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Inner ear lipoma

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ABSTRACT

Intracranial lipomas are uncommon congenital malformations that are often asymptomatic. They are very rarely seen in the inner ear. There are a few case reports in the literature related to intravestibular lipoma. It was also defined in the internal auditory canal and the cerebellopontine angle. We present here a case of an inner ear lipoma that diffusely infiltrated the cochlea, vestibule and the semicircular canals. To our knowledge, this is the first reported case in the literature.

Key words: • *lipoma* • *inner ear* • *computed tomography* • *magnetic resonance imaging*

Intracranial lipomas are uncommon congenital malformations that are often asymptomatic. They are very rarely seen in the inner ear. There are a few case reports in the literature related to intravestibular lipoma (1, 2). We present a case of an inner ear lipoma that diffusely infiltrated the cochlea, vestibule and the semicircular canals. To our knowledge, this is the first reported case in the literature.

Case report

A 30-year-old male presented with a right-sided sensorineural hearing loss that developed recently. Hearing on the left was normal. He complained of intermittent tinnitus. Magnetic resonance imaging (MRI) (1.5 Tesla, Philips Gyroscan Intera, Best, The Netherlands) showed that the right inner ear structures were completely hyperintense on T1-weighted images and contained hypo- and hyperintense areas on T2-weighted images. Fat-saturated T1-weighted images revealed complete saturation of the hyperintensities. No enhancement within these structures was identified on the post-contrast images (Fig. 1). Computed tomography (CT) (Philips Mx 8000) revealed hypoattenuated masses in the vestibule, cochlea, and the semicircular canals. Hounsfield attenuation units on CT images were consistent with fat, measuring -120, -110, and -117 for the vestibule, cochlea, and the semicircular canals, respectively (Fig. 2). The lesion was diagnosed as an inner ear lipoma.

Discussion

Current theory explaining the pathogenesis of intracranial lipomas involves persistence and maldifferentiation of the primordial meninx primitiva, the loose embryonic mesenchymal precursor of the subarachnoid space and meninges (3). Cavitation of the meninx primitiva, occurs simultaneously with the formation of the inner ear in the 4- to 6-week embryo (4, 5). Direct incorporation of the meninx primitiva or its mesenchymal precursor, into the developing otocyst (auditory vesicle), as it invaginates or folds, could explain the intravestibular, intracochlear and intracanalicular location of lipomas. One case of intravestibular lipoma, in an intramembranous location, was described in the literature (1). Our case is the first one involving the whole structures of the inner ear (i.e., the cochlea, vestibule and semicircular canals). Lipomas may also be located in the internal auditory canal and infiltrate canalicular portions of the eighth cranial nerve. The tumor tends to adhere to the eighth nerve and often completely surrounds it. These strong adhesions to the eighth nerve and, to a lesser extent, to the seventh nerve and the surrounding tissues render surgery more difficult (2, 6). Infiltrative nature of the adipose tissue into the nerve is unique to lipomas of the cerebellopontine angle and the internal auditory canal as opposed to other intracranial lipomas (7). As in our case, surgical

