Case Report

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Surgery Section

Experience in Managing A Rare Case of Sporadic Bilateral Juvenile Otosclerosis

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ABSTRACT

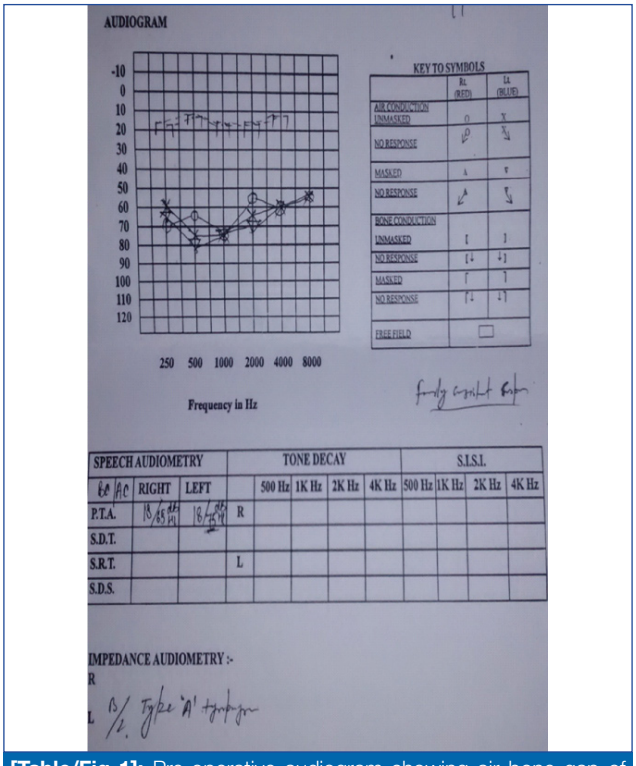
Otosclerosis constitutes a focus of localized new spongy bone formation in the labyrinthine capsule. This progressive conductive type hearing loss is a disease of young and middle aged adults. Studies have shown that the disease is twice as common in women as compared to men. The onset of hearing impairment usually appears after puberty and progresses till adulthood although the histological lesion may actually begins in early childhood or in the preadolescent years. The management modality includes surgery as well as hearing aid trial. The surgery in cases of juvenile otosclerosis however is delayed, because of the surgeon's hesitancy in acknowledging the existence of otosclerosis in such a younger age group. With the developing surgical expertise, sophisticated diagnostic techniques and knowledge about the disease process, diagnosis and treatment can be made reliably in the younger age group.

We are hereby sharing our surgical experience in managing a case of bilateral juvenile otosclerosis that presented to our outpatient department with the chief complaints of bilateral impaired hearing since six years.

Keywords: Exploratory tympanotomy, Juvenile otosclerosis, Laser stapedotomy, Stapedectomy

