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Case Report

"Canalolabyrinthine Schwannoma," A Rare Variant of Intralabyrinthine Schwannoma: A Case Report

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Canalolabyrinthine schwannoma is a rare subtype of neuroma. Only 1 case has been described in the literature. We report the clinical case of a 51-year-old Caucasian female with Meniere's disease-like symptoms. Magnetic resonance imaging showed right VIII cranial nerve schwannoma that had different characteristics from the classical described types. The peculiar features of our case of intralabyrithine canalolabyrinthine schwannoma directed the surgeon and radiologist to tailored considerations for follow-up and therapy.

KEYWORDS: Schwannoma, intralabyrinthine, vertigo, magnetic resonance imaging

INTRODUCTION

Schwannoma is the most common benign neoplasm affecting the internal auditory canal (IAC) and pontocerebellar angle, and it accounts for up 6% of all intracranial tumors [1, 2]. Intralabyrinthine schwannoma (ILS) is a subtype of schwannoma that originates from the perineural Schwann cells of the vestibulocochlear nerve proximal to the membranous labyrinth (cochlea and vestibule), without any outer extension [3]. The prevalence of this disease is still a matter of debate: only 1 of 893 cases has been described in an autopsy study (prevalence of 0.1%) [4]. Further, only 3 of 800 patients who had suggestive symptoms of Meniere's disease and who underwent magnetic resonance imaging (MRI) were found to have ILS (prevalence of 0.4%) [5]. Salzman et al. [6] analyzed 45 cases of ILS confirmed by MRI that were collected over 14 years. Interestingly, they showed that the diagnosis of ILS diagnosis is actually more common than that thought previously. These authors also proposed a classification of the disease on the basis of the anatomical sites affected by the tumor. In particular, they defined the neoplasm as follows:

- Intracochlear schwannoma when it was confined to the loops of the cochlea.
- Intravestibular schwannoma when it was confined to the vestibule with or without extension in the semicircular canals.
- Vestibulocochlear schwannoma when it was extending into the cochlea and was sparing the middle ear and ear canal.
- Transmodiolar schwannoma when it was extending through the modiolus from the cochlea in the inner ear canal through the cochlear nerve.
- Transmacular schwannoma when it was extending from the vestibule to IAC through the lamina cribrosa.
- Transotic schwannoma, when it was extending into the posterior labyrinth, IAC, and middle ear.

Here we describe a case of ILS involving the cochlea, vestibule, and IAC. To the best of our knowledge, this is the second case described in the literature [7].

CASE PRESENTATION

We report a case of a 51-year-old Caucasian female with a history of transmodiolar ILS characterized by fullness sensation in her right ear.

At her 1-year follow-up examination, the patient complained of right subcontinuous tinnitus and rapidly evolving decreased hearing. Four years later, she experienced multiple episodes of objective vertigo associated with ocular nystagmus, neurovegetative symptoms, and loss of consciousness, without an increase in tinnitus. She was treated with 24 mg bid betahistidine; however, this was not effective.

After 1 year from the onset of vertigo, the symptoms of imbalance disappeared.

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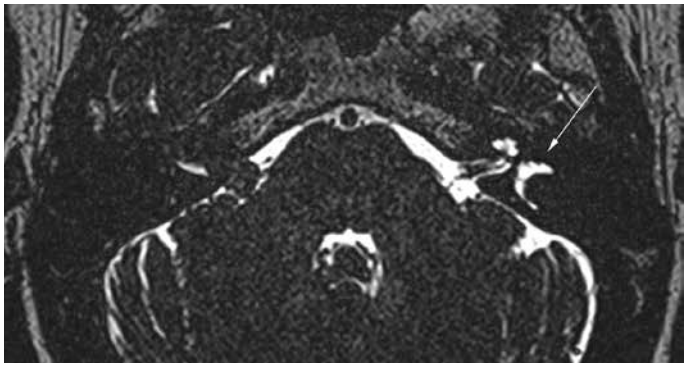

Figure 1. Axial and axial maximum intensity projection (MIP) images obtained with T2-weighted sequences. The characteristic high signal in the left labyrinth (white arrow) is observed. There is no evidence of signal at the site of schwannoma.

