CASE REPORT

Cytophagic histiocytic panniculitis: A diagnostic challenge

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ABSTRACT

Cytophagic histiocytic panniculitis (CHP) was first described in 1980. It classically presents with intermittent recurrent episodes of pyrexia subcutaneous nodules, pancytopenia and liver dysfunction, but most patients have in addition a multitude of other manifestations which confuse the clinical picture. Despite the variable clinical course, the disease is often fatal. Diagnosis is based on histological features. A lobular panniculitis with an infiltrate of cytologically benign cytophagocytic histiocytes in skin nodules is indispensable for establishing the correct diagnosis. We report case of CHP occurring in a 52-year-old male.

INTRODUCTION

Cytophagic histiocytic panniculitis (CHP), a disorder described by Winkelmann et, al and Crotty et, al, is characterized by lobular panniculitis and infiltration with histo lymlogically benign histiocytes showing erythrophagocytosis and lymphophagocytosis.[1, 2, 3] The major clinical features are recurrent fever, multiple panniculitic lesion, anemia, leukopenia and coagulation abnormalities.[4] The clinical course is often fatal, and liver failure, hemorrhagic diathesis, or infection are the main causes of death, but some patients with CHP with a benign clinical course have also been described.[3] In CHP, the following clinical and laboratory features are associated with a poor prognosis: bleeding, mucosal ulcers, lymphadenopathy, fever, hepatosplenomegaly, serosal effusions, anemia, leukopenia, elevated liver enzyme levels, coagulopathy and hypercalcemia.[3] Here, we describe a patient with cytophagic histiocytic panniculitis which could be a diagnostic dilemma.

CASE REPORT

A 52-year-old male presented with high fever and furuncle like lesions on the face and extensive ulcerated nodules on the body. The condition started a month before presentation with a single lesion which started to spread out despite treatment. The patient had a similar episode few months ago, however milder and resolved by treatment, probably antibiotics. There was no history of any systemic illness. On examination he had multiple tender subcutaneous nodules with ulceration. The ulcers were filled with thick yellowish material (Fig. 1). The lesions were very extensive affecting the face, trunk, upper and lower extremities. Full blood count revealed leucocytosis with neutrophilia, lymphopenia and mild anemia. Total bilirubin was elevated and albumin decreased but ALT and AST was normal.

ESR and fasting blood sugar was elevated. Staph aureus grew on blood culture. Coagulation profile showed slightly elevated parameters. ANA, ANCA, Coomb's test (direct and indirect) were negative. Antibodies against HIV 1,2, CMV and EBV were negative as well as hepatitis profile. α_1-antitrypsin was normal. Bence Jones protein in the urine was not detected. Chest X-ray showed mild pleural effusion. Histopathologic examination of a skin biopsy revealed focal infiltration of

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neutrophils in the epidermis and very dense infiltration of neutrophils, lymphocytes and hitiocytes in the dermis.

The subcutaneous fat showed lobular panniculitis with infiltration of neutrophils, lymphocytes and histiocytes (Fig. 2). Few histiocytes showed haemophagocytosis with fragmented erythrocytes, leucocytes or lymphocytes in their cytoplasm (bean bag) appearance (Fig. 3). There was no evidence of vasculitis. According to the above data the diagnosis of cytophagic histiocytic panniculitis was established.

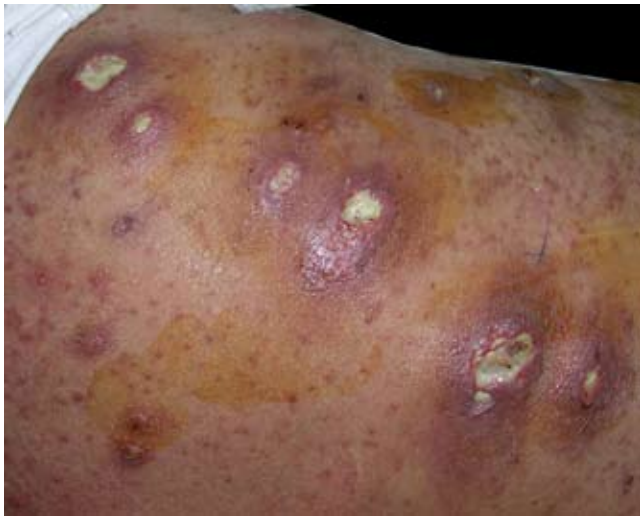

Fig. 1 Ulcerated nodules filled with thick yellowish material.

