Facial Asymmetry in a Newly Born Baby: Diagnostic Challenge!

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ABSTRACT

INTRODUCTION: Facial asymmetry during crying in neonates is an important entity which might be due to an underlying true paralysis of the facial nerve or due to the benign overlooked diagnosis of congenital absence of the depressor angularis oris muscle (DAOM).

CASE REPORT: We report a full-term newborn delivered by normal spontaneous vertex delivery with uneventful pregnancy and Apgar score of 9 and 10 at 1 and 5 minutes respectively. His parents are first-degree cousins with 4 living normal siblings. His birth weight was appropriate for his gestational age. His initial neonatal assessment was normal apart from facial asymmetry during crying in the form of deviation of the angle of the mouth to the left with an otherwise normal facial appearance. The facial asymmetry disappears at rest (without crying). This condition usually poses a diagnostic dilemma. Developmental and traumatic facial paralysis and being part of some syndromes like Mobius and CHARGE syndromes are among the most important differential diagnosis but the disappearance of the facial asymmetry at rest is diagnostic of absent DAOM which is a relatively common but missed diagnosis.

CONCLUSIONS: Pediatricians should be aware about the diagnosis of DAOM in neonates with asymmetric crying face if the face was completely normal at rest with deviation of the angle of the mouth on crying which disappears on rest.

KEYWORDS: Facial asymmetry, neonate, depressor angularis oris muscle

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Introduction

Neonatal asymmetric crying facies (NACF) is a relatively common often misdiagnosed problem. It is mainly caused by the absence of the depressor angularis oris muscle (DAOM) or less commonly the depressor labii inferioris muscle (DLIM), or the compression of the mandibular branch of the facial nerve which innervate those muscles.[1]

The normal appearance of the face apart from the deviation of the angle of the mouth on crying which disappears on rest along with confirmation of the absence or hypoplasia of the DAOM/or DLIM by B-scan ultrasound is diagnostic.[2]

We report a neonate with NACF due to right DAOM absence. The main objective of this report is to shed light on this important benign entity which is usually overlooked and poses a diagnostic dilemma with misdiagnosis with congenital, developmental, and traumatic true facial paralysis with the resultant unnecessary investigations and unjustifiable management. Physicians should have a high index of suspicion for the diagnosis of NACF.

Case Description

MA is a 40-week gestational age neonate who was born by normal spontaneous vertex delivery to gravida 5 para 4 mother with non-significant perinatal history and normal Apgar score of 9 and 10 at 1 and 5 minutes respectively. Parents were first-degree cousins with 4 living normal siblings.

His birth weight was appropriate for his gestational age; 3150 g. Full history taking and body system review revealed no abnormal findings apart from an evident facial asymmetry which is observed during crying in the form of deviation of the angle of the mouth to the left with an otherwise normal facial appearance (Figure 1; Supplemental Material Video 1). The nasolabial folds were normal, and when the infant cries, the forehead wrinkles and both eyes close normally (Figure 1; Supplemental Material Video 1). Facial asymmetry disappears at rest (without crying), suggestive of NACF (Figure 2; Supplemental Material Video 1). Physical examination for known association with major and minor congenital anomalies and deformations came out to be normal with no

Figure 1. The asymmetrical crying face during crying.

Figure 2. The asymmetrical crying face at rest.

dysmorphic features or obvious anomalies. Echocardiographic evaluation was normal.

The absence of the right DAOM was confirmed by B-scan ultrasound which along with the characteristic physical examination, confirmed the diagnosis of NACF.

The condition was explained to the family and the baby was discharged safely to home. His follow-up after 6 months, showed the same picture with the assurance of the parents and appointments for regular follow-up visits.

Discussion

NACF is a relatively common minor anomaly with an incidence of 1 per 160 live births.[3] DAOM hypoplasia or agenesis or less commonly the DLIM (the developmental theory) or injury of one of the peripheral branches of the facial nerve especially the mandibular branch which runs superficially over the mandible in neonates (the traumatic theory) are the underlying causes.[1]

The downward movement of the lower lip is the result of the action of 4 muscles, the DAOM pulls the corner of the mouth downwards, laterally, and everts it, the DLIM which extends from the mandible to the lower lip and depresses it, the mentalis muscle which raises and protrudes the lower lip and the platysma muscle which blends in with the DAOM and assists in its function.[2]

Most of the cases of NACF are caused by the aplasia/hypoplasia of the DAOM and less commonly the DLIM.[1] Another cause is the pressure during delivery on the mandibular branch of the facial nerve which innervate those muscles with the resultant downward deviation of the angle of the mouth to the contra-lateral healthy side due to the unbalanced effect of the healthy well-developed muscle.

