Bangladesh J Otorhinolaryngol 2012; 18(2): 179-182

Case Report

Unilateral internal auditory canal stenosis with progressive facial weakness

Sathiya Murali, Arpana Shekhar, S. Shyam Sudhakar, Kiran Natarajan, Mohan Kameswaran

Abstract:

Internal auditory canal (IAC) stenosis is a rare cause of sensorineural hearing loss. Patient may present with symptoms of progressive facial nerve palsy, hearing loss, tinnitus and giddiness. High resolution temporal bone CT-scan and magnetic resonance imaging (MRI) are the important tools for diagnosis. No specific management has been devised. Here is presentation of a case of unilateral (left) IAC stenosis with profound hearing loss and progressive House Brackmann Grade III-IV facial weakness. The diameter of the IAC was less than 2 mm on high resolution temporal bone computed tomography (HRCT) scan. It was managed by facial nerve decompression by translabyrinthine approach in an attempt to prevent further deterioration of facial palsy.

Key words: *Internal auditory canal stenosis; facial nerve decompression*

Introduction:

Internal auditory meatus (IAM) stenosis is defined as a loss of 3mm or more in the vertical diameter of the internal acoustic meatus, or even as a meatus smaller than 2mm[1]. Inner ear abnormalities may occur in about 20% of the cases of patients who have sensorineural hearing loss[2].

In most cases, it occurs as an isolated congenital manifestation, and in others it may be a part of systemic abnormalities. The major alteration happens due to a constriction caused by impaired bone growth compressing on the nerve bundle. The stenosis may also develop secondary to alterations in the VIII cranial nerve (vestibulocochlear nerve aplasia or hypoplasia)[3]. Similar etiology can explain progressive facial nerve palsy. The embryologic events involved in fetal growth between the 4th and the 8th weeks are crucial for bone growth, and may cause such disease[4]. The labyrinth may be aplastic or deformed or an incomplete cavity. It is not uncommon to have a com-pletely normal labyrinth though[1-3]. An acquired bone disease may also result in stenosis of the internal acoustic meatus (osteomas, osteopetrosis, Paget's disease, etc).

HRCT of temporal bone showing a narrow internal acoustic meatus is the diagnostic tool of choice. MRI scans are helpful in visualization of the IAC structures such as the VIII cranial nerve, which may be aplastic[5].

Madras ENT Research Foundation, No. 1, 1st Cross Street, Off 2nd Main Road, Raja Annamalai Puram, Chennai, Tamil Nadu, India – 600 028.

Address of Correspondence: Dr. Sathiya Murali, Madras ENT Research Foundation, No. 1, 1st Cross Street, Off 2nd Main Road, Raja Annamalai Puram, Chennai, Tamil Nadu, India - 600 028. Email: sathiyadr@gmail.com Phone: 044 24311411

Case Report:

A-13-year old girl presented to us with history of left ear hard of hearing noticed about one year ago of unknown onset. She also had progressive left facial nerve weakness (grade III-IV HB) of 6 months duration. There was no history of tinnitus or any history suggestive of vestibular disturbance. Antenatal, natal and postnatal history was uneventful.

Clinical examination showed normal otoscopic findings with absence of ipsilateral wrinkling of forehead (grade IV). Tuning fork tests were suggestive of unilateral (left-sided) sensorineural hearing loss. Audiological tests included pure tone audiogram which revealed profound hearing loss in the left ear and normal hearing in the right ear. Impedance showed 'A' type tympanogram bilaterally. Otoacoustic emissions were absent on the left side. Electrophysiological studies showed no indication of retrocochlear pathology. Videonystagmography was normal except for the caloric test which showed hypoactive left labyrinth.

