

Patient with diplopia and deteriorating headache

Melpomeni Kosmidou, Eliza Stavride, Ioannis Tsitouridis

Department of Radiology, Papageorgiou General Hospital, Thessaloniki, Greece

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PART A

A 55-year-old female patient was admitted to the hospital with impaired vision, diplopia and headache, progressively deteriorating within the last three months. No previous clinical history was mentioned. Ophthalmologic

examination established the presence of left abducens nerve palsy. Imaging workup included CT and MRI of the brain and temporal bone (Figs. 1-4). Biopsy confirmed the radiologic diagnosis (Fig. 5a-c).

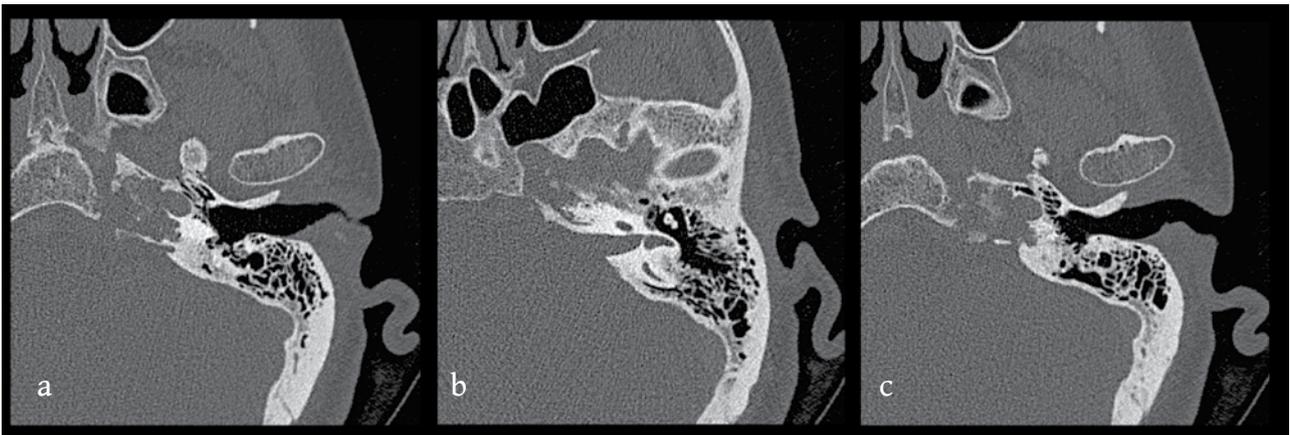


Fig. 1. Axial CT images of the petrous bone.



**CORRESPONDING
AUTHOR,
GUARANTOR**

Corresponding author: Melpomeni Kosmidou, Department of Radiology, Papageorgiou General Hospital, Ring Road, N. Eykarpia, 56403, Thessaloniki, Greece, Email: mel_kosmidou@yahoo.gr

Guarantor: Ioannis Tsitouridis, Department of Radiology, Papageorgiou General Hospital, Ring Road, N. Eykarpia, 56403, Thessaloniki, Greece, Email: tsitouridis1@gmail.com

