Multiple myeloma with cells typically seen in storage diseases

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Abstract: *We report on a rare case of multiple myeloma with atypically large cells containing a great amount of azurophilic inclusions usually seen in storage diseases.*

Keywords: *Multiple myeloma; Plasma cells*

A 45-year-old Brazilian black woman was experiencing weakness and back pain, as well as mucosal pallor. The complete blood count showed a white blood cell count of 9.7 x 10^9/L, red blood cell count of 3.00 x 10^{12}/L, hemoglobin level of 91 g/L and platelet count of 372 x 10^9/L. Rouleaux formation was noticed. Serum protein electrophoresis revealed an M-protein spike of 33.3 g/L in the beta region. The concentrations of the IgG, IgA and IgM immunoglobulins were 52.90 g/L, 0.23 g/L and 0.15 g/L, respectively. The beta2-microglobulin level was 3.99 mg/L. No renal function or calcium metabolism abnormalities were noted as per the following results: creatinine 114.9 µmol/L, serum calcium 2.02 mmol/L and ionized calcium 1.06 mmol/L. There were no skeletal lesions identified by bone radiography, scintigraphy or magnetic resonance imaging. Using an immunochemical method, 40% of free kappa light chain was detected in 1.29 g/24-hour urine protein, which also presented an IgG kappa monoclonal component.

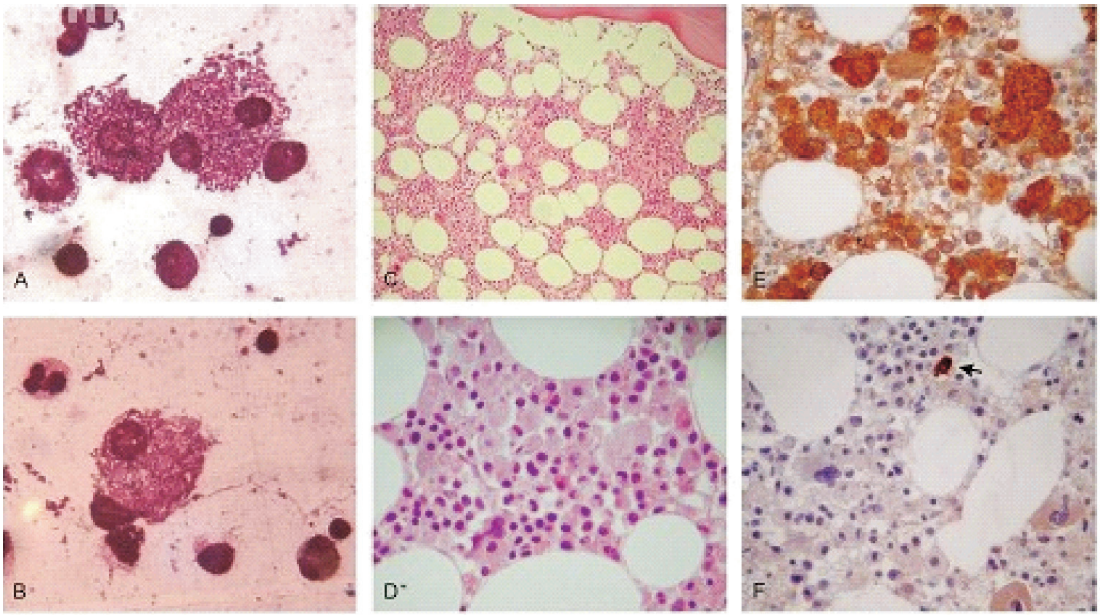


Figure 1 – Atypical large plasma cells with unusual inclusions on May-Grünwald-Giemsa stained bone marrow aspirate films (A and B) and in bone marrow biopsy, HE/100x (C) and HE/400x (D). Positive kappa light chain (E) and negative lambda light chain (F) reactions are show in large cells after immunohistochemical preparation. One normal plasma cell is positive for lambda chain (arrow) (F)

May-Grünwald-Giemsa stained bone marrow aspirate smears showed 7 per cent of typical myeloma plasma cells and 20% of atypical large cells containing a great amount of azurophilic inclusions usually seen in storage diseases (Figure 1A and 1B). Flow cytometric analysis revealed 6.5% of neoplastic plasma cells expressing the following phenotypes: CD19-, CD56-, CD38+++, CD138+++, CD45+, CD117+ and kappa light chain restriction. Cells in the high granularity (SSC) and large size (FLC) region showed the same immunophenotypes. Although no karyotype abnormality was identified by conventional cytogenetic methods, the heterozygous deletion of 13q14.3 was detected using fluorescence in situ hybridization. Hematoxylin-eosin-stained sections in a bone marrow biopsy demonstrated partial replacement with atypical large cells, which accounted for approximately 30% of the nucleated marrow cells. These cells displayed a round, eccentric nucleus and large granular cytoplasm resembling histiocytic cells seen in deposit disease (Figure 1C and 1D). The immunohistochemical study demonstrated a weak reaction for CD68 and a strong positive kappa light chain reaction on the large cells, while lambda light chain reaction was positive only in a few normal plasma cells (Figure 1E and 1F). This type of inclusion is rare in multiple myeloma cells with most of the reported cases being IgA or IgG myeloma but with all having kappa light chains.[1] Our patient achieved complete remission after treatment with VAD-regimen chemotherapy followed by autologous peripheral blood stem cell transplantation and remains event-free at more than five years.

References

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