

CASE REPORT

Large tympanojugular lesion revealing multiple myeloma

Christian Martin, Mamadou Birame Faye, Pierre Bertholon, Jean Michel Prades.

Otorhinolaryngology - Head&Neck - and Plastic Surgery Department - Bellevue Teaching Hospital Saint-Etienne - France.

INTRODUCTION

Paragangliomas, also known as chemodectomas, are by far the most common tumors of the jugular fossa. Neurinomas and meningiomas are less often encountered in this location and may be difficult to differentiate from chemodectomas [1]. We are aware of only five reports of plasmacytoma of the jugular fossa [1-3]. We report a new case in which plasmacytoma of the jugular fossa was the inaugural manifestation of multiple myeloma.

CASE-REPORT

A 45-year-old man was referred to our department for management of a 6 to 8-month history of otalgia, vertigo, pulsatile tinnitus, and hearing loss on the right side. Otoscopy showed a red mass located behind the drum and extending into the external auditory canal. The audiogram suggested conductive hearing loss of 40 dB on average.

Computed tomography (CT) disclosed lysis of the entire mastoid and basal occipital region on the right side and of the right lateral mass of C1; the lesion involved the sigmoid sinus, caused lysis of the jugular foramen, and extended into the middle ear to the incudostapedial joint (Figure 1A and 1B). By magnetic resonance imaging (MRI), the lesion was seen as a large, homogenous, petrous mass measuring 6 cm in

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Corresponding author: Prof. Christian Martin,

Service d'ORL et de chirurgie cervico-faciale et plastique, CHU de Bellevue

Boulevard Pasteur

42055 Saint-Etienne CEDEX 02 - France

e-mail: christian.martin@chu-st-etienne.fr

length and 4 cm in width. The mass was centered on the sigmoid sinus and jugular bulb; it involved the infralabyrinthine region anteriorly and the squamous portion of the occipital bone posteriorly but was apparently contained by the dura mater (Figure 2A). Despite the extradural and fairly posterior location of the lesion, the heterogeneous appearance on T2-weighted MRI scans (Figure 2B) and the increased vascularity shown by MRI angiography (Figure 2C) suggested a racemose tympanojugular glomus vascularized by branches from the external carotid artery, most notably the ascending pharyngeal artery and occipital artery (Figure 2D).

Surgical excision after angiography and embolization was performed rather than a simple biopsy, which could have corrected the diagnosis. The Fisch infratemporal type A approach was used, with ligation of the lateral sinus and jugular bulb, as well as limited transposition of the facial nerve. The external auditory canal was left open in order to preserve hearing. Postoperatively, a fistula discharging cerebrospinal fluid developed. A second surgical procedure was performed, during which the external auditory canal was closed and the cavities filled with abdominal fat. Acute renal failure with normal urinary output developed rapidly, requiring dialysis.

Immunohistological studies of the operative specimen showed an IgG kappa light chain myeloma (Figure 3). Protein immunoelectrophoresis identified a free monoclonal kappa light chain, as well as Bence-Jones proteinuria (4 g/24 h). A bone marrow specimen was found to contain 16.5% of dystrophic multinucleate plasma cells. Radionuclide bone scanning disclosed a solitary focus of mildly increased uptake in the right temporal bone. Prognostic laboratory tests showed the following results: serum calcium, 2.62 mmol/L; normochromic normocytic anemia with a hemoglobin level of 11.5 g/L; moderate b2-microglobulin elevation (2.4 mg/L); and normal serum C-reactive protein level.

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Figure 1A: Preoperative computed tomography scan of the right petrous bone, axial section: *large mass in the jugular fossa with lysis of the inferior aspect of the tympanal and spread to the middle ear and external auditory canal*

