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IP Journal of Otorhinolaryngology and Allied Science

Journal homepage: https://www.joas.co.in/

Case Report

Absent stapedius muscle, tendon and pyramid – An anomaly revisited

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ARTICLE INFO

Article history:
Received 14-12-2022
Accepted 21-12-2022
Available online 04-01-2023

Keywords:
Stapedius muscle
Tendon
Pyramid
Intraoperative

ABSTRACT

Congenital absence of stapedius is an exquisite entity, with an incidence of 0.5% and very few cases reported in live patients. We herein report a case of a middle-aged female presented with bilateral ear discharge and decreased hearing for more than a year, which, on examination, showed a large central perforation bilaterally, and an audiogram preoperatively revealed an average hearing loss of 45 dB and air-bone gap of 35dB bilaterally. Right endoscopic type I tympanoplasty was done, which unveiled absent stapedius muscle, tendon, and pyramid on the right side. The absent stapedius should circumspect the operating surgeon to look for other associated anomalies such as tympanosclerosis, fixed footplate and aberrant course of the facial nerve.

1. Introduction

Congenital middle ear anomalies are uncommon (1 in 10,000 live births). The stapedius muscle contributes significantly to acoustic reflex in response to loud sounds and protects the inner ear from acoustic trauma. Various anomalies of the stapedius unit have been reported, such as double or ectopic stapedius, ossified tendon, or absence of stapedius. Congenital absence of stapedius is an *extremely rare entity,* with an *incidence of 0.5%* and very few cases reported in live patients. These are often accompanied by other middle ear anomalies, such as congenital aural atresia, aberrant facial nerve or dysplastic ossicular chain, fixed footplate or tympanosclerosis.[1–5]

2. Discussion

A middle-aged female presented with bilateral ear discharge for two years and decreased hearing in both ears for

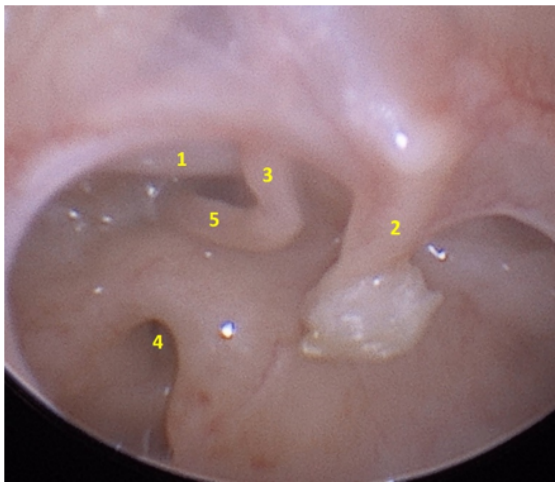

Fig. 1: Absent pyramid, stapedius muscle and tendon in the right ear(1 - tympanic segment of facial nerve, 2 – handle of malleus, 3 – long process of incus, 4 – round window, 5 – posterior crura of stapes)

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https://doi.org/10.18231/j.ijoas.2022.032

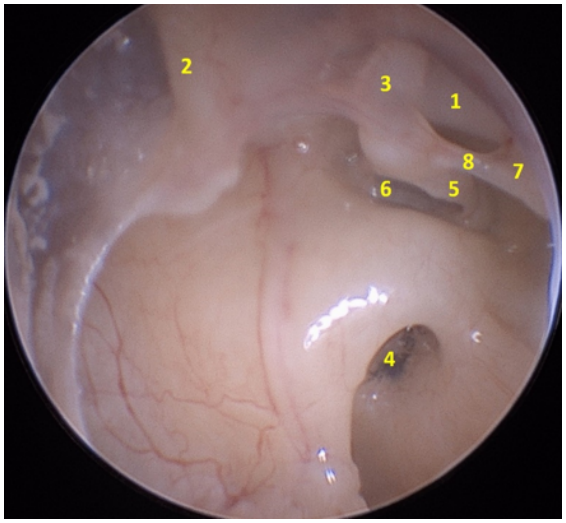

Fig. 2: Normal pyramid, stapedius muscle and tendon in the left ear(1 - tympanic segment of facial nerve, 2 – handle of malleus, 3 – long process of incus, 4 – round window, 5 – posterior crura of stapes, 6 – stapes footplate, 7 – pyramid, 8 – stapedius muscles and tendon)

one year. On examination, a large central perforation was present bilaterally. The audiogram revealed bilateral moderate conductive hearing loss with an average hearing loss of 45 dB and an air-bone gap of 35 dB. Right endoscopic type I tympanoplasty was done. The stapedius muscle, tendon, and pyramid were absent on the right side and present on the left side. (Figures 1 and 2) The round window reflex was present on the right side. The patient had no complaints of hyperacusis. The graft healed well six weeks postoperatively, and hearing improved.

The case reported by Magnuson et al. had an absent stapedius with a well-developed pyramid but an aberrant facial nerve and absent footplate, and oval window. However, in our patient, it was unilateral, asymptomatic, and without any other anomalies. In his case series of patients with absent stapedius tendon, Nassiri et al. (2021) found congenital stapes footplate fixation in all eight patients.[6,7] However, this was not seen in our case, as the footplate was mobile.

Stapes is the first ossicle to appear and is first seen at around the fifth week of gestation. Conventionally, it was believed that stapes have a dual origin, with its suprastructure originating from the second arch and footplate from the otic capsule. However, this theory has been rejected, and stapes is known to originate from the second arch. The posterior belly of the digastric muscle gives off fascicles which pass through the stylomastoid foramen into the tympanic cavity and gives stapedius muscles through the neck of stapes. This also explains the similar innervation of the digastric and stapedius posterior belly and their common origin from the second pharyngeal arch.[4,5,8,9]

Preoperative identification of this entity is *challenging* even with High-Resolution Computed Tomography. Due to this and its rare incidence, the *diagnosis of absent stapedius* is almost always made *intraoperatively.* The pyramid plays a vital role and gives bony projections, i.e. the chordal ridge and the ponticulus. These landmarks divide the superior retrotympanum into facial recess, posterior tympanic sinus, lateral tympanic sinus and sinus tympani. The absence of the pyramid can lead to the distorted anatomy of these recesses. Thorough knowledge and evaluation of this should be done preoperatively on radiology by the operating surgeon as these are the most common sites for residual disease.

3. Results and Conclusion

1. Absent stapedius muscle is a rare entity with an incidence of 0.5%.
2. This is usually associated with other middle ear anomalies and can present with hyperacusis and conductive hearing loss. However, in our case, it was unilateral, asymptomatic, and isolated (without any other anomalies).
3. The absent stapedius should alert the operating surgeon to look for other associated anomalies such as tympanosclerosis, fixed footplate and aberrant course of the facial nerve.

4. Source of Funding

None.

5. Conflict of Interest

None.

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Cite this article: Bhardwaj A, Sood R, Malhotra M, Priya M, Ravichandran N, Yadav AC, Prasath R, Dave J. Absent stapedius muscle, tendon and pyramid – An anomaly revisited. *IP J Otorhinolaryngol Allied Sci* 2022;5(4):138-140.