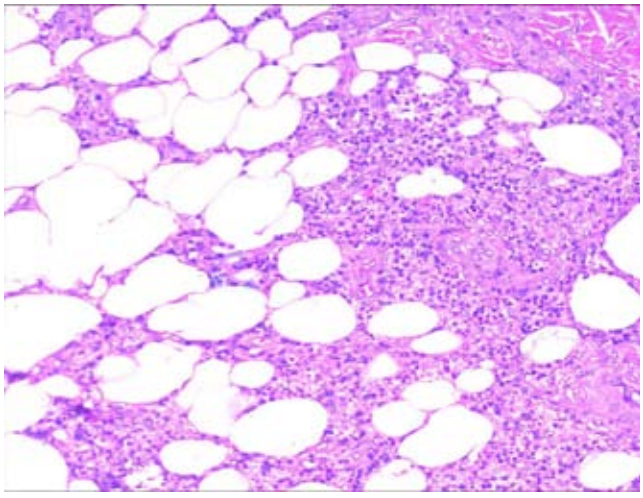

Fig. 2 Lobular panniculitis with fat tissue necrosis.

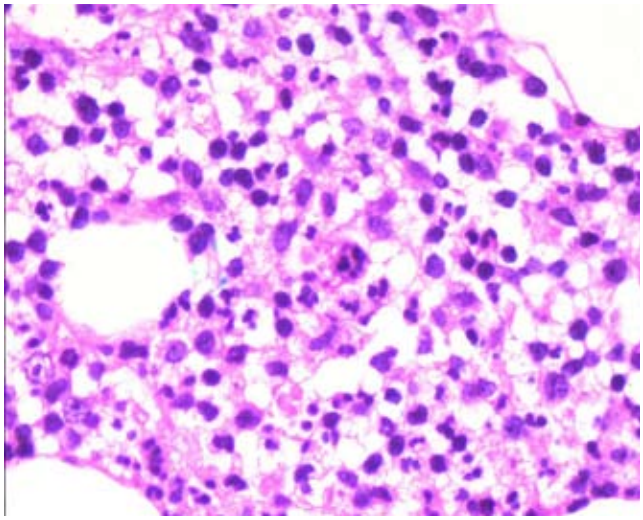

Fig. 3 Some histiocytes show "bean-bag"appearance.

DISCUSSION

Cytophagic histiocytic panniculitis (CHP) was first described by Winkelmann and Bowie in 1980.[1] It classically presents with intermittent recurrent episodes of pyrexia and self healing panniculitis. It may run a benign chronic course, but rapid aggressive disease has been reported.[5] Rapid deterioration and death is usually due to the development of the haemophagocytic syndrome and subsequent hemorrhagic diathesis.[2] It seems that CHP includes two distinct enteties: subcutaneous panniculitis-like T-cell lymphoma (SPTCL) and the authentic CHP.[5] Histologically, CHP is characterized by a lobular panniculitis

with an infiltrate of small, benign, mature lymphocytes and benign histiocytes.[1] Cytophagic histiocytic panniculitis and the entities in the SPTCL group are the major differential diagnoses in cases of recurrent self-healing panniculitis where pancreatic disease, infection, vasculitis, connective tissue diseases, tuberculosis, sarcoidosis, trauma, α_1-antitrypsin deficiency or injection of substances into the panniculus have been excluded.[5] As for the treatment , most cases have been reported to achieve temporary remission with chemotherapy, immunosuppressive agents or radiotherapy, but

when elapses occur they are followed by rapidly fatal or indolent course.[7] The prognosis of CHP is variable.[4] Patients with SPTCL generally have a poor prognosis, even when treated with corticosteroids or chemotherapy.[5]

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