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Case Report

Extradural petromastoid calcifying pseudoneoplasm of the neuraxis (CAPNON): Case report and literature review

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ABSTRACT

Background: Calcifying pseudoneoplasm of the neuraxis (CAPNON) is a very rare tumor with fewer than 70 cases reported in the literature. In general, this tumor occurs intracranially either within the brain parenchyma or in an extra-axial location, but it has also been described within the spine as an extra-axial lesion.

Case Description: We describe an unusual case of intracranial-extradural CAPNON involving the mastoid region. This may be only the second such case reported in the literature, as one patient with CAPNON has been reported involving the sinonasal region. Our patient was managed with surgical resection through a translabyrinthine approach with good early result.

Conclusions: We describe an unusual case of extradural CAPNON involving the mastoid bone. It appears that when located extradurally, this tumor may have a predilection for the bony sinuses. This little-known, generally benign entity can mimic more common lesions such as meningiomas, and should be considered in the differential diagnosis of skull base tumors, particularly when associated with heavy calcification.

1. Introduction

Calcifying pseudoneoplasm of the neuraxis (CAPNON) is a very rare tumor with fewer than 70 cases reported in the literature [1–3]. In general, this tumor occurs intracranially either within the brain parenchyma or in an extra-axial location, but it has also been described within the spine as an extra-axial lesion [1,4]. One exceptional case has been reported involving an extradural lesion of the skull base with anterior fossa sino-nasal extension [5]. We describe a second intracranial-extradural case involving the mastoid region. This little-known, generally benign entity can mimic more common lesions such as meningiomas, and should be considered in the differential diagnosis of skull base tumors, particularly when associated with heavy calcification [6,7].

2. Case description

We describe a 39-year-old woman in good health with no significant previous medical history. She presented with a three-week history of sudden hearing loss in her right ear with associated intrusive right-sided tinnitus. Audiogram demonstrated a mild-moderate asymmetric high frequency sensorineural hearing loss on her right side with a pure tone average of 22 dB. The patient had normal hearing in her left ear. An MRI of her brain (Fig. 1A–D) revealed subtle contrast enhancement of the dura along and just inferior to the right internal auditory canal (IAC).

A CT scan of the temporal bone (Fig. 1E, F) showed a focal circumscribed lucent abnormality with an internal calcified matrix located in the region of the pars nervosa of the jugular foramen on the right side. The patient was initially treated with steroids, which improved her tinnitus, but symptoms recurred immediately upon cessation of the steroids with no change in her asymmetric high frequency hearing loss.

The patient underwent an infracochlear Giddings-Brackmann approach with open biopsy of the lesion, which did not return a diagnosis. She was managed conservatively for 3 months, but sensorineural hearing loss progressed to severe with decline in pure tone average to 55 dB in the affected ear. Follow-up MRI and CT demonstrated progression of the lesion, which now involved the IAC and cochlea, with

