Right Sided Facial Nerve Palsy, Bilateral Microtia with Polydactyly in an Infant – A Rare Case Report

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INTRODUCTION

Congenital facial palsy (CFP) is facial palsy of the seventh cranial nerve present at birth. It is commonly believed to be either developmental or originally acquired. Facial palsy of developmental origin is associated with other anomalies including ear, eye or cardiac anomalies. But it is rarely associated with polydactyly. We report a 10-month-old female infant who had right CFP with bilateral microtia and polydactyly.

Congenital facial nerve palsy unilaterally manifesting as weakness of entire face on one side due to infranuclear cause is a rare occurrence in paediatric population.¹ The facial palsy of lower motor neuron type involves weakness of same side of muscles of face of one side. Facial nerve as it emerges out of cranium traverses through facial canal medial to mastoid process and divides into multiple branches. It supplies ear, autonomic fibres for lacrimal gland, salivary glands, and motor supply to the face. Any pathology, congenital or acquired causing compression of facial nerve in this pathway can lead to facial palsy. Microtia is a congenital condition that varies from minor structural defects to full ear absence in severity and may occur as an individual defect or as part of a syndrome. Polydactyly is the most common hereditary limb anomaly characterized by extra fingers or toes. Here, we report a case presented with bilateral microtia and right-side facial nerve palsy.

PRESENTATION OF CASE

A 10-month-old female infant born out of non-consanguineous marriage was brought with the complaint of facial asymmetry and deformity of both ears since birth. The child was born prematurely via Caesarean section at 34 weeks of gestation with birth weight of 1.2 kg without birth asphyxia. The mother had polyhydramnios and gestational hypertension. There was no history suggestive of intrauterine infection or drug intake during pregnancy. In view of very low birth weight and prematurity, the baby was kept in neonatal intensive care unit (NICU) for 25 days. At birth, the child had small pinna bilaterally and facial deformity. At 1 month of age, patient was diagnosed to have retinopathy of prematurity for which she was operated at 3 months of age. There was no family history of congenital malformations.

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DOI: 10.14260/jemds/2021/285

How to Cite This Article:

*Chaudhary S, Injeti G, Taksande A, et al.
Right sided facial nerve palsy, bilateral
microtia with polydactyly in an infant – a
rare case report. J Evolution Med Dent Sci
2021;10(18):1353-1355, DOI:
10.14260/jemds/2021/285*

*Submission 17-12-2020,
Peer Review 06-03-2021,
Acceptance 12-03-2021,
Published 03-05-2021.*

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Her weight was 7.4 kg, height of 65 cm and head circumference was 42 cm. Her vitals were: Heart rate: 110 / min, respiratory rate: 36 / min, blood pressure: 84 / 56 mmHg and Spo2: 98 %. On general examination, facial deformity, bilateral small pinna (microtia, grade III) (Figure 1), right preaxial polydactyly were present. Neurological examination was suggestive of right sided lower motor neuron facial palsy suggested by inability to close the right eye completely and loss of a nasolabial fold on the same side (Figure 2). Absence of forehead wrinkles and asymmetric faces with the right lower lip being pulled down while crying was also seen. Left eye closure and nasolabial fold were normal. There was no other cranial nerve palsy and the remainder of the neurological examination was normal. Respiratory, cardiovascular and abdominal examination were normal. Spine and genitalia were normal. On investigations, haemoglobin was 11.2 gm %, total leukocyte count 7,000 / cumm, platelet count 1.55 lakhs and peripheral smear showed normocytic normochromic RBCs. Liver function test and renal function test were normal. Brainstem evoked response audiometry showed VTH wave formation at 100 dB in both ears suggestive of moderate sensory neural hearing loss. Abdominal ultrasonography, transthoracic echocardiogram, cranial ultrasonography and ophthalmological examination were normal.

Figure 1.
Small Pinna
on Both Sides

Figure 2.
Right Side Facial Palsy

DISCUSSION

An uncommon disorder with an occurrence of 2.1 in 1,000 live births is congenital facial nerve paralysis.² It is typically associated with birth injury in 78 percent of cases.³ Congenital facial palsy is clinically defined as facial palsy of the seventh

cranial nerve which is present at birth or shortly thereafter. The facial nerve is compromised and compressed due to particular anatomical course making it difficult to navigate and sustain inflammatory response in a tight space. This makes unique value proposition for trauma to the nerve and following weakness. The 7th cranial nerve may not be developed properly due to agenesis or partial development of temporal bone. The adjoining area of mastoid process and ear cartilage is also affected. Kulkarni et al.³ and Mahali RR⁴ have reported case on facial nerve palsy with ear anomaly. Other congenital anomalies associated include cleft palate, hypoplastic maxilla, renal defects and heart defects. Association of ear-pinna defects and facial palsy has been reported in 10 - 15 % of patients. Absence of pinna occurs in around 1 in 20000 live births. Other syndromes associated with ear deformity and facial nerve palsy like velo-cardio-facial, Moebius syndrome, Poland's syndrome, Goldenhar's syndrome, Lambert and Townes Brocks syndrome.^{5,6} Velo-cardio-facial syndrome has vertebral and cardiac defects, Moebius syndrome includes abducent nerve palsy; Poland syndrome has absence of pectoralis major muscle; Goldenhar syndrome involves entire half of the face along with cardiac defects.⁷ Lambert syndrome includes brachial dysplasias, cholestasis, inguinal hernia and club foot and Townes Brocks syndrome includes imperforate anus, thumb malformations.⁸ Our case report is different from others, in this report along with facial nerve palsy and bilateral microtia, patient also had polydactyly.

FINAL DIAGNOSIS

Cranio Facial Palsy with bilateral anotia and external auditory canal atresia with polydactyly in an infant.

Financial or other competing interests: None.

Disclosure forms provided by the authors are available with the full text of this article at jemds.com.

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