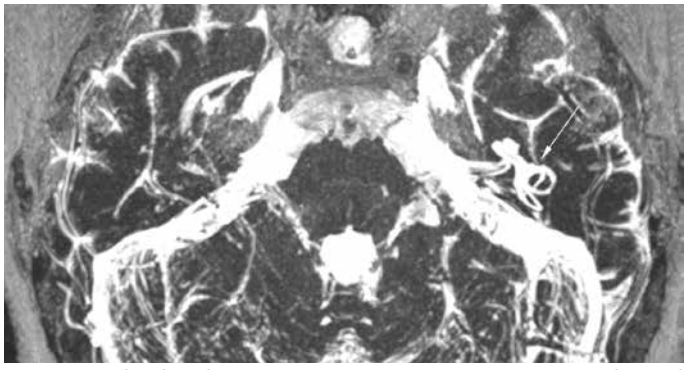

Figure 2. Axial and axial maximum intensity projection (MIP) images obtained with T2-weighted sequences. The characteristic high signal in the left labyrinth (white arrow) is observed. There is no evidence of signal at the site of schwannoma.

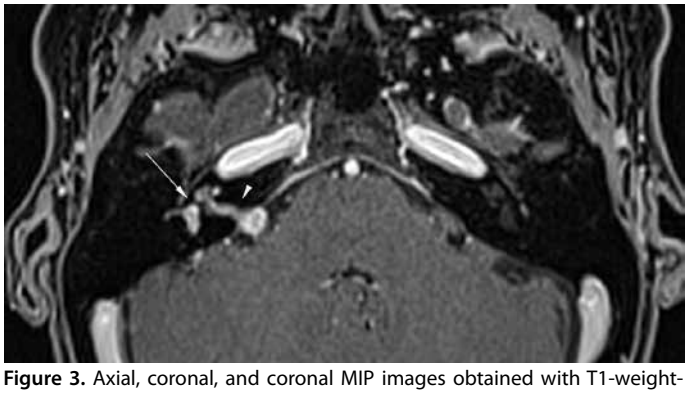

Figure 3. Axial, coronal, and coronal MIP images obtained with T1-weighted sequences with a paramagnetic contrast medium show high signal in the right labyrinth, i.e., the site of schwannoma (white arrow), with an extension in IAC (arrowhead). There is no evidence of signal in the right middle ear. On the controlateral side, there is no signal enhancement in any ear structure.

Otoscopic examination showed normal tympanic membrane anatomy, whereas pure-tone audiometry demonstrated anacusia of the right ear and normoacusia of the left ear. Tympanogram showed a normal type A pattern.

Gadolinium-enhanced MRI revealed a schwannoma affecting the right VIII cranial nerve with the epicenter in the pontocerebellar angle and intracanal extension. In addition, IAC was mildly dilated compared with the left side. The lesion had a transverse diameter of 13 mm and antero–posterior diameter of 7 mm. Moreover, the neoplasm was associated with asymmetrical signal intensity of the inner ear and particularly with a hypointense signal in T2-weighted images (Figures 1, 2) and with gadolinium-based contrast agent (GBCA) enhancement in T1-weighted images (Figures 3-5).

