AN UNUSUAL CASE OF ORBITAL PLASMACYTOMA

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CASE REPORT

A 61-year-old female Caucasian patient presented in 1995 with a monoclonal gammopathy of uncertain significance. Investigations at that time included plasma paraprotein and immunoglobulin measurements and a radiological skeletal survey. The paraprotein concentration was 12 g/l, and the immunoglobulin concentrations were IgG-18 g/l, IgM-1·5 g/l and IgA-1·0 g/l. A skeletal survey did not show any lytic lesions.

In 1999, the paraprotein concentration had risen to 44 g/l and the patient was found to have multifocal osteolytic lesions in her bones. A bone marrow asperate showed that 80% of the marrow cells were plasma cells confirming a diagnosis of multiple myeloma. She was treated with melphalan by mouth and had received five courses by 2000. By March 2000, her paraprotein concentration had risen gradually to 89 g/l. Later that year she was re-admitted with headache and bone pain, predominantly in the thoracic spine. She was found to have an anaemia, a high plasma viscosity and a raised paraprotein concentration. She was hydrated and treated with ABCM chemotherapy consisting of doxorubicin, carmustine, melphalan and cyclophosphamide cycles every six weeks, with melphalan and cyclophosphamide by mouth between intravenous cycles. The treatment was discontinued because of a low platelet count. In October 2001, she was commenced on thalidomide by mouth with a good response in that the paraprotein concentration fell to 22 g/l.

She was soon readmitted with symptoms and signs of anaemia, a chest infection and high plasma viscosity. Sputum cultures showed *Aspergillus fumigatus*, and she received AmBisome for three weeks. The treatment was then changed to itraconazole orally. She continued on thalidomide and oral dexamethasone, and subsequently developed cholestatic jaundice when all treatment was discontinued.

She was then re-admitted with frontal headaches, predominantly on the right side, in March 2002. Sinus radiographs showed mucosal thickening suggestive of chronic sinusitis and a CT scan of the brain suggested soft tissue swelling anterior to the left eustachian tube. Ear nose and throat examination, however, showed no abnormality in this region. After two months she presented with worsening right eye pain and deteriorating vision. She was found to have a relative afferent pupillary defect, and was blind in her right eye.

An MRI scan of the brain showed a destructive lesion about 2 cm in diameter, eroding the floor of the anterior cranial fossa above and medial to the orbital apex, extending upwards slightly to indent the brain in the region of the right *gyrus rectus*, with local oedema. The orbital apex was filled with 'low signal' material obscuring the optic nerve (see Figures 1 and 2). Coronal sections showed extension through the medial wall of the posterior part of the orbit, the orbital apex being filled with tumor tissue (see Figures 3 and 4).

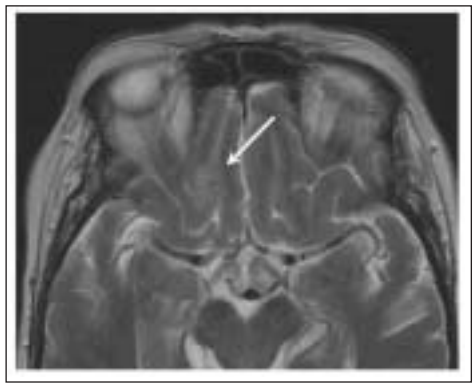

FIGURE 1

Axial T2 weighed section showing oedema in the right gyrus and medial orbital gyrus (arrow).

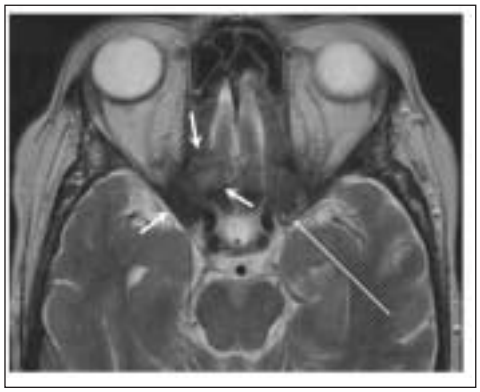

FIGURE 2

Axial T2 weighed section showing 2 cm tumour in the floor of the anterior cranial fossa (short arrows). Note that the optic nerve, lying within its CSF sheath is clearly seen on the left (long arrow), but the right orbital apex is full of low signal material and the optic nerve cannot be seen.

Loss of vision was clearly due to optic nerve compression by a myeloma deposit invading the right orbital apex. Local radiotherapy was given but the

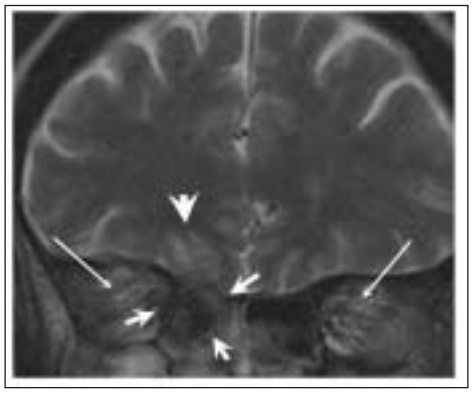

FIGURE 3

Coronal T2 weighed section showing a 2 cm heterogenous low signal tumour transgressing the floor of the anterior cranial fossa and invading the medial wall of the orbit (short arrows), with localised oedema (arrow head). The optic nerves are clearly seen at this point (long arrows).

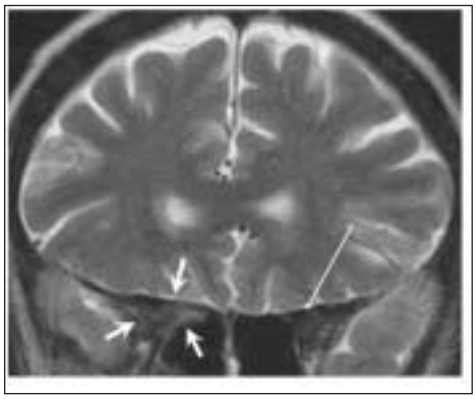

FIGURE 4

Coronal T2 weighed section showing tumour in the right orbital apex. The optic nerve is only seen on the left (long arrow).

patient died. The clinical picture at the time of death was of a progressive intracranial space-occupying-lesion.

DISCUSSION

Monoclonal gammopathy of uncertain significance is characterised by M protein in the plasma serum without any clinical symptoms. The frequency of transformation into multiple myeloma over five years is 5%, over ten years is 15% and over 15 years is 30%. For patients who respond to initial therapy, myeloma growth-rate measured by M protein doubling time, increases progressively with each subsequent relapse. As remissions become shorter, the bone marrow is more compromised and patients eventually develop pancytopenia. Occassionally, myeloma cells differentiate and an extramedullary plasmacytoma develops. Myeloma cells are still sensitive to chemotherapy but their regrowth is too rapid to respond to treatment.

Plasmacytoma is a tumour of predominantly mature cells. Solitary intracranial plasmacytoma is exceedingly rare. There are reports of 14 cases of solitary dural and four cases of intracranial plasmacytomas. Orbital manifestation is rare. Patients present late, usually with proptosis or decreased visual acqiuty. On imaging, the lesions appear smooth and often cause lytic destruction of the orbital bone. The correct diagnosis and management are difficult. The imaging of choice is magnetic resonance imaging. The lesion requires biopsy for proof. In our patient's case biopsy was not feasible because the disease was too advanced.

ACKNOWLEDGEMENTS

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