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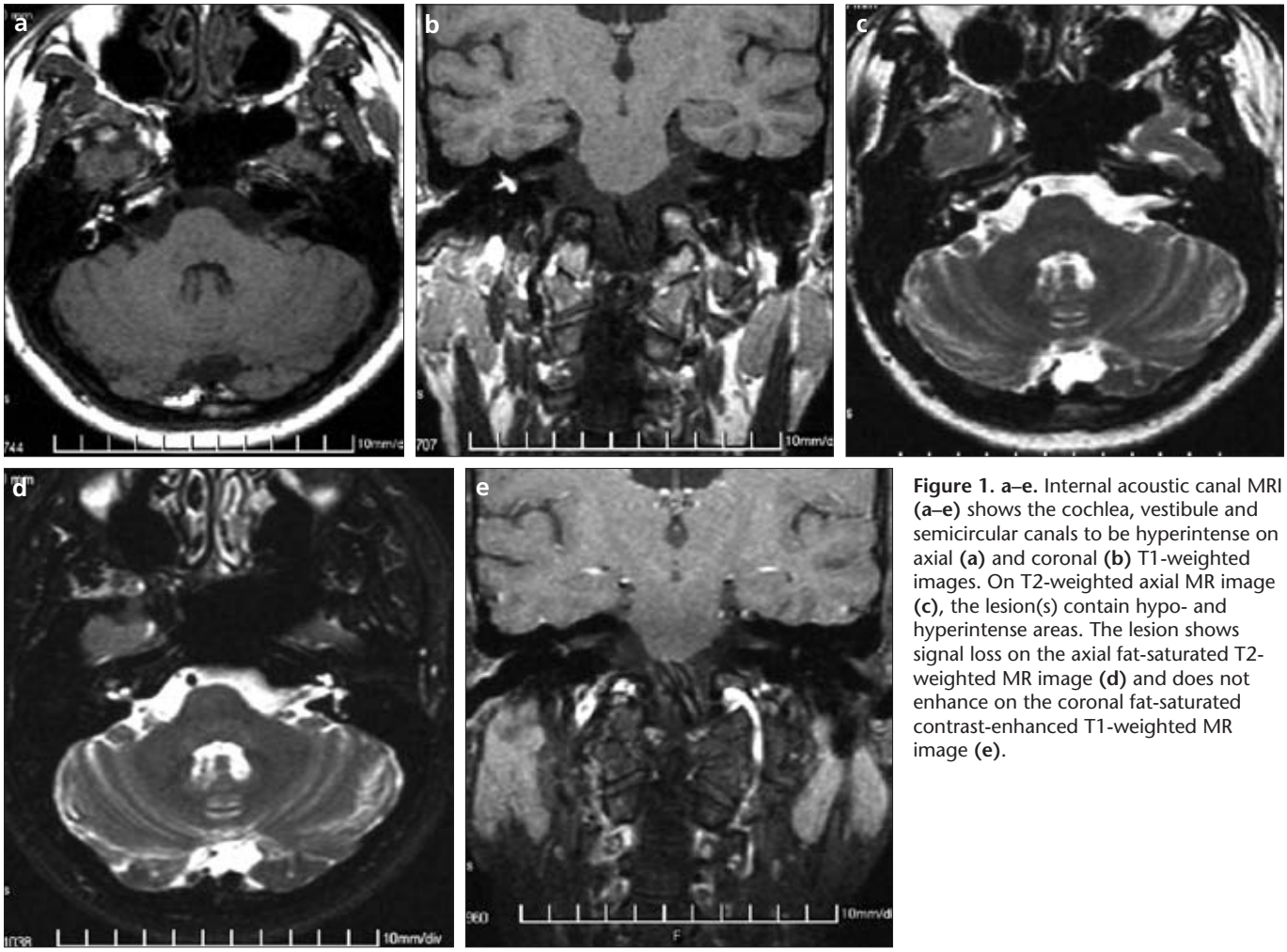

Figure 1. a–e. Internal acoustic canal MRI **(a–e)** shows the cochlea, vestibule and semicircular canals to be hyperintense on axial **(a)** and coronal **(b)** T1-weighted images. On T2-weighted axial MR image **(c)**, the lesion(s) contain hypo- and hyperintense areas. The lesion shows signal loss on the axial fat-saturated T2-weighted MR image **(d)** and does not enhance on the coronal fat-saturated contrast-enhanced T1-weighted MR image **(e)**.

