Temporal bone neoplasia: a rare entity

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DESCRIPTION

A previously healthy 33-year-old pregnant woman (32 weeks of gestational age) presented to our emergency department with right peripheral facial paralysis, diplopia and ipsilateral hemifacial anaesthesia. Also, she presented a 3-month unilateral progressive hearing loss and headache episodes. On clinical examination, she presented a grade II House-Brackmann right peripheral facial paralysis and the right otoscopy (figure 1) revealed a tympanic opacity suggestive of a reddish mass in the middle ear. Tonal audiometry documented a right conductive hypoacusia with 50 dB air-bone gap with a type b tympanogram.

On the same day, she underwent a temporal bone CT in a low radiation field with plumbic apron (figure 2A,B), and an MRI without gadolinium (figure 3A,B,C) revealed a right temporal bone infiltrative lesion extending from the petroclivus to the cerebellopontine angle and middle ear cavity. A right tympanotomy biopsy was performed and histology showed a plasmocytoma. After thoraco-abdomino-pelvic MRI staging, no other lesions were identified, and it was staged as a solitary plasmacytoma. Caesarean delivery was planned at 35 weeks by a multidisciplinary team in order to start radical stereotactic radiotherapy (46 Gy in 4,5 weeks). Afterwards, the patient reported important symptomatic improvements with substantial neoplasm regression, with lesion regression in a new MRI (figure 4A,B,C) and without other remaining suspicious lesions in the Positron Emission Tomography (PET) scan. Presently, she and her baby are fine, and the patient is in the normal follow-up regimen.

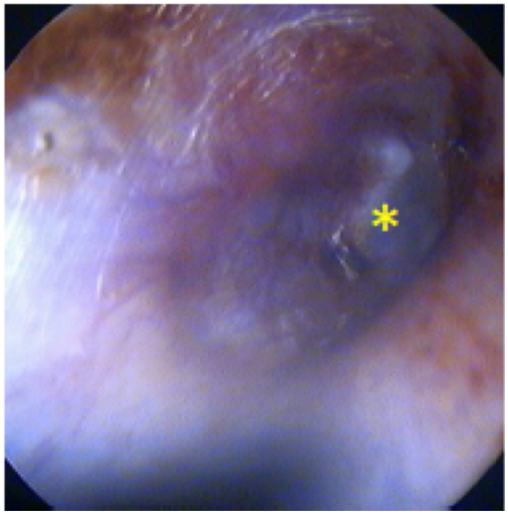

Figure 1 Right otoscopy showing a reddish opacity filling all the tympanic cavity. (*)—manubrium of malleus.

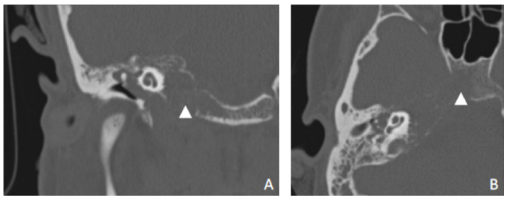

Figure 2 CT images of the right temporal bone. Coronal (A) and axial (B) showing an infiltrative lesion extending from middle ear to apex of temporal bone with extensive cortical erosion and petroclivus destruction. Note the white arrow in both images.

Plasma cell (PC) neoplasms represent a spectrum of diseases characterised by clonal proliferation and accumulation of immunoglobulin-producing differentiated B cells.[1] The spectrum includes benign common conditions, such as monoclonal gammopathy of unknown significance, as well as rare disorders such as Castleman's disease or indolent conditions such as Waldentröm's macroglobulinaemia and, finally, the more common malignant entity, multiple myeloma, and a more aggressive form, PC leukaemia.[1] There are two rare important variants of myeloma—solitary bone plasmacytoma (SBP), when it arises in an intraosseus location, which was what happened in our clinical case; and solitary extramedullary plasmacytoma (SEP), when it arises within the soft tissue.[1] Both are associated with a survival of ≥10 years,[1] accounting for approximately 2–5% of all PC disorders[2 3] and are characterised by a localised proliferation of neoplastic monoclonal PCs, with no radiological evidence of additional skeletal lesions, absence of signs and symptoms of multiple myeloma (hypercalcaemia, renal insufficiency, anaemia and/or bone lesions) and a normal bone marrow examination or having less than 10% clonal PC infiltration.[2] More than 80% of solitary plasmacytomas show up in head and neck area.[3] Meanwhile, it is rare at skull base and, particularly, in temporal bone. Usually, the first-line treatment is stereotactic radiotherapy, specially at skull base.[4] SBP may recur in other bony sites or evolve into myeloma. SEP rarely recur or progress.[1]

The diagnosis is very difficult due to unspecific symptoms and large spectrum of presentation.[4] As we can see in the selected images (figure 3), plasmacytoma presented on MRI as an isointense lesion on T1 and T2, and in post-radiotherapy period (figure 4) as decreased cystic cavity filled with CSF, with hyposignal on T1 and hypersignal on T2 due to a neoplasia substitution to a new empty cavity that is filled with CSF.

