

Otolaryngology

**EXOSTOSIS OF INTERNAL AUDITORY CANAL: CASE REPORT** 

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| ABSTRACT In contrast to the relatively common finding of temporal bone exostoses in the external auditory canal, the finding of such formations in the internal auditory canal is extremely area. This report deals with a formation addressent 17 years addressent 17 yea |   |

**ADSTRACT** formations in the internal auditory canal is extremely rare. This report deals with a female adolescent, 17 years of evolving with rapidly progressive bilateral hearing loss 2 years ago, with no associated abnormality.

KEYWORDS : Sensorineural, hearing loss, exostosis, internal auditory canal.

## Introduction

Exostoses are sessile lamellar growths, commonly multiple and bilateral, almost always in a medial position in the external auditory canal, being the most common bone alteration of the temporal bone<sup>1</sup>. It is frequently found in cold water athletes, with chronic exposure to water being their main risk factor<sup>2</sup>. The appearance of exostosis in the internal auditory canal is extremely rare and of unknown etiology, and only 8 cases are described by the year 2015<sup>3,4</sup>. The rarity and gravity of the case prompted the authors to publish this report.

# **Case Report**

# - History

A 17-year-old female patient presented progressive bilateral hearing loss 2 years ago, with no triggering factors and no association with any other audiological, vestibular, or neurological symptomatology. He denied comorbidities. There was no previous history of ototoxic use, otological diseases, meningitis, exposure to noise, acoustic trauma or family history of deafness.

### - Examination

Otorhinolaryngological and neurological exams were normal. Rinne negative test bilaterally and Weber test without lateralization. The tonal audiometry showed a lowering of the audiometric thresholds of sensorineural characteristic from the frequency of 500 Hz bilaterally, with progressive worsening in the subsequent frequencies, reaching deep loss at 2000 Hz and undetectable threshold starting at 4000 Hz. Vocal discrimination for both monosyllables and disyllables was very poor in both ears. The tympanometry was normal bilaterally, with curve A, and absence of stapedial reflexes. Laboratory tests including complete blood count, thyroid function, rheumatoid factor, antinuclear factor were normal. Computed tomography of the ears and mastoids (Image 1) showed bone density lesions in the two internal, nonpedicled, broad-based, non-pedicular auditory conduits with significant narrowing of both ducts. The nuclear magnetic resonance of the inner ears and the angle-blunt angle (Image 2) corroborated the tomographic findings, with significant extrinsic compression of the neural structures of the ducts bilaterally.





## Image 2

#### - Follow-up

It was opted for clinical follow-up of the patient, with expectant management. There was no interest on the part of the patient to perform any surgical intervention, and she was referred for auditory rehabilitation through a bilateral sound amplification apparatus.

#### Discussion

In 1984, Smelt was the first to report a case of internal auditory canal exostosis in a necropsy finding of an asymptomatic patient<sup>3</sup>. Since then, another 7 cases have been published and are available for analysis. Exostosis of the internal auditory canal is a rare entity with unknown etiopathogenesis characterized by progressive loss of hearing due to extrinsic bone compression of the eighth pair, usually bilateral, and may be associated with tinnitus, dizziness or even asymptomatic or incidental finding<sup>4</sup>.

In most cases, the diagnosis was presumed by means of clinical, tomographic and nuclear magnetic resonance findings. Otorhinolaryngological and neurological examinations are normal, and the audiogram, in general, shows lowering of the tonal thresholds to varying degrees, with a sensorineural component<sup>6</sup>. Computed tomography evidences bone growths, uni or bilateral, of a regular, sessile, broad-based feature with no bone marrow content, resulting in stenosis and compression of the neural structures of the conduit. Magnetic resonance imaging in T1 presents a hypointense lesion pronounced in the conduit, in contact with the neural pathway, and in T2 it shows very low signal intensity, compatible with bone content or calcification<sup>7</sup>.

The histopathological diagnosis is difficult to perform, given the difficulty and high risk of biopsy of the internal auditory canal<sup>3</sup>. Only two cases of this disease had histopathological confirmation because

41

they underwent surgery<sup>4</sup>. It presents as parallel concentric layers of subperiosteal bone, without fibrovascular channels and with abundant osteocytes, with absence of bone marrow, unlike osteomas<sup>1</sup>.

Differential diagnosis is made with intracanalicular vestibular swastioma (bilateral cases more frequent in patients with type II neurofibromatosis), osteoma, Paget's disease, fibrous dysplasia, malformation, otosclerosis and vitamin A deficiency<sup>3,5</sup>. In practice, the imaging study by CT and MRI has made the diagnosis due to the difficulty of performing a biopsy.

Treatment should be individualized. In conditions of severe hearing loss or intense dizziness, one may opt for surgical intervention, as has been done in cases of bilateral osteomas<sup>8</sup>. Most of the cases have been managed clinically, with auditory rehabilitation, use of medications (corticosteroids) or surgery<sup>4,9,10</sup>. Doan *et al* reported a case submitted to surgery by posterior fossa access in a patient with deep unilateral hearing loss in the right ear, with good postoperative evolution after decompression of the eighth pair by means of the removal of bone excess with drill<sup>9</sup>. Baik *et al* submitted a patient with disabling vertigo and bilateral exostosis to surgery via unilateral retrossigmoid craniotomy, with an important improvement in the condition<sup>10</sup>. The other cases were conducted in an expectant manner, based on clinical data and patient choice<sup>4</sup>, as in this case reported here.

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