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CASE REPORT

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Multiple Myeloma with Histiocytosis

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ABSTRACT

Myeloma is a disease characterized by abnormal clonal plasma cells that produces paraprotein. The coincidence of myeloma and histiocytosis is a rare clinical manifestation. This case report describes a 50-year-old female with back pain, anemia, increased globulin and hypercalcemia due to multiple myeloma. Bone marrow cytomorphology revealed an increased number of plasmocytes and histiocytosis.

Keywords: myeloma, histiocytosis.

INTRODUCTION

Multiple myeloma is a rare disease caused by an abnormal proliferation of clonal plasma cells in the bone marrow that produce paraproteins. Signs and symptoms of multiple myeloma are characterized by organ involvement: hypercalcemia, renal impairment, anemia and bone lesions (1).

Histiocytosis in multiple myeloma is a rare finding, It is usually related with crystal storage histiocytosis, an uncommon phenomenon in disorders associated with monoclonal gammopathy due to paraprotein accumulation in the intracellular lysosome of macrophages (2).

This case report aims to describe the case of a patient with multiple myeloma and histiocytosis. ❑

CASE PRESENTATION

A 55-year-old female presented with back pain that had progressively worsened in the last month. She had routinely taken non-steroid anti-inflammatory drugs for pain as well as omeprazole and sucralfate. She also complained of ab-

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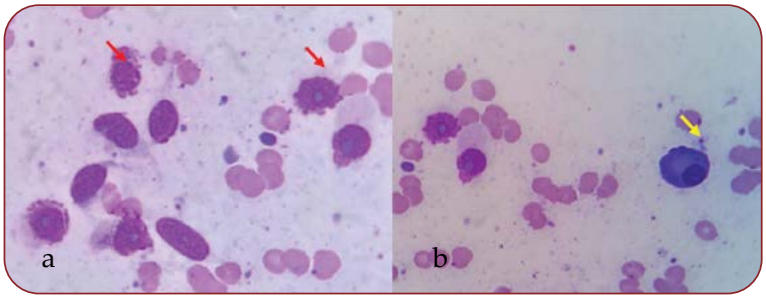

FIGURE 1. Bone marrow aspirate in Hematoxylin Eosin, 400x: (a) red arrow – histiocyte; and (b) yellow arrow – plasmocyte

dominal pain, discomfort, decreased appetite and fatigue. Unintentional weight loss of 5 kg occurred in the last month too. The woman looked pale and moderately ill.

Laboratory findings showed normocytic normochromic anemia (Hb 8.9 g/dL) and an increase in the erythrocyte sedimentation rate by 150 mm. The blood smear showed rouleaux formation. Plasma protein showed markedly increased globulin 8.83 g/dL (N: 1.80-3.90) with low albumin. Further studies discovered high calcium (11.4 g/dL) and monoclonal IgA lambda. The bone survey revealed multiple lytic lesions in the costae, parietal, femur and vertebrae. The bone marrow was hypercellular, with increased plasmocyte number (12.0%) and histiocytosis (24.5%) and suppressed erythropoiesis and thrombopoiesis. The patient was treated with thalidomide and dexamethasone, since at that time bortezomib was unaffordable for her. After two cycles, she got severe pneumonia due to bacterial infection. Her condition rapidly deteriorated during hospitalization. She died three months after being diagnosed with multiple myeloma. ❑

DISCUSSION

In this patient, histiocytosis can be reactive due to multiple myeloma. The most common finding is related with crystal storage histiocytosis (2). Plasma cell dyscrasias linked to CSH-associated with M paraprotein had been reported (2). The mechanism of crystal formation is not clearly understood. Several possible mechanisms include amyloid aggregation, overproduction of abnormal paraprotein, substitution of amino acids in the paraprotein structure and reduced degradation rate of paraproteins. The study conducted by Lebeau *et al* (3) showed that the possible mechanism was an amino acid substitution in the paraprotein structure, which led to hydrophobic interactions and conformational alteration of monoclonal protein. The structural changes in the paraprotein lead to crystallization and/or prevent degradation (4).

Shamanna *et al* (5) reported a case series of 13 patients with crystal storing histiocytosis and a median age of 60 years (range, 33-79). Bone marrow was the most frequently involved site related to plasma cell myeloma (4/13), followed by the gastrointestinal tract (3/13) and lymph node (2/13). Involved serum paraprotein was as follows: kappa in 50% of cases and lambda in 40% of cases. Singh Sachdeva *et al* (2) reported non-crystal storage histiocytosis. The incidence of this condition was rarely reported in the literature. There is no specific treatment for histiocytosis in myeloma patients. Therapy is directed to the underlying disease (2). ❑

CONCLUSION

The coincidence of myeloma with histiocytosis is an uncommon finding. The underlying mechanism remains unclear. ❑

Conflicts of interest: none declared.
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References

1. **van de Donk NWCJ, Pawlyn C, Yong KL.** Multiple myeloma. *The Lancet* 2021;397:410-427.
2. **Singh Sachdeva M, Das R, Ahluwalia J, Varma N.** Reactive histiocytosis: A diagnostic dilemma in multiple myeloma. *Indian J Pathol Microbiol* 2010;53:577-578.
3. **Lebeau A, Zeindl-Eberhart E, Müller EC, et al.** Generalized crystal-storing histiocytosis associated with monoclonal gammopathy: molecular analysis of a disorder with rapid clinical course and review of the literature. *Blood* 2002;100:1817-1827.
4. **Braunstein MJ, Petrova-Drus K, Rosenbaum CA, et al.** Plasma Cell Myeloma Presenting With Amyloid-Laden Crystal-Negative Histiocytosis. *Am J Clin Pathol* 2020;154:767-775.
5. **Kanagal-Shamanna R, Xu-Monette ZY, Miranda RN, et al.** Crystal-storing histiocytosis: a clinicopathological study of 13 cases. *Histopathology* 2016;68:482-491.