

# Macrofocal multiple myeloma with frontal plasmacytoma

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## DESCRIPTION

A 27-year-old man presented with a 5-month history of left-sided forehead swelling. The patient had no other symptoms. Examination was unremarkable except for pallor and swelling of the left frontal bone displacing the orbit in a downward and outward direction (**figure 1A, B**).

The patient had normochromic normocytic anaemia (haemoglobin 8.7 g/dL) with normal total leucocyte count (3900/ $\mu$ L) and platelet count (200 000/ $\mu$ L). His renal parameters (urea 29 mg/dL, creatine 0.66 mg/dL) as well as serum calcium level (9.2 mg/dL) were normal. Positron emission tomography CT showed intense fluorodeoxyglucose uptake in left frontal region accompanied by multiple lytic lesions (**figure 2A–D**). Fine-needle aspiration from forehead swelling was suggestive of plasmacytoma (**figure 3**). Bone marrow showed 8% plasma cells. There was no paraproteinaemia, based on normal serum/urine protein electrophoresis and serum

immunofixation. A serum  $\beta$ 2-microglobulin (2.1 mg/L) and serum free light chain study was normal. Diagnosis of macrofocal multiple myeloma (ISS-I, C-R-A+B+) was made.

The patient received six cycles of chemotherapy (dexamethasone 40 mg weekly, thalidomide 100 mg daily) followed by thalidomide maintenance owing to financial constraints and non-affordability for autologous haematopoietic stem cell transplantation. His forehead swelling disappeared following three cycles (**figure 1C, D**).

Presence of extramedullary plasmacytoma (commonly involving soft tissue, lungs, pleura, genitourinary system and skull) is associated with poor outcome.<sup>1</sup> Macrofocal multiple myeloma is a distinct entity (with multiple lytic lesions and no bone marrow involvement) with superior rates of survival.<sup>2</sup> Orbital plasmacytomas are treated differently by various authors, using radiotherapy, local dexamethasone injection or systemic chemotherapy.<sup>3</sup>

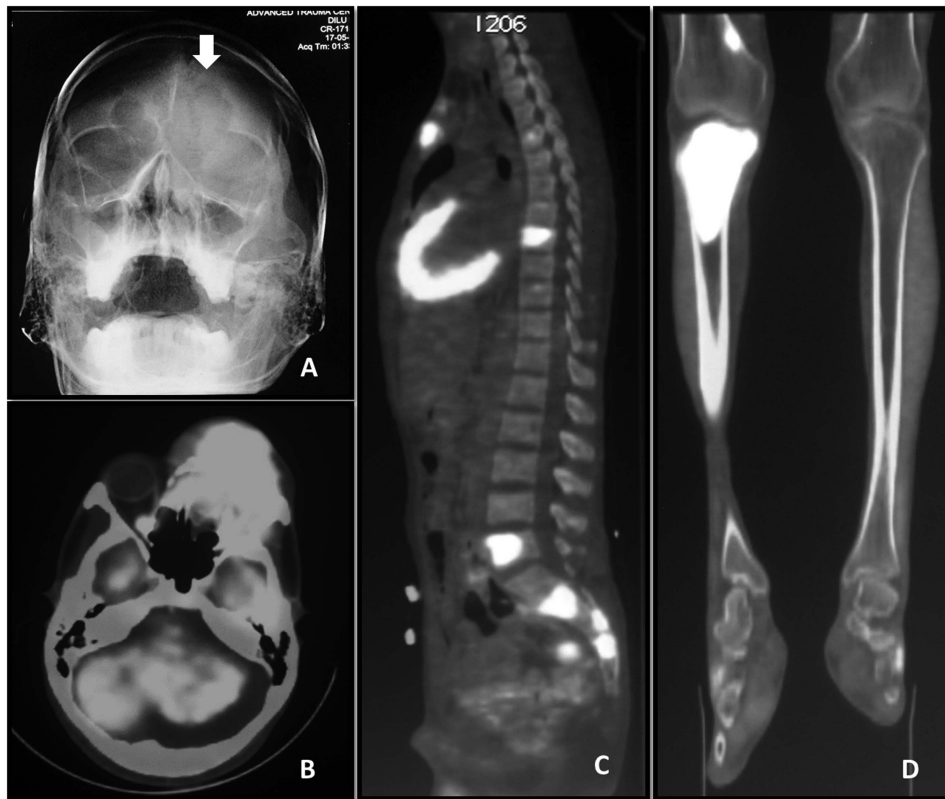


**Figure 1** (A) Frontal view showing protuberant mass in left forehead causing downwards and outwards displacement of orbit pretherapy. (B) Left lateral view showing protuberant mass in left forehead causing downwards displacement of orbit pretherapy. (C and D) Frontal and left lateral views showing resolution of forehead swelling postchemotherapy.

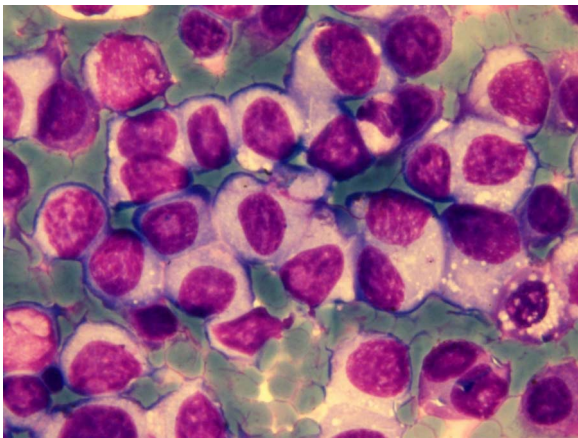


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**Figure 2** (A) X-ray of the skull (Water's view) showing increased scalloping of frontal sinus of left as compared with right side (pretherapy). (B–D) Positron emission tomography CT showing intense fluorodeoxyglucose uptake in the heterogeneously enhancing soft tissue mass lesion in the left frontal, manubrium sterni, multiple vertebral bones and right head of the tibia (pretherapy).



**Figure 3** Fine-needle aspiration from the forehead swelling showing infiltration by plasma cells suggestive of plasmacytoma (H&E staining—oil immersion— $\times 1000$ ).

### Learning points

- ▶ Macrofocal multiple myelomas have improved rates of survival owing to low tumour burden despite multiple lytic bone lesions.
- ▶ Macrofocal multiple myelomas have higher response rates to primary treatment.
- ▶ Patients are treated either with radiotherapy or chemotherapy in extramedullary plasmacytomas and macrofocal multiple myeloma.

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**Patient consent** Obtained.

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