Review of the clinical and pathological panoply of systemic mastocytosis.

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Abstract

Mastocytosis is a rare disease with varied presentation, myriad symptomatology and variable prognosis. Most patients present with cutaneous disease and mediator-related symptomatology with a small subset having systemic disease (systemic mastocytosis, SM). A subset of the latter develops synchronous or metachronous haematologic neoplasms (SM-AHN), most commonly chronic myelomonocytic leukaemia (CMML). Advanced systemic mastocytosis (ASM) is seen in a relatively small number of patients and is usually associated with organ dysfunction, and may present with hepatosplenomegaly, lymphadenopathy and ascites with progression to leukaemic transformation (mast cell leukaemia/acute myeloid leukaemia) occurring in a few patients. This paper discusses the clinical and pathologic features of the entire spectrum of SM in adults.

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