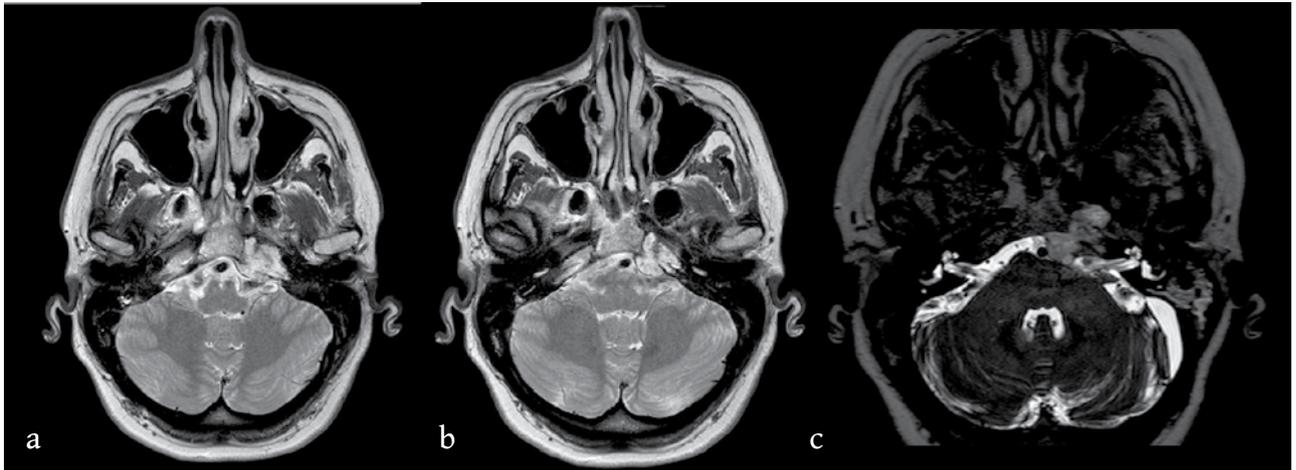


Fig. 2. *a, b.* Axial T2-weighted MR images. *c.* T2-weighted 3D-DRIVE MR image.

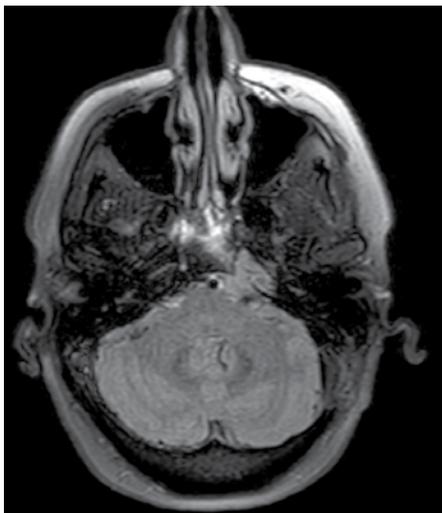


Fig. 3. Axial FLAIR MR image.

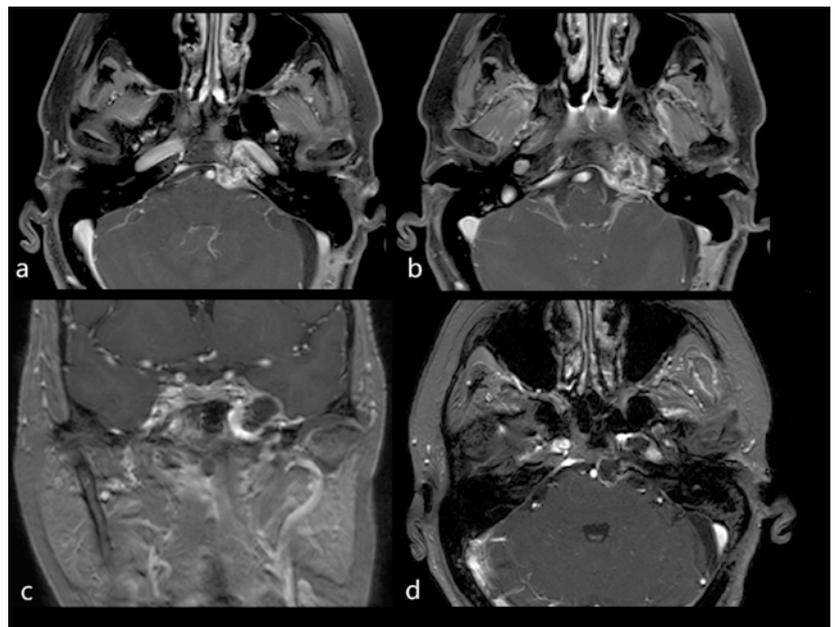


Fig. 4. Axial (*a, b, d*) and *c.* coronal contrast enhanced fat suppressed T1-weighted MR images.