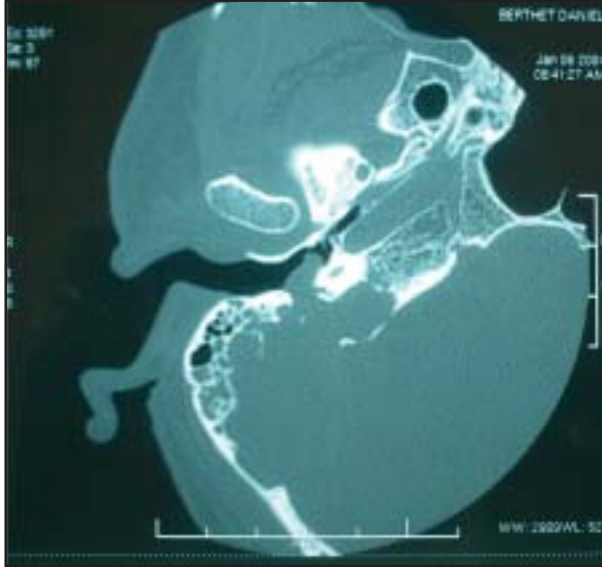
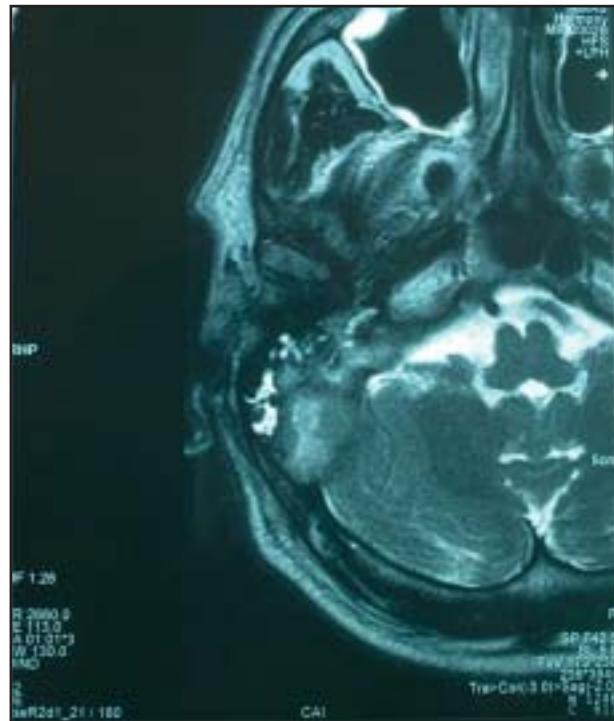
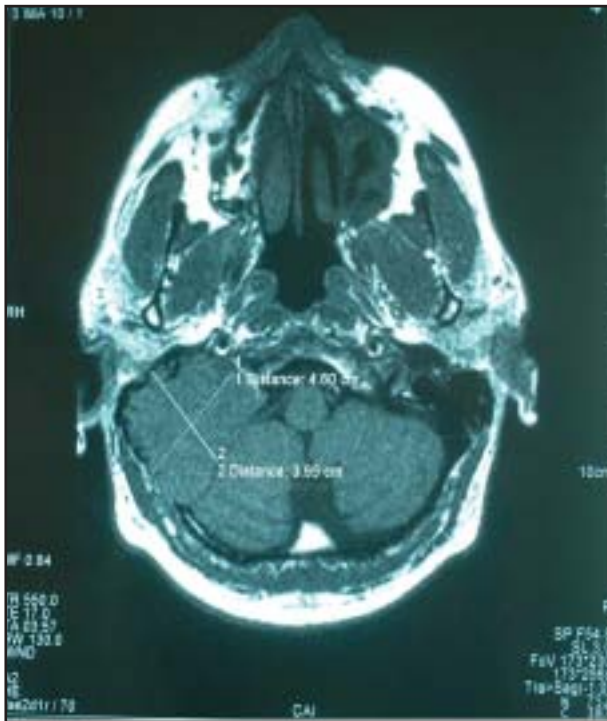


Figure 1B : Preoperative computed tomography scan of the right petrous bone, axial section: *large mass in the jugular fossa with lysis of the mastoid and base of the right occipital bone, as well as spread to the lateral sinus.*



Figures 2A and 2B. Preoperative magnetic resonance imaging, axial section: *large homogeneous lesion in the petrous bone generating intermediate signal on T1-weighted sequences (Figure 2A) and areas of moderately high signal on T2-weighted sequences (Figure 2B). The lytic hypervascular lesion measures 6 cm in length and 4 cm in width. It is centered on the jugular bulb and sigmoid sinus and spreads to the infralabyrinthine region anteriorly and the squamous portion of the occipital bone posteriorly but does not seem to breach the dura mater. These features are suggestive of a racemose tympano-jugular glomus.*



