

Caso Clínico / Radiological Case Report

EXTENSIVE INNER EAR AND FACIAL CANAL LIPOMA - A CASE REPORT*EXTENSO LIPOMA DO OUVIDO INTERNO E CANAL DO FACIAL – CASO CLÍNICO*Mariana Cardoso Diogo¹, Amets Sagarrabay², Carla Conceição¹

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Abstract

Inner ear lipomas are rare benign congenital lesions, most frequently presenting at the internal auditory canal and labyrinth. Lipomas of the facial nerve are even less frequent. We present a case of a lipoma involving all internal ear structures and VII nerve canal and extending intracranially and extracranially, presenting with facial palsy and ipsilateral hearing loss. To our knowledge this is the most extensive case of inner ear lipoma described.

Key-words

Inner ear lipoma; Inner ear lipochoristoma; Facial nerve lipoma; Sensory hearing loss; Facial nerve palsy.

Resumo

Os lipomas do ouvido interno são lesões congénitas benignas raras, localizadas mais frequentemente no canal auditivo interno. Lipomas do nervo facial são ainda mais raros. Apresentamos um caso de lipoma a envolver a totalidade das estruturas do ouvido interno, canal do nervo facial e com extensão intra- e extracraniana, manifestando-se com paresia facial e hipoacusia ipsilaterais. Tanto quanto nos foi possível averiguar, este é o caso mais extenso de lipoma do ouvido interno descrito na literatura.

Palavras-chave

Lipoma do ouvido interno; Coristoma do ouvido interno; Lipoma do nervo facial; Hipoacusia neuro-sensorial; Paresia facial

Introduction

Inner ear lipomas are rare benign congenital lesions, believed to derive from the persistence and maldifferentiation of the mesenchymal precursor of the meninges (mininx primitiva) that becomes trapped inside the otic capsule during early embryonic development¹. In most cases they present extension along the vestibulocochlear nerve to the internal auditory canal, with some presenting with cerebellopontine angle (CPA) masses. Lipomas of the facial nerve are even less frequent with only a few cases published. We present a case of a lipoma involving all internal ear structures and VII nerve canal and extending intracranially and extracranially. To our knowledge this is the most extensive case of inner ear lipoma described. Institutional board review was waived for a single retrospective case report.

Clinical Case

A five-month-old female patient was referred to our center due to congenital left peripheral facial nerve palsy and ipsilateral moderate to severe sensorineural hearing loss (SNHL). She had passed the newborn hearing screening test. Her development was otherwise unremarkable for her age and she presented no other neurological deficits. Computed tomography (CT) was initially performed, followed by magnetic resonance imaging (MRI). On CT a dilatation of the facial nerve canal, particularly of the geniculate ganglion region and the third (mastoid) portion was identified. The internal auditory canal (IAC) was mildly abnormal (Figure 1B), with a large falciform crest. Round well-marginated low-density masses were identified emerging from the porus acusticus into the CPA, and in the temporal fossa (figure 2), adjacent to the dehiscent geniculate ganglion region, and also extracranially, protruding from the stylomastoid canal (Figure 3). A small mass could also be identified protruding in the posterior

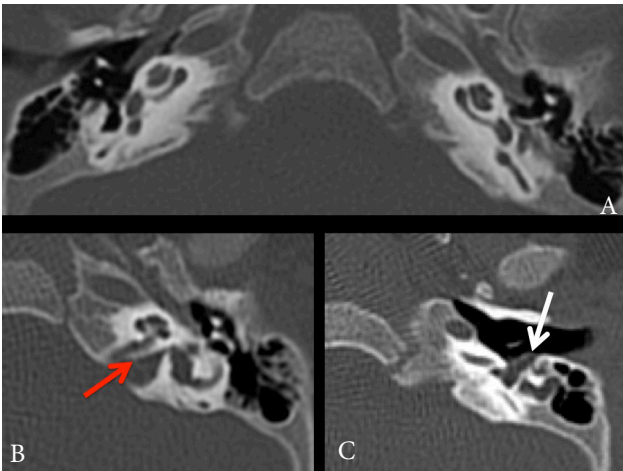


Figure 1 – Axial CT scan, bone window, at the level of the cochleae (A), depicting a relatively normal and symmetrical anatomy; left ear, at the level of the internal ear canal (B), showing an enlarged canal and the prominent falciform crest (red arrow); and at the level of the epitympanum (C) depicting the lipomatous mass extending into the posterior middle ear (white arrow).