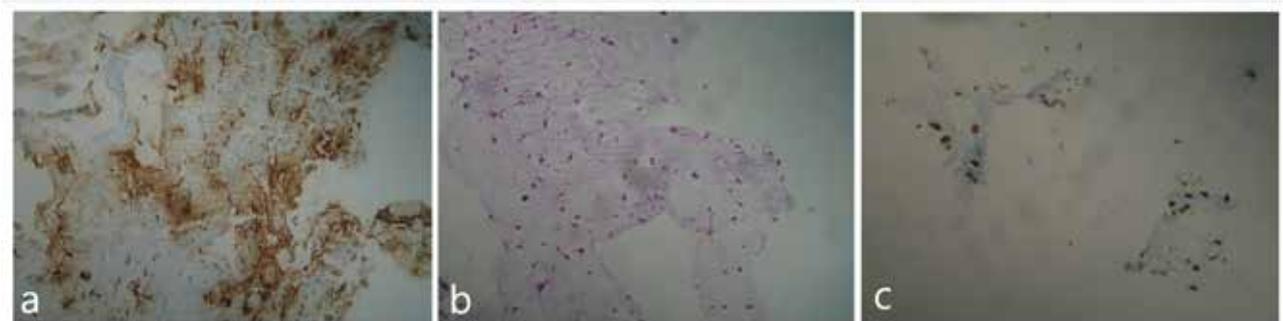


Fig. 5. High-power view (*a*) and immunohistochemistry (*b, c*).

PART B

Diagnosis: Chondrosarcoma of the petrous apex

Chondrosarcoma of the skull base represents a rare entity accounting for 0.15% of all intracranial tumours and 6% of all skull base lesions [1]. Although chondrosarcomas usually arise de novo, the association with disorders such as Ollier's disease, Maffucci syndrome and Paget's disease has been reported [2]. These neoplasms typically manifest in the 3rd-5th decade of life, without demonstrating sex predilection. The apex of the petrous bone lies in a complex anatomic region that contains a number of critical neural and vascular structures [3]. Therefore, chondrosarcomas arising in this area cause variable and occasionally severe symptoms, such as headache, vertigo, ocular symptoms and hearing loss, as a result of compression or direct invasion of important anatomic structures, the most relevant of which are cranial nerves, brainstem, or the internal carotid artery. Imaging studies play an im-

portant role in assessing this occult to clinical assessment anatomic region, detect a lesion early and accurately and also contribute to treatment management. Imaging findings on CT include a destructive petrous apex mass showing in the matrix arcs and rings of calcification, which reflect the chondroid nature of the tumour [3]. On MRI, the lesions have low to intermediate signal intensity on T1-weighted (T1WI) images and high signal intensity on T2-weighted (T2WI) images relative to that of brain tissue [3]. Enhancement of the tumour may vary. According to the enchondral development of the osseous areas along the synchondrosis, the origin of chondrosarcoma could be explained by the degeneration of remnants of this enchondral cartilage [1-2, 4]. Alternative aetiological explanations for tumours in these sites have included metaplasia of fibroblasts and origin from primitive multipotent mesenchymal cells [5].

