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Case Report

Congenital Absence of Stapedial Tendon and Pyramidal Eminence - Incidental Finding During Tympanoplasty - A Rare Entity

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Abstract

Introduction: With 9 indexed reports absence of stapedius muscle and its tendon contributes to a rare congenital anomaly. Patient with this condition present with non progressive conductive hearing loss on pure tone audiometry but the condition cannot be diagnosed with PTA or even HRCT temporal bone.

Discussion: Thus it becomes one of the differential diagnoses for conductive hearing loss with unremarkable tympanic membrane. Diagnosis can be made only via performing exploratory tympanotomy or during tympanomastoid surgery in cases with perforated tympanic membrane it can co exist with certain acquired or genetic conditions of middle or inner ear. Lack of standardized or evidence based protocol make its diagnosis challenging. The theory behind its origin is absence or regression of separation of cranial mesenchyme of second pharyngeal arch (stapedial anlage) from reichert's cartilage. Here, we present a case of 14-year-old female with congenital absence of stapedius muscle and tendon. She presented with congenital, right sided ear discharge, non progressive, moderate conductive hearing loss, and was diagnosed with this condition at surgery. She underwent type 1 tympanoplasty and was diagnosed incidentally during surgery. The Ossicular chain and round window reflex was intact. Apart from the clinical presentation and its management, the present report also emphasises the plausible theories on relevant embryology explaining such an unusual event.

Conclusion: Awareness of variation of this type is important for surgeons specially while operating for Ossicular chain fixation or discontinuity.

Keywords: Exploratory Tympanotomy; Congenital Middle Ear Anomalies; Hearing Reconstruction; Reichert's Cartilage; Stapedial Anlage

Introduction

Conductive hearing loss in children is usually acquired. The most common aetiology is otitis media with effusion but it may be the result of chronic otitis media (mucosal or squamosal). In the presence of severe congenital deformity of external ear, associated abnormalities of Ossicular chain are common but in isolation they are rare and often have a delayed diagnosis, particularly if unilateral [1].

Congenital malformations of the middle ear have been described in association with various head and neck anomalies. The isolated middle ear anomalies may present with only conductive hearing loss and are rarely encountered during middle ear surgery or during surgical explorations [2,3]. Stapedius muscle extends from the wall of the pyramidal eminence; its tendon passes forward through the apex of the pyramid to the neck of the stapes [4,5]. The stapedius and the tensor tympani muscle contract together in a reflex response for sounds of high intensity, and the stapedius pulls the footplate of the stapes for decreasing the amplitude of vibrations at the oval window [6]. It also prevents the excessive movement of the stapes [6-8]. The stapedius muscle may be doubled or ectopic [6,8,9] and its tendon may ossificate [9] or muscular unit may be absent. The incidence of the absence of the tendon of stapedius is 0.5%. There are scarce literature reports on the absence of the stapedius muscular unit [10,11]. The absence of this muscular unit can be associated with other anomalies or pathological conditions. Here we report a case of CSOM with incidental finding of absent stapedial tendon and pyramidal eminence.

Case Report

We present here a case of 14 year old female patient who presented with complaint of right sided ear discharge since childhood and decreased hearing. On examination we found that right ear had central perforation with intact tympanic membrane on left. Pure tone audio entry revealed conductive hearing loss of 45db in right ear. On HRCT temporal bone inner ear and mastoid system was normal with no anomaly. This patient was planned for right sided type 1 endoscopic Tympanoplasty. After freshening the margins once the tympanomeatal flap was elevated, Ossicular chain was intact but the stapedial tendon and pyramid was found absent. Also as per the Durcan., *et al*. classification of facial nerve variation in middle ear here in this case it was type 1 that is inferiorly located facial nerve. As the Ossicular chain was intact type 1 Tympanoplasty was performed in this patient.