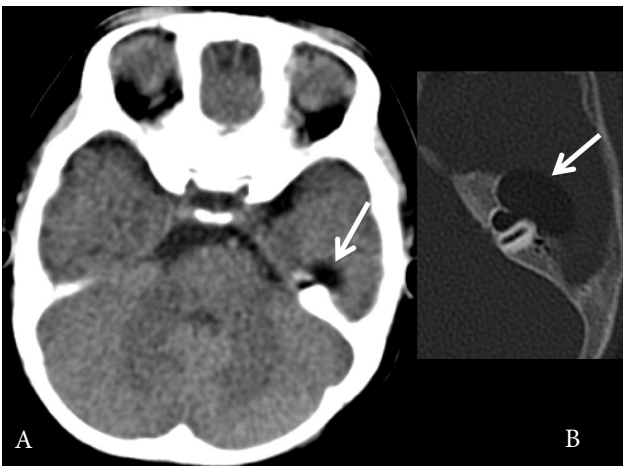


Figure 2 - CT scan performed at 5 months, axial scans in soft tissue (A) and bone (B) windows depicting the temporal fossa mass (white arrow), extending directly from the inner ear structures. Notice the hypodensity corresponding to fat density.

wall of the tympanic cavity (Figure 1C). An abnormal density (-112 HU) was identifiable in the vestibule, facial canal and IAC, in apparent continuity with the intracranial and extracranial masses. The lesion was better depicted on MRI (Figures 3, 4), and shown to further involve the cochlea and semicircular canals. It presented high signal intensity (SI) on T1 and T2-weighted images, without enhancement after gadolinium injection. A lipomatous lesion was suspected, but given the exuberance and size of the mass, a neoplastic lesion could not be completely excluded and the middle fossa component was surgically excised. Histopathological analysis was compatible with a lipoma.

Follow-up exams were performed annually for two years and biannually thereafter. The lesion remained imagiologic and clinically stable for over 6 years, with the patient maintaining a normal development. Given this stability, she currently has no indication for further imaging studies in the absence of new symptoms.

Discussion

We present the CT and MR imaging findings in a child with a lipoma involving the IAC, inner ear structures and facial

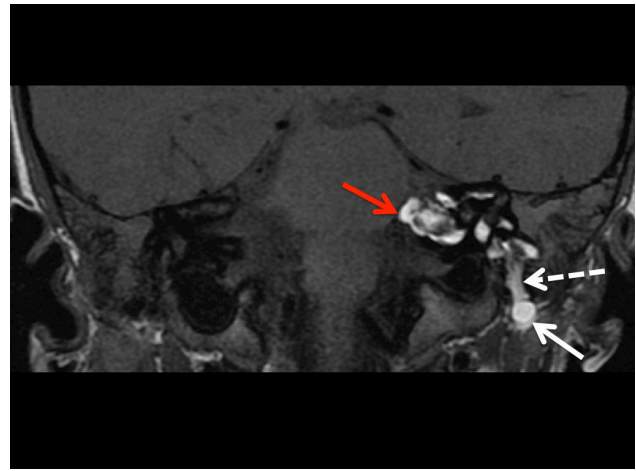


Figure 3 – Coronal T1 image showing the extension into the cerebellopontine angle (red arrow), the 3rd mastoid portion of the facial canal (dashed arrow), extending to the extracranial space inferiorly (white arrow)