Histologically, three subtypes are recognised: clas-

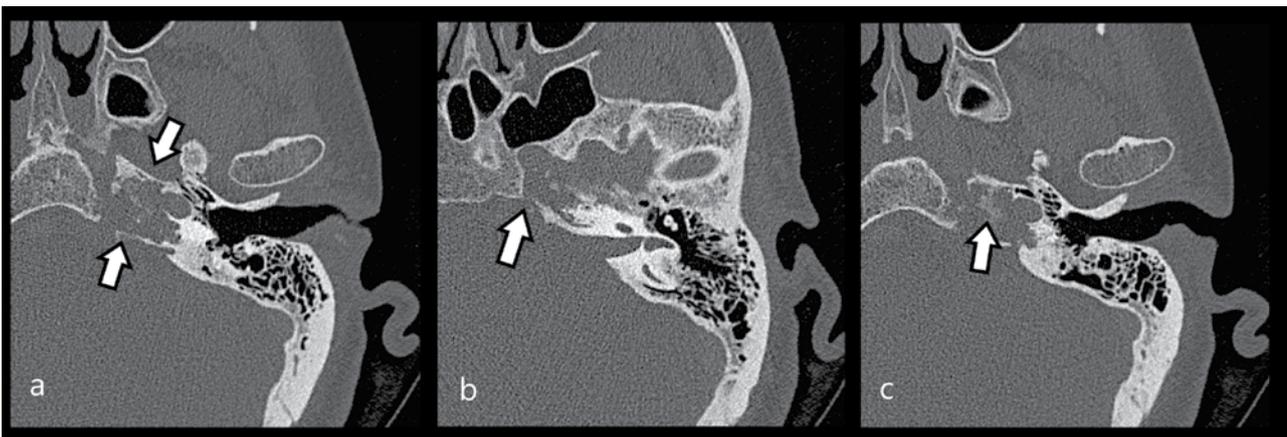


Fig. 1. Axial CT images of the petrous bone: **a.** an osteolytic mass of the petrous apex involves the left internal carotid artery canal and the jugular vein canal (arrows). **b.** the lesion extends to the petroclival fissure (arrow). No bone remodelling is seen. **c.** internal calcifications are depicted clearly (arrow).



KEY WORDS

Chondrosarcoma; Skull base; Petrous apex lesions; Cross sectional imaging

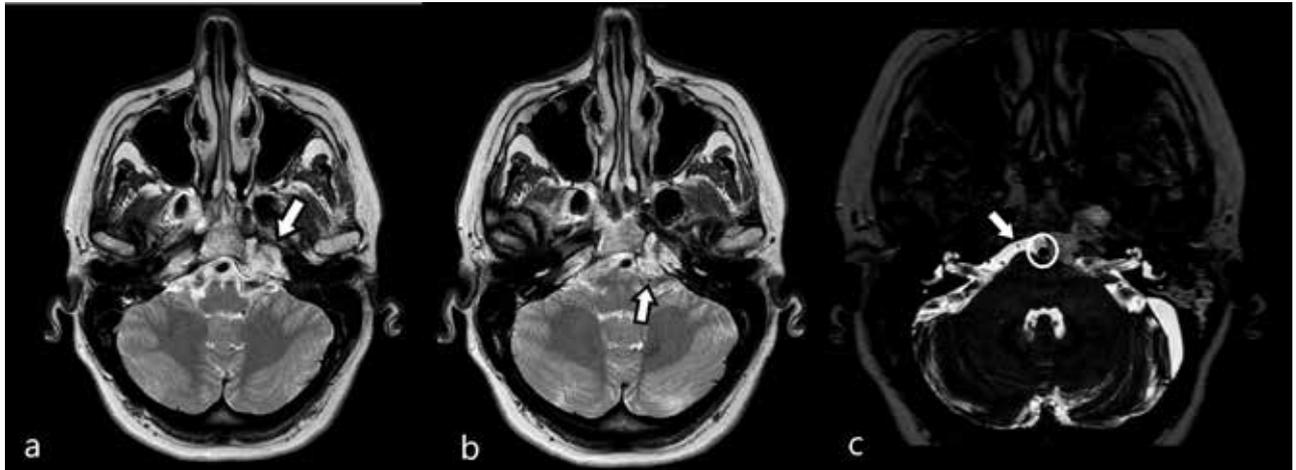


Fig. 2. Axial T2-weighted MR images. **a.** a moderately hyperintense mass involves the left part of the petrous apex, in close proximity to the left internal carotid artery. The lesion extends anteriorly and superiorly to the left Meckel's cave (arrow). **b, c.** Posteriorly the tumour extends to the left prepontine cistern (arrow in **b**), at the level of Dorello's canal, abutting but not displacing the basilar artery (circle in **c**) and causes compression to the left abducens nerve. Arrow in **c** depicts the normal right VI nerve.



