Congenital Hypoplasia of Depressor Angularis Oris Muscle

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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 male neonate was born by spontaneous vaginal delivery to a 22 years old primigravida mother at term with uneventful antenatal and perinatal period. Birth weight was 2.4 kg. There was no history of birth trauma. He had a vigorous cry and was closing his eyes satisfactorily. The face was symmetrical while the neonate was quiet or sleeping, however on crying, the right corner of the mouth drew right and downward, while left corner did not move (Fig. 1). Clinical evaluation revealed normal vital parameters. The frontalis, orbicularis oculi, zygomaticus and mentis muscles functioned adequately. Extra ocular movements were intact. There was a palpable thinning of the left lower lip near its left margin. Further evaluation revealed a deformed left pinna and apex beat on right side in the 5th intercostal space. Dextrocardia was confirmed by chest radiograph and electrocardiography. Echocardiography did not reveal any structural anomaly except dextrocardia. The neonate was diagnosed as a case of asymmetric crying facies due to congenital hypoplasia of left depressor angularis oris muscle and dextrocardia. There was no neurological deficit. Systemic examination was essentially within normal limits.

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

Fig. 1 : Photograph showing symmetrical face during quiet awake stage (above) and while crying. The right angle of mouth was pulled down to right side and downward due to hypoplasia of depressor angularis oris muscle on left side (below)

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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]. Our case was associated with cardiac anomalies in the form of dextrocardia. 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 a variable deletion on chromosome 22q11. The deletion within chromosome region of 22q11 may occur in patients with dysmorphic and cardiologic 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. 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.

Conflicts of Interest

None identified

References

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