Signet-ring cells, typical of muciparous adenocarcinoma, are seldom observed in other solid tumors or in lymphoproliferative disorders. Two cases of myeloma with signet-ring morphology have been reported so far. 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 x 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 bone-eaten aspect, due to small multiple lytic lesions of the skull, vertebrae, pelvis and femora. Immunophoresis disclosed monoclonal 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, and the presenting symptoms initially suggested a misleading diagnosis of solid tumor.

References