Some reports showed that neonates with NACF have 3.5-folds higher risk of association with other major congenital anomalies like cardiac,[4-8] cervicofacial[3] (auricular, mandibular hypoplasia),[9,10] neurological (corpus callosum agenesis, brain cyst, hydrocephalus),[9] genitourinary (cryptorchidism, hypospadias, vesicoureteral reflux, hydronephrosis),[9,10] gastrointestinal (megacolon, imperforate anus, esophageal atresia, inguinal hernia),[9,10] skeletal (syn/poly/clinodactyly, cortical thumb, and hemivertebra),[9,10] and genetic syndromes (4P deletion, trisomies 21or 18, Klinefelter, VATER, or Griscelli).[11-13] Other reported associations with NACF include hemihypertrophy,[14] cystic lymphangioma,[15] collodion baby,[16] and pulmonary agenesis.[17]

Minor anomalies like strawberry hemangioma, anal/preauricular tags, pilonidal sinus, accessory nipple, and single horizontal palmar crease were also described.[9,10]

Ear and feet deformations like overfolded helix, cup ear, protruding earlobes, and pes valgus/or varus respectively were also described.[9]

If abnormal cardiovascular findings are detected, it is essential to do a further genetic evaluation to roll out chromosome 22q11 microdeletions which have a known association with NACF.[4,7,8]

The main importance of NACF lies in the diagnostic dilemma it poses with the common misdiagnosis as traumatic, congenital, or developmental true facial paralysis with the later might be isolated or part of syndromes like Mobius, CHARGE, Goldenhar, hemifacial macrosomia, and hereditary developmental facial paresis.[18]

Physicians should be aware of NACF syndrome and apply its suggested diagnostic criteria to avoid unnecessary workup and to provide the appropriate management plan.[2]

The diagnostic criteria for the NACF secondary to DAOM aplasia or hypoplasia include the following[2]: (1) one-side downward movement of the corner of the mouth while the opposite side does not move during crying, but at rest, the face appears symmetric, (2) palpable thinning of the lateral portion of the lower lip on the affected side (usually present), (3) normal and symmetric forehead wrinkling, closure of eyelid, nasolabial fold depth, frowning, tearing, and nostril dilatation with respiration, (4) normal conduction time and nerve excitability study results, (5) ultrasonographic demonstration of hypoplasia or agenesis of DAOM.

B-scan ultrasound is the recommended radiological assessment for DAOM.[19] Although computed tomography (CT) scan and magnetic resonance imaging (MRI) can be used, it is difficult to obtain accurate views with those modalities with the added risk of radiation on CT scan use.[1,2]

It is also important to mention the diagnostic criteria of NACF secondary to compression of the mandibular branch of the facial nerve during delivery which include the following: (1) suggestive perinatal history (large baby, multiple births, difficult labor, or forceps delivery), (2) suggestive physical examination of mandibular asymmetry and maxillary mandibular asynclitism, (3) electromyographic evidence of mandibular branch compression with abnormal excitability and conduction.[2]

Family counseling about the problem, its workup, outcome, and management is essential. Patients with NACF fulfilling the above mentioned diagnostic criteria and have no associated physical abnormalities need only observation with no further work up.[2]

Those with mandibular branch compression usually resolve spontaneously within few months while those with muscle agenesis or hypoplasia might be less noticeable with age when the functions of other facial muscles and smiling dominate the child's facial expressions[12] but if no appreciable improvement noticed, plastic surgery correction might be needed.[3]

Conclusions

NACF is a minor relatively common condition. Physicians' awareness about condition is crucial to avoid unnecessary investigations and management.

Author Contributions

NK, MO: Diagnosed the patient.
NK, MO, RA, NA: collected patient's data.
NK, MO, SA, LS, ME: reviewed literature, drafted the manuscript, critically analyzed the data.
All authors reviewed and approved the manuscript for final publication.

Ethical Approval

This study was approved by the research & ethical committee of Alhada Armed Forces Hospital, Taif, Saudi Arabia.

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Supplemental Material

Supplemental material for this article is available online.

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