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Case Report

Unusual association of cirrhosis and sickle cell anemia revealed by hemolytic anemia: A case report

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Abstract

Hepatobiliary complications of sickle cell disease are rare, cirrhosis remains very exceptional, especially in heterozygous forms of the disease. We report the case of a 19-year-old patient whose etiologic investigation of hemolytic anemia revealed heterozygous sickle cell disease complicated by hepatic cirrhosis. The diagnosis of cirrhosis was made due to the presence of signs of hepato-cellular insufficiency, portal hypertension syndrome and hepatic dysmorphia on imaging. The etiological assessment was negative. The liver biopsy was not performed due to the risk of bleeding. The interest of this observation is to evoke hepato-biliary complications (in particular cirrhosis) in patients with sickle cell anemia, in order to avoid a pejorative evolution burdened with serious complications.

Introduction

Sickle cell disease is an autosomal recessive genetic disease resulting from a mutation in the gene responsible for the formation of hemoglobin. It is characterized by rigid red blood cells in the shape of a "sickle" responsible for several complications: delayed development of the child, vaso-occlusive crises, a predisposition to bacterial infections and hemolytic anemia. 80% of cases are diagnosed in sub-Saharan Africa [1].

Hepato-biliary complications of sickle cell anemia are relatively rare, ranging from laboratory abnormalities of liver function in asymptomatic patients to hepatic cirrhosis [2,3]. They occur mainly in patients with the homozygous form, and more rarely in heterozygous forms of the disease [4]. We report the case of a 19-year-old patient whose etiologic assessment of hemolytic anemia revealed heterozygous sickle cell disease associated with cirrhosis.

Observation

Patient information

A 19-year-old male patient with no particular personal or family history. Has complained for 2 years of an anemic syndrome characterized by asthenia and skin paleness associated with cutaneous jaundice with normal colored urine and stools, without other associated signs.

Clinical findings

Clinical examination found a conscious, hemodynamically and respiratory stable patient (Heart rate: 84 beats per minute, respiratory rate: 18 cycles per minute), Temperature: 37.5°C, with paleness and jaundice, abdominal examination revealed abdominal swelling (Figure 1), splenomegaly goes past the umbilicus, with hepatomegaly, without dullness of the flanks.

Diagnostic assessment

Laboratory results showed pancytopenia consisting of

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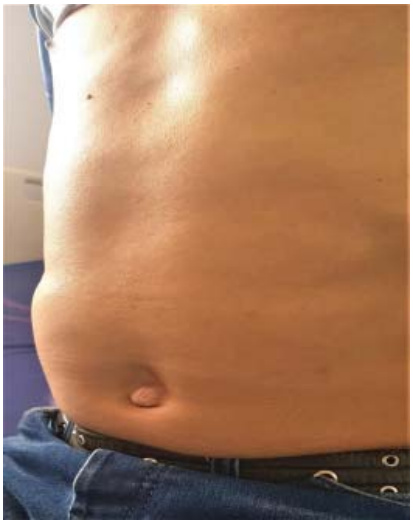

Figure 1: Abdominal swelling related to splenomegaly.

normochromic normocytic regenerative anemia (hemoglobin level = 11.0 g / dl, mean corpuscular volume = 88 fl, mean corpuscular hemoglobin content = 30 pg, reticulocyte count = 221,000), leukopenia (White blood cells = 3125 / l) and thrombocytopenia (Platelet count = 41,000 / mm3). Bilirubin was elevated predominantly conjugated (Total Bilirubin = 41, Conjugated Bilirubin = 39, Unconjugated Bilirubin = 2). Lactate Dehydrogenase (LDH) was elevated to 624 U / L. Hemoglobin electrophoresis revealed the profile of heterozygous sickle cell disease. The rest of the biological assessment showed a low prothrombin level (49%), hypoalbuminemia at 27 g / L. hepatic transaminase, alkaline phosphatase, and gamma glutamyl transferase levels were normal.

The abdominal ultrasound performed revealed a splenomegaly measuring 22 cm, associated with hepatomegaly with a heterogeneous echotexture and irregular surface, the portal vein was dilated (diameter measuring 16 mm), with no visible peritoneal effusion. Eso-gastro-duodenal fibroscopy showed stage III esophageal varices with portal hypertensive gastropathy. Other tests including a blood ionogram, renal function, Alpha-Fetoprotein (AFP) test and an electrocardiogram did not reveal any abnormalities.