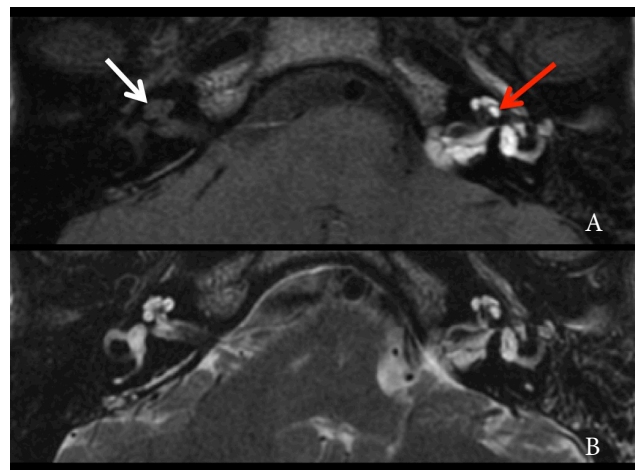


Figure 4 – Axial MRI images at the level of the cochleae. On T1-weighted image (A) the high signal intensity of all inner ear structures (red arrow) can be identified and is very distinct from the contralateral (normal) side (white arrow). On the T2-weighted image (B) signal intensity of fat is similar to the endolymph and perilymph and the abnormalities are not as clear.

canal, protruding intracranially to the CPA and middle cranial fossa, and extracranially through the stylomastoid foramen. Inner ear lipoma is a rare congenital lesion, often associated with a simultaneous lipoma in the cerebellopontine angle². They are believed to arise from the persistence of the mininx primitiva, with its incorporation into the developing otocyst, as it invaginates or folds in the early stages of embryonic development¹⁻³.

On CT lipomas have a density in the range of fat (around -100 HU). They may enlarge and slightly remodel the bone, but show no infiltrative or aggressive characteristics, and clear cortical margins of surrounding bone should be seen at all times. On MR they present as non-enhancing lesions that follow the signal intensity of fat tissue in all sequences, suppressing completely with fat saturation. In the inner ear, they might be overlooked on T2W images as their typical high signal can mimic that of endolymph/perilymph (Figure 4B). In smaller (focal) inner ear lipomas main differential diagnosis are intralabyrinthine schwannomas, that may also present in the CPA – it is essential to perform T1 sequences before gadolinium, to avoid mistaking T1 high signal intensity (SI) with contrast enhancement. A more diffuse T1 high SI

of the inner ear structures can also be present in cases of haemorrhage or very high protein content. FS sequences are essential in these cases.

In contrast to intracranial lipomas, which are usually asymptomatic, most patients with inner ear lesions present with SNHL¹. Facial palsy is observed in only 5% of patients². The exact etiology of the SNHL cannot be entirely explained by the mass effect of the tumor on the VIII nerve, as a significant percentage of patients develop progressive hearing loss after birth, while lipoma size remains stable. It has been proposed that this may be secondary to toxic effect on cochlear hair cells of lipid material breakdown in the intralabyrinthine space¹. Our patient presented with peripheral facial palsy and progressive SNHL, probably of mixed origin - nerve compression and infiltration and cochlear dysfunction. She exhibited no signs of vestibular dysfunction despite the semicircular canals involvement.

These lesions remain stable throughout life and any change in clinical complaints or imaging characteristics should warrant an adequate investigation. Otherwise, a follow up exam one year after diagnosis, to ensure that there isn't significant change in size, appears to suffice⁴. There is no described advantage in long term radiological follow up.

References

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Surgical intervention in patients with intravestibular lipoma is associated with high morbidity and should be avoided, making correct imaging diagnosis paramount^{3,4}.

We believe that the intracranial extensions observed in our patient's case are part of the same lesion, in continuity with the VII and VIII cranial nerves through the porus acusticus into the CPA, from the geniculate ganglion following the GSPN as it enters the middle cranial fossa, and exiting the skull base inferiorly with the facial nerve through the stylomastoid foramen. The imaging characteristics are undoubtedly of an adipose mass and the stability of the lesion attests to its benignity. The histopathological results confirm the diagnosis.

To our knowledge there is only one other case of a lipoma involving all structures of the inner ear reported in English literature⁵. Our report further adds involvement of the facial nerve canal in its entirety and of the IAC.

In conclusion, inner ear lipoma is a very rare pathology, with typical density and signal characteristics on both CT and MRI. Knowledge of this entity is essential to avoid unnecessary invasive procedures or imaging follow-ups, even when lesions are imagiologically exuberant, such as in the case presented.

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