CASE REPORT

UNILATERAL EXTERNAL AUDITORY CANAL ATRESIA WITH CONGENITAL FACIAL NERVE PALSY: A CASE REPORT

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ABSTRACT: Congenital facial palsy is generally due to developmental anomaly or acquired cause. Unilateral congenital facial palsy due to developmental defect is most often associated with inner ear abnormalities. We report a rare case of unilateral congenital facial palsy with atresia of pinna and external auditory canal and normally developed middle ear and inner ear structures.¹

KEYWORDS: Congenital facial nerve palsy, External auditory canal atresia, Bell's palsy, Microtia.

INTRODUCTION: Congenital facial nerve palsy associated commonly with inner ear abnormalities. The presentation of congenital facial palsy with external ear canal atresia and normal middle and inner ears is a rare presentation.

CASE REPORT: A 17years old male patient presented to ENT OPD with facial asymmetry, absence of right ear since birth. Patient did not seek any medical advice since birth. There is no history of Intrauterine birth infection, Prolonged labour and there is no premature delivery.

ON EXAMINATION: Patient has Right atresia of external auditory canal with preauricular tag, Right LMN Type of facial palsy with Bells phenomenon. Left ear is normal on examination and other cranial nerves on examination are normal.

Tuning Fork Test: Weber is lateralized to right ear.

C.T SCAN TEMPORAL BONE:

Right Ear:

- Hypoplasia and hypopneumatisation with sclerosis of right mastoid air cells.
- Right external auditory meatus is not made out- atresia.
- Soft tissue density is seen in the mesotympanum.

Left Ear:

- External auditory meatus is normal.
- Left mastoid air cells are normal.
- Left middle ear cavity structure appears normal.

DISCUSSION: According to Bobby R. ALFORD study of 118 cases of aural atresia prevalence of ear abnormalities with facial palsy is seen in 13% of cases where as inner ear abnormalities are seen in 22% of cases. Incidence of facial palsy in live births is 0.8-2.1/1000 live births of these 88% are associated with difficult labour.² developmental cause of facial palsy is mobeiuss syndrome where facial palsy is associated with lateral rectus palsy. Unilateral aural atresia associated with facial palsy is rare congenital facial palsy may cause multiple problems in new born like difficulty in nursing, eyeclosure,

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development of speech, expression of emotion, mastication. The physical examination is intended to exclude other syndromes like extra ocular muscle palsy, cleft palate and internal organs. Another congenital abnormality associated with facial palsy is that of maxilla like cleft palate, hyoplastic mailla, duplication of palate.

Atresia, microtia with facial palsy are associated with inherited defects or acquired embyopathies owing to intrauterine infection like TORCH or exposure to toxins.

Embryonic insult due to toxins is severe can also produce abnormalities of other organs in addition to atresia like inner ear abnormalities

Deformities of canalisation of auditory canal are associated with deformities of pinna. The time for intervention of atresia along with facial palsy is debatable.

Sometimes facial palsy is associated with wide spectrum of congenital anomalies that involve structures from Ist and IInd arch Goldenhar syndrome is oculo-auriculo-vertebral dysplasia with congenital facial palsy.³

Most frequent cause of congenital facial palsy is birth trauma related to difficult delivery. Risk factors include forceps delivery, birth weight >3.5kg, primiparity, injury to facial nerve vertical segment by the posterior blade over the mastoid.⁴ Facial nerve is susceptible to trauma as it exits from stylomastoid for a men where soft tissue compression can lead to damage of facial nerve (CHARGE Syndrome). Deformities of canalisation of auditory canal can lead to deformities of pinna.⁵

In addressing congenital facial palsy, some medical professionals advise initial surgery during preschool to avoid psychosocial problems associated with physical abnormality. However waiting till adolescence when facial growth is mature and child is able to understand the risks and benefits of surgery has merit.⁶

Muscle transplantation for facial palsy is effective but better results are observed in case of traumatic facial palsy and reconstruction of auditory canal and pinna has to be taken when planning for muscle transplantation surgery with facial palsy.⁷

In this case as middle ear and inner ear is well developed the case is planned for reconstruction of ear canal and pinna. Facial palsy cannot be corrected as patient has reported late.

Fig. 1: Frontal view-Congenital facial palsy right side with aural atresia showing deviation of angle of mouth.

Fig. 1

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Fig. 2: Lateral view showing Right aural atresia with preauricular tag.

Fig. 3: Bell's phenomenon.

Fig. 2

Fig. 3

Fig. 4: CT SCAN picture showing Right external canal atresia.

Fig. 5: CT SCAN picture showing Right middle ear and inner ear normal with atresia of external auditory canal.

Fig. 4

Fig. 5

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