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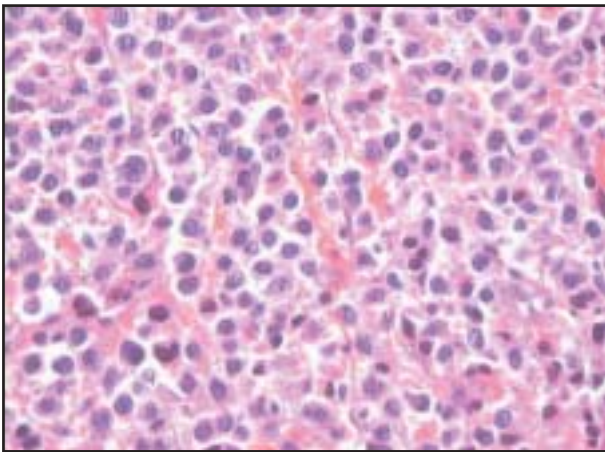
Figure 2 C: Magnetic resonance imaging-angiography with gadolinium injection: massive gadolinium enhancement of the hypervascular petrous lesion invading the sigmoid sinus.



Figure 2 D: Preoperative angiography: lesion whose vascular supply comes from the ascending pharyngeal artery, occipital artery, and posterior auricular artery.



Figure 3: Histological study of the operative specimen, original magnification x40: diffuse proliferation of tumor cells exhibiting plasmacytoid features.



Tests were negative for chromosome 13 deletion and chromosome 14 translocation.

The tumor burden was small according to the Durie Salmon classification system. Myeloablative induction chemotherapy with vincristine, adriamycin, and dexamethasone was given once a month, followed by an autologous peripheral stem-cell transplant.

DISCUSSION

Multiple myeloma is a rare hematological malignancy characterized by monoclonal plasma-cell proliferation in the bone marrow. Men older than 60 years of age are predominantly affected [4]. Bone pain is the most common presenting symptom, whereas renal failure and amylosis are less often inaugural [2]. Plasma-cell tumors as the initial manifestation are exceedingly rare; they generally develop in the sinonasal cavities, nasopharynx, liver, vertebrae, cervical lymph nodes, thorax, and genitourinary system [2,4-5].

Unusual features in our patient include the young age and isolated tympanojugular tumor masquerading as a chemodectoma. We are aware of only five previous reports of plasmacytoma arising in the jugular fossa; the tumor was solitary in 4 patients and revealed multiple myeloma in 1 patient [1-3]. Increased vascularity and osteolysis are features shared by jugular-fossa plasmacytomas and chemodectomas. In addition, several other tumors can arise near the jugular foramen. Neurinomas of sensorimotor nerves are located more posteriorly and often extend into the posterior subparotid space; they generate a homogeneous signal on T1-weighted MRI scans, although large neurinomas may be cystic. Meningiomas of the lateral skull base are far less common and usually produce no

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symptoms. Their blood supply often comes from the internal carotid artery, most notably via the middle meningeal artery; vascularization via the ascending pharyngeal artery is exceedingly rare. Imaging studies show enlargement of the jugular foramen, calcifications, and sclerosis [1,3].

Immunophenotyping is now crucial to the diagnosis of plasma cell granuloma, malignant lymphoma, malignant melanoma, granulocytic sarcoma, esthesioneuroma, and anaplastic carcinoma [5]. When the diagnosis is in doubt in a patient with a highly vascular tympanojugular mass, the appropriateness of a biopsy is a matter of debate given the risk of bleeding. Transtympanic biopsy is no longer performed. However, a diagnostic transmastoid biopsy may be safe and may supply valuable preoperative information [1].

The prognosis of multiple myeloma remains bleak despite advances in chemotherapy. Median survival ranges from 2 to 3 years [4-5]. Hope is being placed in the development of monoclonal antibodies directed against the myeloma cells.

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CONCLUSION

Involvement of the skull base by multiple myeloma is uncommon but not exceptional. Cases masquerading as tympanojugular chemodectoma, in contrast, are exceedingly rare, and we are aware of only one other reported case. The prognosis remains grim.

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