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Case Report

Cochlear Erosion due to a Facial Nerve Schwannoma

Elke Loos, Laura Wuyts, Tony Puls, Bert De Foer, Jan W. Casselman, Anja Bernaerts, Robby Vanspauwen, Erwin Offeciers, Joost van Dinther, Andrzej Zarowski, Thomas Somers

Department of ENT-HNS, European Institute for Otorhinolaryngology-Head and Neck Surgery and Skull Base Surgery, Sint Augustinus Hospital, GZA, Antwerp, Belgium (EL, RV, EO, JvD, AZ, TS)
Department of Radiology, Sint Augustinus Hospital, GZA, Antwerp, Belgium (LW, BDF, AB)
Department of ENT-HNS, AZ Herentals, Herentals, Belgium (TP)
Department of Radiology, AZ St. Jan Hospital, Bruges, Belgium (JWC)

ORCID IDs of the authors: E.L. 0000-0003-4005-2457; L.W. 0000-0002-9219-7079; T.P. 0000-0002-8091-0826; B.D.F. 0000-0003-3594-3597; J.W.C. 0000-0002-8970-6979; A.B. 0000-0002-8652-7967; R.V. 0000-0001-8044-9358; E.O. 0000-0002-3181-9823; J.v.D. 0000-0002-5275-7852; A.Z. 0000-0002-8811-0655; T.S. 0000-0003-0739-6215.

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Facial nerve schwannomas are rare benign neoplasms. We report a case of a 60-year-old woman who initially presented with vestibular complaints. Magnetic resonance imaging (MRI) revealed a facial nerve schwannoma centered on the right geniculate ganglion extending in the labyrinthine segment. The patient consulted again after 2 months because she developed a sudden and severe right-sided sensorineural hearing loss. MRI showed no progression or pathological enhancement in the membranous labyrinth. A cone beam computed tomography (CT) of the temporal bone was performed and revealed a large erosion at the region of the geniculate ganglion in open communication with the middle turn of the cochlea. This case report demonstrates the importance of CT in facial nerve schwannomas for evaluating the impact on the surrounding structures.

KEYWORDS: Facial nerve, neuroma, sensorineural hearing loss, skull base

INTRODUCTION

Facial nerve schwannomas are rare benign neoplasms that can emerge anywhere along the course of the seventh cranial nerve. There are no large data available regarding its symptoms, imaging modalities, prognosis, and treatment because of its low prevalence. Magnetic resonance imaging (MRI) is the most frequent imaging modality used in facial nerve schwannomas. We present a case of a 60-year-old woman with a facial nerve schwannoma causing cochlear erosion, highlighting the possible cause of hearing loss in facial nerve schwannomas, and the importance of additional computed tomography (CT) for evaluating the impact on the surrounding structures.

CASE PRESENTATION

A 60-year-old woman was referred with vertigo and falling to the left side, ongoing for several months, but without hearing loss, tinnitus, or otalgia. On audiogram, a mild symmetrical high-frequency sensorineural hearing loss was revealed. Videonystagmography showed a horizontal semicircular canal areflexia on the right side. MRI of the posterior fossa (3T MAGNETOM Skyra; Siemens, Munich, Germany) was performed with a dedicated hydrops protocol to investigate endolymphatic hydrops. Although a stronger enhancement was seen in the right-sided labyrinth, the saccule, and utricle remained separated on both sides; therefore, vestibular hydrops was excluded. Surprisingly, MRI demonstrated a facial nerve schwannoma centered on the right geniculate ganglion and extending in the labyrinthine segment (Figure 1a and b). A conservative approach was adapted. The patient consulted again after 2 months. She developed a severe sudden right-sided sensorineural hearing loss with a pure tone average of 98 dB HL and no measurable bone conduction. MRI was repeated. However, there was no pathological enhancement seen in the membranous labyrinth. She was treated with intravenous corticosteroids and CO_2 inhalation, but there was no recovery of hearing loss. A cone

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Corresponding Address: Thomas Somers E-mail: Thomas.Somers@GZA.BE

