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CASE REPORT

Hairy cell leukaemia presenting with ascites, pleural effusion and increased CA 125 serum level

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ABSTRACT

The body cavities are rarely involved in hairy cell leukaemia. Here we report a patient who had pancytopenia, hepatosplenomegaly, massive haemorrhagic ascites, pleural effusion at the left hemithorax and increased CA 125 serum level at the time of initial diagnosis. Laparoscopy showed multiple nodular white, opaque lesions on the omentum and on the parietal peritoneum. Laparoscopic biopsy of these lesions, and a bone marrow biopsy revealed a diffuse cellular infiltrate of tartrate-resistant acid phosphatase staining mononuclear cells. These mononuclear cells with irregular cytoplasmic protrusions were also found in the peripheral blood, in the ascites fluid and in the pleural effusion. The patient was treated with cladribine 0.1 mg/kg/day with continuous infusion for seven days. Three months after the treatment, the patient achieved a complete remission with normalisation of the peripheral blood count, bone marrow findings, CA 125 serum level, with no detectable ascites and/or pleural effusion.

KEYWORDS

Ascites, CA 125, hairy cell leukaemia, peritoneal hairy cell infiltration

INTRODUCTION

Hairy cell leukaemia (HCL) is a chronic lymphoproliferative disorder characterised by splenomegaly, cytopenia and the presence of malignant B-cells with hair-like protrusions in peripheral blood, bone marrow, spleen and liver.[1] Rarely the patients may present with marked leucocytosis, spontaneous rupture of the spleen, cryptococcal meningitis, massive splenomegaly due to hairy cell infiltration but with normal peripheral blood and bone marrow findings at the time of initial diagnosis,[2-4] but not with massive ascites and pleural effusion. The occurrence of massive ascites and pleural effusion in HCL has been reported as a complication during the course of the disease.[4,5]

Here, we report a patient who presented with massive haemorrhagic ascites and haemorrhagic pleural effusion at the left hemithorax, an elevated CA 125 serum level and a multiple nodular infiltration of the peritoneum with hairy cells at the time of initial diagnosis. To our knowledge, haemorrhagic ascites and pleural effusion with elevated CA 125 level have not been reported previously as initial findings in this disorder.

CASE REPORT

A 49-year-old man was referred to our department because of pancytopenia and ascites. Three months prior to admission he started to experience easy fatigue, abdominal swelling and slight shortness of breath. On examination, the spleen and the liver were palpable 10 cm and 6 cm below the costal margins, respectively. In addition to these findings, he had massive ascites and pleural effusion at the left hemithorax. No superficial lymph node enlargements were noted. An X-ray of the chest showed elevation of the diaphragm and pleural effusion at the left hemithorax. A computed tomography confirmed the presence of hepatosplenomegaly, ascites and pleural effusion. There was no lymphadenopathy.

Peripheral blood count revealed haemoglobin (Hb) 4.6 mmol/l, platelets 86 x 10^9/l, and white blood cell count (WBC) 4.1 x 10^9/l. The differential showed 36% neutrophils, 20% lymphocytes and 44% mononuclear cells with irregular cytoplasmic protrusions. The bone marrow was aspirated with difficulty, and the sample showed typical hairy cells. A bone marrow biopsy revealed a diffuse cellular infiltrate of tartrate-resistant acid phosphatase (TRAP) stained mononuclear cells. These cells were B cells, staining with the pan B-cell marker CD20 (*figures 1A-C*).

Figure 1A. *Neoplastic infiltration in bone marrow trephine biopsy, haematoxylin and eosin x 20*

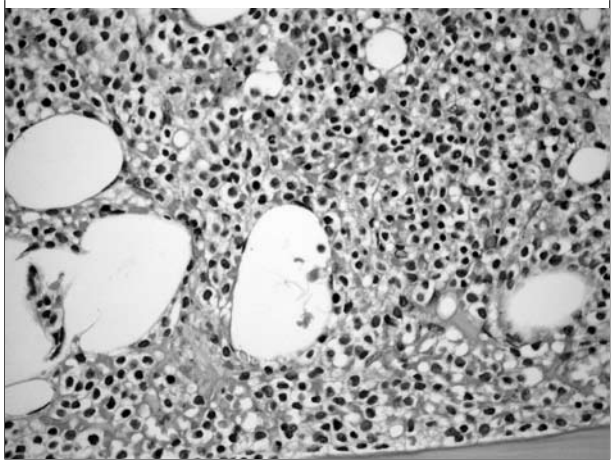


Figure 1B. *Neoplastic infiltration in the bone marrow with CD20 immune reactivity, anti-CD20 primary antibody (L 26), aminoethylcarbasol (AEC) chromogen x 20*