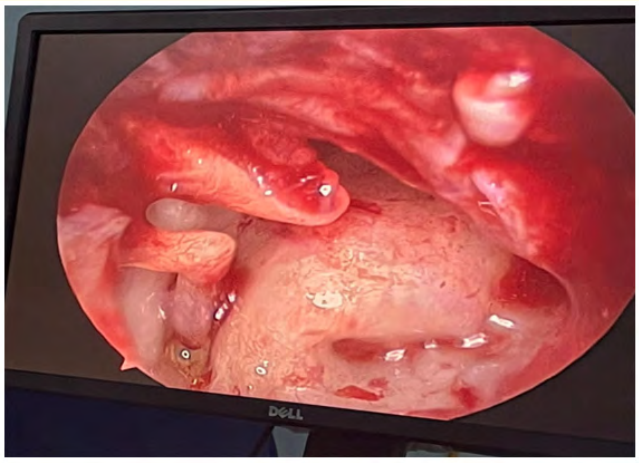

Figure 1

Discussion

The first and second branchial arches give origin to the development of Ossicular chain and their attachment [2,10,12]. Posterior belly of digastrics muscle give rise to the fascicule of stapedius muscle. The digastric muscle after reaching the eminence of the mastoid process, gives fibers of muscle into the tympanic cavity, passing through the stylomastoid opening and approaching the neck of stapes to form the stapedius muscle. Hence both muscles are being innervated by the facial nerve [4]. The stapedial reflex is important for protection against hazardous levels of noise, and for improving better speech in the presence of background noise [4,6,7,11]. A number of variations or anomalies of the stapes bone have been described [2,13]. The absence of the stapedius tendon appears to be a rare congenital malformation of the middle ear.

Hough reported five cases of total absence of the stapedius muscle in his series [8].

Magnuson and Har-El found the absence of the stapedius muscle; in their case, few tendinous fibers connected the stapes head to a well-developed pyramidal process [14].

Other reports showed that the isolated absence of the total stapedius can be considered as a relatively rare variant.

Kanona H., *et al*. 2015 [15] presented a case of Left sided progressive hearing loss in a 13-year-old girl, since 2 years her PTA: Conductive hearing loss in the left ear with PTAv at 68 dB HL. HRCT temporal bones: Suggestive of the anomalies on retrospection at surgery: No connection between stapes suprastructure and footplate, fixed footplate, absent stapedius tendon.

Congenital middle ear anomalies include isolated syndromes within the broad category of oculo-auriculo- vertebral spectrum of 1st and 2nd arch (hemifacial microsomia, branchio-otic/branchio-oto-renal spectrum disorders, Goldenhar syndrome, etc.,) [16], or might be associated with related syndromic disorders like Pierre-Robin, Treacher-Collins, Crouzon, Mobius, Fanconi, Vater and Klippel-Feil.

The second pharyngeal (hyoid) arch enlarges by the sixth intra-uterine week to overlap the third, fourth and sixth arches [17]. The Reichert's cartilage- part of the second arch mesenchyme- forms the styloid ligament, manubrium of the malleus, long process of incus, the stapes suprastructure, and the body and lesser horn of the hyoid bone. Muscular component of this arch gives rise to the muscles of facial expressions, the stapedius, stylohyoid, and posterior belly of digastric- all innervated by the facial nerve.

There are conflicting reports in literature regarding the development of stapes footplate, suprastructure and stapedius muscle and tendon. Traditionally, stapes originates from two sources- its footplate from the otic capsule (derived from neuro-ectoderm), and the suprastructure from the Reichert's cartilage mesenchyme [15].

However, recent studies contradict this hypothesis [18,19]. Both the footplate and suprastructure are now said to originate from stapes anlage, the cluster of signaled embryonic cells derived from the cranial mesenchyme of the second pharyngeal arch. The stapedius muscle/tendon unit originates from the internal part of interhyale, a localized mesenchymal condensation separating cranial mesenchyme of second arch with Reichert's cartilage [19].

According to this theory, the stapes anlage, interhyale, and the Reichert's cartilage are compartmentalized components of the second pharyngeal arch mesenchyme arranged cranio-caudally, which are anatomically linked but with distinct derivatives. The present concept can suitably explain the occurrence of congenital absence of the stapedius muscle/tendon unit by the fact that the internal part of the interhyale might be either absent or have undergone regression, independent of the stapes anlage [19].

The stapedius muscle responds primarily to high intensity and low frequency sound. When the incident sound wave reaches 20 dB above the acoustic reflex threshold (normal: 70-100 dB sound pressure level), the attenuation reflex reduces the intensity of sound transmitted to the organ of Corti by ~15 dB [20]. Therefore, the stapedius muscle/tendon unit is pivotal in maintaining the physiology of hearing and vocalization. One of the challenges in managing congenital absence of stapedius muscle/tendon unit lies in establishing the diagnosis prior to surgical intervention.

Without specialized imaging facilities and with low index of suspicion, definitive diagnosis for such rare congenital anomaly can be reached only at surgery. Interestingly, this condition might co-exist with other middle/inner ear diseases, like Tympanosclerosis CSOM (as in the present case), or Otosclerosis, as noted by Dalmia D and Behera SK [21]. There may also be other co-existing congenital disorders that may contribute to the conductive deafness. For example, absent stapedius tendon might be associated with lack of communication between the footplate and suprastructure [15]. Management in such situations should aim at addressing the associated disease(s) besides the primary anomaly.

