Case Report

Pediatric Education and Research
Volume 4 Number 1, January - April 2016
DOI: https://dx.doi.org/10.21088/per.2321.1644.4116.7

Congenital Hypoplasia of Depressor Angularis Oris and Orbicularis Oculi Muscle

G.S. Aher*, Sunil Mhaske, Urmila Gavali***, Liza Bulsara******

*Professor, Dept of OBGY, **Professor & Head, ***Assistant Professor; ****Resident, Dept. Of Paediatrics, Padmashree Dr. Vithalrao Vikhe Patil Medical College & Hospital, Near Govt. Milk Dairy, Vilad Ghat, Ahmednagar – 414111.

Abstract

Congenital hypoplasia of depressor angularis oris muscle (CHDAOM) is one of the rare causes of asymmetric crying facies in newborn. Major congenital anomalies have been reported to be associated with this facial defect in 45-70% cases [1, 2]. We report a case of CHDAOM in a neonate.

Keywords: Congenital Anomaly; Facial Defect; Asymmetric Crying; Depressor Angularis Oris; Orbicularis Oculi.

Introduction

Congenital hypoplasia of depressor angularis oris muscle (CHDAOM) is one of the rare causes of asymmetric crying facies in newborn. Major congenital anomalies have been reported to be associated with this facial defect in 45-70% cases [1,2]. We report a case of CHDAOM in a neonate.

Case Report

A 21 years primigravida was admitted to our hospital with labour pains. On examination her vitals were stable. On P/A examination patient was having cephalic presentation and head was 4/5th palpable. She was having 1-2 uterine contractions per 10 min lasting for 10-15 seconds. On P/V examination cervix was 3cm dilated, presenting part was vertex with right occipito posterior position. All routine investigations including USG were normal. Patient was taken was for emergency LSCS for CPD. A female neonate of 2.5 kg was delivered. She had a vigorous cry. The face was asymmetrical with drooping of right eyelid while the neonate was quiet or sleeping, however on crying, the left corner of the mouth drew left and downward, while right corner did not move *(Photo 1).* Clinical evaluation revealed normal vital parameters. The frontalis, zygomaticus and mentis muscles functioned adequately. Extraocular movements were intact on nonaffected side. USG of facial nerve at birth & at 72 hours were normal and there was no history of birth trauma which excluded

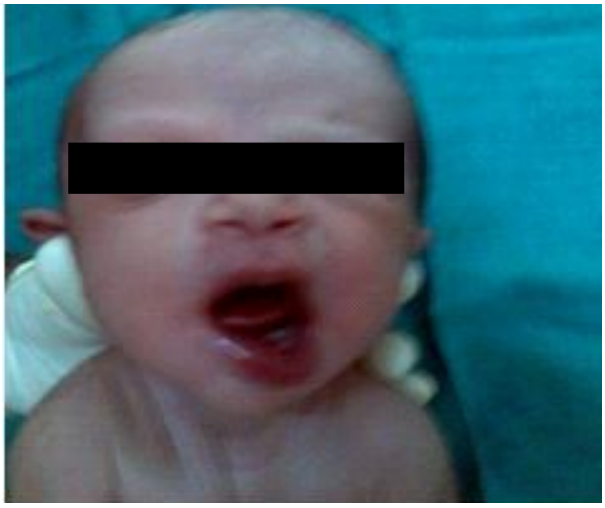

Photo 1: On crying, the left corner of the mouth drew left and downward, while right corner did not move

Corresponding Author: G.S. Aher, Professor & Head, Dept. of OBGY; Padmashree Dr. Vithalrao Vikhe Patil Medical College & Hospital, Near Govt. Milk Dairy, Vilad Ghat, Ahmednagar – 414111.
E-mail: drgsaher@gmail.com

facial nerve palsy as a cause of facial asymmetry.

The neonate was diagnosed as a case of asymmetric crying facies due to congenital hypoplasia of right depressor angularis oris and orbicularis oculi muscle. There was no neurological deficit and no other congenital anomalies associated with it. Patient was discharged on 8th post operative day.

