

Acute Cutaneous Flare and Hemophagocytosis: An Unusual Presentation of Adult Anaplastic Large Cell Lymphoma

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Abstract

Hemophagocytic lymphohistiocytosis (HLH), also known as hemophagocytic syndrome, is a disorder characterized by the pathologic activation and proliferation of histiocytes, mainly in the bone marrow, liver, spleen and lymph nodes. Although HLH is frequently associated with lymphomas, it is a rare presentation of anaplastic large cell lymphoma (ALCL). We report a case of anaplastic large cell lymphoma (ALCL) in an adult manifested as generalized cutaneous nodules and fulminant hemophagocytosis.

Keywords: Hemophagocytic lymphohistiocytosis; Anaplastic large cell lymphoma

Introduction

Hemophagocytic lymphohistiocytosis (HLH) is an aggressive and potentially life-threatening disease most often affecting infants from birth to 18 months of age, but cases in older children and adults have been reported [1]. It is due to cytokine dysfunction causing immune dysregulation. HLH can be inherited or acquired; the latter being most commonly associated with infections, autoimmune disorders, hematological malignancies or immunocompromised states [2].

Case Report

A 54-year-old African American man was with a history of chronic ulcerating skin lesions presented with fever, weight

loss, and worsening cutaneous ulcers. Two years prior to his presentation, he developed multiple areas of hyperpigmented skin nodules and extensive exophytic plaques. Multiple skin biopsies had shown activated lymphocytes without atypia.

Physical examination showed diffuse hyperkeratotic scaly papules with central ulceration and yellow drainage. Multiple subcentimeter lymph nodes were palpable in the cervical, axillary and inguinal areas. Hemoglobin was 7.8 g/dL, leukocytes $2.0 \times 10^9/L$ and platelets count $47 \times 10^9/L$. Serum ferritin level was 27,591 ng/mL (reference range (15 - 400 ng/mL)). Plasma fibrinogen level was 150 mg/dL (reference range (210 - 634 mg/dL)). A computed tomography scan revealed a homogeneous enlarged liver measuring 21 cm, and enlarged inguinal lymph nodes, the largest measuring $3.6 \times 1.2 \times 2$ cm.

The patient was empirically treated with antibiotics and high dose steroids daily. A bone marrow biopsy demonstrated histiocytic hyperplasia with hemophagocytosis (Fig. 1a), and a diffuse infiltration of atypical lymphoid cells that were CD3-, CD4+, CD30+, ALK-, CD20- and CD163+ (Fig. 1b-d) favoring a diagnosis of anaplastic large cell lymphoma (ALCL). The patient status worsened despite empiric steroid therapy, and he expired of multiorgan failure four weeks after hospitalization.

Discussion

This is an unusual case of anaplastic large cell lymphoma (ALCL) that presented as generalized cutaneous nodules and fulminant hemophagocytosis. The fever along with pancytopenia, elevated ferritin, hypofibrinogenemia, and hemophagocytosis in the bone marrow confirmed the diagnosis of HLH in our patient. This was the initial presentation of the underlying lymphoproliferative disease. HLH secondary to malignant lymphoma usually manifest with a fulminant clinical course masking the underlying malignancy. Consequently, the possibility of a masked malignant disease should always be considered and ruled out by means of biopsies of involved sites and bone marrow.

HLH secondary to ALCL is extremely uncommon in adults, and only few cases of pediatric HLH secondary to ALCL have been reported [3]. As for the cutaneous aspect

Manuscript accepted for publication July 20, 2012

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doi: <http://dx.doi.org/10.4021/jh35e>

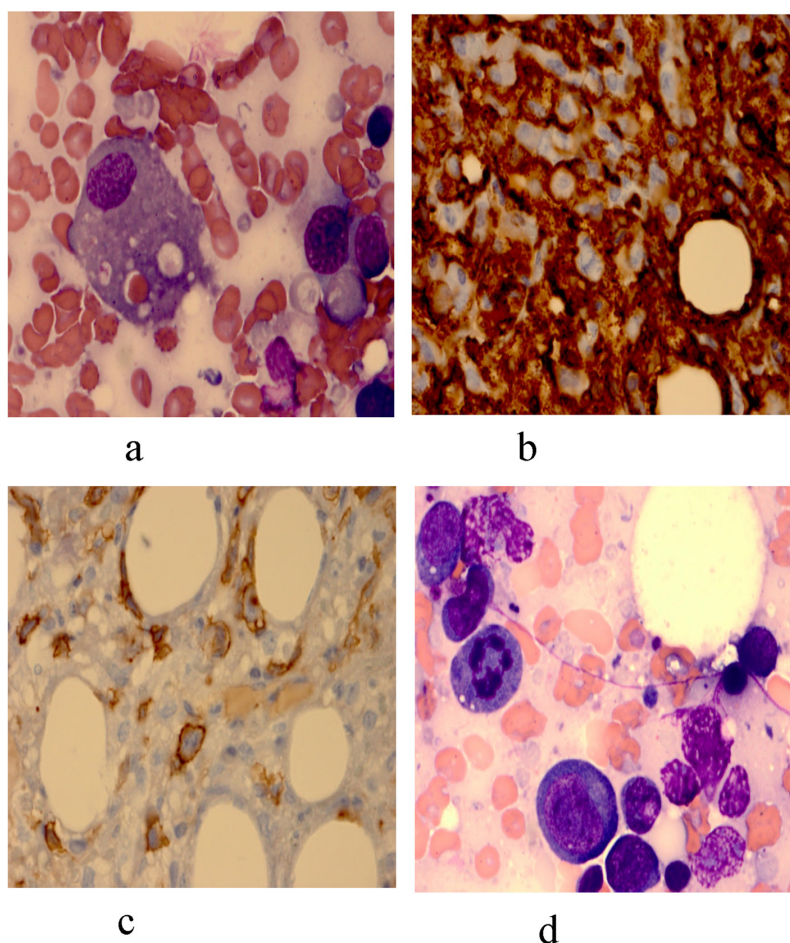


Figure 1. (a): Bone marrow aspirate revealing hemophagocytosis; (b): Histiocytic hyperplasia highlighted by CD163 immunostain; (c): The lymphoid cells marked with CD30 immunostain; (d): Anaplastic lymphoma cells, one in mitosis.

of this disease, a single case of diffuse hyperpigmented eruption as a manifestation of systemic ALCL has been described in the literature [4]. Unusual cutaneous lesions in presence of features of bone marrow failure should alert the internist about the possibility of an underlying malignancy.

Disclosure Statement

All authors have no disclosures.

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