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Case Report

The Hepatologist in the Haematologist Field: A Rare Cause of Non-Cirrhotic Portal Hypertension

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Abstract

An 81 year-old gentleman was admitted to the emergency department of our hospital due to hematemesis and melena, and endoscopic evidence of portal hypertension. Due to presence of ascites and esophageal varices he was referred to gastroenterology care. His history included systemic mastocytosis, and biochemistry showed marked cholestasis with normal liver function tests. Clinical work-up was not consistent with liver disease-related portal hypertension, but rather hepatic involvement of systemic mastocytosis and the development of non-cirrhotic portal hypertension. This clinical case highlights that although systemic mastocytosis is uncommon, the subsequent development of liver involvement and non-cirrhotic portal hypertension are complications of the disease that need always be considered.

Keywords: Non-cirrhotic portal hypertension; Systemic mastocytosis; Bleeding; Ascites

Introduction

Non-Cirrhotic Portal Hypertension (NCPH) is a disease characterized by portal hypertension with near normal hepatic venous pressure gradient and relatively preserved liver function. Patients with NCPH usually present with signs or symptoms of portal hypertension such as gastro-oesophageal varices, variceal bleeding, and splenomegaly. In this context, ascites and liver failure can occur in association with other precipitating factors. This condition has a vascular origin and is classified anatomically on the basis of the site of resistance to blood flow as: pre-hepatic, post-hepatic and hepatic. In this last case NCPH is subdivided into pre-sinusoidal, sinusoidal and post-sinusoidal [1]. Mastocytosis is a rare myeloproliferative disorder characterized by an abnormal proliferation of mast-cells with infiltration and accumulation in one or more organ and systems and is a rare cause of hepatic sinusoidal NCPH [2].

Case Presentation

An 81 year-old gentleman was admitted to the emergency department of our hospital due to hematemesis and melena. An upper digestive endoscopy was performed, showing medium-large gastro-oesophageal varices with cherry-red spots, and portal hypertensive gastropathy. Medical treatment for variceal bleeding was started, and the patient was transferred to our unit with a diagnosis of variceal bleeding in cirrhosis. Accurate history revealed that the patient suffered from chronic myelo-monocytic leukemia and systemic mastocytosis. Laboratory work-up performed upon admission showed marked cholestasis (alkaline phosphatase=1.013 U/L, gamma-glutamyl transpeptidase=271 U/L) with normal amino transferases and fairly preserved hepatic synthetic function (albumin=33.2 g/L), while laboratory tests were negative for all known causes of liver disease. On examination the patient was asymptomatic with normal vital parameters, the abdomen was distended with mild ascites, and no signs of spontaneous bacterial peritonitis atdiagnostic paracentesis. Abdomen ultrasonography showed mildly coarse hepatic pattern, ascites, and relevant splenomegaly (165 mm). Although no liver biopsy was performed due to patient refusal, on the basis of the available data absence of known causes of liver disease, cholestasis and clinically significant portal hypertension without signs of hepatic dysfunction we hypothesized a diagnosis of NCPH due to infiltration of the liver by his hematological disease.

Discussion

In systemic mastocytosis, NCPH can be related to the microscopic infiltration of the portal tracts by abnormal mast-cells, leading to deposition of sub-endothelial collagen due to mast-cell factors, causing fibrosis and obliteration of sinusoids, determining increased portal pressure in a context of preserved liver function. This condition is more frequently diagnosed in young people, although some cases are described in elderly people [2]. In the largest series published in elderly patients with mastocytosis, Rouet, et al. reported the presence of ascites or other signs of portal hypertension and abnormal liver tests in 26.4% and 32.1% of patients, respectively [3]. Recently, some cases of variceal bleeding in patients with NCPH due to systemic mastocytosis even with bleeding as the first manifestation of this hematologic disease were reported, thus encouraging the clinicians to "think outside the box" when dealing with unusual cases of portal hypertension [4,5]. This clinical case highlights that although systemic mastocytosis is uncommon, the subsequent development of liver involvement and NCPH are complications of the disease that need always be considered.

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