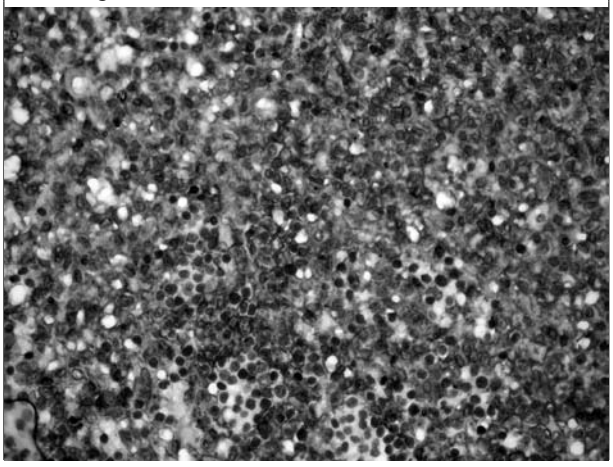
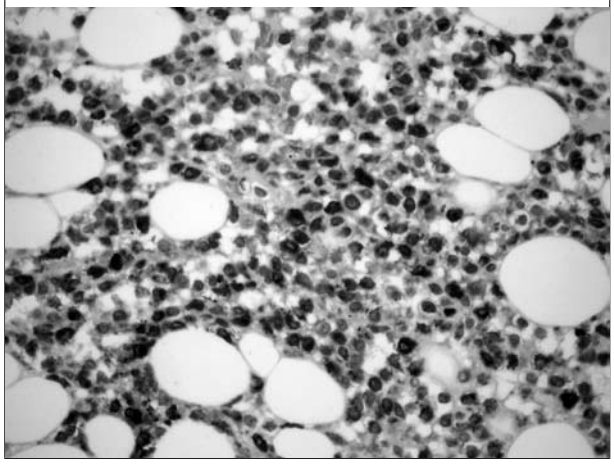


Figure 1C. *Neoplastic infiltration in the bone marrow biopsy with TRAP immune reactivity, AEC chromogen x 40*



Immunophenotyping of the peripheral blood demonstrated the presence of HCL-specific antigen expression (CD19 89%, CD20 60%, $CD11^c$ 84%, CD25 88% and CD103 78%).

In the haemorrhagic ascites, total protein was 3.6 g/l with 1.9 g/l albumin, Hb 0.6 g/l, WBC 0.9 x 10^9/l, with 60% typical hairy cells using Wright's stain. Pleural effusion was similar in appearance and in chemistry to the ascites fluid. Bacterial culture of both fluid samples remained sterile. The liver function studies were normal except for an elevated polyclonal γ-globulin level (1.6 g/l). Serum total protein and albumin levels were normal (5.5 g/l and 2.6 g/l, respectively). The CA 125 serum level was found to be elevated (272 IU/ml; normal 0 to <30 IU/ml).

The laparoscopic examination showed multiple nodular white, opaque lesions on the omentum and on the parietal peritoneum (*figure 2*). Laparoscopic peritoneal biopsy of these lesions revealed a dense infiltration with TRAP positive hairy cells. These cells were positive for the B-cell marker CD20 (*figures 3A-C*).

The patient was treated with cladribine 0.1 mg/kg/day with continuous infusion for seven days. The result obtained with this treatment was very satisfactory. The ascites and pleural effusion subsided with cladribine therapy. The patient is still asymptomatic and has no detectable hepatosplenomegaly, ascites and/or pleural effusion with a normal CA 125 serum level, normal bone marrow findings and peripheral blood levels (Hb 9.6 g/l, platelets 310 x 10^9/l, WBC 5.8 x 10^9/l) after three months of therapy.

Figure 2. *Multiple nodular lesions on the peritoneum as seen in laparoscopy*

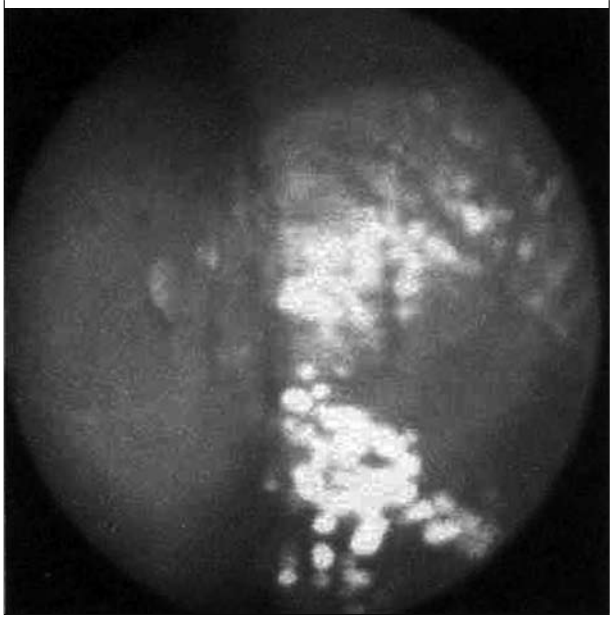


Figure 3A. *Peritoneal biopsy: Mesothelial surface and neoplastic lymphoid infiltration, haematoxylin and eosin x 40*

