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Liposarcoma of the Petrous Bone: Unusual Localization of Sarcoma: About Case

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ABSTRACT

Malignant tumors of the ear are uncommon and may arise from multiple anatomical compartments, leading to heterogeneous clinical presentations and management strategies. Lesions of the external auditory canal (EAC) can mimic refractory otitis externa, warranting biopsy when symptoms persist despite appropriate medical therapy. We report the case of a 55-year-old woman with a history of thigh liposarcoma previously treated with radiotherapy who presented with hearing loss and right temporo-occipital headaches. Otoscopic examination revealed an exophytic mass obstructing the right EAC. MRI demonstrated a 46 × 32 mm mixed osseous–soft tissue mass involving the right petrous temporal bone and posterior skull base, with extension into the tympanic cavity and EAC, invasion of adjacent soft tissues including the parotid space, and cervical epidural extension to C4 causing mild spinal cord compression without radiologic myelopathy. Vascular involvement was prominent, with tumor thrombus in the right transverse/sigmoid sinus and internal jugular vein, collateral venous extension, and encasement/invasion of the right vertebral artery, which remained patent. Biopsy confirmed dedifferentiated liposarcoma with a minimal heterologous osteosarcomatous component, consistent with the patient's known disease. Given the extent and critical skull base involvement, the patient was treated with definitive radiotherapy. This case highlights the diagnostic challenge of EAC masses, the importance of early biopsy in atypical or treatment-resistant presentations, and the complexity of achieving local control for head and neck sarcomas where wide surgical margins are often not feasible.

KEYWORDS

External auditory canal; Petrous temporal bone; Skull base; Dedifferentiated liposarcoma; Tumor thrombus; Internal jugular vein; Lateral sinus thrombosis; Head and neck sarcoma; Radiotherapy

MAIN ARTICLE

INTRODUCTION:

Malignant tumors of the ear encompass lesions arising from distinct anatomical compartments composed of heterogeneous tissue types, which accounts for their wide variability in clinical presentation and, consequently, in therapeutic management.

In the external auditory canal (EAC), neoplastic lesions often mimic the symptoms of otitis externa. Therefore, a biopsy should be systematically performed in any case of otitis externa that fails to respond to appropriately conducted medical therapy. In this site, beyond the usual cutaneous malignancies, tumors may include adenocarcinomas and adenoid cystic carcinomas originating from ceruminous and other glandular cells of the auditory canal.

Osseous tumors of the petrous temporal bone are most commonly primary (sarcomas) or secondary (bone metastases from lung, gastrointestinal, renal, or breast carcinomas).

Sarcomas are malignant non-epithelial neoplasms derived from mesenchymal (supporting) tissues and may arise in soft tissues or bone. Substantial advances in histopathologic diagnosis—particularly in tumor classification and grading—now allow treatment to be tailored to specific sarcoma subtypes. Histologic grade is the key prognostic factor, strongly associated with metastatic risk and overall survival. Diagnosis relies on biopsy, the technique and approach of which are well standardized. The quality of surgical management is the principal determinant of local control. In head and neck sarcomas, achieving local control is especially challenging because of the proximity of critical anatomical structures. Therapeutic planning should therefore prioritize the feasibility of wide excision, favoring free-flap reconstruction when needed, and should consider the role of chemotherapy and, in selected cases, preoperative radiotherapy to improve the likelihood of negative margins. Adjuvant chemotherapy has not demonstrated a survival benefit in head and neck sarcomas, except for rhabdomyosarcoma (RMS) and Ewing sarcoma. Radiotherapy—particularly in the postoperative setting—is widely indicated for these locations.

CASE REPORT:

A 55-year-old female patient, followed for a thigh liposarcoma treated with radiotherapy, presented with hearing loss and right temporo-occipital headaches.

Otologic examination revealed an exophytic mass in the right external auditory canal; the remainder of the ENT examination was unremarkable.

(Figure 1)

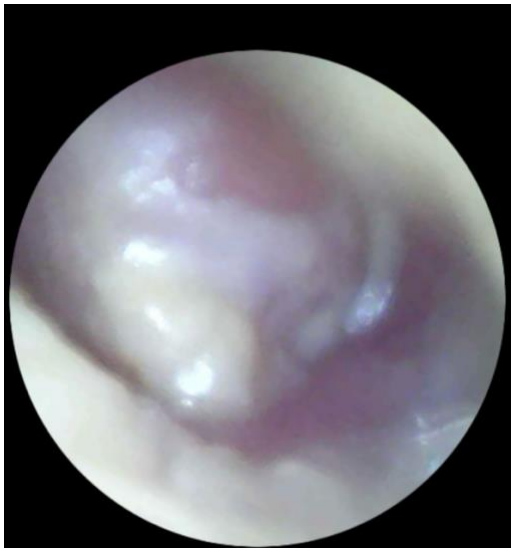

Figure 1 : An exophytic mass in the external auditory canal.