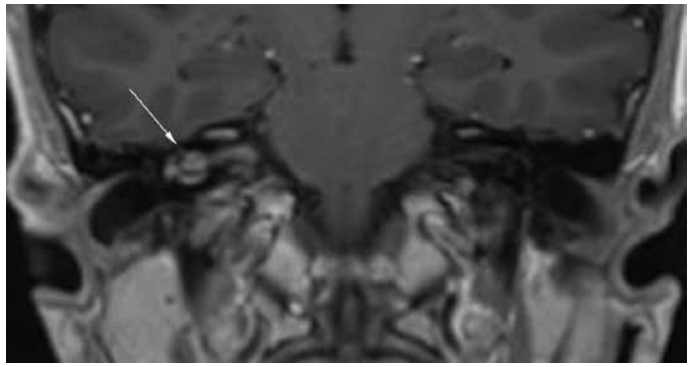

Figure 4. Axial, coronal, and coronal MIP images obtained with T1-weighted sequences with a paramagnetic contrast medium show high signal in the right labyrinth, i.e., the site of schwannoma (white arrow), with an extension in IAC (arrowhead). There is no evidence of signal in the right middle ear. On the controlateral side, there is no signal enhancement in any ear structure.

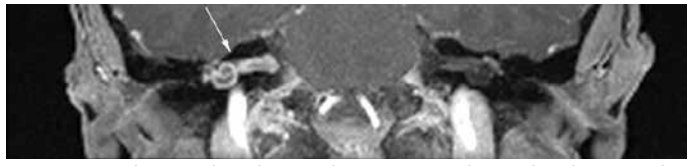

Figure 5. Axial, coronal, and coronal MIP images obtained with T1-weighted sequences with a paramagnetic contrast medium show high signal in the right labyrinth, i.e., the site of schwannoma (white arrow), with an extension in IAC (arrowhead). There is no evidence of signal in the right middle ear. On the controlateral side, there is no signal enhancement in any ear structure.

There were no signs of compression on the brain stem at the root entry and exit zones of the VIII cranial nerve and no contrast enhancement of the middle ear bilaterally.

The left facial–stato–acoustic fascium and left cochleo–vestibular structure showed a normal morphology and signal intensity.

The diagnosis of canalolabyrinthine schwannoma was made considering all these features.

DISCUSSION

We present a rare case of canalolabyrinthine schwannoma.

Magnetic resonance imaging has revolutionized the diagnosis and treatment modalities of schwannoma [8]. It enables easy identification of the canalolabyrinthine subtype by high-resolution imaging. Typically, canalolabyrinthine schwannoma does not appear as a high-intensity signal on T2-weighted images, while labyrinthine fluid appears hyperintense. Conversely, the lesion becomes slightly more intense than the normal fluid in unhenanced T1-weighted images [9]. After GBCA injection, however, homogeneous contrast enhancement of the labyrinthine structures is observed [6].

Magnetic resonance imaging also enables the differential diagnosis between schwannoma and labyrinthitis [10]. Acute labyrinthitis is characterized by less pronounced enhancement that gradually decreases and disappears during follow-up. Conversely, schwannoma

is associated with persistent enhancement that does not change or that increases during follow-up. In subacute and chronic labyrinthitis, calcification or fibrosis often replaces the labyrinth fluid, but the edges of the lesion are not well defined [9].

Serial MRI detects any growth of the neoplasm (even if rare) and represents the key step in the management of schwannoma according to Kennedy et al. [1]. In our case, serial MRI revealed no significant growth over 3 years of follow-up. Surgery is advisable in case of tumor growth as well as in case of ILS that develops inside the internal ear canal (transmacular, transmodiolar, or transotic) if patients experience intractable vertigo symptoms [6]. The transotic approach would be preferred because of the implication of cochlea. Vestibular nerve section is deemed unnecessary because of the absence of vestibular symptoms [1].

CONCLUSION

Our case of canalolabyrinthine schwannoma does not fit any classification. Clinical presentation may pose challenges in the differential diagnosis of canalolabyrinthine schwannoma from Meniere's disease because of the similarity of symptoms of these two conditions. Magnetic resonance imaging enables early and accurate staging of canalolabyrinthine schwannoma and aids surgeons in making treatment decisions.

Informed Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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