ijlpr 2022; doi 10.22376/ijpbs/lpr.2022.12.5.L78-81

International Journal of Life science and Pharma Research

ISSN 2250-0480

Case Report

Facial Nerve Palsy

A Case of Congenital Facial Nerve Palsy with Microtia

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Abstract: Congenital facial nerve palsy is defined as the presence of seventh cranial nerve palsy at birth or shortly thereafter. It occurs due to paralysis of muscles of facial expressions. It is mostly developmental or due to birth trauma. It is rare to be born with unilateral facial nerve palsy with associated ear anomaly, and hence determining the etiology is pivotal in accessing the prognosis and for selecting the right treatment for better outcome. In this case, a child is born with unilateral left -sided lower motor neuron type facial palsy, along with left sided microtia with no history of birth trauma or any associated congenital anomalies. Therefore, it is of developmental origin and does not fit into any syndromic diagnosis with an magnetic resonance imaging brain showing normal facial nerve anatomy. Child had little improvement in facial expressions with better eye closure with regular physiotherapy. Facial reanimation surgeries can be done either in preschool or adolescent period depending on the child's condition. In this case, the child is being managed conservatively until he is fit enough to perform facial reanimation surgeries.

Key Words: Congenital, Facial Nerve Palsy, Microtia, Developmental, Conductive Hearing Loss, Facial Reanimation Surgeries.

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Received On 30 April, 2022

Revised On 1 July, 2022

Accepted On 8 July, 2022

Published On 1 September, 2022

Funding This research did not receive any specific grant from any funding agencies in the public, commercial or not for profit sectors.

Citation Dr.S.Sundari, and Dr.R.Srinithi , A Case of Congenital Facial Nerve Palsy with Microtia.(2022).Int. J. Life Sci. Pharma Res.12(5), L78-81 http://dx.doi.org/10.22376/ijpbs/lpr.2022.12.5.L78-81

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1. INTRODUCTION

Congenital facial palsy is seen in 8-14% of all cases of facial nerve palsy.[1] It may be due to defect in the motor nucleus of facial nerve or facial nerve itself (neurogenic), defective neuromuscular junction, defective facial muscles, or it may be idiopathic. It may also be caused due to trauma during forceps delivery which may result in temporary or permanent facial palsy. Clinical characteristics include deviation of angle of mouth on crying, loss of forehead wrinkles, lagophthalmos, drooling of saliva, loss of nasolabial fold. causes of congenital facial weakness including congenital fibrosis of the extraocular muscles (CFEOM) type 3A, brain stem dysgenesis, CHARGE syndrome, hereditary congenital facial paresis, Moebius syndrome, and oculo-auriculo-vertebral spectrum. Congenital myopathies causing facial weakness include, central core disease, Carey-Fineman-Ziter syndrome, myotubular myopathy, and myotonic dystrophy type 1. Other causes include asymmetric crying facies, Marden Walker syndrome, and Nablus mask like facial syndrome. It carries many difficulties for the newborn due to difficulties in feeding and closing eyes. It is usually associated with anomalies of the external ear along with hearing loss[3,4].This case report is about 9 months old child who was presented with unilateral seventh cranial nerve palsy, which was present at birth along with unilateral microtia and conductive hearing loss in both ears.[5,6]Investigations play an important role in differentiating the causes of facial nerve palsy, which includes MRI brain with constructive interference in steady state sequence that accurately describes the anatomy of cranial nerves which could not be clearly seen with normal MRI brain. Brainstem evoked response is gold standard in assessing the hearing loss in these children with microtia. Other investigations like Echocardiogram, Ultrasound abdomen and other blood investigations are done to rule out associated congenital anomalies. Child is followed up from birth till present, to prevent any complications arising due to facial palsy like vision abnormalities due to inadequate eye closure, speech difficulties due to associated hearing loss, and to avoid psychosocial problems that may arise when a child enters school. Hence it is a multidisciplinary approach involving general paediatrics plastic surgeon, ophthalmologist, speech therapist along with developmental psychologist. There are many differential diagnoses in congenital facial weakness in newborns. Hence it is important for the treating physician to have a broad idea on the causes of facial palsy to narrow down the diagnostic approach. It carries poor prognosis when compared to facial palsy due to birth trauma in achieving complete recovery, and surgery is not usually performed on these infants at the time of birth. Timing of surgeries and selection of candidates for surgery is assessed on an individual basis, with lots of new modalities of reanimation surgeries being done to achieve good outcomes. Parents should be counseled regarding the prognosis of the disease and the treatment modalities available for treating the child. Genetic counseling should be given if it is due to familial causes. Since the child has facial palsy from birth, it has been found that the child has better adaptive responses to the facial weakness and they show compensatory expressive behavior in the form of verbal and non verbal communication. This study highlights the importance of diagnosing the etiology of facial palsy and planning the management in infancy for future better outcomes.

