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CASE REPORT

Peripheral blood hemophagocytosis in an unusual lymphoma

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Key Clinical Message

We describe a patient with fever, pancytopenia, and hepato-splenomegaly associated with the finding of neoplastic lymphoid cells and histiocytes with hemophagocytosis in the peripheral smear; the diagnostic features were suggestive for a biological overlap between a large B-cell lymphoma with intravascular involvement and the Asian variant of intravascular B-cell lymphoma.

Keywords

Diffuse large B-cell lymphoma, hemophagocytic syndrome, intravascular lymphoma, peripheral blood smear examination.

Introduction

A 80-year-old Caucasian man was hospitalized because of fever and pancytopenia (hemoglobin 6.8 g/dL, Mean Corpuscolar Volume 93 fL, reticulocytes 1.8%, platelet 27,000 per cubic millimeter, leukocyte 1910 per cubic millimeter). The physical examination revealed hepato-splenomegaly but no peripheral lymphoadenopathy. There was no evidence of Central Nervous System involvement. Serological tests for Epistein-Barr virus, Cytomegalovirus, Hepatitis B and C virus, and Human Deficency virus were negative. The lactate dehydrogenase (LDH) level was markedly elevated (>1000 UI/L); hyperferritinemia (1100 ng/mL) and hypertriglyceridemia (434 mg/dL) was also found. The peripheral smear showed occasional large lymphoid cells with basophilic cytoplasm (Fig. 1A and B) and histiocytes with hemophagocytosis (Fig. 2A–D). Bone marrow biopsy revealed large atypical $CD20^{+}CD10^{-}$ $CD5^{-}$ lymphoid cells with a nodular and intrasinusoidal pattern of involvement and histiocytes with hemophagocytosis. A total body CT scan, confirmed hepato-splenomegaly, showed a pleural effusion and the presence of thoracic and abdominal lymphoadenopathy with a long axis less than 1.5 cm, with the exception of a mediastinal node with a long axis of 2.5 cm. Chemoimmunotherapy (bendamustine + rituximab) was given; after six cycles, the patient had a complete response, confirmed with whole-body CT scan and PET/CT findings. The clinical picture was suggestive of a lymphoma-associated hemophagocytic syndrome (HPS), as found in the intravascular

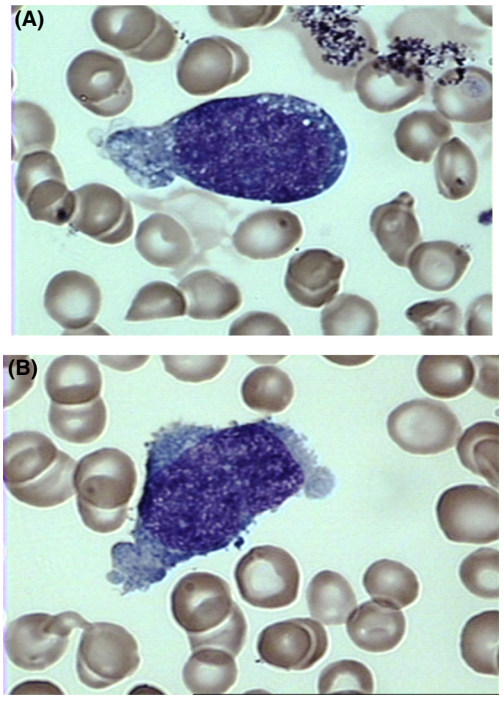

Figure 1. Lymphoblast in peripheral blood. (A, B)

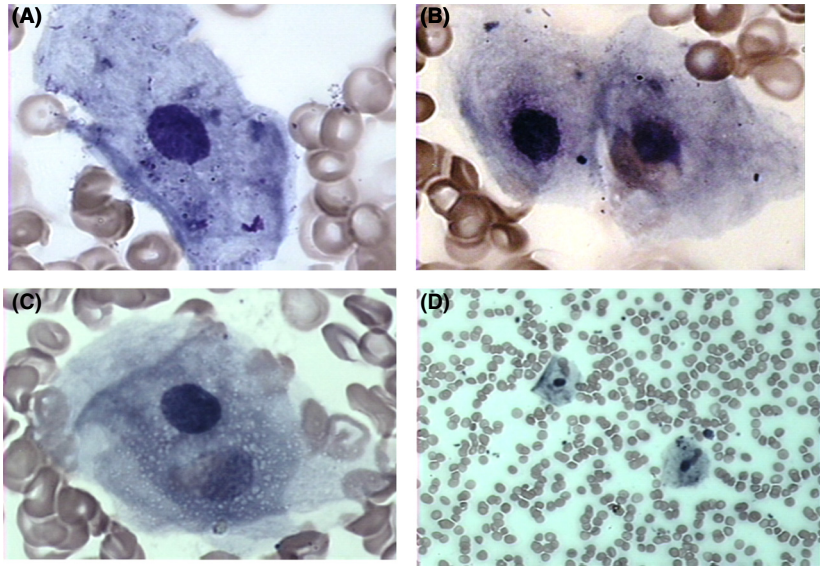

Figure 2. (A, C) Histiocytes with hemophagocytosis in peripheral blood. (B, D) Histiocytes in peripheral blood.

B-cell lymphoma (IVBL). IVBL is a rare and aggressive subtype of extranodal diffuse large B-cell lymphoma, characterized by proliferation of neoplastic lymphoid cells exclusively within the lumina of small vessels. The clinical manifestations of this disease are variable: there are some differences between patients diagnosed in Asian and Western countries [1]. CNS and skin involvement are more often seen in the Western population, whereas Asian patients show HPS with fever, hepato-splenomegaly, bone marrow involvement, and cytopenia. The "Asiant variant" is rarely reported also in Caucasian patients [2–5]. Therefore the true difference between the two forms is the presence or absence of hemophagocytosis, rather than geographic region of the patients. Peripheral blood involvement is uncommon in all subgroups; atypical lymphoid circulating cells may be occasionally observed [6]. The diagnosis is often a real challenge and many cases are discovered in postmortem examination. In IVBL the lymphoma cells are found exclusively within the lumen of small blood vessels and lymphoadenopathy is usually absent. In the case of this patient, we found a mixed intrasinusoidal and nodal pattern of involvement by lymphoma cells and a lymphoadenopathy, however small, was present. Therefore, we may consider this case as a biological overlap between a large B-cell lymphoma with intravascular involvement and the Asian variant of intravascular large B-cell lymphoma: the presence of a HPS was the pathogenetic factor characterizing the clinical picture. A similar finding was previously described by others [7]. Besides, in this patient, we found the unique feature of histiocytes with hemophagocytosis also in peripheral blood: this is the first report of such a peculiar finding. A careful examination of the peripheral smear has allowed an early diagnosis and effective treatment.

Conflict of Interest

None declared.

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