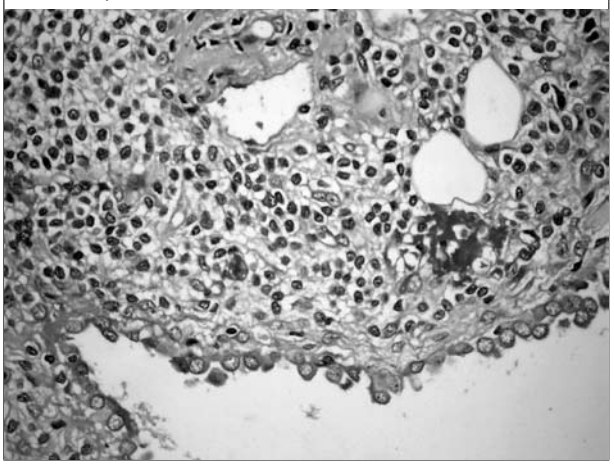


Figure 3B. *CD20 positive neoplastic lymphoid infiltrations in the same biopsy as in figure 3A, AEC chromogen x 40*

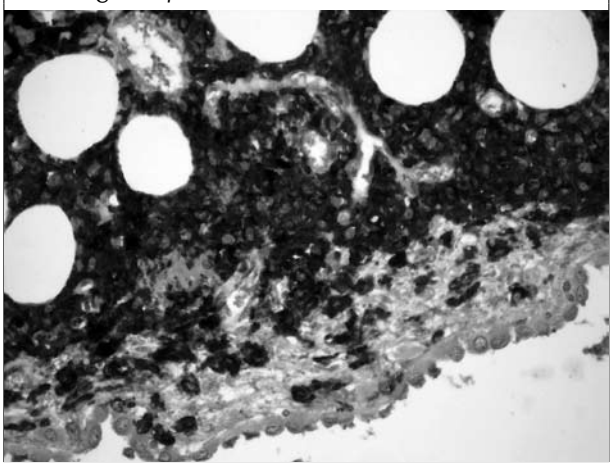
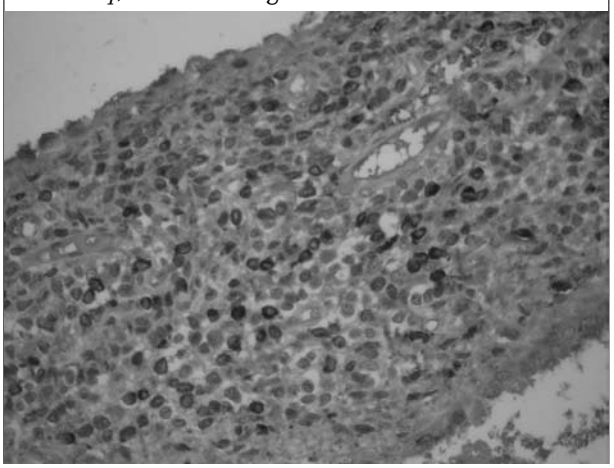


Figure 3C. *Neoplastic cells with TRAP immune reactivity, AEC chromogen x 20*



DISCUSSION

This report describes a case of HCL. Clinically, the patient presented with massive ascites, pleural effusion at the left hemithorax and hepatosplenomegaly. Ascites and pleural effusion are unusual manifestations of HCL at the time of initial diagnosis. Several case reports of HCL describe ascites, but in each, there was histologically proven cirrhosis of the liver.[6,7] Only three patients with HCL have been reported in the literature who developed ascites during the course of their illness.[4,5] In one of these patients, chylous ascites appeared 16 months after the diagnosis of hairy cell leukaemia.[5] In the other two patients, massive ascites and pleural effusion developed 10 and 13 months after the diagnosis.[4] Ascites was not chylous. All of the patients had massive retroperitoneal and abdominal lymphadenopathy. According to the authors, the cause of the massive ascites in these patients showed a similarity to that seen in lymphomas and was most likely related to the extensive lymphadenopathy.

To our knowledge, massive ascites and pleural effusion, in association with elevated CA 125 serum levels, as the initial manifestation of HCL, has not been reported previously. In our experience, this is the first patient in whom the multiple nodular peritoneal infiltration was documented by laparoscopy, and the diagnosis of HCL was confirmed by laparoscopic biopsy.

CA 125 is a glycoprotein antigen expressed in the celomic epithelium. It is a tumour marker used for the diagnosis and monitoring of epithelial ovarian carcinoma.[8,9] This marker has also been found to be increased in patients with serosal effusions, derived from non-neoplastic inflammatory disease,[10,11] in advanced non-Hodgkin's lymphoma,[12,13] and in acute leukaemia[14] with serosal involvement. Serial CA 125 measurements may be of value in monitoring response to chemotherapy in these patients.

CA 125 elevation in our patient is most likely to be due to a serosal reaction caused by the leukaemic infiltration. After the cladribine treatment, our patient showed a complete haematological and clinical response with normalisation of the peripheral blood cell count, the CA 125 serum level, disappearance of the ascites, the pleural effusion and, hepatosplenomegaly.

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