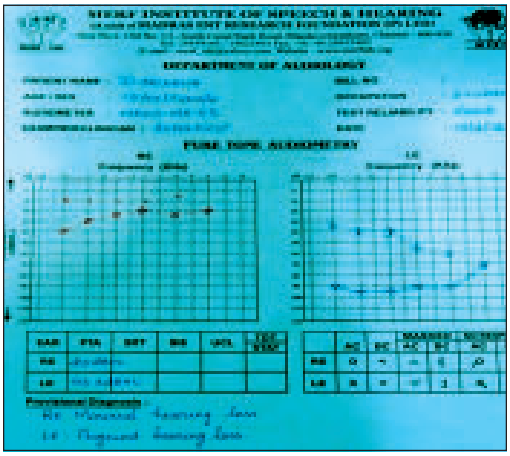

Figure 1: *Pure Tone Audiometry: Left ear – Profound hearing loss.*

HRCT scan of temporal bones revealed narrowing of left internal auditory meatus measuring only 2 mm in diameter with normal dimensions on the right side. No other congenital anomalies of inner ear were found. MRI showed hypoplastic left vestibulocochlear nerve. Facial nerve conduction study showed reduced CMAP (compound myogenic action potential) on the left side suggestive of left facial neuropathy. Hence in order to prevent further progression of facial weakness, it was decided to decompress the facial nerve at the IAM by translabyrinthine approach. After all the necessary preoperative workup, the patient was taken for surgery under general anaesthesia.

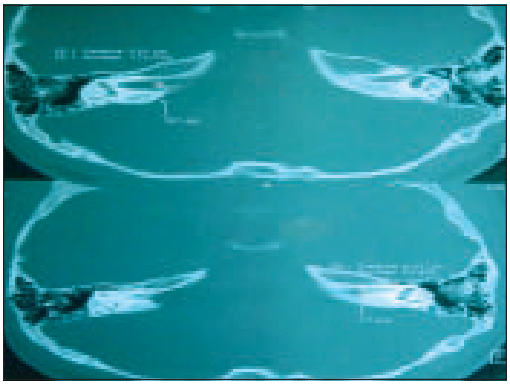

Figure 2: *CT scan of temporal bone showing narrowing of left internal acoustic canal.*

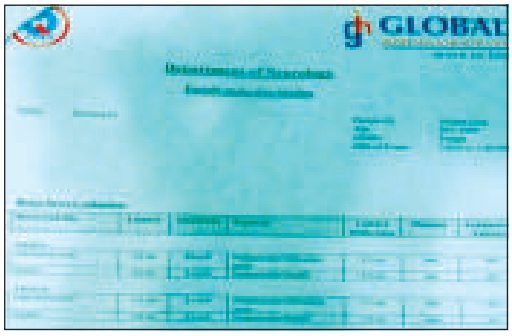

Figure 3: *Nerve conduction study- showing Left facial neuropathy*

Operative procedure:

Under GA, postaural incision was made. Inferiorly based musculoperiosteal flap

elevated. Radical mastoidectomy was done. Three canal labyrinthectomy was performed. Internal acoustic meatus approached and the stenotic segment decompressed preserving the vestibulocochlear and facial nerve. Continuous facial nerve monitoring was done during the procedure. A small CSF leak was encountered. It was closed by an airtight seal with harvested fascia lata and tissue glue. Temporalis muscle was used to obliterate the cavity. A wide meatoplasty was performed and the wound was closed in layers. Postoperative period was uneventful and the patient was discharged on the seventh day. Sutures were removed on 10th post operative day. Wound had healed well. Four weeks after surgery clinical improvement in facial nerve function was noticed and at the end of 3 months the facial nerve function had clinically improved to HB grade II.

