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BENCE JONES MYELOMA WITH SIGNET RING-LIKE PLASMA CELLS

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Signet-ring cells, typical of muciparous adenocarcinoma, are seldom observed in other solid tumors or in lymphoproliferative disorders.[1] Two cases of myeloma with signet-ring morphology have been reported so far.[2,3] To the best of our knowledge, the following is the first case of signet-ring cell Bence-Jones myeloma.

A 69-year-old man presented with cachexy, dehydration, liver enlargement and multinodular goiter. Blood tests revealed leukocytosis (12.9×10^9/L), increased serum creatinine (130 μmol/L) and lactic dehydrogenase (534 U/L). Proteinuria (1 g/L), hypercalcemia (2.86 mmol/L) and hypogammaglobulinemia (7.81 g/L) were also present. Serum concentrations of tissue polypeptidic antigen (177 U/L), thyroglobulin (261 μmg/L), and β2-microglobulin (5.39 mg/L) were increased, while carcinoembryogenic antigen, α-fetoprotein, calcitonin, CA 199 and prostate-specific antigen were normal. Briefly, chest X-ray, esophagogastroduodenoscopy, echography and needle aspirate of the thyroid, and echography and CT scan of the abdomen did not detect any solid tumor. Skeletal roentgenograms revealed a diffuse worm-eaten bone aspect, due to small multiple lytic lesions of the skull, vertebrae, pelvis and femora. Immunophoresis disclosed monoclonal λ light chains in the urine and serum. Iliac bone marrow smear and trephine biopsy demonstrated an interstitial infiltrate of small lymphoplasmacytic elements and several larger cells that contained a broad single cytoplasmic vacuole displacing the nucleus to the periphery, conferring a signet ring-like appearance (Figure 1, A,B,C). Only λ light chains were documented in both cell types by immunohistochemistry. The lymphoplasmacytes displayed an intense diffuse cytoplasmic pattern; signet-ring cells showed a similar reaction at the periphery of vacuoles and scanty positivity inside (Figure 1, D).

Bence-Jones myeloma, esophagitis and non-toxic multinodular goiter were diagnosed. The patient experienced temporary benefit from treatment with clodronic acid, melphalan, prednisone. He died eighteen months later of cerebral hemorrhage.

This case represents a rare morphological variant of an equally rare disorder. Both the peculiar signet-ring aspects, probably the result of an accumulation of material from defective immunoglobulin assemblage,[4] and the presenting symptoms initially suggested a misleading diagnosis of solid tumor.

References

1. McCluggage WG, Bharucha H, El-Agnaf M, Toner PG. B cell signet-ring cell lymphoma of bone marrow. J Clin Pathol 1995; 48:275-8.
2. Chen KTK, Ma CK, Nelson JW, Padmanabhan A, Brittin GM. Clear cell myeloma. Am J Surg Pathol 1985; 9:149-54.
3. Eyden BP, Banerjee SS. Multiple myeloma showing signet-ring cell change. Histopathology 1990; 7:170-2.
4. Caligaris Cappio F, Cavo M, De Vincentiis A, et al. Peripheral blood stem cell transplantation for the treatment of multiple myeloma: biological and clinical implications. Haematologica 1996; 81:356-75.

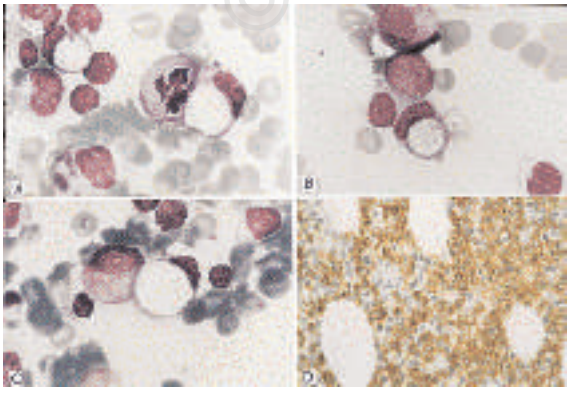

Figure 1. A, B, C: varying signet-ring aspects of plasma cells in a bone marrow smear (May-Grünwald-Giemsa). Original magnification 1,250×; D: lymphoplasmacytes and signet-ring cells stained for λ light chains in bone marrow section (ABC complex method). Original magnification 400×.

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