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REVIEW ARTICLE

SYNCHRONOUS SMOLDERING MULTIPLE MYELOMA AND NIEMANN PICK IN AN ELDERLY PATIENT

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ABSTRACT

Here we report a case of synchronous smoldering multiple myeloma and Niemann Pick in a 54-year-old man.

Key words:

Multiple Myeloma- Niemann Pick.

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INTRODUCTION

A 54-year-old man was referred to our hematologic department for exploration of splenomegaly discovered following abdominal pain. No concept of alcoholism or smoking during interrogation and his family history was negative for hematologic malignancies or Niemann Pick. On physical examination, he showed splenomegaly and hepatomegaly 5 and 4 cm, respectively below the costal margin. The white blood cell count was 5100/mm³ comprising 77% granulocytes, 17% lymphocytes, and 6% monocytes. The hemoglobin was 9.9g/dl and the platelet count was 286 000 /mm³. He had normal serum calcium, renal and liver function tests. Hepatitis B and C serology were negative. Abdominal ultrasound revealed homogeneous hepatomegaly without focal lesion or vascular anomaly, heterogeneous splenomegaly at 25 cm with hypo echoic nodular images and absence of intra-abdominal lymphadenopathy. Serum protein electrophoresis showed a monoclonal protein of 15 g/l at the level of the Beta 2 fraction, characterized as IgA-kappa by immunofixation. No monoclonal protein was detected in the urine. Free light chain assay showed an elevated kappa light chain, a normal lambda light chain, and an abnormal free light chain ratio.

Given the monoclonal protein and the presence of hepatosplenomegaly, the diagnosis of MM complicated by amyloidosis was the first suspected diagnosis leading us to perform bone marrow examination (BM). The medullogram objectified a rich marrow with the presence of numerous abnormal histiocytes (Figures 1a-b). Further examination of these cells shows vacuoles, a foamy appearance of the histiocytes and sometimes intense blue granulations with the May-GrünwaldGiemsa stain, a "sea blue" appearance reminiscent of Niemann Pick cells (Figure 1 c-d). Associated with this histiocytic infiltration, a medullary plasmacytosis made up of 35% of dystrophic plasma cells was observed. An appearance of flamed plasma cell was observed called Undritz Cells. These are cells with pink or red cytoplasm reflecting the intracytoplasmic accumulation of IgA carbohydrate. The conventional cytogenetic BM study revealed a normal karyotype. Further evaluation on FISH didn't reveal the translocation t(11; 14) gene rearrangement and the del17p. In order to know if the patient was indication or not for anti-myeloma therapy, magnetic resonance imaging (MRI) was performed showing absence of bone lesions and the international staging system (ISS) was equal to 1 (β 2microglobuline < 2,5 mg/l and albumin \geq 35g/l).

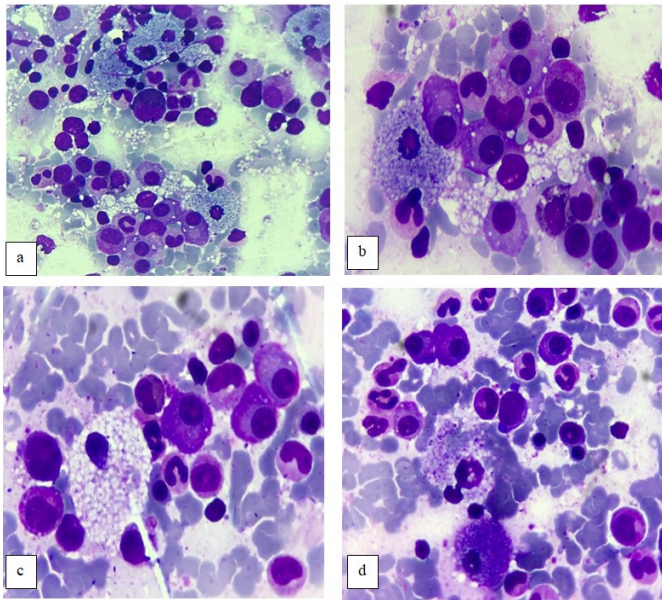


Figure 1. Medullary smear stained by May Grunwald Giemsa, seen at magnification (x40(a); x100 (b,c,d)), showing the presence of sea blue histiocytes and dystrophic plasma cells

According to this finding, the patient had no indication for anti-myeloma therapy and he is currently undergoing frequent laboratory monitoring to assess the evolution of symptomatic myeloma. In summary, we report a synchronous smoldering MM and Niemann Pick discovered in an elderly patient [1].

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