Discussion

Congenital hypoplasia of depressor angularis oris muscle causes facial asymmetry, especially when infant cries [3]. The incidence of CHDAOM is approx 3-6 / 1000 live birth and is often confused with facial nerve palsy [1,2]. The depressor angularis oris muscle (DAOM) originates from the oblique line of the mandible and extends upward and medially to the orbicularis oris. It attaches to the skin and the mucous membrane of the lower lip. The depressor angularis oris muscle is innervated by two branches, buccal and mandibular branch. The DAOM draws the lower corner of the mouth downward and everts the lower lip. Hence on crying, angle of mouth and mandible are pulled down on normal side due to unopposed action of DAOM, while no movement on hypoplasia side. The lower lip on the affected side looks thinner because of the lack of eversion and muscle agenesis. The cause for agenesis of the muscle is not known. These patients have symmetrical forehead wrinkling, eye closure and nasolabial fold depth. The diagnosis may be confirmed by electrophysiological studies. The facial nerve conduction time to the mentalis and orbicularis oris muscle are normal [4]. It is usually associated with cardiac, gastro-intestinal, genito-urinary anomalies and other malformations [1,2,5]. The common anomalies seen are congenital heart disease (44%), head and neck (48%), skeletal (22%) and genitourinary tract anomalies (24%) [1]. CATCH 22 is a medical acronym for cardiac defects, abnormal facies, thymic hypoplasia, cleft palate, hypocalcemia and avariable deletion on chromosome 22q11. The deletion within chromosome region of 22q11 may occur inpatients with dysmorphologic and cardiological syndromes; DiGeorge syndrome, velo-cardiofacial syndrome and conotruncal anomaly face syndrome[6-8]. This condition should be differentiated from other causes of facial asymmetry at birth like intra-uterine position and pressure over stylomastoid foramen during labor, which may cause facial paralysis. This is a benign condition and mainly a cosmetic problem. It does not interfere with feeding or speech. The best time for diagnosis is careful physical examination of newborn and if present, neonate should be screened for associated anomalies.

Conclusion

CHDAOM is a rare cause of asymmetric crying facies which needs to be differentiated from facial nerve palsy. The importance of recognizing CHDAOM lies in the fact that there is strong association of this anomaly with other significant anomalies. In an isolated anomaly, no treatment is required because the asymmetry is not noticeable in a grown up child.

References

1. Lin DS, Huang FY, Lin SP, et al. Frequency of associated anomalies in congenital hypoplasia of depressor angularisoris muscle: a study of 50 patients. Am J Med Genet. 1997; 71: 215-8.
2. Caksen H, Odabas D, Tuncer O, et al. A review of 35 cases of asymmetric crying facies. Genet Couns. 2004; 15: 159-65.
3. Caksen H. Asymmetric crying facies. Indian Pediatrics. 2000; 37: 1385.
4. Martin GMA., Arguelles F, Roche HMC, Omenaca F, Lopez TJM, Ortigado A. Facial asymmetry with crying: neurophysiological study and clinical account of this entity. An EspPediatr. 1998; 49: 436-7.
5. Narang M, Goyal JP. Uncommon manifestations of Klippel Feil Syndrome. Indian Pediatrics. 2006; 43:265-6.
6. Akcarus M, Gunes T, Kurtoglu S, et al. Asymmetric crying facies associated with congenital hypoparathyroidism and 22q11 deletion. Turk J Pediatr. 2004; 46; 191-3.
7. Akcakus M, Ozkul Y, Gunes T, et al. Associated anomalies in asymmetric crying facies and 22q11 deletion. Genet Couns. 2003; 14: 325-30.
8. Lahat E, Heyman E, Barkav A, Goldberg M. Asymmetric crying facies and associated congenital anomalies: prospective study and review of literature. J Child Neurol. 2001; 16: 778.