The diagnosis of cirrhosis was made based on a set of arguments including signs of hepato-cellular insufficiency (low prothrombin level and hypoalbuminemia), portal hypertension syndrome (splenomegaly, dilated portal vein, esophageal varices, portal hypertensive gastropathy) and hepatic dysmorphia on imaging. The etiological assessment comprising the viral markers of chronic hepatitis B and C and the autoimmune test was negative, the copper test ruled out Wilson's disease, and serum ferritin was normal. The liver biopsy was not performed due to the risk of bleeding.

Therapeutic intervention

The therapeutic management of cirrhosis consisted of putting the patient on non-cardioselective beta-blockers and ligation of the esophageal varices to prevent bleeding complications, and to monitor the patient for other complications of the cirrhosis. Regarding sickle cell disease, the therapeutic attitude included preventive measures against vaso-occlusive crises: avoiding hypoxia (intense effort, altitude), dehydration, stress, fever, and taking stimulants and alcohol.

Follow-up

The follow-up of the patient over a period of 6 months was marked by clinical improvement, marked by the improvement of the anemic syndrome and the regression of the jaundice. During this period, the patient never had any hemorrhagic complication or ascites decompensation related to the cirrhosis.

Discussion

Hepato-biliary manifestations during sickle cell disease are relatively rare, resulting from multiple physiopathological mechanisms: consequences of hemolytic anemia, multiple transfusions, vaso-occlusion of the hepatic vessels, etc [5]. They mainly occur in patients with homozygous, and more rarely in heterozygous forms of the disease [4].

The hepato-biliary involvement in sickle cell anemia varies from laboratory abnormalities of hepatic function (elevation of hepatic transaminases, hyperbilirubinemia, increase of alkaline phosphatase and gamma glutamyl transferase) in asymptomatic patients at stages of insufficiency and of hepatic cirrhosis [2,3].

The etiologies of hepatic cirrhosis in patients with sickle cell disease are multiple and variable including tissue hypoxia secondary to vaso-occlusion of hepatic vessels, gallstones secondary to chronic hemolysis, chronic viral hepatitis or iron overload. Due to multiple transfusions... [5] However, the data in the literature underline the importance of vascular lesions and the important role of sickling in hepato-biliary lesions in the majority of patients with sickle cell disease, as reported by Charlotte, et al. [6] and Maher, et al. [7].

After eliminating in our patient chronic viral hepatitis, iron overload, autoimmune hepatitis, it is similar that his hepatic cirrhosis is secondary to the sickle cell disease itself.

Post-mortem histological results from a series of 70 patients with sickle cell disease showed Kupffer cell erythrophagocytosis in 91% of cases, sinusoidal dilation of red blood cells in 71% of cases and iron deposits in 47% of cases. [8] Other less frequent abnormalities also noted in this study including focal necrosis in 35% of cases, portal fibrosis in 20% of cases, regeneration nodules in 20% of cases and cirrhosis in 16% of cases. Similar histological results were found in other series [9-12].

Abdominal ultrasound in patients with sickle cell disease may show gallstones and hepatic and pancreatic hyperechogenicity due to iron deposits [13]. Computed Tomography (CT) can show several abnormalities in patients with a homozygous form, including diffuse hepatomegaly, a hepatic infarction or abscess, gallstones, an atrophic and calcified spleen, etc. In addition, patients with heterozygous forms of the disease usually present with splenomegaly. Other

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results may also be found in heterozygotes including a splenic infarction or abscess, or even splenic rupture secondary to extensive infarction [4].

The link between liver damage (especially cirrhosis) and increased mortality in patients with sickle cell disease remains controversial to this day. Although some authors have shown no increased mortality from liver damage in patients with sickle cell disease [14]. Others implicated cirrhosis as the main factor in death in 11% of patients [15].

Conclusion

Cirrhosis is a rare liver complication of sickle cell anemia. Many factors can contribute to its etiology, in particular ischemia, viral hepatitis linked to transfusion, iron overload, etc.

Early recognition of this complication in patients with sickle cell disease will allow optimal management and prevent progression to decompensation and complications.

Authors' contributions

All authors participated in the conception, drafting the work, critically revised the manuscript, approved the final version to be published, and agree to be accountable for all aspects of the work.

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