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Case Report Open Access

Congenital Asymmetric Crying Face with Bilateral Microtia

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Introduction

Congenital asymmetric crying faces is a minor congenital anomaly due to absence or hypoplasia of the depressor anguli oris muscle on one side of the mouth which manifest in a newborn at birth [1]. Microtia is a congenital malformation characterized by total or partial absence of the whole auricle or any of its components, varying from a small auricle to the total absence (anotia) [2]. We present a case of congenital asymmetric crying face with bilateral microtia.

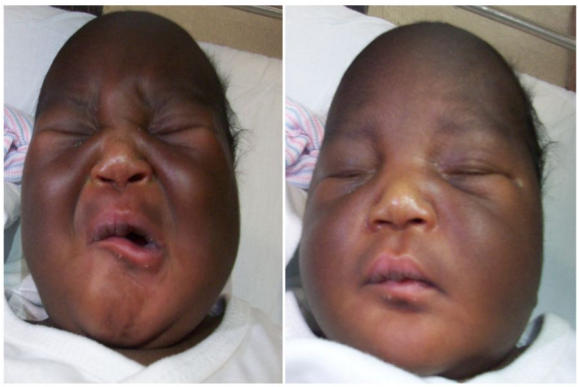

Figure 1: Physical examination of Face.

We performed an assessment of a newborn girl with a birth weight of 4020 gr, head circumference of 35 cm, chest circumference of 34.5 cm, abdominal circumference of 34 cm and length of 50 cm. The Apgar score was 8 and 9 at 1st and 5th minute. The infant was born to a 40 y/o African mother, multigravida (6 pregnancies, 5 full term deliveries), after 39 weeks of gestation. She had a history of gestational diabetes in her last pregnancy, that was controlled with diet only and the baby was born without any anomalies. She did not have a family history of diabetes. The mother attended our facility for 2 prenatal visits and the prenatal course was significant for poorly controlled gestational diabetes, as she was non-compliant with prescribed treatment of Metformin 500 mg two times daily. At 10 weeks of gestation where the Finger stick glucose test was 255 mg/dL and Hemoglobin A1C was 11.8%. At 39 weeks of gestation the Finger stick glucose test was 155 mg/dL and Hemoglobin A1C was 9%. The mother denied any symptoms including polyuria, fatigue, blurred vision, increased thirst or hunger, weight loss or gain. She had a prenatal fetal sonogram at 16 weeks that did not show any evidence of anatomical anomalies. At time of delivery an Ultrasound screen did not detect any evident anatomical abnormalities. She was also seen by a genetic counselor and failed to keep up the subsequent appointment, but she was seen by a cardiologist for a fetal cardiac echo at 18 weeks which showed no fetal congenital heart anomalies.

The infant physical examination was performed at birth soon after delivery. It was a surprise for the mother and the staff who delivered the baby as it was remarkable for a large for gestational age infant who, when grimacing or crying had a downward pulling of the mouth on the left side while the right side remained raised and did not move down. The face looked symmetrical at rest, forehead was broad and slopping but its movement was not affected, eye closure on both sides and sucking were normal (Figure 1). A bilateral microtia with low set ears was noted, with the right ear showing a vestigial pinna and nodule and the left ear with larger but underdeveloped pinna (Figure 2). The examination of the tongue, palate, genital organs and extremities was normal and no other abnormalities were detected. The initial finger stick glucose test performed on the baby was 54 mg/dL and subsequently feedings and glucose monitoring were normal. The infant postnatal renal and head ultrasounds were normal. The infant failed the newborn hearing screening test on both ears and also failed the ABR test. The only test we did not perform was the chromosomal assay due to refusal of the mother.

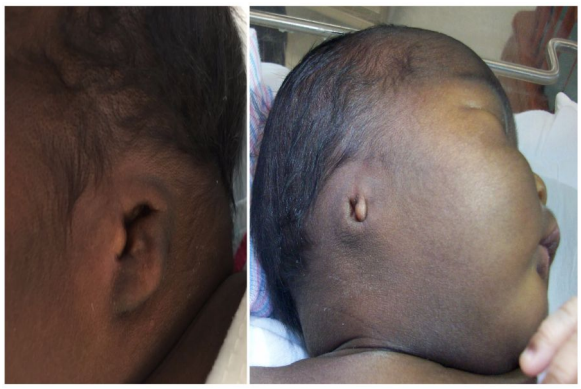

Figure 2: Physical examination of Ear.

Discussion

Congenital hypoplasia of depressor angulioris muscle can occur singly or associated with other anomalies. It causes facial asymmetry when the infant cries or grimaces; when infant is calm and not crying facial features are symmetrical. The incidence of this anomaly is 3-6/1000 live birth [3]. Typical clinical presentation includes lower lip asymmetry during crying or grimacing while forehead wrinkling, nasolabial fold depth and eye closure remain intact. The diagnosis can be confirmed by electrophysiological studies showing a normal

conduction time of the facial nerve to the mentalis and orbicularis oris muscle. It can be associated with congenital anomalies affecting the cardiovascular, head and neck, skeletal, genitourinary, chromosomal and gastrointestinal system [3]. Cardiovascular and head and neck anomalies have been the most common anomalies. Cardiovascular anomalies associated with this condition include ventricular septal defect, tetralogy of Fallot, patent ductus arteriosus, coarctation of the aorta and atrial septal defect; these were ruled out in our case. Head and neck anomalies associated with this condition include maxillary and mandibular hypoplasia, auricular malformation, low set ears and auditory dysfunction [3]. Caksen et al. reviewed 35 cases of asymmetric cyring facies and only one infant in the study was born to a diabetic mother [4]. Our patient presented with low set ears with bilateral microtia with no external auditory canal with auditory dysplasia.

The frequency of Microtia in different parts of the world ranges from 0.4 to 5.5/10000 newborns [2]. It can occur bilaterally or unilaterally where the right ear is more often affected [5]. It can be an isolated finding or part of syndromes including Goldenhar syndrome, congenital rubella syndrome, trisomy 21, and diabetic embryopathy [6]. Infants of diabetic mothers (IODM) are at an increased risk of Oculo-Auriculo-Vertebral Syndrome associated with hearing loss, cardiac, renal and limb anomalies. Ewart-Toland et al study found microtia present in 52% on IODM [7].

The type of diabetes in pregnancy, also associated risk factors increase the risk of fetal anomalies. The incidence of fetal congenital anomalies is 2 to 3 times more frequent in IODM than the general population affecting almost all organ system in the body. Our patient did not have coexisting cardiac anomalies as documented by fetal echocardiogram, which ruled out Cayler Cardio-Facial syndrome. The infant failed the hearing test, and post-natal renal ultrasound was normal. The infant was discharged and will be followed in the General pediatrics clinic and with the Institute of Reconstructive Plastic and ENT Surgical team.

Conclusion

Congenital asymmetric crying face noted at birth without any other anomalies may occur as a single entity due to in utero position of the fetus with pressure over the mandible which compresses the branch of facial nerve. If associated with other anomalies, like in our case with microtia, it can be a part of a genetic syndrome affecting multiple organ systems. Thus, a cardiovascular and renal evaluation via echocardiogram and renal ultrasound are warranted. Infant of diabetic mothers should be particularly assessed for craniofacial anomalies as maternal diabetes is known to have teratogenic effects.

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