

Solitary Plasmacytoma of the Frontal Bone.

Case Report and Review of Literature

Vijay K Kak, MS, FRCS(Eng & Edin), FAMS*

Khalifa A Borashid, MD (Eng & Edin)*

Ashru K Banerjee, MD, FAMS, FRCPath**

ABSTRACT

A case of solitary plasmacytoma of the frontal bone was presented. The patient underwent total excision of the lesion and acrylic cranioplasty. Post-operative radiotherapy was not given. He remains free of local recurrence or evidence of systemic myeloma after a follow-up of 12 months. Relevant literature was reviewed.

Plasma cell neoplasms are categorised as solitary plasmacytoma of bones (SPB), extramedullary plasmacytoma (EMP) and multiple myeloma (MM). These are generally considered to represent different manifestations of the same disease. SBP accounts for less than 10% of all plasma cell malignancies^{1,2}. These commonly occur in flat bones containing red marrow, such as the spine, pelvis, ribs, skull, sternum and mandible^{3,4}. Localisation of SBP in the skull vault is distinctly rare, with only 28 cases having been reported in literature⁴⁻¹⁸.

We report one patient with solitary plasmacytoma of the frontal bone who was managed by primary surgical excision and acrylic reconstruction.

THE CASE

A 52 year old male was admitted to Salmaniya Medical Centre, Bahrain with a midline swelling over the vertex of two weeks duration. He had been having recurrent headaches for the past five years. Examination revealed a tender bony swelling in the midline in posterior frontal region. There were no other clinical abnormalities.

X-rays of the skull showed a large osteolytic area in the midline in posterior frontal region, without marginal sclerosis (Fig 1). A CT scan showed a large, intradiploic, uniformly enhancing tumour mass in the mid-posterior frontal region, destroying the inner table of the skull and depressing the underlying dura (Fig 2). Routine blood and urine examinations, serum calcium, phosphorus, alkaline phosphatase and liver function tests were normal.

At surgery, the tumour and all infiltrated bone were excised. The dura was not involved. The skull defect was reconstructed with an acrylic cranioplasty. Histological examination of the tumour was consistent with the diagnosis of plasmacytoma (Fig 3). This was further supported immunohistochemically by positive staining of the tumour cells for monoclonal kappa light chain.

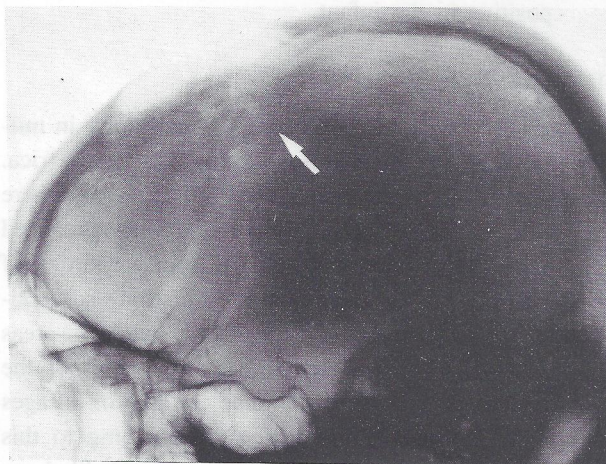


Figure 1: Lateral skull radiograph showing a large posterior frontal osteolytic area, without marginal sclerosis.

*Consultant Neurosurgeon
Department of Surgery
Salmaniya Medical Centre
State of Bahrain

**Consultant Pathologist
Department of Pathology
Salmaniya Medical Centre
State of Bahrain

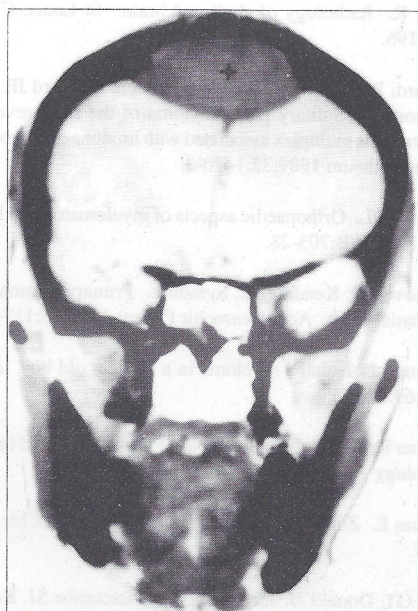


Figure 2: Contrast enhanced CT scan showing a large, intradiploic, uniformly enhancing mass destroying the inner table of skull and depressing the underlying dura.

