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Congenital Hypoplasia of Depressor Angularis Oris Muscle With Atrial Septal Defect: A Rare Case Report

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Abstract
An asymmetric crying facies of newborn is usually of a major concern to parents and treating pediatrician. Hypoplasia of depressor angularis oris musle is one of the common causes of such an asymmetric crying facies. It is moreover a cause of concern because in many cases there is increased incidence of other congenital anomalies especially cardiovascular malformations. A close differential diagnosis of this condition includes partial peripheral facial nerve palsy. Congenital hypoplasia of depressor angularis oris is a condition which is not expected to improve on its own and hence will invariably require surgical correction for cosmetic purposes. Its presence in any newborn make it necessary that treating physician look for any congenital heart disease, head and neck anomalies and genitourinary anomalies. Here we report a case of congenital hypoplasia of depressor angularis oris muscle with Ostium secondum type of atria septal defect.

KeyWords : Depressor angularis Oris, Assymetric crying facies, Atrial Septal Defect

Introduction

An asymmetric crying facies in a newborn is a major cause of concern for treating pediatrican because of its association with other congenital anomalies like cardiovascular, genitourinary, musculoskeletal and head neck face anomalies. Asymmetric crying facies is also associated with CATCH 22 (cardiac defect, abnormal facies, thymic hypoplasia, cleft palate, and hypocalcemia) and VACTERL (vertebral

anomalies, anal atresia, cardiac malformations, tracheoesophageal fistula, renal anomalies, limb abnormalities) anomalies. Congenital hypoplasia of depressor angularis oris muscle is usually noticed in post natal period when baby cries. During crying there is deviation of angle of mouth to opposite side. Though this is subtle manifestation but its identification is critical as in these patients treating pediatrician must rule out associated anomalies.

Case Report:

A full term male child delivered by normal vaginal delivery was admitted in NICU in view of neonatal hyperbillirubinemia on Day-5 of life. On admission the baby was active with no signs of encephalopathy. Complete blood count was done which was within normal limits. Serum billirubin estimation was done. Total billirubin was 24.6 mg/dl. Indirect billirubin was 22.2 while direct billirubin was 2.4. There was no h/o Rh incompitability. Since baby was active and there were no signs of encephalopathy or infection phototherapy was started and breast feeding was continued. A repeat billirubin next day showed decreased billirubin levels. After 2 days of therapy baby became unicteric. Repeat billirubin levels showed Total Billirubin 7.6 mg/dl, Direct billirubin 0.6 and indirect billirubin to be 7 mg/dl. During NICU stay it was noticed that there was deviation of the angle of mouth of the baby on left side. During crying baby was able to close both the eyes. In view of deviation of angle of mouth to left side and baby's ability to close both the eyes a provisional diagnosis of right sided hypoplasia of depressor angularis oris muscle was made.

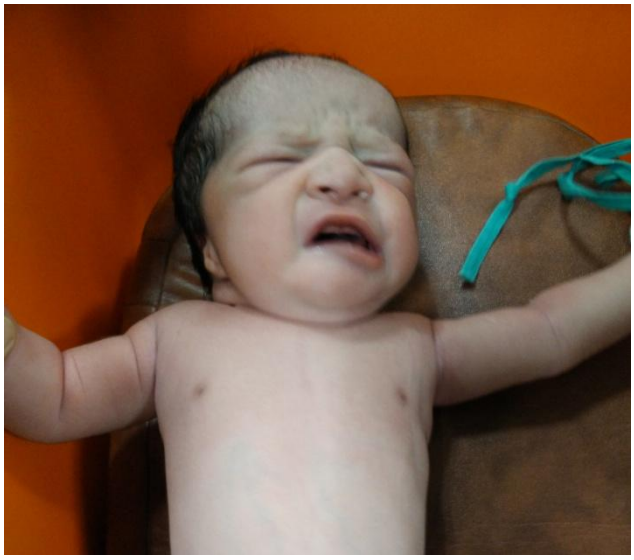

Fig:1- Assymetric crying facies. Note ability of newborn to close his eyes.

Since Hypoplasia of depressor angularis oris muscle is known to be associated with cardiovascular abnormalities 2 D-Echo was done which showed ostium secondum type of atrial septal defect. Other anomalies like genitourinary and musculoskeletal anomalies were ruled out.

Counselling of parents was done regarding the need for future surgical intervention for facial asymmetry. The need to follow up with pediatric cardiologist was also conveyed. Baby was discharged on day 9 of life with an advice to follow up after 1 week.

Discussion:

Congenital hypoplasia of depressor anguli oris is one of the causes of asymmetric crying face In a neonate. clinical feature of children with congenital hypoplasia of depressor anguli oris includes typical clinical picture, which includes asymmetry during crying while forehead wrinkling, nasolabial fold depth, and eye closure remain intact and equal on both side. In facial nerve palsy the forehead wrinkling, nasolabial fold depth and eye closure are also affected [1]. The diagnosis is based upon the typical clinical features in absence of history of birth trauma. The diagnosis can be confirmed by electromyography [2]. Though functional outcome of this condition is poor but surgery is used for cosmetic purposes. The congenital hypoplasia of depressor angularis oris muscle is associated with many congenital anomalies like cardiofacial syndrome, velocardiofacial syndrome, CATCH 22 (cardiac defect, abnormal facies, thymic hypoplasia, cleft palate, and hypocalcemia), VACTERL (vertebral anomalies, anal atresia, cardiac malformations, tracheoesophageal fistula, renal anomalies, limb abnormalities), and Trisomy 18 [3]. The cardiac anomalies associated with hypoplasia of depressor angularis oris includeTetrology of fallot, atrial septal defect, ventricular septal defect, patent ductus arteriosus and coarctation of the aorta. Associated congenital heart diseases increases the morbidity and mortality in the form of congestive cardiac failure [4]. The maxillofacial anomalies include auricular malformation, maxillary or mandibular hypoplasia, low set ears, and auditory dysfunction [5]. The deletion within chromosome region of 22q11 may occur in patients with dysmorphologic and cardiological syndromes; DiGeorge syndrome, velo-cardiofacial syndrome and conotruncal anomaly face syndrome [6] The etiopathogenesis of congenital hypoplasia of depressor angularis oris has not been established. Various causative factors like intrauterine causes, antenatal viral infection , and heredity have been suggested as causative factor [7,8]. Most accepted theory is that this condition is multifactorial [9,10].

Conclusion:

Any Neonate presenting with asymmetric crying facies should be thoroughly investigated for presence of other congenital anomalies specially congenital heart diseases.

Conflict Of Interest: None

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