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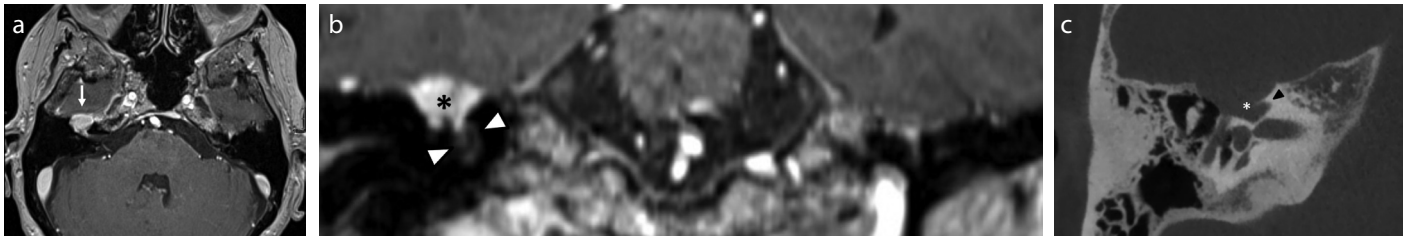

Figure 1. a-c. (a) Axial gradient echo T1-weighted image. The facial nerve schwannoma (arrow) is centered on the geniculate ganglion region on the right side with extension in the labyrinthine segment. (b) Coronal multiplanar reconstruction of a 3D gradient echo T1-weighted sequence at the level of the geniculate ganglion demonstrates the communication between the facial nerve schwannoma (asterisk) and the adjacent cochlea (arrowheads).

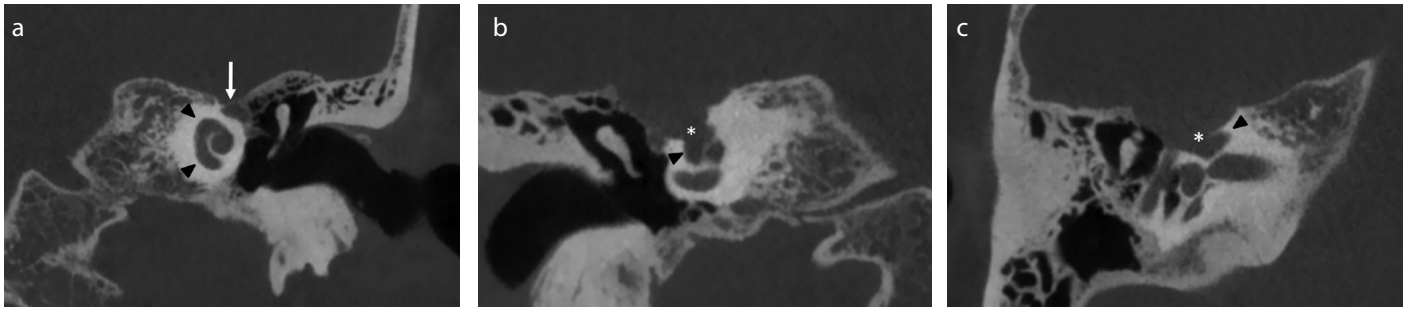

Figure 2. a-c. (a) Reformatted coronal CBCT reformation at the level of the geniculate ganglion on the left side. Note the normal size of the geniculate ganglion (arrow) and the distinctly separated cochlea (arrowheads). (b) Reformatted coronal CBCT reformation at the level of the basal turn of the cochlea on the right side. Sharply delineated erosion (asterisk) at the region of the geniculate ganglion in open communication with the middle turn of the cochlea. The arrowhead indicates the apical turn of the cochlea. (c) Reformatted axial CBCT at the level of the upper part of the second turn of the cochlea (arrowhead) on the right side, demonstrating the erosive lesion (asterisk) between the geniculate ganglion and the cochlea.

CBCT: cone beam computed tomography

beam computed tomography (CBCT) of the temporal bone (NewTom 5G XL Cone Beam CT; Verona, Italy) was performed after 1 week and revealed a large (±2.8 mm) erosion at the region of the geniculate ganglion in open communication with the middle turn of the cochlea (Figure 2a, b, and c). Consent was obtained from the patient for publication of this case study.