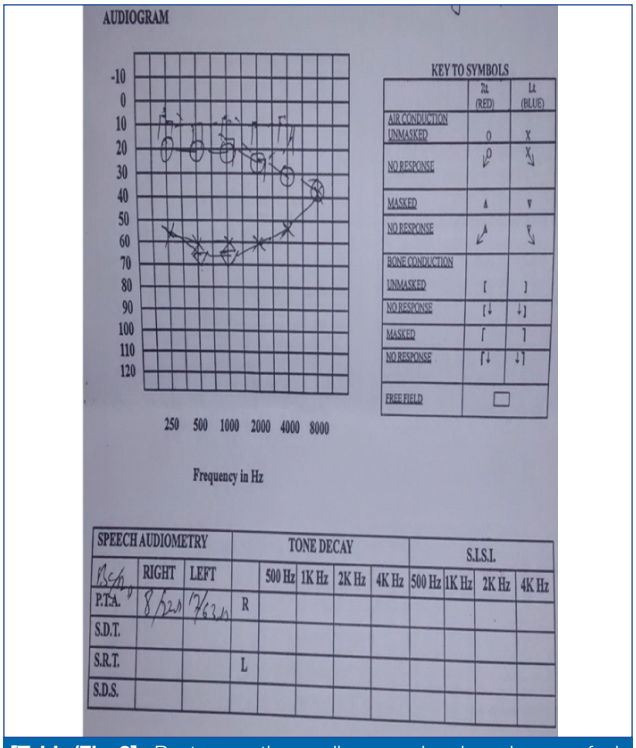
CASE REPORT

An 18-year-old boy residing in foothills of Himalaya in Kashmir was referred to our center for audiometric confirmation of his bilateral impaired hearing and its management. The boy's parents noticed that he was having an insidious onset, gradually progressive hearing difficulty since he turned 12. There was no history of ear discharge, earache, physical or acoustic trauma during childhood. The primary otological examination showed bilateral intact tympanic membrane, which was mobile on segalisation. The tuning fork tests showed bilateral negative Rinne test and an inconsistent Weber response. With a preliminary diagnosis of middle ear effusion an otoendoscopy was done that revealed bilateral absolute normal looking tympanic membrane, thus challenging the diagnosis. The impedance audiometry showed bilateral type A curves and four frequency pure tone audiogram (0.25, 0.5, 1 and 2 kHz) revealed an air bone gap of 47 dB on the right side and 57 dB on the left side [Table/Fig-1]. A differential diagnosis of juvenile otosclerosis v/s congenital ossicular fixation was kept and his parents were briefed regarding a trial of surgical treatment. Family history also revealed similar complaints in cousin brothers. An informed consent for surgery including the possibility of dead ear was taken and the boy was planned for right exploratory tympanotomy under general anaesthesia. A



[Table/Fig-1]: Pre-operative audiogram showing air bone gap of 47 db in right ear.

transcanal approach was planned and the mobility of malleus was confirmed. Intraoperatively, there was dehiscent fallopian canal and a low lying facial nerve overlying superior part of the oval window with a whitish obliterative focus completely covering the stapes footplate. The mobility of stapes was assessed by gentle palpation using an angled pick and was found to be completely absent. This was due to the presence of otosclerotic foci covering the footplate.This confirmed the final diagnosis of juvenile otosclerosis. Using a 0.75 mm Karl-Storz® drill at speed of 2500 rpm, firstly the blue lining of stapes footplate was done. Carbon dioxide laser was then used for creating fenestra in the footplate and a 0.6 mm Teflon audio® piston was neatly inserted [VIDEO 1]. Extubation was uneventful and post operatively there was subjective hearing improvement, noted in the recovery room. Facial nerve and vestibular functions were normal and patient was discharged after 24 hours on tapering doses of oral steroid over four weeks. The six weeks post-operative pure tone audiogram showed closure of air bone gap within 15 dB in the operated side [Table/Fig-2]. The patient was explained due precautions for the operated ear and has been kept under follow-up for the surgery of contralateral side after a year.



[Table/Fig-2]: Post operative audiogram showing closure of air bone gap of 14 dB in right ear.

DISCUSSION

The rarity of juvenile otosclerosis is evidenced by the paucity of reports on children who undergo surgery but what is more significant is that 15% of operated patients had developed the hearing losses before the age of 18 and almost 92% bilateral [1,2]. In 1944, the histologic study done by Guild SR et al., showed an incidence of less than 1% in children aged below five years and 4% among those aged between 5 to 18 years [3].

In otosclerosis the otospongiotic bone invades the margins of the oval window, resulting in the ankylosis of stapes footplate. This results in progressive hearing loss [4]. Although the development of otosclerotic lesion and its aetiology remains obscure, but the literature review says that there exists a strong familial predisposition of almost 50%. Owing to this hereditary tendency, the highest incidence of otosclerosis is seen among the whites, while lowest is seen among the members of black race. It is relatively common in Indians and rare in the Chinese population [5].

Patient had history of normal hearing during childhood and at the begining of school, which substantiates the fact that it was not a case of congenital otosclerosis. The right ear was preferred for surgery because of the ease of doing laser surgery on right side by the right handed surgeon. Juvenile otosclerosis patients have a higher incidence of obliterative focus, upto a tune of 22% and thus requiring footplate drilling during surgery [6]. Although, in the reported literature, 27% to 41% of the juvenile otosclerosis cases required drilling as in our case, but there are other literature reviews indicating higher need for foot plate drilling upto 45.2% in over 18 year of age patients [2,7]. Thus, the surgical risks appear to be lesser with the juvenile group, and by deferring the surgery till adulthood the chances of drilling and resultant footplate complications increases. Also various studies on juvenile otosclerosis surgery have already shown that the risk of sensorineural hearing loss post surgery is less than 10% [8,9].

CONCLUSION

There shall be no contraindication to perform surgery in juvenile otosclerosis, as the results are as good as or better than if the surgery is delayed.

The follow-up audiometry is more easily obtained in children and almost 50% patients have more than 5-years follow-up.

Deferring the surgery until later in life will not reduce the incidence of delayed hearing loss post surgery and should not be a factor in considering postponement of surgery until maturity.

The longer the foot plate pathology, greater are the chances of obliterative focus and need of drill out procedures, thus inviting complications.

ATTATCHMENTS

[Video-1]

https://youtu.be/oYZcRmvNkKc

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