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Solitary plasmacytoma of the skull base presenting with unilateral sensorineural hearing loss

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Abstract
Solitary plasmacytoma of the skull base is a rare entity with only a few reported cases in the literature. We review the literature and present our experience with this lesion that produced ipsilateral sensorineural hearing loss, vertigo and ipsilateral sixth nerve palsy.

Key words: Plasmacytoma; Skull base

Introduction

The term plasma cell tumour embraces a clinical spectrum that includes multiple myeloma, solitary plasmacytoma of bone and extramedullary plasmacytoma (EMP) (Cotran *et al.*, 1989). EMP accounts for three per cent of all plasma cell tumours and one per cent of all head and neck tumours. Eighty per cent of EMP occur in the head and neck (Batsakis, 1979).

Solitary EMP of the skull base is extremely rare. EMP has been reported in the nose, paranasal sinuses, nasopharynx and tonsil (Russel and Rubinstein, 1969; Batsakis, 1979), some cases involving soft tissue only with others showing destruction of bone in the skull base.

Case report

A 62-year-old female patient was referred to the Central Middlesex Hospital with a three-month history of diplopia and progressive vertigo. The positive examination findings included right lateral rectus palsy and a Weber test that lateralized to the left. Ophthalmic, neurological and general physical examination was otherwise unremarkable. The full blood count, urea and electrolytes, liver function tests, ECG and chest X-ray were all normal.

A pure tone audiogram showed a right-sided sensorineural hearing loss of 80–90 dB (Figure 1). The computed tomography (CT) scan showed a large mass with local bony destruction involving the apex of the right petrous temporal bone, right mastoid process with extension down to the first two cervical vertebrae (Figure 2).

The patient was referred to a neurosurgeon who performed open biopsy at the level of the second cervical vertebra via a posterior approach. Histopathological findings were consistent with plasmacytoma, showing sheets of neoplastic plasma cells, with characteristic nuclear morphology and basophilic cytoplasmic rim (Figure 3).

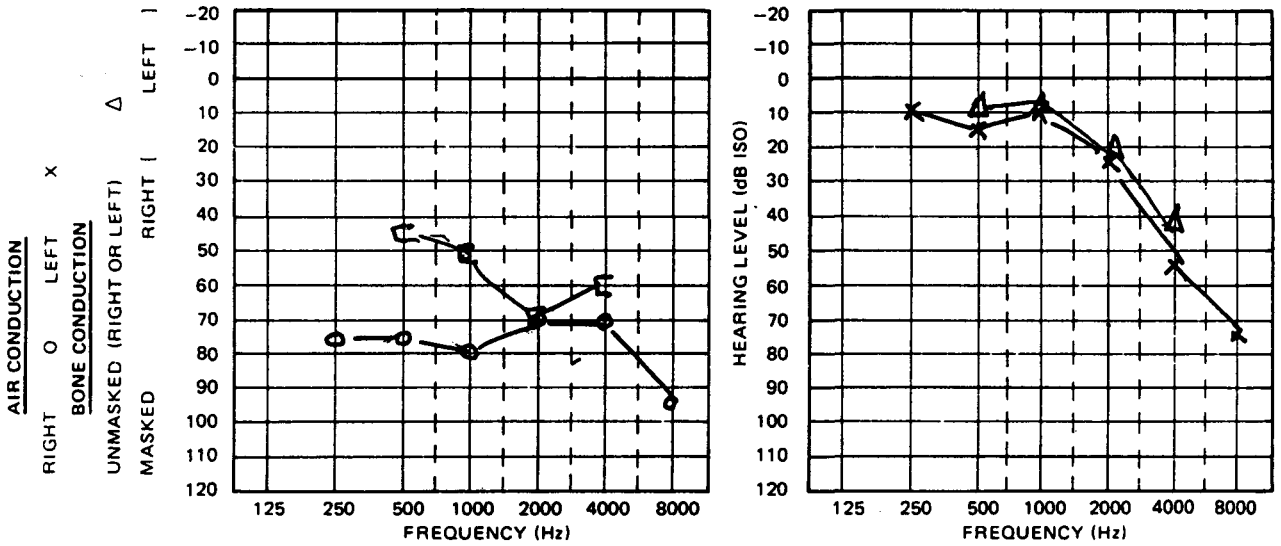

FIG. 1
Pure tone audiogram.

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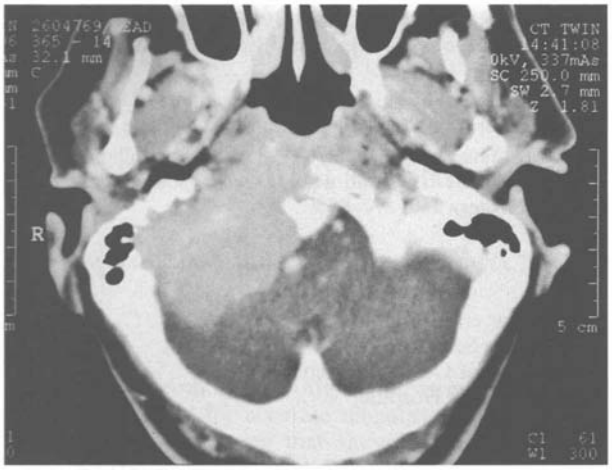

FIG. 2
Axial contrast CT scan of the posterior cranial fossa.