An MRI of the temporal bones/skull base demonstrated a 46 × 32 mm osseous and soft-tissue lesion involving the right petrous temporal bone and the posterior skull base, with invasion of adjacent soft tissues, the right parotid space, and the right cervical epidural space extending to C4, causing mild spinal cord compression without imaging evidence of compressive myelopathy. Notably, there was extension into the tympanic cavity and the external auditory canal.

The lesion was also associated with vascular invasion, with an in situ tumor thrombus involving the right transverse/sigmoid (lateral) sinus and right internal jugular vein, extending into additional collateral veins, with cranio-caudal progression of internal jugular vein involvement. There was also encasement/invasion of the right vertebral artery, which remained patent and opacified.

No other enhancing osseous lesions were identified within the limits of the examined field.

(Figure 2)

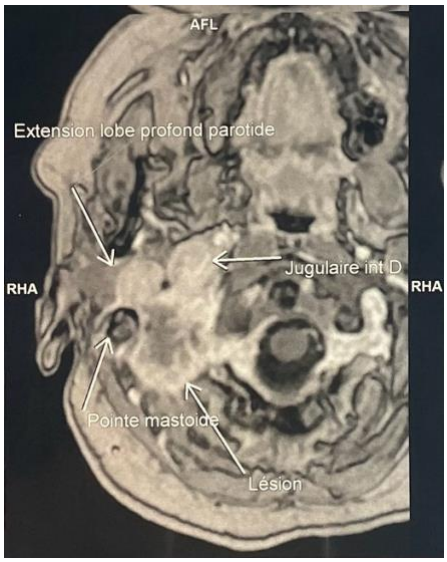

Figure 2 : A soft-tissue lesion involving the right petrous temporal bone.

A biopsy of the mass was performed, showing a dedifferentiated liposarcoma with a minimal heterologous osteosarcomatous component, consistent with the patient's known diagnosis. The treatment consisted of exclusive radiotherapy.

DISCUSSION:

Sarcomas are malignant non-epithelial tumors arising from supporting tissues (connective and/or bone), excluding neoplasms of the central nervous system and the hematopoietic system. The World Health Organization (WHO) classification [1, 2] stratifies mesenchymal tumors into four prognostic groups: (i) histologically benign tumors; (ii) intermediate tumors that are locally aggressive but non-metastasizing; (iii) intermediate tumors with a risk of metastasis; and (iv) malignant tumors with a high metastatic potential. The last two categories correspond to sarcomas in the strict sense; however, highly locally aggressive entities (e.g., desmoid-type tumors) are often included in sarcoma series because they raise similar locoregional management issues.

Well-differentiated liposarcoma (also termed atypical lipomatous tumor) is composed of adipocytes separated by fibrous septa containing atypical cells; the lipoma-like variant closely mimics normal adipose tissue morphologically. Unlike other liposarcoma subtypes,

this entity typically shows amplification of MDM2, CDK4, and HMGA2 within the 12q13–15 region. Metastatic spread is generally not expected, and the lesion is therefore considered an intermediate-malignancy tumor. Dedifferentiated liposarcoma may be present at initial diagnosis or develop following recurrence of a well-differentiated liposarcoma; it behaves more aggressively and carries a high risk of metastasis.

In the head and neck, liposarcoma can occur at virtually any site, with a predominance of cervical and subcutaneous locations, followed by pharyngolaryngeal involvement [3, 4]. A male predominance has been reported, with a median age around 57 years [3].

The most favorable outcomes are observed in well-differentiated tumors, and de novo metastatic presentations are uncommon. Prognosis appears better for head and neck primaries than for tumors arising at other sites [3]. Surgery is the cornerstone of treatment. For well-differentiated liposarcomas, a wide excision can provide effective local control [4]; similar results may be achieved in small or myxoid tumors, provided that close follow-up is feasible and that salvage surgery remains an option in case of recurrence. For other histologic subtypes, postoperative radiotherapy is recommended, particularly in patients with high-grade disease, incomplete resection (R1 or R2), large tumors, or anatomically complex extension that limits surgical margins [5]. The metastatic risk is highest in poorly differentiated and pleomorphic forms.

CONCLUSION:

Petrous temporal bone liposarcoma is an exceptionally rare malignant soft-tissue neoplasm arising from adipose tissue within the temporal bone. It is an aggressive cancer that requires specialized management, typically based on wide surgical excision, often combined with radiotherapy, given the high risk of local recurrence.

ACKNOWLEDGEMENTS

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