

Bone marrow necrosis with Charcot-Leyden crystals in a patient with idiopathic hypereosinophilic syndrome

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A 22-year old man presented with a 20-day history of fever, asthenia, weight loss, and bone pain. He showed liver enlargement and interstitial pulmonary edema. The leukocyte blood count was $26.7 \times 10^9/L$, with 34% eosinophils ($9.1 \times 10^9/L$) and no blasts. The bone marrow examination demonstrated increased cellularity with 90% eosinophilic forms (Figure 1). Cytogenetic studies were normal and the BCR/ABL fusion was not detected. No conditions causing reactive eosinophilia were found, and the diagnosis of idiopathic hypereosinophilic syndrome was established.¹ Twenty-four hours later, the patient experienced a severe clinical deterioration. A second bone marrow examination revealed severe necrosis, with few remaining eosinophils, and abundant Charcot-Leyden crystals (Figure 2). These structures were bipyramidal and measured about 15-40 μm in length and 2-10 μm in width; they were hexagonal at transversal section. The patient was treated with prednisone, hydroxyurea and vincristine, and responded favorably in a few days. During the long term follow-up of thirteen years the patient received maintenance treatment with very low doses of prednisone and hydroxyurea and led a normal life. He maintained his eosinophil levels between 0.3 and $1.4 \times 10^9/L$. Charcot-Leyden crystals have been described in focal accumulations of eosinophils such as granulomas associated with parasitic diseases, sputa of patients with bronchial asthma and myeloid leukemia.^{1,2} Ultrastructural and biochemical studies have demonstrated that these structures are derived from the granules of the eosinophil leukocytes. The main constituent of the crystals is a protein of the carbohydrate-binding

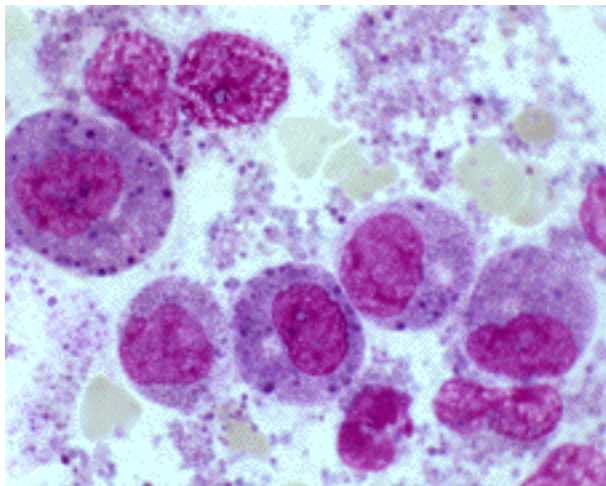


Figure 1. First bone marrow aspirate (May-Grünwald-Giemsa, x 1,000). High predominance of eosinophilic forms, mainly myelocytes and metamyelocytes, with different degrees of degranulation; the granules are dispersed in the background.

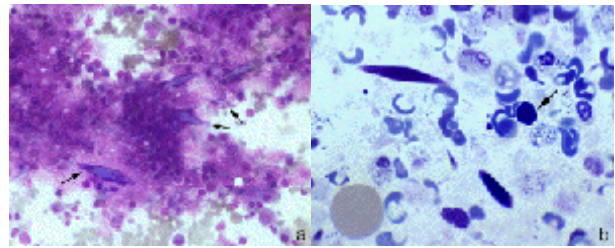


Figure 2. Second bone marrow aspirate. a) Cytological smear stained with May-Grünwald-Giemsa (x 200): the marrow is necrotic, with only a few cells that are preserved, and shows a number of Charcot-Leyden crystals of different sizes (arrows); b) Thick resin epoxy section stained with toluidine blue (x 400): the crystals reveal their characteristic bipyramidal shape; a transversally sectioned crystal shows the typical hexagonal contour (arrow).

family of galectins that has lysophospholipase activity. It has also been demonstrated that this protein can crystallize to form the bipyramidal hexagonal Charcot-Leyden crystals.

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