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ASYMMETRIC CRYING FACIES: A CLINICAL ODDITY

Neonatology

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ABSTRACT

Background – Congenital hypoplasia of depressor angularis oris muscle (CHDAOM), which is often confused with unilateral facial nerve palsy, is a rare congenital disorder presenting with asymmetric crying facies in a newborn. Accurate diagnosis of this condition is imperative in order to ensure further screening to rule out associated anomalies and appropriate management. **Clinical description** – We report a late preterm male neonate who was noticed to have facial asymmetry selectively during episodes of crying. During quiet or sleeping state, the face was symmetrical. Clinical examination did not reveal any other facial abnormality or any evidence of facial nerve palsy. **Management** – Associated anomalies were ruled out by thorough evaluation. In view of isolated CHDAOM, parents were explained regarding benign course of the condition. The neonate was discharged after an uneventful hospital stay. **Conclusion** – This case report emphasizes the importance of a detailed clinical examination in the diagnosis of CHDOAM. CHDOAM, if occurring in isolation, is a benign condition and does not require any intervention. Parental counselling is the cornerstone of management. However, the diagnosis of CHDAOM should raise suspicion for other associated congenital anomalies and warrants a thorough evaluation.

KEYWORDS

Congenital hypoplasia of depressor angularis oris muscle, Asymmetric crying facies, Facial nerve palsy

A neonate with asymmetric crying facies, an uncommon condition, can present as a diagnostic challenge. Congenital hypoplasia of depressor angularis oris muscle (CHDAOM), which is often confused with unilateral facial nerve palsy, is a rare congenital disorder presenting with asymmetric crying facies in a newborn. However, just by keen observation, facial nerve palsy can be ruled out. Accurate diagnosis of this condition is imperative in order to ensure further screening to rule out associated anomalies and appropriate management. This article emphasizes the importance of clinical observation while approaching such cases.

Clinical description

A male neonate was born via Cesarean section to a 29-year old G2P1L0 mother at 36^{+4}-week gestation with a birthweight of 2.5 kg. Baby cried soon after birth and had normal Apgar scores. Antenatal period and family history were unremarkable, and there was no history of birth trauma or instrumentation. As soon as baby started crying, facial asymmetry in form of downward and outward deviation of left corner of mouth was noticed (Figure 1, Video 1). However, during quiet or sleeping state, the face was symmetrical (Figure 1). This asymmetry reappeared on crying. Breastfeeding was commenced soon after birth, and no difficulty was observed in sucking. On further examination while crying, nasolabial folds, eye closure, and forehead wrinkling were noted to be symmetrical (Figure 1, Video 1). However, thinning of right lower lip was noted. No other facial abnormalities were detected. His vital parameters and systemic examination findings were within normal limits. There were no neurological deficits.

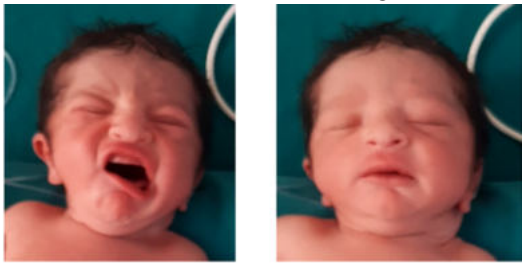

Figure 1 Asymmetric crying facies

Management and Outcome

A diagnosis of CHDAOM was considered in index case. Associated anomalies were ruled out by echocardiography, x-ray spine and ultrasound cranium. In view of isolated finding, parents were explained regarding benign course of the condition. The neonate was discharged after an uneventful hospital stay.

DISCUSSION

The term "asymmetric crying facies" describes facial asymmetry which becomes evident only during crying state. Facial features are symmetrical during quiet or sleeping state. Possible etiologies include facial nerve compression due to birth trauma or abnormal fetal position, congenital absence of facial nerve nucleus, impingement of marginal mandibular branch of facial nerve (congenital facial paralysis), and CHDOAM.

CHDAOM is a rare entity with an incidence of 3-6 /1000 live births. It classically presents as asymmetric crying facies and closely mimics facial nerve palsy. [1] Majority of cases (approximately 80%) involve left side of the face. [2] It most commonly presents during neonatal period.

The function of DAOM is to evert the ipsilateral lower lip and depress the ipsilateral angle of mouth. Unilateral hypoplasia of DAOM leads to pulled down corner of mouth and mandible on the normal side during crying due to unopposed action of DAOM on unaffected side. Lack of eversion and muscle hypoplasia leads to apparent thinning of lower lip on the affected side. This asymmetrical appearance, which resembles lower facial drooping, may give an impression of facial nerve palsy. However, in contrast to facial nerve palsy, this condition is characterized by symmetrical nasolabial folds, eyelid closure, and forehead wrinkling indicating normal facial nerve functions.

The underlying etiology of CHDAOM is largely unknown. Diagnosis is mostly established by clinical examination with focus on key finding such as presence of symmetrical nasolabial folds, eyelid closure, and forehead wrinkling which differentiate it from facial nerve palsy. Nerve conduction studies may reveal normal facial nerve.[3]

The major clinical significance of CHDAOM lies in its association with multiple other congenital anomalies which necessitates thorough examination and screening, if clinically indicated. Coexisting congenital anomalies can occur in up to 45-70% of the cases. Reported anomalies most commonly involve head and neck region (45-50%), cardiovascular system (40-50%), genitourinary (24%), and musculoskeletal (22%) system. [2,4] Cardiac anomalies are the major reason behind mortality and morbidity. The involvement of genitourinary, central nervous system and gastrointestinal system is

less frequent. Syndromic associations include Cayler cardiofacial syndrome, Digeorge syndrome, velocardiofacial syndrome, CATCH 22, VACTERL association, and Trisomy 18. [5–7] An abnormal clinical examination warrants detailed assessment for these anomalies. CHDOAM, if occurring in isolation, is a benign condition and does not require any intervention. It is predominantly a cosmetic problem, and gradually becomes less noticeable with age. Feeding and speech mostly remain unaffected. Parental counselling is the cornerstone of management. However, the diagnosis of CHDAOM should raise suspicion for other associated congenital anomalies and warrants a thorough evaluation and comprehensive screening to rule out aforementioned additional anomalies.

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