Fig. 3. Axial FLAIR MR image. The mass has an intermediate signal intensity (arrow). Cerebrospinal fluid-equivalent signal intensity in the mass reflects the variably mineralised chondroid matrix nature of the lesion.

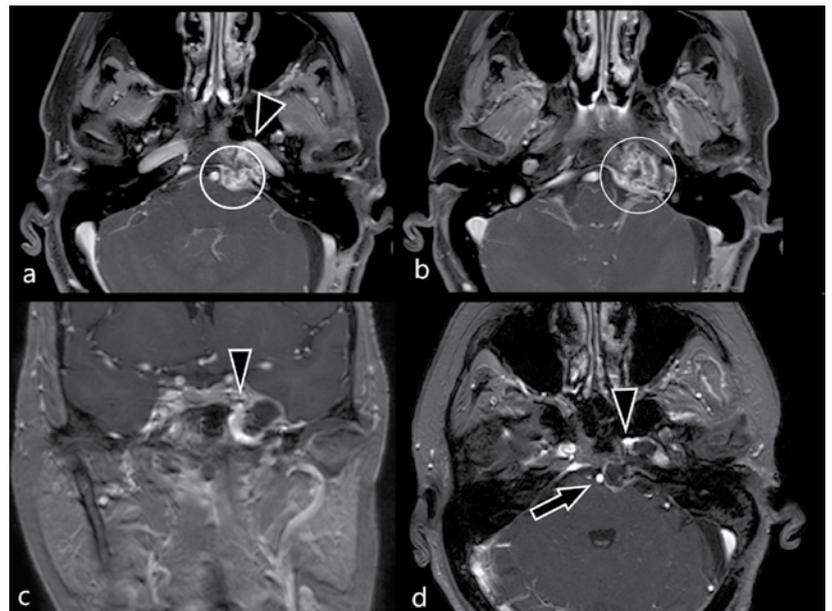


Fig. 4. Axial (**a, b, d**) and coronal (**c**) contrast enhanced fat suppressed T1-weighted MR images show inhomogeneous enhancement of the lesion (circles in **a, b**). Axial (**a, b and d**) and coronal (**c**) images depict a clear encasement of the petrous segment of the internal carotid artery (arrowheads). Axial image (**d**) shows the mass abutting the basilar artery without displacing or encasing it (arrow).

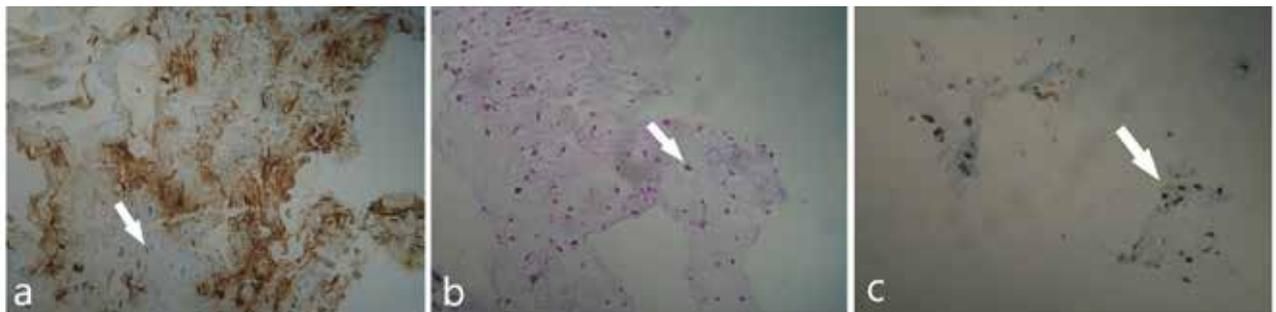


Fig. 5. High-power view (**a**) and immunohistochemistry (**b, c**). High-power view (**a**) shows hyaline differentiation. Mostly uniform chondrocytes with hyperchromatic nuclei lie in lacunae in the chondroidstroma. Little cells demonstrate irregular distribution and mild nuclear pleomorphism (arrow). Immunohistochemistry (**b, c**). The neoplastic cells express S-100 protein. Nuclei are demonstrated with a haematoxylin counterstain (arrow in **b**). Positive staining of tumour cells for Vimentin (arrow in **c**).