Bone scan did not reveal any evidence of skeletal lesions. Urine electrophoresis revealed kappa light chains (Bence-Jones proteins) of 1.5 g/l. Serum electrophoresis showed a small increase in alpha-2 globulins but no M-band.

The patient underwent a course of craniocervical beam radiotherapy which controlled the lesion locally. She proceeded to develop multiple myeloma, confirmed on bone marrow trephine biopsy (showing similarly staining plasma cells) and 24-hour urinary collection proteinuria of 2.24 g per day with Bence-Jones protein. There was no circulating paraprotein, renal impairment or hypercalcaemia. The second skeletal survey revealed widespread lytic lesions affecting ribs, proximal femur and skull.

She was treated with intravenous pamidronate and commenced on a chemotherapeutic regimen of melphalan and prednisolone (eight courses).

Twelve months following initial presentation the repeat CT scan showed no evidence of local recurrence, the 24-hour urinary protein excretion had fallen to 0.19 g per day and repeat bone marrow trephine showed only an occasional plasma cell; confirming the response to chemo-radiotherapy. The patient's general health was considered reasonable.

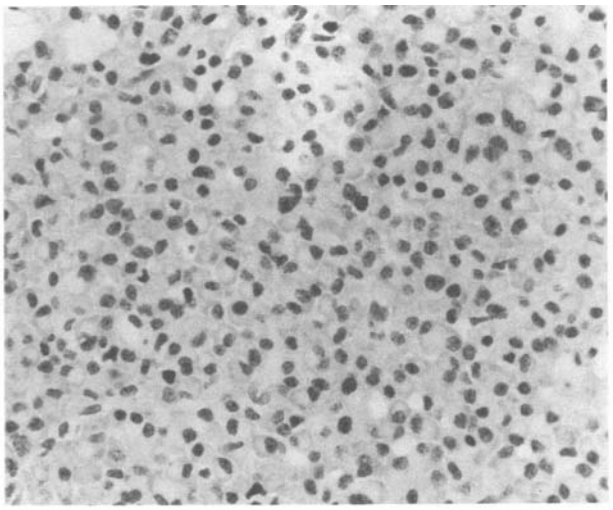

FIG. 3
Medium-power photomicrograph showing sheets of plasma cells with rounded, variably sized nuclei and faint basophilic rim of cytoplasm (H & E; × 375).

Discussion

EMP of the skull based is thought to arise from the mucosal lining of the middle ear and mastoid air cells (Wax *et al.*, 1993) and usually presents with cranial nerve palsies. The abducent and the vestibulo-cochlear nerves are the most frequently affected (Clarke, 1954). Other cranial nerves can also be affected by larger lesions. EMP may present with otalgia, headache or a post-auricular mass (Lavine *et al.*, 1979; Marks and Brooks, 1985; Shone, 1985; Funakubo and Kikuchi, 1994). However, the symptoms are often non-specific and late presentation is common (Marais *et al.*, 1992).

Investigation of EMP entails serum and using electrophoresis, full blood count, electrolytes, calcium and bone marrow studies. Low levels of paraprotein may be detected in plasmacytoma, disappearing after treatment. Persistence of this paraprotein is suggestive of occult myeloma (Nofsinger *et al.*, 1997).

The diagnosis of EMP is based on histopathological evidence of plasmacytoma and the absence of clinical, histological or radiological evidence of multiple myeloma (MM). This distinction between EMP and MM is very important, as the treatment and prognosis are different.

The 10-year survival is 70 per cent (Kost, 1990). The main prognostic indicator for EMP is progression to multiple myeloma (a fatal disease with a mean survival of two to three years). Twenty to 30 per cent of EMP progress to MM.

Histologically plasmacytomas may be similar to other small cell tumours (melanoma, undifferentiated carcinoma, lymphoma, pituitary adenoma) and benign inflammatory plasma cell tumours. However, immunohistochemically they are monoclonal and Congo red staining may reveal amyloid (Nofsinger *et al.*, 1997).

Radiotherapy is the standard treatment for EMP due to the tumour's high degree of radiosensitivity (Nofsinger *et al.*, 1997). Surgical excision is also potentially curative (Bindal *et al.*, 1995), but is reserved as second-line therapy.

EMP may recur (Nofsinger *et al.*, 1997) and continued vigilance is necessary to detect early the progression to multiple myeloma.

Conclusion

EMP is a distinct entity along the clinical spectrum of plasma cell tumours with different presentation, treatment modalities and prognosis. Solitary EMP of the skull base is a rare clinical entity. We discuss a patient who presented with ipsilateral sensorineural hearing loss, abducent nerve palsy and unsteadiness caused by a large skull base plasmacytoma which responded to radiotherapy. She subsequently progressed to multiple myeloma which at 12-month follow-up has responded to chemotherapy leaving the patient in a reasonable state of general health.

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