The patient's serum proteins, including immunoelectrophoresis, were normal. There was no Bence Jones proteinuria. Bone marrow aspiration and biopsy showed no evidence of systemic myeloma. A complete radiological skeletal survey was normal.

The patient did not receive radiotherapy and has been under regular follow-up. After nine months, there was no evidence of local recurrence or systemic myeloma.

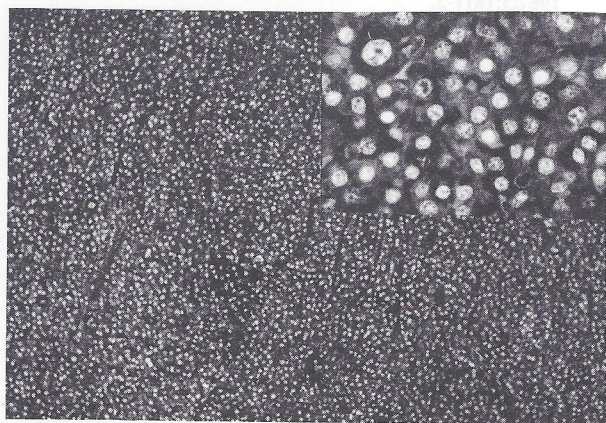


Figure 3: Microphotograph of the tumour showing diffuse sheets of monomorphic cells. Inset shows features of plasma cells with mild variation of nuclear morphology. (Haematoxylin & eosin, x 120; inset x 400)

DISCUSSION

SBP is an uncommon neoplasm constituting only 3% of all plasma cell malignancies¹⁹. There is a strong male preponderance, in contrast to MM, which affects both sexes equally^{1,2,20}. The mean age for SBP is about a decade lower than that for MM^{1,4,20}. It mainly affects the axial skeleton. The skull is a very rare site, with most lesions being described in the parietal or occipital regions.

The criteria for the diagnosis of SBP were summarised by Bataille and Sany²¹ in a review of 114 cases, and include:

- presence of a solitary bone lesion on x-rays
- histological confirmation of plasma cell neoplasia
- absent (or less than 10%) plasma cells in the bone marrow involvement
- absence of anaemia, hypercalcaemia or renal involvement.

The detection of a monoclonal component in the serum of urine does not exclude the diagnosis of a solitary plasmacytoma.

Skull plasmacytomas tend to be large, irregular, septate and expanding lesions, associated with a soft tissue mass. They rarely cause pressure over the brain²² or superior sagittal sinus¹⁷. X-rays generally reveal a lytic lesion involving both the tables of skull with little or no sclerosis and a paucity of periosteal reaction^{3,4,9}. The CT appearances are those of a slightly hyperdense extradural lesion with homogenous enhancement and focal calcifications⁹.

The majority of cases of SBP of skull have been managed by surgery and radiotherapy^{8,9,13,15}. Seven patients, including the present case, were treated by total surgical excision alone^{5,6,9,11}, reserving radiotherapy in case of local recurrence. Gherardi et al¹¹ described a patient with solitary plasmacytoma of the skull, who presented with mononeuritis multiplex. Removal of the plasmacytoma resulted in clinical improvement and clearance of the vasculitis and immune complex deposits in vessel walls.

SBP is a distinct entity with an unclear relationship to MM^{1,2,20,23}. Its natural behaviour is different from EMP, with a lower median survival and a higher conversion rate to MM^{1,2,19,23}. Lesion size, total serum protein levels

and the presence of a monoclonal spike in serum may be of prognostic significance in identifying SBP, which will ultimately progress to MM².

The treatment of choice for SBP is radiotherapy^{1,2,23,24}; chemotherapy perhaps delays the conversion of SBP to MM². Most reported cases of SBP of the skull have been managed by surgery and radiotherapy^{8,13,15}. However, surgical excision alone with complete removal of the lesion is an alternative method of treatment, with radiotherapy used only in cases of recurrence^{5,6,9,11}. This present case has also been managed by total excision of the lesion with cranioplasty.

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