Hypoplasia of Internal Acoustic Meatus

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Introduction

Hypoplasia of internal acoustic meatus (IAM) is a rare condition that may be associated with agenesis/hypoplasia of vestibulocochlear nerve (VCN) and facial nerve with or without associated labyrinthine malformation. We report a case of unilateral hypoplasia of IAM with hypoplasia of VCN but well developed inner ear.

Case Report

A 20 year old son of a serving warrant officer presented with complaints of bilateral hearing loss alongwith otorrhoea and otalgia of left ear of 3 weeks duration. There was no history of tinnitus or vertigo, preceeding major illness, exanthematous fever, head injury, local aural trauma, exposure to loud noise, ototoxic drugs or specific sexually transmitted diseases. He denied any family history of hearing loss, diabetes mellitus, hypertension or tuberculosis. General and systemic examinations were normal. ENT examination revealed normal tympanic membrane (TM) on right side. The left TM showed a small central perforation which was actively discharging. Tuning fork tests showed no response on right side while on left side Rinne's test was negative and absolute bone conduction was normal. Weber's test showed lateralization to left side. Otoneurological examination showed no abnormality. Facial nerve and other cranial nerves (except VCN) were normal. His speech and higher mental functions were normal. There was no sensory/motor deficit or signs of cerebellar dysfunction. Pure tone audiometry (PTA) revealed profound sensori-neural hearing loss (SNHL) on right side with mild conductive hearing loss on left side.

He was prescribed local and systemic antibiotics, analgesics and decongestants. The perforation of left TM healed within 02 weeks, earache subsided and hearing in left ear improved. A repeat PTA showed profound SNHL on right side with normal air and bone conduction thresholds on left side. A caloric test performed at this stage showed canal paresis on right side and normal responses on the left side.

In view of unilateral SNHL he was investigated further. Routine examinations of blood and urine were normal. Blood

sugar and blood urea were also normal. Serological tests for syphilis were negative.

HRCT of temporal bones revealed a uniformly narrowed IAM on right side (diameter 1.2mm) with well developed cochlea, vestibule and semicircular canals (SCC). On left side IAM and inner ear were normal. There were no radiological features suggestive of Paget's disease, fibrous dysplasia or osteopetrosis indicating that narrowing of IAM was congenital rather than acquired (Fig.1). MR study showed non visualization of VCN on right side in porus acousticus / intracanalicular horizontal parts consistent with congenital agenesis (Fig.2).

The patient was explained the congenial nature of his disease and reassured against any intracranial mass lesion. He confirmed that he did face some hearing difficulty in the right ear prior to this episode of aural discharge but became aware of the severity of hearing loss only after developing problem in hearing in left ear. In view of normal speech and normal hearing on left side no further intervention was felt necessary.

Discussion

Normal IAM has an average length of 12mm and vertical diameter of 2 to 8mm with an average diameter of 4mm. Diameter of < 2mm qualifies as narrow IAM. Variation in diameter between the two sides does not exceed 1-2mm. Narrowed IAM can be congenial (hypoplasia) or acquired as in Paget's disease, fibrous dysplasia, osteopetrosis etc.

Congenital hypoplasia of IAM is a rare entity which may be associated with agenesis/hypoplasia of VCN and facial nerve with or without associated maldevelopment of labyrinth. In cases of maldeveloped labyrinth the IAM besides being narrow is also tapering laterally. In a series of 35 consecutively investigated patients of unilateral SNHL over a period of 2 ½ years, Bamiou et al [1] found only one case showing markedly narrow IAM while Kokat et al [2] reported 2% incidence of narrow IAM among 126 patients with inner ear

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Fig. 1 : CT showing narrow IAM on right side

malformations diagnosed with CT of temporal bones over a period of 4 years.

VCN agenesis/hypoplasia as in our case, frequently accompanies IAM hypoplasia. According to Stjernholm and Muren [3], if the diameter of IAM is $< 1.4\text{mm}$, possibility of cochlear nerve abnormality should be considered. Studies also suggest that hypoplasia of IAM is secondary to VCN agenesis/hypoplasia and not vice-versa [4,5].

Normal development of the labyrinth can occur despite a narrow IAM and VCN agenesis [6]. Some amount of cochlear function can also occur in these cases as demonstrated by O'Leary and Gibson [7] on electrocochleography. However, in bilaterally hypoplastic IAM, MR studies must be undertaken to exclude VCN agenesis/hypoplasia prior to cochlear implant as in these cases the cochlear implant may fail or the technique may need to be modified.

Conflicts of Interest

None identified

Fig. 2 : MR image showing non visualization of vestibulocochlear nerve on right side

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