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Case report

Systemic mastocytosis with plasma cell dyscrasia: Report of a case

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Abstract

Systemic mastocytosis (SM) comprises a heterogeneous group of disorders characterized by infiltration of bone marrow and other tissues by neoplastic mast cells. A subset of patients with SM has associated hematologic malignancy usually of myeloid origin and comprises an entity termed systemic mastocytosis with associated clonal hematological non-mast cell lineage disease (SM-AHNMD) by the current WHO classification. Reports of clonal lymphoid malignancies associated with SM are rare. We describe a patient who was simultaneously diagnosed with indolent SM and a plasma cell dyscrasia fitting the definition of monoclonal gammopathy of undetermined significance (MGUS). We also discuss the pathologic interaction between the neoplastic mast cells of SM and the lymphoid/plasma cell malignancy when these two entities coexist.

Keywords: Systemic mastocytosis; Plasma cell dyscrasia; *KIT* mutation; Pathogenesis

1. Introduction

Systemic mastocytosis (SM) is characterized by abnormal collection of neoplastic mast cells in the bone marrow and other tissues. The criteria for diagnosis of SM have been developed by the World Health Organization (WHO). The major criterion for diagnosis is the histologic demonstration of multifocal dense infiltrates of mast cells in the bone marrow and/or in other extracutaneous organs. Minor criteria include the atypical mast cell morphology, aberrant expression of CD2 and/or CD25 on bone marrow mast cells, presence of *KIT* mutations at codon 816 (mostly D816V) in an extracutaneous organ, and a persistently elevated serum tryptase level (>20 ng/ml). Diagnosis of systemic mastocytosis is made when one major and one minor criteria or three minor criteria are fulfilled [1,2].

A subset of patients with SM has associated hematological malignancies and this entity is termed systemic mastocytosis with associated clonal hematological non-mast cell disease (SM-AHNMD) by the WHO. Of the hematological malignancies that are associated with SM, the majority are of myeloid origin and include acute myeloid leukemia, myeloproliferative disorders and myelodysplasia [3–5]. Although extremely rare, lymphoid and plasma cell malignancies including chronic lymphocytic leukemia, Hodgkin's Lymphoma and multiple myeloma have also been reported in association with SM. [6–10]. We describe a patient who was concurrently diagnosed with SM and monoclonal gammopathy of undetermined significance (MGUS) and discuss the association of SM with lymphoproliferative disease.

2. Case report

An 84-year-old man with renal failure of unclear etiology and longstanding anemia was seen with complaints of recent rapid weight loss of 20 pounds. Physical examina-

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tion revealed erythematous, maculopapular hyperpigmented lesions consistent with urticaria pigmentosa involving the trunk and pretibial regions. Lymphadenopathy and hepatosplenomegaly were absent. Laboratory studies revealed hemoglobin of 9 gm/dL, white blood cell (WBC) count of 10.3×10^9/L and platelet count of 490×10^9/L. Serum chemistry showed a creatinine of 4.3 mg/dL. Serum protein electrophoresis showed IgG lambda monoclonal gammopa-

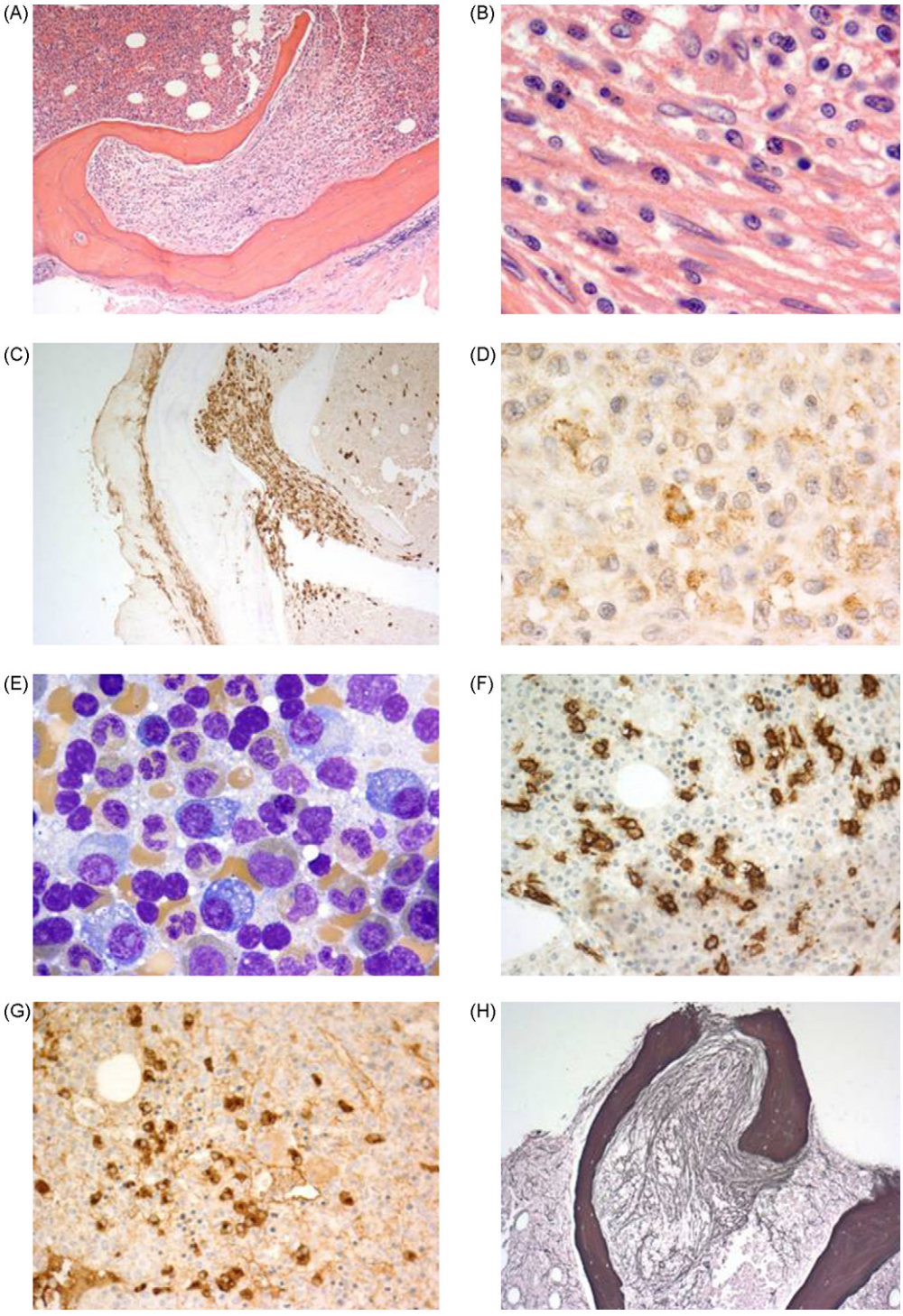

Fig. 1. Bone marrow biopsy section stained with hematoxylin and eosin showing markedly hypercellular bone marrow with paratrabecular mast cell aggregate (A, ×100). Spindle shaped atypical mast cells are shown at higher magnification (B, ×400). Immunohistochemically, the mast cells are positive for CD117 (C, ×100) and show aberrant CD2 expression (D, ×400). Bone marrow aspirate stained with Wright Giemsa stain showing increased numbers of plasma cells (E, ×500). Bone marrow biopsy showing increased plasma cells comprising approximately 10% of the cellular elements and are highlighted by CD138 staining (F, ×400). These plasma cells showed lambda monotypic staining pattern (G, ×400). 4+ Reticulin fibrosis was present (H, ×100).