A case of hairy cell leukemia resembling asymptomatic chronic liver disease on presentation

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Abstract

Hepatic infiltration in hairy cell leukemia is quite common but results of liver test are usually normal. A 44-year-old man with a history of alcohol abuse was evaluated for persistent elevation in serum aminotransferases. A percutaneous liver biopsy showed an extensive mononuclear cell infiltrate in the sinusoids and this finding led to the diagnosis of hairy cell leukemia. Liver involvement in hairy cell leukemia is discussed. Hairy cell leukemia should be considered as a potential cause of persistent increase in liver-related enzymes. (Acta gastroenterol. belg., **1998**, 61, **483-484**).

Key words : hairy cell leukemia, liver enzymes, sinusoidal infiltration.

Introduction

Hairy cell leukemia is a rare lymphoproliferative disorder with a generally indolent course, accounting for approximately 2% of all cases of leukemia (1). Hepatic involvement is quite common but results of liver tests are usually normal (2). Chronic liver disease was suspected in an asymptomatic patient who presented with thrombocytopenia and persistent abnormalities in liver transaminases. A liver biopsy showed a mononuclear cell infiltrate in the sinusoids and this finding led to the diagnosis of hairy cell leukemia.

Case report

A 44-year-old man was in a good state of health until late 1995 when he presented with unspecific dyspepsia. A barium esophagogram showed a sliding hiatal hernia and a laboratory work-up revealed an elevation in liver-related enzymes and thrombocytopenia. Antacids were started, symptomatic control being achieved and he was referred to the Liver Unit for further evaluation. Medical history was unremarkable except for alcohol abuse (more than 100 g/day) several years previously. Physical examination showed a well nourished man without stigmata of chronic liver disease. The liver and spleen were not felt. Laboratory evaluation revealed a hemoglobin of 14.9 g/dl, leukocyte count of 5.4 $\times$ 10^9/L (53% neutrophils ; 38% lymphocytes ; 8% monocytes), platelet count 73 $\times$ 10^9/L, prothrombin activity of 73% and ESR of 6 mm per hour. His serum AST was 56 U/L (normal 5-37 U/L), serum ALT was 84 U/l (normal 5-41 U/L), and serum GGT was 91 U/L (normal 11-49 U/L). Results of serum protein electrophoresis revealed the following values : albumin, 4,9 g/dL (normal 3-5 g/dL) ; α_1-globulin, 0,2 g/dL (normal 0,2-0,4 g/dL) ; α_2-globulin, 0,5 g/dL (normal 0,4-0,7 g/dL) ; ß-globulin, 0,7 g/dL (normal 0,7-0,9 g/dL) ; and γ-globulin, 1 g/dL (normal 0,9-1,5 g/dL). His serum IgG was 1290 mg/dL (normal 800-1700 mg/dL), serum IgA was 262 mg/dL (normal 85-490 mg/dL), and serum IgM was 131 mg/dL (normal 50-370 mg/dL). Values for total bilirubin, alkaline phosphatase, copper, ceruloplasmin, iron, transferrin and ferritin levels were all within the normal range. Screening for hepatitis virus B and C, human immunodeficiency virus infection and autoimmune liver disorders was negative. Abdominal ultrasound showed mild hepatosplenomegaly and an upper gastrointestinal endoscopy disclosed no varices. Sections from a transjugular liver biopsy were striking by the presence of a mononuclear cell infiltrate in the sinusoids, being mainly B-cells (CD 20 +), and mild sinusoidal congestion (fig. 1). Occasionally the nuclei of these cells was surrounded by a clear rim of abundant cytoplasm giving a "halo" appearance. Stains for tartrate-resistant acid phosphatase in blood and bone marrow were positive. In addition, immunophenotyping were also consistent (CD 103 +) with the diagnosis of hairy cell leukemia.

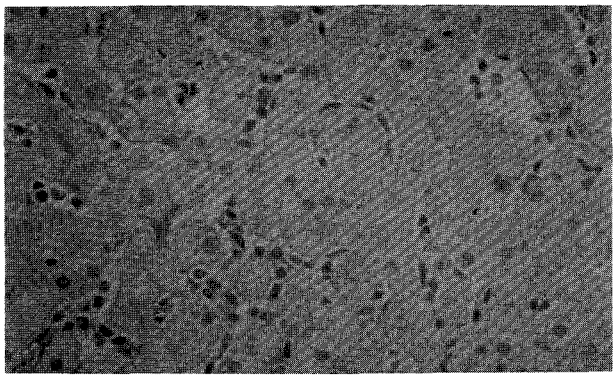

Fig. 1. — Hairy cell leukemia in the liver. Hepatic sinusoids filled with leukemic cells. Hepatocellular changes are minimal (H.E. $\times$ 250).