2. CASE REPORT

A 9 month old child born out of non-consanguineous parents presented to Sree Balaji medical college and hospital, Chennai, India with unilateral left sided facial nerve palsy along with left sided microtia which was present since birth of the child.[7] Child had complaints of deviation of mouth to right side since birth and was noted by the mother while crying and smiling. He also had difficulty closing the left eye completely. No difficulty in feeding and drooling of saliva seen. Antenatal history was uneventful and there was no history of maternal intrauterine infections. Child was born out of Lower Segment Caesarean Section with a birth weight of 2.5 kilograms. Baby cried immediately after birth with no history of any birth trauma or Neonatal intensive care unit admission.. Developmentally the child was normal with no significant family history. On examination, the child had left lower motor neuron type facial palsy manifested as deviation of angle of mouth to right side with inability to close left eye. Left side nasolabial fold was absent along with the absence of forehead wrinkles on the left side. Other cranial nerve examination was done with normal extraocular movements and other systems were normal. Child had no nystagmus or limb and cardiac defects. Basic blood investigations like complete blood count, peripheral smear and liver function tests were normal. MRI BRAIN with Constructive interference in steady state (CISS) sequence of facial nerve showed both side -normal trackers of intracranial and meatal segments with left hypoplastic pinna. Immittance audiometry showed middle ear pathology with both sides absent acoustic reflex. Auditory brainstem responses(ABR) showed right ear mild conductive hearing loss and left ear showed moderate conductive hearing loss. Echocardiogram and ophthalmic examination were normal. There were no associated congenital anomalies. He was managed conservatively with physiotherapy for improved facial muscles strengthening and he was regularly followed up to prevent development of corneal ulceration by instilling frequent eye drops and eye patches.

Fig 1: Left Sided Lower Motor Neuron Type Facial Palsy

Fig 2: Microtia – Deformed Pinna Left

3. DISCUSSION

Congenital facial palsy occurs due to developmental or acquired causes[7,8]. It most commonly occurs as a result of birth in children. Its anatomical position makes it more prone for injury due to inflammatory responses.[9,10] Facial nerve palsy occurs in isolation or as part of syndromes with associated congenital anomalies. It can be unilateral or bilateral, complete or incomplete associated with anomalies of ears, eyes, heart andlimbs. It can be part of syndromes like Moebius syndrome, Poland syndrome, velo cardio facial syndrome, Lambert syndrome, Townes Brocks syndrome, Goldenhar syndrome.[11] Microtia is an important diagnostic marker for presence of hearing loss in the child. Incidence of conductive and sensorineural hearing loss in children with microtia are 96% and 8% respectively.[12] Microtia and atresia of external auditory canals can be part of several syndromes or acquired due to intrauterine infections like syphilis and rubella or due to toxin exposure or ischemic injury which results in hemifacial microsomia. Facial nerve palsy severity is graded by many scoring systems and most commonly used scale is House-Brackmannaccording to which our child falls under mild grade.[13] Investigations should be done to identify the cause of palsy and for detecting associated anomalies.[14,15] Computed tomography(CT) scan of temporal bone can be considered if paralysis occurs due to trauma and if it is not resolving. Electrophysiology of facial nerve function is used to identify the extent of injury and for planning surgeries. BERA (brainstem evoked response audiometry) can be done to assess the extent of hearing loss.[16]Initial management should focus on eye care by instilling artificial eye drops. Steroids use in congenital facial nerve palsy is not proven to be beneficial in cases of developmental origin.[17] Surgical explorations like direct neurorrhaphy and nerve graft have good outcomes if done early within 72 hours of insult in case of trauma. In cases of developmental cause, facial reanimation surgeries can be done at either before child enters school or in adolescent period with main aim of attaining good facial symmetry at rest and during voluntary expressions with spontaneous emotive movements.[18-20] Hearing impairment should be corrected by hearing aids or surgeries depending on the child's condition. Eye protection surgeries are indicated for extreme corneal ulceration.[21]

4. CONCLUSION

Congenital facial nerve palsy with ear anomalies due to developmental origin has less chance to attain entirely normal appearance of face. This case report highlight the significance of diagnosing the condition early in life to prevent any complications arising out of it. Timely management enables in our case a better prognosis in future in achieving better facial features. Research is being done to find the best timing for facial reanimation surgeries to attain good prognosis. Hence, it is pivotal for treating doctors to identify the basic etiology and to decide on appropriate timely intervention for better future outcomes.

5. AUTHORS CONTRIBUTION STATEMENT

Dr.S.Sundari conceptualized and gathered the data with regard to this work. Dr.R. Srinithi analyzed these data and necessary inputs were given towards the designing of the manuscript. All authors discussed the methodology and results and contributed to the final manuscript.

6. CONFLICT OF INTEREST

Conflict of interest declared none.

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