Short reports

Different guises of plasmacytoma—from skin to bone

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Abstract

A case is reported of a patient with plasmacytoma which assumed different clinicopathological features over seven years. The tumour first presented as a subcutaneous mass which was misdiagnosed as anaplastic carcinoma. Complete response to radiotherapy ensued but recurrence took the form of an intracerebral tumour. Further complete response to "palliative" corticosteroids was followed by osseous lesions three years later, when the diagnosis of plasmacytoma was confirmed. This report illustrates the highly variable clinicopathological presentations of plasmacytoma, including the rare primary cutaneous manifestation.

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Plasmacytoma is a localised collection of neoplastic plasma cells. These can occur during the course of multiple myelomatosis or as an isolated lesion (solitary plasmacytoma). The latter commonly presents as lesions in bones or the upper respiratory tract.¹ Primary cutaneous plasmacytoma without associated marrow plasmacytosis is very rare with few published cases.²⁻⁴ We describe an unusual case of plasmacytoma, presenting as sequential isolated mass lesions in three different sites over seven years.

Case report

A 61 year old man presented in 1988 with a painless lump about 4 cm in diameter in the scalp, which had started as a small subcutaneous nodule and progressively increased in size over two years. An x ray picture of the skull did not show an osteolytic lesion. A

computed axial tomogram showed a subcutaneous mass with some erosion on the skull bone underneath. Fine needle aspiration of the mass was carried out and anaplastic small cell carcinoma was diagnosed. No other primary site was detected. He was treated with palliative radiotherapy, which resulted in complete disappearance of the mass. He presented again in 1990 with right-sided hemiparesis. A computed tomogram showed a mass in the left cerebral hemisphere. There was no associated bony lesion. In view of the original diagnosis of anaplastic small cell carcinoma, the lesion was regarded as a secondary deposit and the patient was treated palliatively with dexamethasone without histological examination. However, complete neurological recovery ensued. A computed tomogram performed a year later showed no detectable cerebral lesion. The patient defaulted on follow up.

He presented again in 1993 with bilateral knee pain which he had had for two weeks. Physical examination showed localised tenderness. An x ray picture showed osteolytic lesions affecting the right proximal tibia and left distal femur. A ⁹⁹technetium bone scan showed increased uptake in these areas. The biopsy specimen of the osteolytic lesion showed plasmacytoma. Bone marrow aspirate and a trephine biopsy specimen showed normal marrow with no increase in plasma cells. Local radiotherapy was given to the osteolytic lesions with good clinical response. He was asymptomatic during the latest follow up four months later.

Pathology

The haematological findings are summarised in the table. Blood counts, serum globulin, and immunoelectrophoresis had been normal throughout the clinical course. However,

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Haematological and immunological findings

Time	Haemoglobin (g/l)	White cell count ($\times 10^9/l$)	Platelet ($\times 10^9/l$)	Serum globulin (g/l)	IgG	IgM	IgA	Immuno electrophoresis	Immunofixation
1988	115	9.0	280	NA	NA	NA	NA	NA	NA
1990	133	10.4	322	32	NA	NA	NA	NA	NA
1993	106	15.5	514	33	1644	160	300	Normal	Monoclonal IgG λ

Serum globulin (normal range 24-36 g/l); IgG (mg/dl, normal range 700-1850 mg/dl); IgA (mg/dl, normal range 90-450 mg/dl); IgM (mg/dl, normal range 50-300 mg/dl).

Figure 1 Osseous lesion showing monotonous sheets of plasma cells. Nuclei showing "cogwheel" chromatin arrangement can be seen (arrowheads) (haematoxylin and eosin).

Figure 2 Fine needle aspiration of scalp lesion, showing sheets of malignant cells with extensive autolysis. In retrospect, some cells (arrowheads) also possess "cogwheel" chromatin arrangement (haematoxylin and eosin).

using the more sensitive immunofixation technique, serum monoclonal IgG λ was found in the latest serum sample.

Histological examination of the osteolytic tibial lesion showed a monotonous proliferation of polygonal malignant cells with occasional interspersed capillaries. The malignant cells showed a distinct cell border, abundant pyroninophilic cytoplasm, an occasional perinuclear Golgi zone, eosinophilic Russell's bodies, and frequent mitosis averaging 1.5 per high power field. Nuclei were round and eccentric, showing chromatin clumping with a cogwheel appearance (fig 1). Immuno-

histochemical staining showed that the cells were positive for leucocyte common antigen, vimentin, and monotypic λ light chain, and negative for epithelial membrane antigen (EMA). The overall features were those of a plasmacytoma.

A retrospective review of the fine needle aspirate of the scalp lesion showed malignant cells arranged in small clusters. The tumour cells were similar in morphology to those of the latest biopsy specimen (fig 2). Immunohistochemical staining showed that the cells were positive for leucocyte common antigen (LCA).

Discussion

This case is unusual in that it presented as a rare primary cutaneous plasmacytoma, which later relapsed as intracerebral and osseous lesions, without associated marrow plasmacytosis. Although the intracerebral lesion was not proved histologically, given the clinical history and the very good response to dexamethasone, it is probably justified to make that presumptive diagnosis.

This case also illustrates the pitfalls in the diagnosis of plasmacytoma, particularly when it occurs in an unusual site such as the skin and in the absence of a previous history of myelomatosis. Cytologically, the neoplastic plasma cells can assume various forms, ranging from anaplastic large cells or signet ring cells, which may mimic metastatic carcinoma, to tadpole-like cells with rosette formation, resembling peripheral neuroepithelioma.⁵ The clue to cytological diagnosis is the presence of a cogwheel chromatin pattern and prominent Golgi zone in at least some of the tumour cells. In contrast, poorly differentiated carcinoma and anaplastic carcinoma tend to show more nuclear membrane irregularities, while small cell carcinoma of lung usually has inconspicuous nucleoli and scanty cytoplasm. With the increasing use of fine needle aspiration cytology for the diagnosis of solid tumours, it is important to recognise these features so as not to miss the diagnosis. Histologically, confusion with poorly differentiated carcinoma because of similar histological appearances could occur, compounded by the fact that on immunophenotyping, plasmacytoma might be negative for LCA, but positive for EMA⁶ and occasionally cytokeratin.^{7,8} Demonstration of monotypic expression of immunoglobulin light chain will help to establish the diagnosis.⁹ Because plasma cell neoplasms frequently express EMA and B cells tend to lose LCA when they reach the plasma cell stage, an incorrect diagnosis of carcinoma might be made if a plasma cell tumour exhibited EMA in the absence of LCA. As EMA is not very specific to epithelial cells,¹⁰ it should not be used as the sole

marker of epithelial origin in tumour diagnosis. Immunohistochemical study should be used as a supplement to morphological diagnosis; all diagnostic conclusions require the correlation of immunohistochemical findings with morphology. This distinction is important, because localised plasmacytoma is usually very amenable to treatment and has a much better prognosis than metastatic carcinoma or multiple myeloma,^{11,12} as shown in this case. Finally, all isolated plasmacytomas should have long term follow up, because recurrence may occur in distant sites or as myelomatosis.

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