In the present cases, the middle ear stapedius tendon with pyramidal eminence was found absent.

Conclusions

Congenital absence of stapedius muscle/tendon unit is a rare second pharyngeal arch anomaly that usually presents as non progressive hearing impairment. It is often associated with other middle/inner ear pathologies, and even with multiple congenital disorders affecting the hearing. As with our case it presented with inactive mucosal type of chronic otitis media and intact Ossicular chain patient was treated with type 1 tympanoplasty. Understanding the embryology of this anomaly is crucial in determining the pathogenesis and planning the management, and more importantly, considering it as a potential differential diagnosis of non progressive conductive deafness with intact tympanic membrane. The present clinical illustration revisits this unusual clinical entity through presentation of a new patient,

and discussion on its embryologic perspectives and management aspects in terms of diagnosis and surgical reconstruction of hearing mechanism.

Bibliography

1. Scott-Brown WG., *et al*. "Congenital middle ear abnormalities". In: Scott-Brown's Otorhinolaryngology: Head and Neck Surgery. eighth. Boca Raton: CRC Press (2019): 107.
2. Herman HK and Kimmelman CP. "Congenital anomalies limited to the middle ear". *Otolaryngology–Head and Neck Surgery* 106 (1992): 285-287.
3. Reiber ME and Schwaber MK. "Congenital absence of stapes and facial nerve dehiscence". *Otolaryngology–Head and Neck Surgery* 116 (1997): 278.
4. Williams PL., *et al*. "Gray's Anatomy". 38th ed. New York: Churchill Livingston Publishers (1995): 263, 284, 1375-1376.
5. Causse JB., *et al*. "Stapedius tendon reconstruction during stapedotomy: Technique and results". *Ear Nose and Throat Journal* 76 (1997): 256-8, 260-269.
6. Abdul-Baqi KJ. "Objective high-frequency tinnitus of middle-ear myoclonus". *Journal of Laryngology and Otology* 118 (2004): 231-233.
7. Wynsberghe DV., *et al*. "The senses". In: Human Anatomy and Physiology. 3rd ed. New York: McGraw-Hill Inc.; (1995): 512.
8. Hough JV. "Congenital malformations of the middle ear". *Archives of Otolaryngology* 78 (1963): 335-343.
9. Kurosaki Y., *et al*. "Congenital ossification of the stapedius tendon: Diagnosis with CT". *Radiology* 195 (1995): 711-714.
10. Hoshino T and Paparella MM. "Middle ear muscle anomalies". *Archives of Otolaryngology* 94 (1971): 235-239.
11. Hough JV. "Malformations and anatomical variations seen in the middle ear during the operation for mobilization of the stapes". *Laryngoscope* 68 (1958): 1337-1379.
12. Schuknecht HF and Gulya AJ. "Anatomy of the Temporal Bone with Surgical Implications". Philadelphia, PA: Lea-Febiger; (1986): 240-269.
13. Teunissen EB and Cremers WR. "Classification of congenital middle ear anomalies. Report on 144 ears". *Annals of Otology, Rhinology, and Laryngology* 102.8 Pt 1 (1993): 606-612.
14. Magnuson T and Har-El G. "Middle ear anomalies". *Otolaryngology–Head and Neck Surgery* 111 (1994): 853-854.
15. Kanona H., *et al*. "A rare stapes abnormality". *Case Reports in Otolaryngology* 2015 (2015): 387642.
16. Dutta M. "The ectopic accessory parotid system with congenital cheek fistula: An overview and current update". *Laryngoscope* 127.6 (2017): 1351-1360.
17. Patel PK., *et al*. "Head and neck embryology" (2021).
18. Rodríguez-Vázquez JF., *et al*. "Development of stapedius muscle and unilateral agenesia of the tendon of the stapedius muscle in a human fetus". *Anatomical Record (Hoboken)* 293.1 (2010): 25-31.
19. Rodríguez-Vázquez JF. "Development of the stapes and associated structures in human embryos". *Journal of Anatomy* 207 (2005): 165-177.
20. Brask T. "The noise protection effect of the stapedius reflex". *Acta Otolaryngology* 360 (1979): 116-117.
21. Dalmia D and Behera SK. Congenital absence of stapedius muscle and tendon: Rare finding in two cases". *Indian Journal of Otology* 23 (2017): 43-45.