DISCUSSION

In this case report, we present a case of a facial nerve schwannoma on the geniculate ganglion and labyrinthine segment, with erosion of the middle turn of the cochlea, resulting in vestibular dysfunction and a severe right-sided sensorineural hearing loss.

Facial nerve schwannomas are rare benign neoplasms that can arise anywhere along the course of the seventh cranial nerve. Most previous studies describe the involvement in more than one segment [1]. The geniculate ganglion and the tympanic portion are the most involved segments. Almost half of patients with a facial nerve schwannoma present with hearing loss, whereas only one-third present with a facial nerve paralysis. Other symptoms can include tinnitus, otalgia, and vestibular dysfunction. Occasionally, otoscopy can reveal a mass behind the eardrum [2].

Sensorineural hearing loss may be caused by cochlear erosion (labyrinthine or tympanic segments) or compression of the cochlear nerve within the internal auditory canal or brainstem (intracranial or canalicular segments). Another possible cause of sensorineural hearing loss could be through the ototoxic effect of secreted factors that could cause spiral ganglion and neuron degeneration, as well as hair cell loss [1].

This case study illustrates the need for careful evaluation of the adjacent bony structures on CT, complementary to the findings on MRI. In facial nerve schwannomas, MRI demonstrates a fusiform, solid mass with, when they are large, possible intramural cystic changes. Schwannomas demonstrate an intermediate to low signal on T1-weighted images and are hyperintense on T2-weighted images. They show no diffusion restriction on diffusion-weighted images.

The radiological appearance of schwannomas on CT comprises fusiform expansion of the facial nerve canal with well-circumscribed and smooth margins due to long-standing bony compression [3]. This differentiates them from hemangioma, which has a characteristic amorphous honeycomb appearance and/or internal bony spicules with more aggressive bony changes, seen as irregular margins at CT imaging.

Schwannomas on the geniculate ganglion often extend into the adjacent labyrinthine and anterior tympanic segments and can extend in the middle cranial fossa. Large, schwannomas of the perigeniculate segment and tympanic segment may cause erosion of the petrous bone, cochlea, semicircular canals (lateral), and ossicles [4, 5]. A facial nerve schwannoma of the geniculate ganglion with erosion of the cochlea can be mistaken for an intralabyrinthine schwannoma. However, intralabyrinthine schwannomas do not extend beyond or destroy the bony wall of the cochlea.

CONCLUSION

This case study demonstrates the importance of CT in facial nerve schwannomas for evaluating the impact on the surrounding structures and the differentiation with a hemangioma. MRI remains the most important imaging technique for diagnosing facial nerve schwannomas, but high-resolution CT of the temporal bone best visualizes the course of the nerve within the fallopian canal to the stylomastoid foramen and can be necessary to evaluate bony erosion.

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REFERENCES

1. Remenschneider AK, Gaudin R, Kozin ED, Ishai R, Quatela O, Hadlock TA, et al. Is the Cause of Sensorineural Hearing Loss in Patients With Facial Schwannomas Multifactorial? Laryngoscope 2017; 27: 1676-82. [CrossRef]
2. Wiggins RH 3rd, Harnsberger HR, Salzman KL, Shelton C, Kertesz TR, Glastonbury CM. The Many Faces of Facial Nerve Schwannoma. AJNR Am J Neuroradio 2006; 27: 694-9.
3. Mundada P, Purohit BS, Kumar TS, Tan TY. Imaging of facial nerve schwannomas: diagnostic pearls and potential pitfalls. Diagn Interv Radiol 2016; 22: 40-6. [CrossRef]
4. Chung SY, Kim DI, Lee BH, Yoon PH, Jeon P, Chung TS. Facial nerve schwannomas: CT and MR findings. Yonsei Med J 1998; 39: 148-53. [CrossRef]
5 De Foer B, Nicolay S, Vercruysse JP, et al. Imaging of cholesteatoma. In: Lemmerling M, De Foer B, eds. Temporal bone imaging. New York: Springer Verlag, Berlin, Heidelberg; 2015: 69-87. [CrossRef]