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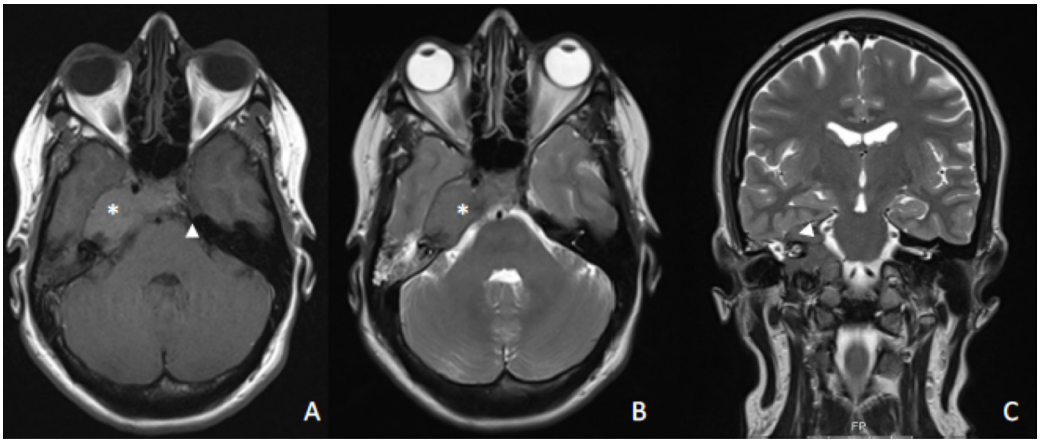

Figure 3 Isointense lesions in both T1 and T2, homogeneous and centred on the right petrosal apex. (A) Axial T1-weighted magnetic resonance without gadolinium showing an infiltrative (*) isointense mass, related to brain tissue, with compression of right trigeminal emergence and temporal lobe remodelling with extension from the temporal bone to clivus. Note the left normal hypointense space (white arrow), filled with cerebrospinal fluid (CSF), related to normal trigeminal left emergence from the midbrain. (B) Axial T2-weighted magnetic resonance without gadolinium showing the same slice in (A). Note the isointense signal from the referred (*) lesion and the hyperintense retention liquid in the mastoid cells. (C) Coronal T2-weighted magnetic resonance without gadolinium showing an isointense lesion (rotated white arrow) filling the right cerebellopontine angle and extension to internal acoustic meatus, with an otic capsule involvement.

This entity can mimic other lesions like endolymphatic sac tumour, paraganglioma or chondrosarcoma.[4] To narrow the list of differential diagnosis, the imaging study by high-resolution CT and MRI with contrast and a frozen biopsy are essential .[3 4] In the CT scan, we can see an infiltrative soft-tissue mass opacity of the temporal bone. In MRI, we can see an isointense mass relative to brain tissue on T1 and T2, without diffusion restriction or flow voids, commonly seen in paraganglioma or haemorrhagic clusters characteristic of endolymphatic sac tumour.[4] In chondrosarcoma, the lesion is more heterogeneous and hyperintense on T2.[4]

Contributors MSB wrote the article and selected the images and made the last revision of the article. ASM coworked in the selection of the images and carried out revisions of the manuscript. DAM coworked in the selection of the images and critically reviewed the manuscript. JR performed the neuroradiological imaging study and commented and selected the best images to this 'Images in…' article.

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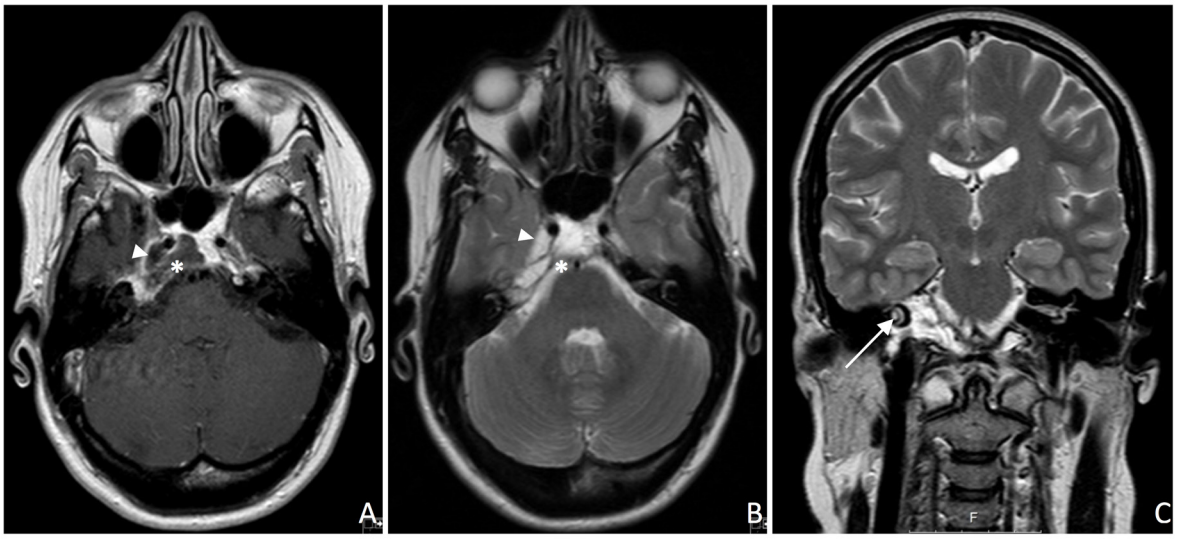

Figure 4 Axial (A) T1 post-gadolinium and (B) T2-weighted magnetic resonance. Post-radiotherapy MRI shows substantial lesion regression, with substitution of the isointense signal by a space related to a cystic tumorous cavity filled with liquid with hyposignal on T1 and hypersignal on T2 (*). Note the remodelling decrease of the right temporal lobe. The cistern of Gasser's ganglion (white arrow) is only seen in the post-radiotherapy MRI, because previously it was compressed just like the otic capsule (long white arrow) seen on (C) coronal T2-weighted magnetic resonance.

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Learning points

- The diagnosis of temporal bone plasmacytoma requires a high suspicion index due to its unspecific symptoms.
- Stereotactic radiotherapy is the gold standard treatment modality.

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