF and associated congenital anomalies concluded that the unilateral hypoplasia of a depressor anguli oris muscle is generally an isolated finding and very rarely associated with the other birth defect [10].

The surgical management includes plastic reconstructive procedures on the lower lip by the affected side, plication of the orbicularis oris muscle, transposition of the orbicularis muscle, cheiloplasty or wedge excision, fascia lata sling, and digastric muscle transfer. A procedure like selective neurectomy of the marginal mandibular branch of the opposite side can be done for the weakening of the muscle of the lower lip on the non-affected side [11]. Two cases of ACF was reported to be successfully treated with botulinum toxin type A (BTX-A) injection. Treatment with BTX-A gives a temporary correction up to 6 months. BTX-A is known to be a presynaptic neuromuscular

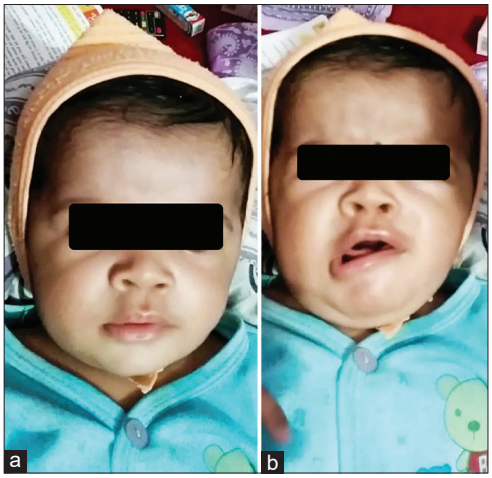

Figure 1: Asymmetric crying facies during (a) sleep/resting position; (b) crying

Table 1: Diagnostic criteria for ACF [9]

Due to nerve compression	Due to depressor anguli oris muscle agenesis or hypoplasia
A thorough medical history should obtain information about: • Multiple births • Difficult labor and delivery • Primiparity • Large baby • Uterine tumors • Forceps delivery • Polyhydramnios (possible) • Mandibular asymmetry and maxillary mandibular asynclitism reveals on physical examination • Abnormal values for excitability and conduction of the mandibular branch of the facial nerve seen on electromyography.	• One-side descending development of the edge of the mouth while the contrary side does not move during crying, however, at rest the face looks symmetric • Palpable thinning of the lateral segment of the lower lip on the affected side • Normal and symmetric forehead wrinkling, closure of eyelid, nasolabial fold depth, frowning, tearing, and nostril dilatation with respiration • Normal conduction time and nerve sensitivity study results • Ultrasonography exhibits of hypoplasia or agenesis of depressor anguli oris muscle

ACF: Asymmetric crying facies

blocking agent that produces selective muscle weakness when injected intramuscularly in minimal dose. Such treatment options may provide time for surgical interventions to be planned [12]. In conclusion, meticulous physical examination of the newborn and genetic testing is important for the early diagnosis of ACF. USG and electrodiagnostic testing could be helpful for the differential diagnosis of ACF from congenital facial nerve dysplasia. In instances, where asymmetry of the jaws and maxillary-mandibular asynclitism of the gums is there, this gives a clue for nerve compression and other anomalies may not be found.

CONCLUSION

After accurate diagnosis of ACF, it is important to provide information about the disease to parents regarding prognosis and future associated risk. If associated anomalies like a defect in the heart or kidneys found, then the child should be referred to a concerned specialist for the management.

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