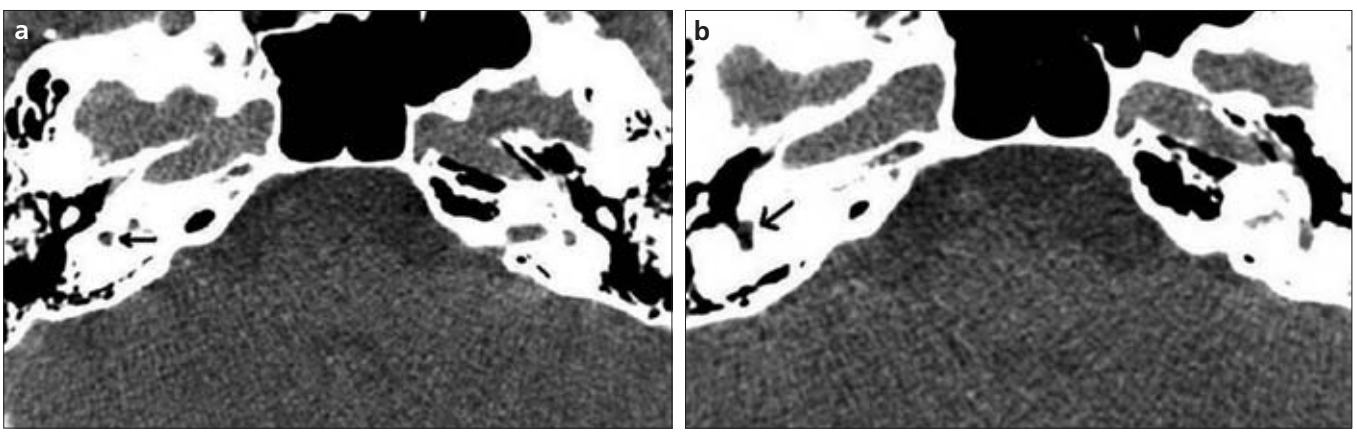

Figure 2. a, b. Internal acoustic canal CT images show hypoattenuated areas in the right semicircular canals *(arrows)*.

intervention in patients with an inner ear lipoma should be avoided.

MRI and CT findings are essential in evaluating an inner ear lipoma and making correct diagnosis. Imaging of lipoma features hyperintensity on T1-weighted MR images. Signal intensity on T2-weighted MR images is variable and suppressed completely with fat saturation. Alternatively, Hounsfield attenuation units consistent with fat are also diagnostic on CT. Lipomas are nonenhancing lesions. Additional sources for hyperintensity within the vestibule on T1-weighted MR images include hemorrhage or highly proteinaceous fluid. These are easily differentiated from a lipoma because of their lack of signal intensity suppression with fat saturation (8). Our patient had a hyperintense cochlea, vestibule, and lateral, posterior and superior semicircular canals on T1-weighted MR images. Because all of them showed hypointen-

sity on T1-weighted MR images with fat saturation, we thought that these lesions represented a lipoma.

Our patient had a right-sided sensorineural hearing loss. The hearing loss was recently recognized and it correlated with the side of lipoma. In general, intracranial lipomas are asymptomatic, whereas lipomas of the inner ear are associated with hearing loss of the same side (6). Mass effect from the lipoma or chemical toxicity to the cochlea may cause hearing loss. According to the literature, a lipoma within the membranous labyrinth may be related to a toxic cause of sensorineural hearing loss. Because lipid materials or breakdown products could be transported via the membranous labyrinth to the cochlea, they then make a toxic effect on the cochlear hair cells. This could also explain the high-frequency nature of the hearing loss in that the hair cells along the basal turn would be most affected because of reflux of the toxic material through the ductus reuniens. Huang reported a patient with intravestibular lipoma who had vertigo, daily nausea and vomiting, tinnitus, ear fullness, and complete hearing loss in the affected ear (1). These findings are related to endolymphatic hydrops, which might have been due to the mass effect from the lipoma or to an unrelated history of mumps labyrinthitis in the reported patient (1). Our patient, on the other hand, had only tinnitus as a vestibular symptom although the vestibule and semicircular canals were diffusely infiltrated by the tumor.

In conclusion, inner ear lipoma is a very rare pathology that has characteristic features on CT and MRI, and it must be taken into consideration in the differential diagnosis of inner ear pathologies.

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