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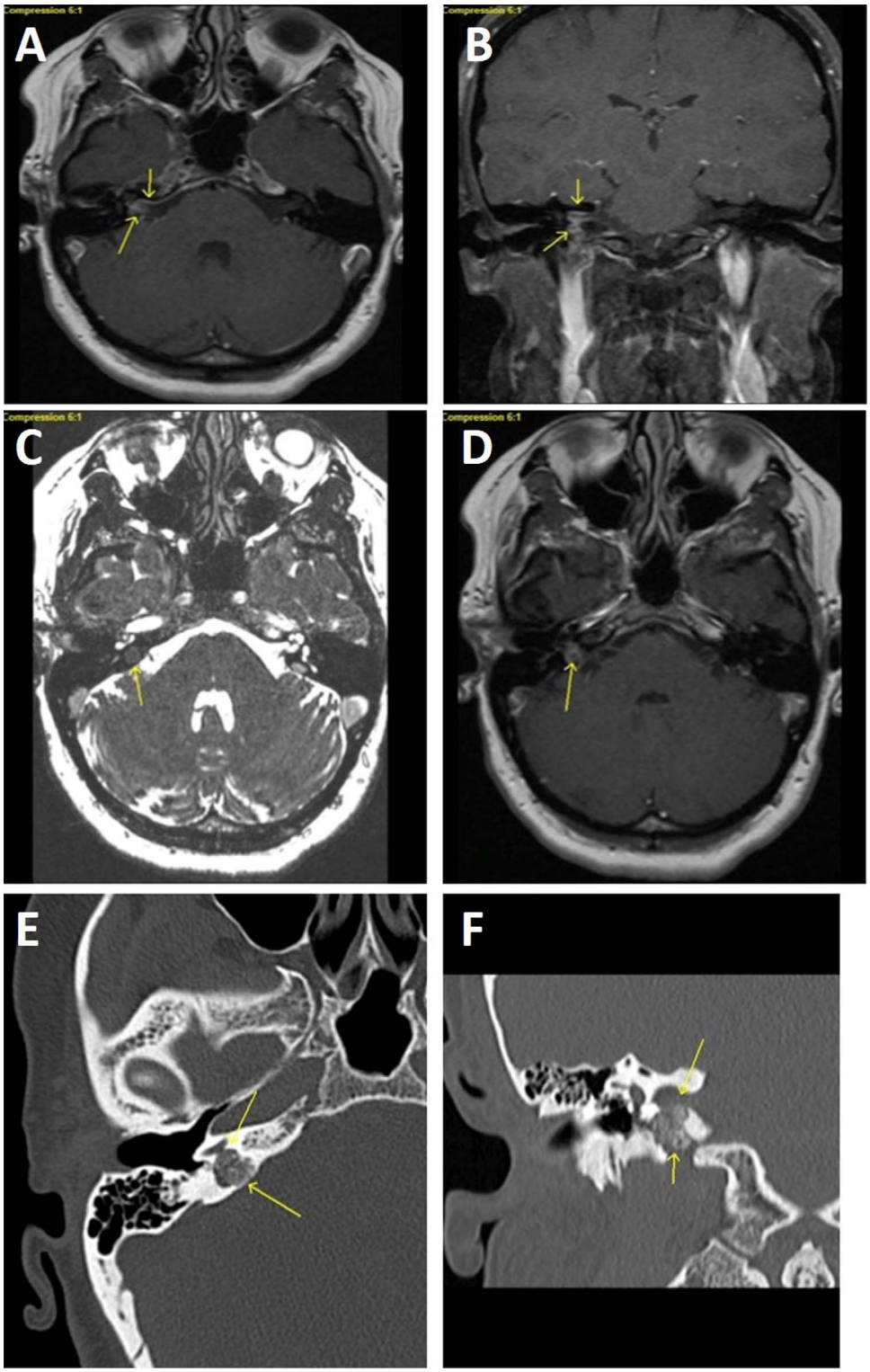

Fig. 1. A: Axial T1 post-contrast image demonstrates linear enhancement along the walls of the right internal auditory canal (arrows). B: Coronal T1 post-contrast image demonstrates linear enhancement along the walls of the right internal auditory canal contiguous with an enhancing lesion just inferiorly in the right petrous apex (arrows). C: Axial FIESTA T2-weighted sequence demonstrates a faintly hyperintense lesion in the right petrous apex adjacent to the basal turn of the cochlea (arrow). D: Axial T1 post-contrast image demonstrates enhancement within the lesion (arrow). E: Axial temporal bone CT image demonstrates a circumscribed lucent lesion with calcified internal matrix in the right petrous apex adjacent to the basal turn of the cochlea (arrow). F: Coronal temporal bone CT image demonstrates the lucent lesion with internal calcified matrix extending from the inferior aspect of the right internal auditory canal down to the pars nervosa of the right jugular bulb (arrows).

new erosion of the carotid canal in the petrous apex. She was offered a translabyrinthine approach for resection based on the location of the tumor inferior to the IAC and superior to the jugular bulb.

2.1. Intraoperative management

At the time of the resection, an extradural lesion involving the mastoid bone was encountered above the jugular bulb and was removed up to the inferior border of the IAC (Fig. 2). The involved bone was moth-eaten in appearance, gray-white, and softer than normal bone. Postoperatively, the patient's symptoms of imbalance and vertigo completely resolved; postoperative CT and MRI showed complete surgical resection of the lesion with no residual enhancing tissue; the dural sinuses were patent (Fig. 3). She experienced an expected loss of hearing on the right side due to resection of the labyrinth for exposure of the lesion. She has been fitted with a contralateral routing of sound (CROS) hearing aid.