Extramedullary Plasmacytoma Associated with Multiple Myeloma

Sir,

I read with interest the case report “Solitary plasmacytoma of skull bone” by T Das, V Chaudhary published in Feb. 2002 issue of JAPI. We present here a rare case of extramedullary plasmacytoma involving frontal and ethmoid sinuses associated with multiple myeloma.

A 60 years old male presented with chief complaints of gradually increasing painful swelling on the forehead in the midline for the period of one year. There was no other significant history. General examination revealed pallor. Regional examination revealed an approximately 6x6 cm sized, soft to firm, tender swelling over forehead in the midline extending superiorly upto the hairline and inferiorly upto the nasal bridge. The swelling was fixed to the underlying skull bone. No other significant finding was noted.

He was investigated thoroughly. The skull radiograph showed a large soft tissue swelling over frontal region with destruction and erosion of the underlying bone (Fig. 1). CT scan of head showed expansile swelling involving bilateral ethmoid and frontal sinuses with bony destruction of their wall and spreading into the adjacent structures. Biopsy of the swelling taken by nasal endoscopy showed sheets of plasma cells suggestive of plasmacytoma. Other investigations revealed lytic lesion involving right sided 7th rib near its posterior end, 24% plasma cells on examination of bone marrow aspiration, monoclonal band on serum protein electrophoresis and haemoglobin of 11.5 gm%. Other investigations like total and differential blood count, ESR, serum calcium, phosphorus, alkaline phosphatase, urea, creatinine, uric acid were within normal limit. Urine examination did not show proteinuria and Bence Jones protein, skeletal survey of other bones was within normal limit.

From the above clinical and laboratory features the diagnosis of extramedullary plasmacytoma associated with multiple myeloma was made using Durie and Salmon criterion.1

Extramedullary plasmacytoma (EMP) represents 4% of plasma cells tumours. It is classified as either primary EMP when there is absence of coexisting multiple myeloma or secondary EMP when it is associated with multiple myeloma. In 90% cases they occur in the head and neck region, the commonest sight being subepithelial lymphoid tissue of paranasal sinuses and nasopharynx.2 In our case the EMP presented as midline forehead swelling arising from the underlying frontal and ethmoid sinuses. This implies that EMP should be considered in the differential diagnosis of head and neck swelling. Other sights of EMP are gastrointestinal tract, central nervous system, urinary bladder, thyroid, breast, testis, parotid gland and lymph nodes.

On finding plasma cell tumour in extramedullary sight by histopathological examination, extensive search is to be done by bone marrow examination, total skeletal survey and appropriate blood and urine examination to find out the presence of associated multiple myeloma as the treatment and prognosis differs. The diagnosis of primary EMP is based on the absence of coexisting multiple myeloma whereas its presence implies secondary EMP. Our case was a case of secondary EMP since systemic evaluation revealed the presence of coexisting multiple myeloma. In the secondary EMP, the EMP usually occurs late in the course of multiple myeloma.3 But in our case the EMP manifested itself in the initial presentation of multiple myeloma which is a rare finding. Primary EMP has an indolent course and a very low threshold for conversion into multiple myeloma whereas the prognosis in secondary EMP depends on the stage of the associated multiple myeloma.

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Received : 10.6.2002; Accepted : 7.11.2002
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