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Case Study

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CHRONIC LIVER INVOLVEMENT IN SICKLE CELL DISEASE: A CASE REPORT

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ABSTRACT

We present the case of a 30-year-old male with sickle cell disease who developed an acute vaso-occlusive crisis occurring in chronic hepatopathy. Admitted with abdominal pain, jaundice and fever, precipitated by septic arthritis. The laboratory assessment showed elevations of the liver enzymes, cholestasis associated with hyperbilirubinemia (predominantly unconjugated), slight thrombocytopenia and low prothrombin rate. Abdominal ultrasound and cross-sectional imaging demonstrated: fine bile ducts and cholelithiasis, hepatomegaly, dysmorphic liver, splenomegaly and moderate peritoneal effusion. A liver transplant and exchangetransfusion were necessary for severe hepatocellular insufficiency and edema-ascitic syndrome. But the patient died during transplantation planning.

KEYWORDS: Sickle cell hepatopathy, Sickle hepatic crisis, Exchange Transfusion, Liver Transplantation.

INTRODUCTION

Sickle cell anemia (SCA) is an inherited genetic disease affecting the chains of hemoglobin. Red blood cells are deformed and take a sickle shape. They therefore have difficulty in circulating and can cause clots in the vessels, causing many complications, such as acute chest syndrome, stroke, osteonecrosis, sickle nephropathy, and hyposplenism. Hepatic involvement in SCA appears to be common. The incidence of sickle hepatopathy is difficult to determine, as abnormalities in liver function tests are common in patients with SCA. Liver disease may result from viral hepatitis and iron overload due to multiple transfusions of blood products or due to disease activity causing varying changes in vasculature. The clinical spectrum of disease ranges from ischemic injury due to sickling of red blood cells in hepatic sinusoids, pigment gall stones, and acute/chronic sequestration syndromes.

In this report, we describe a case of vaso-occlusive crisis occurring on chronic hepatopathy in a 30-year-old patient.

CASE PRESENTATION

A 30-year-old male had been followed-up for sickle cell disease (SCD; genotype HbSS) from the age of 12 years. His past medical history included asymptomatic gallstone disease and osteomyelitis of the right leg at the age of 23 years. He had no history of alcohol excess.

He presented to our emergency department with abdominal pain, jaundice and fever, precipitated by septic arthritis. At clinical examination, he was found to be icteric, febrile at 38,5°C, with painful hepatomegaly with splenomegaly and moderate ascites. The laboratory assessment showed elevations of the liver enzymes aspartate aminotransferase (314 UI/l) and alanine aminotransaminase (154 UI /l), cholestasis : alkaline phosphatase (541 UI/) and gamma-glutamyl-transferase (309 UI /l), associated with hyperbilirubinemia (predominantly unconjugated) (318 mg /l). Blood count showed deep anemia : hemoglobin (5,1 g/dl), slight thrombocytopenia : platelets (146000 / mm3) and low prothrombin rate (54%). Abdominal ultrasound and cross-sectional imaging demonstrated: fine bile ducts and cholelithiasis, hepatomegaly, dysmorphic liver, splenomegaly and moderate peritoneal effusion.

It should be noted that the viral serology and liver autoantibody screen were normal.

Given the results of the evaluation, a diagnosis of an acute sickle liver in the context of an acute vaso-occlusive crisis occurring in chronic hepatopathy was made.

Treatment included intravenous hydration and analgesia associated the treatment of septic arthritis: surgical drainage and antibiotics. Liver biopsy has not been practiced because is a relative contraindication for the high risk of bleeding and liver rupture (low prothrombin time).

About one month after initial presentation, infectious syndrome have improved contrary a rate of bilirubin who did not return to the pre-episode baseline value. Hyper

bilirubinemia gradually deteriorated (660 mg / l). The right upper quadrant pain did not improve. The deterioration in liver function got worse (prothrombin rate : 27% and factor V : 22%) with occurrence of renal impairment (creatinine : 19 mg /l). The situation was that of an uncommon hepatological complication of a common hematologic condition. A liver transplant and exchangetransfusion were necessary for severe hepatocellular insufficiency and edema-ascitic syndrome. But the patient died during transplantation planning.

DISCUSSION

Sickle cell anemia is inherited disorders of hemoglobin (Hb) structure characterized by chronic hemolytic anemia, frequent bacterial infections, and recurrent episodes of sickling of erythrocytes within the vasculature resulting in ischemia, infarction, and fibrosis.[1] The incidence is estimated to be between 300,000 and 400,000 neonates globally each year, the majority in sub-Saharan Africa.[2] Abnormal liver function tests are common in patients withsickle cell anemia, even in the absence of liver disease.[3] His prevalence is estimates of 10%.[4] Sickle cell hepatopathy characterized by extreme hyperbilirubinemia and either mild or severe hepatic dysfunction.[5] The prevalence of cirrhosis in autopsy series is up to 30%[1], indicating that chronic liver disease is an important consideration in this group of patients such as our case report.

In this report, the patient developed progressive chronic liver disease, culminating in decompensated cirrhosis. The natural history of this chronic phase of sickle hepatopathy is not well defined, probably secondary to recurrent intrahepatic sickling.[6]

An important question that remains unanswered is whether any specific intervention in the patient with established chronic liver disease will modulate the risk of progression to end- stage liver disease.[6]

Hydroxyurea who increases HbS levels, no difference was noted in the frequency of hepatic sequestration crises compared with untreated controls.[7] Further work is needed on exploring the preventative effects of hydroxyurea on organ damage including sickle hepatopathy. But the role of liver transplant not defined.[8] Poor results reported in early case series. Because she may carry a high risk of graft loss due to vascular problems.[9] This results have improved, to some degree, in later studies.[10] Recommendations for perioperative management of SCA include regular transfusional management to maintain HbS fraction <20-30%.[11] However, good vital organ function is crucial for liver transplantation surgery itself, this precludes many SCA patients who have already developed significant sickle-related complications of the heart, lungs, kidneys, and brain, concomitant with their sickle hepatopathy.[12]

CONCLUSION

Hepatic dysfunction is a common finding in patients with sickle cell disease. The clinical spectrum of sickle cell disease ranges from mildliver function test abnormalities in asymptomatic patients, to dramatic clinical crises with marked hyperbilirubinemia and liver failure. However, the nature of liver dysfunction in SCA has not been characterized, nor has the natural history and pathogenesis of the liver disease been fully defined. This lack of knowledge prevents specific monitoring and deciding when, how, and in whom to intervene. Finally, it is necessary to insist that patients approaching end-stage liver disease be referred to centers where joint hematologic and hepatological assessment can be performed and patients offered the full range of treatment options.

ABBREVIATIONS

SCD: Sickle cell disease SCA: Sickle cell anemia Hb: hemoglobin

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