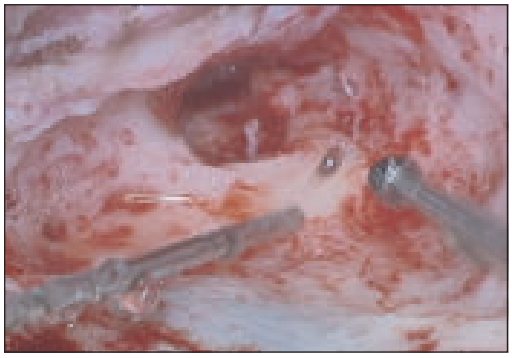

Figure 4: *Three canal labyrinthectomy*

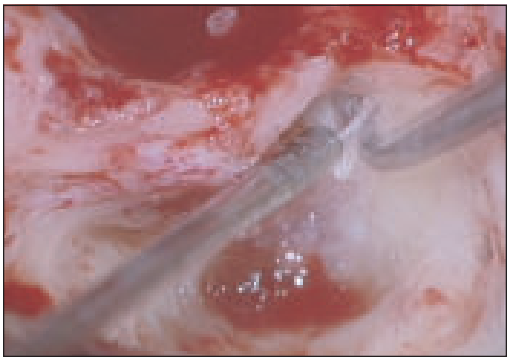

Figure 5: *Removal of stenotic segment of Internal acoustic meatus.*

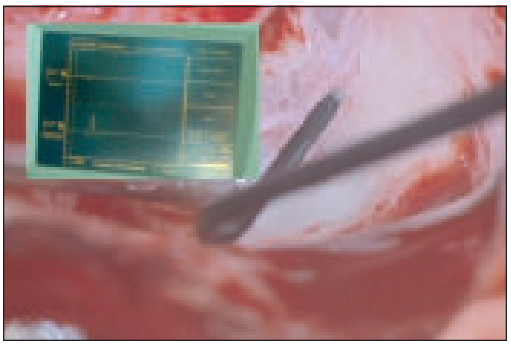

Figure 6: *Intra-operative facial nerve monitoring- Mastoid cavity with IAM opened up with probe tip touching the facial nerve.* ***Inset:*** *Facial nerve monitor*

Figure 7: *3 months post operative picture.*

Discussion:

Facial nerve palsy and sensorineural hearing loss (SNHL) due to internal auditory canal (IAC) stenosis with hypoplasia of the cochleovestibular nerve is a rare disorder. The physician should investigate the patient's obstetric, natal and post-natal history in order to rule out malformations during this period[5].

The diagnosis of the IAC stenosis requires both high resolution computed tomography scan (HRCT) and magnetic resonance imaging (MRI). An internal auditory canal measuring less than 2 mm in vertical dimension by high resolution CT temporal bone is called stenotic.

Approximately 12% of patients with congenital sensorineural hearing loss have radiographic evidence of inner ear

abnormalities including IAC stenosis. IAC stenosis without any other abnormalities is extremely rare. In addition to primary causes of IAC narrowing, an osseous lesion such as an exostosis or an osteoma can cause secondary narrowing of IAC. Also Paget's disease, otosclerosis and fibrous dysplasia can cause narrowing of the canal[6-8].

Internal auditory canal stenosis can be an important cause of sensorineural hearing loss, facial nerve palsy, and vestibular dysfunction. In a study 4 out of 7 patients had vestibular function loss[1]. But this patient with acquired IAM stenosis showed no features suggestive of vestibular dysfunction.

Surgery of the internal auditory canal occupies an increasingly important place each year and it would therefore; seem useful to review the main indications for this surgical approach as they are today. They may be classified as follows: 1. (a) surgery for the internal auditory canal stenosis itself, (b) surgery of facial nerve, (c) surgery of the vestibular nerve (neurectomy for vertigo), (d) Cochlear neurectomy for subtotal deafness with severe symptoms such as tinnitus or intolerable distortion. 2. Opening the internal auditory canal may also be necessary during the course of a more extensive operation as on (a) the petrous temporal bone, (b) the posterior cranial fossa, and (c) the middle cranial fossa[8].

As the case was of a young lady with progressive facial palsy still in its initial stages, facial nerve decompression via trans-labyrinthine approach was planned for. As hearing loss was unilateral with completely normal hearing in the opposite ear, it was decided to manage the patient conservatively with periodical audiological follow up.

Conclusion:

Internal acoustic canal stenosis is a rare disorder. It may present as facial neuropathy, progressive hearing loss or vestibular dysfunction. Imaging plays an invaluable role in the diagnosis.

References:

1. Baek SK, Chae SW, Jung HH. Congenital internal auditory canal stenosis. J Laryngol Otol 2003; 117: 784-7.
2. Nakamura K, Koda J, Koike Y. Stenosis of the internal auditory canal with VIIth and VIIIth cranial nerve dysfunctions. ORL 1999; 61: 16-8.
3. Guirado CR. Internal auditory meatus malformations. Rev Laryngol 1992; 113: 419-21.
4. Ivair Massetto Junior, Alfredo Rafael Dell'Aringa et al. Internal acoustic meatus stenosis – Case report. Rev Bras Otorrinolaringol 2008; 74(2): 318.
5. OLeary SJ, Gibson WP. Surviving cochlear function in the presence of auditory nerve agenesis. J Laryngol Otol 1999; 113: 1008-10.
6. Vrabec JT, Lambert PR, Chaljub G. Osteoma of the internal auditory canal. Arch Otolaryngol Head Neck Surg 2000; 126: 895–898.
7. M. Portmann. Surgery of the internal auditory canal. J Laryngol Otol 2003 Oct; 117(10): 784-7.
8. Hamersma H. Osteopetrosis (marble bone disease) of the temporal bone. Laryngoscope 1970; 80: 1518–1539.