HOSPITAL CHRONICLES 2015, 10(4): 235–236

IMAGES IN MEDICINE

A Case of an Unusual Relapse of Multiple Myeloma

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ABSTRACT

Extramedullary relapse constitutes an uncommon manifestation of multiple myeloma, but central nervous system involvement as the only manifestation of relapse appears even less common. A 50-year-old man with a history of multiple myeloma achieved complete remission after autologous hematopoietic stem cell transplantation. Fifteen months later, he presented with central nervous relapse with no signs of systemic disease.

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KEY WORDS: *multiple myeloma; autologous hematopoietic stem cell transplantation; central nervous system relapse*

ABBREVIATIONS
MM = multiple myeloma
CNS = central nervous system

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Manuscript received March 3, 2015; Revised manuscript received August 6, 2015; Accepted August 24, 2015

A 50-year-old man with a history of IgA-lambda multiple myeloma (MM) achieved complete remission after high dose chemotherapy followed by autologous hematopoietic stem cell transplantation. Fifteen months later, he presented with a hoarse voice, numbness and cauda equina syndrome. Complete blood count and a peripheral blood smear at this time showed normal hemoglobin (13.6 g/dL; normal range 13.5-17.5 g/dL), leukocyte count (8.6×10^9/L; normal range 4-11×10^9/L), and platelet count (169×10^9/L; normal range 150-350×10^9/L). There was no evidence of a monoclonal component in the blood and urine as measured by serum protein electrophoresis and immunofixation. Serum kappa free light chains were 9.74 mg/L (normal range 3.3-19.4 mg/L), lambda free light chains 5.07 mg/L (normal range 5.71-26.3 mg/L) and free kappa/lambda ratio 1.92 (normal range 0.26-1.65). In addition, the serum calcium level (2.3 mmol/L; normal range 2.3-2.74 mmol/L) and renal function (creatinine, 57.4 μmol/L; normal range 53-106 μmol/L) were normal.

Microscopic analysis of the cerebrospinal fluid revealed infiltration by abnormal plasma cells (Fig. A); some cells contained mitotic features (thick arrow), others contained intranuclear Dutcher bodies (arrowhead) and some were binucleated (arrow). Immunophenotypic evaluation revealed the presence of plasma cells using the markers CD38 and CD138 (Fig. B), while lambda light chain clonality was confirmed (Fig. C). Spinal cord magnetic resonance imaging showed multiple epidural nodules. Osteolytic lesions were not detected.

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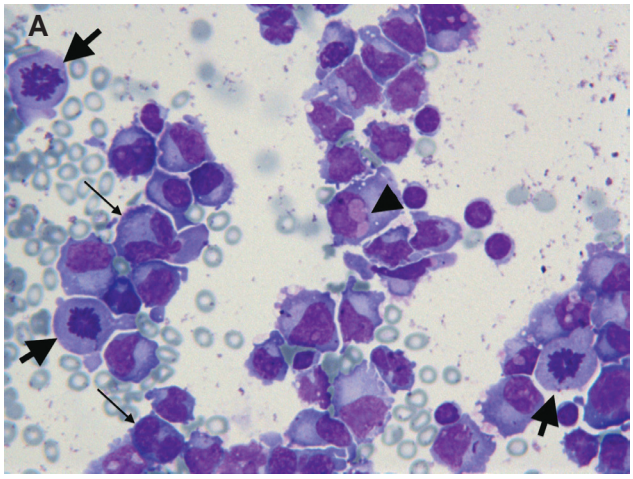
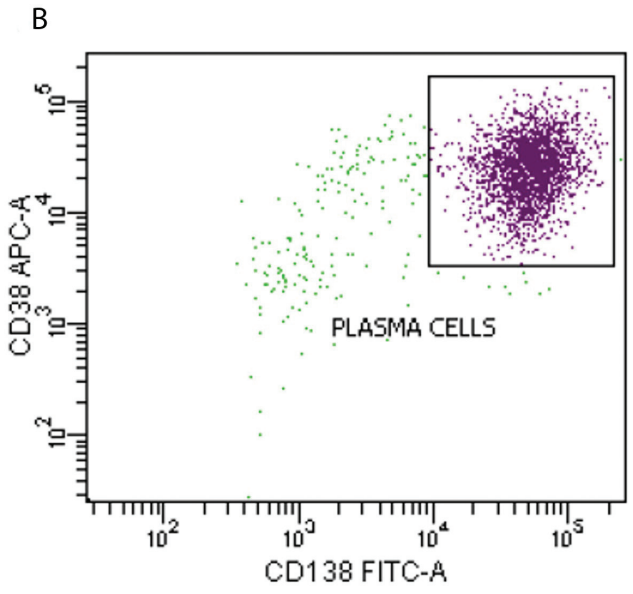
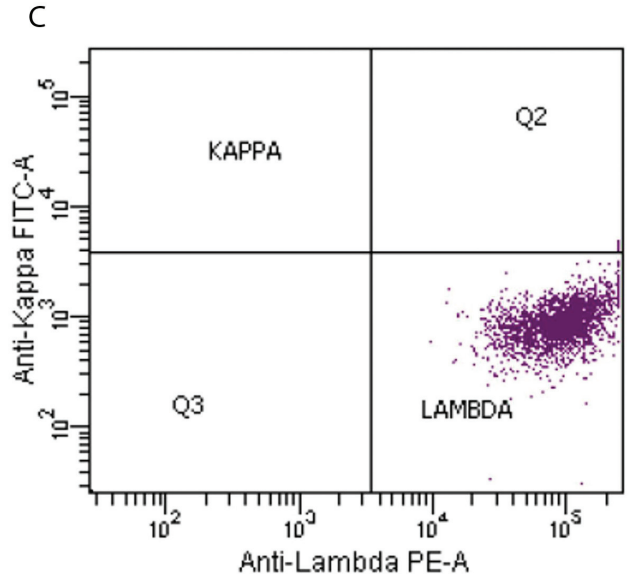
Neurological manifestations frequently accompany MM, and these manifestations may occur in the setting of metabolic disorders, hyperviscosity syndrome, spinal cord or nerve root compression, amyloidosis or peripheral neuropathies due to chemo-

The authors declare no conflict of interest







therapy toxicity.[1] On the contrary, central nervous system (CNS) involvement in MM is extremely rare. It may present with solitary or multiple intraparenchymal plasmacytomas due to extention of lesions of the skull and/or leptomeningeal disease with abnormal cerebrospinal fluid findings in advanced stages of the disease.[2,3] Cases of isolated CNS relapse of MM after autologous hematopoietic stem cell transplantation are very few in the literature.[1,4,5] Factors that reliably predict MM relapse in the CNS are uncertain.[4]

In recent years, multiparametric flow cytometry has become mandatory in the clinical management of hematological malignancies, both for diagnostic and monitoring purposes. Flow cytometry gives the ability to identify plasma cells among other hematopoietic cells, characterize aberrant plasma cell phenotypes and confirm clonal nature of plasma cells.[6,7] Central nervous system MM portends a poor survival prognosis and currently has limited treatment options. The efficacy of immunomodulatory drugs and proteasome inhibitors in such cases has to be demonstrated in future clinical trials.[8,9]

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