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Discussion

Asymptomatic chronic liver disease was suspected in our patient as he had a history of alcohol drinking, persistent increase in liver-related enzymes and low platelet count. Liver biopsy findings showed an extensive sinusoidal infiltration by mononuclear cells and subsequent studies confirmed the diagnosis of hairy cell leukemia.

Hairy cell leukemia is a rare B-cell disorder which affects males more often than females, with a peak incidence in the fifth and sixth decades (3,4). Patients typically present with splenomegalia and a chief complaint of abdominal fullness or discomfort (1,5). Other patients experience mostly fatigue, weakness or weight loss or present with bacterial or opportunistic infection secondary to severe granulocytopenia and monocytopenia, whereas in about one-fourth, the disease is incidentally discovered during evaluation for an unrelated cause (1). In addition, about two-thirds of the patients have moderate to severe pancytopenia at the time of diagnosis, but in some cases hematologic parameters are normal or there is isolated thrombocytopenia (3). Splenomegaly is palpable in about 85 percent of all patients at diagnosis and this is the most prominent physical finding (1,3). Enlargement of superficial lymph nodes is rare and, when present, is minimal and localized, although mesenteric and retroperitoneal adenopathy often can be detected on computed tomography of the abdomen (6). The cytochemical demonstration of acid phosphatase which is resistant to treatment with tartrate in blood and bonemarrow is a useful diagnostic procedure as this is quite sensitive for hairy cell leukemia (~ 99%) and is also fairly specific (5). In addition, although there is no single antibody that can identify an antigen unique to the hairy cell, CD 103 is the most useful marker for distinguishing hairy cell leukemia from other B-cell leukemias (1).

Abnormalities of the liver in hairy cell leukemia seem unique among hepatic localizations of blood malignancies (7) being diffuse mononuclear cell infiltration throughout the hepatic sinusoids and in the portal areas a constant feature (2,7). Angiomatous lesions made of periportal and intralobular cavities lined by a layer of hairy cells and filled with red blood cells and hairy cells have also been observed (7). These angiomatous lesions and the presence of splenic pseudosinuses which are structures lined by hairy cells lacking the endothelial cells and ring fibers, of normal splenic sinuses appear to be quite specific for hairy cell leukemia as they are not seen in other types of chronic leukemia (8). Identification of hairy cells on sections stained with hematoxilin and eosin appears to be difficult without a prior knowledge of the disease or without special processing of tissue for examination (tartrate-resistent acid phosphatase activity in methacrylate-embedded sections) (2). However, it has been proposed that the presence of many mononuclear cells assuming clear cell pattern ("halo" appearance) infiltrating the sinusoids, sinusoidal congestion and sinusoidal beading is strongly suggestive of hairy cell leukemia (2,9).

Liver involvement in hairy cell leukemia is rarely associated with serum biochemical abnormalities. Nonetheless, mild increases in liver enzymes have been noted (2,4,7). In the series of Yam *et al.* (2) serum AST was increased in only 2 out of 18 and serum ALT in 1 out of 3 patients tested. Despite extensive histologic involvement there was little or no deterioration of hepatic function (2). However, a case of relapsing hairy cell leukemia presenting as fulminant hepatitis has been reported (10).

Another alternative of diagnosis might be the presence of opportunistic liver infections, because patients with this disorder seem to be prone to infection with mycobacteria. Tuberculosis was found in 8% of hairy cell leukemia patients in a recent series (11), while disseminated atypical mycobacterial infection ocurred in 5% of them in another large series (12). However, these alternative explanations for the liver abnormalities are unlikely in our patient as he was asymptomatic and the presence of fever and chills were constant features of mycobacterial infection in all patients at diagnosis (11,12).

In conclusion, it should be kept in mind that hairy cell leukemia on presentation is a potential cause of persistent abnormalities in liver-related enzymes. Because the early unimpressive nature of its clinical and laboratory data, and its generally indolent course, the disease can be discovered during routine studies for chronic liver disease. As in our case, liver histopathologic features could became the key for the diagnosis.

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