sis, myxoid and mesenchymal [6]. Tumour grading is determined by the classification of the World Health Organisation into three grades. Grade I tumours are well differentiated and grade II tumours are moderately differentiated. Grade III lesions however are poorly differentiated and are highly malignant [6]. Prognosis is determined by the type and grade of the tumour. The mesenchymal subtype is an aggressive malignant tumour with high tendency for recurrence, metastasis and increased vascularity, showing the lowest survival rates [6, 7]. Evans et al reported 5-year survival rates of chondrosarcoma grade I to be 90%, grade II to be 81%, and grade III to be 43% [5, 8].

Daniel et al. concluded that several radiographic features of chondrosarcoma may be helpful in assessing its biological behaviour, including the appearance of margins and patterns of calcification on plain radiographs and the size of the soft-tissue mass, patterns of growth and calcification, and presence of necrosis on CT [9].

Differential diagnosis includes other petrous apex lesions, mainly chordoma and chondroidchordoma, lesions that have distinct histological and immunohistochemical characteristics. In immunohistochemical analysis both chordoma and chondrosarcoma express S100 and vimentin. However, the former also expresses epithelial markers such as cytokeratine and epithelial membrane antigen which the latter does not express.

Optimal treatment involves radical surgical resection followed by high-dose radiotherapy in most of the cases [10]. However, radiotherapy may not have an adequate benefit-to-risk ratio in well-resected, low-grade chondrosarcomas [10].

In our patient, CT showed a mass causing bone destruction without remodelling. The matrix showed punctate and flocculent calcifications (Fig. 1a-c). The structures of the middle and inner ear appeared normal. MRI revealed a mass which was moderately hyperintense on T2WI (Fig. 2a-b) and hypointense on T1WI and FLAIR MR images (Fig. 3). There was no restricted diffusion on diffusion sequences. The le-

sion was expansile, occupying the left Meckel's cave anteriorly and superiorly (Fig. 2a) and part of the prepontine cistern posteriorly, encasing the petrous segment of the left internal carotid artery (Fig. 4c-d), the left abducens nerve and causing mass effect to the basilar artery (Fig. 2b-c). After intravenous administration of contrast (Gd-DTPA), the lesion enhanced inhomogeneously (Fig. 4a-b). Findings were suggestive of chondrosarcoma. A radical resection of the tumour was initially planned and the patient underwent surgery. However, only a small part of the tumour was amenable to resection. Histopathologic features of the tumour were consistent with low-grade (grade II) chondrosarcoma. Macroscopically, the specimens had a pale gelatinous appearance. Microscopically, there were round and ovoid cells with eosinophilic cytoplasm and with round, homogeneously vesicular plump nuclei, without the presence of nucleoli. Little cells demonstrated mild atypia and pleomorphism. Neoplastic cells were arranged individually and irregularly in a chondromyxoid stroma. Few of them were located in lacunae (Fig. 5a). Immunohistochemistry expressed positive staining for vimentin and S100 (Fig. 5b-c), but negative staining for cytokeratin AE1/AE3 and CK 8/18. Adjuvant radiation was suggested in order to prevent or delay disease progression. The patient received treatment with radiotherapy with a total dose of 70 Gy in 35 fractions within three months. Currently, 32 months after primary radiotherapy, the patient's status is stable without signs of tumour progression.

In conclusion, skull base chondrosarcoma is a rare entity arising from primitive mesenchymal cells or from the embryonic rest of the cartilaginous matrix of the cranium [10]. Imaging studies play a key role in treatment planning, providing valuable information about the growing pattern and involvement of critical anatomic structures. **R**

Conflict of interest

The authors declared no conflicts of interest.

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