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A Rare Case of Acute Ischemic Hepatitis Secondary to Hepatic Sequestration Crisis in Sickle Cell Disease

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Abstract Acute Ischemic Hepatitis in sickle cell disease could lead to acute hepatic failure. The term Sickle Cell Hepatopathy encompasses a wide range of hepatic manifestations that occur due to this disease itself or secondary to treatment. Although our patient had a homozygous inheritance for the sickle cell gene but never experienced any hepatic complications of the disease. Our patient experienced an acute hepatic sequestration crisis which otherwise usually occurs in the spleen. This case signifies the importance of early evaluation and management of hepatic involvement in sickle cell disease to prevent overt hepatic failure.

Keywords: *sickle cell disease, hepatic sequestration crisis, acute ischemic hepatitis, acute abdominal pain, exchange transfusion*

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1. Introduction

Sickle cell disease (SCD) is the most common inherited hematologic disease in the United States [1]. In combination with other abnormal hemoglobin types, sickle cell disease results and affects a large number of persons in the African and African American communities. Ten percent of African American children have sickle cell trait and 0.2% have SCD [2]. It results from a single amino acid substitution in the globin chain [3]. The effects of this disorder are multisystemic, including involving the gastrointestinal tract particularly the hepatobiliary system in a number of ways. The pathogenic mechanisms in SCD that lead to liver involvement are termed as Sickle Cell Hepatopathy [4], which occurs in roughly 10-40 % of the cases [5]. Various pathogenic mechanisms have been proposed that lead to sickle cell hepatopathy including the rapid breakdown of red blood cells which is termed as 'sequestration crisis' that leads to the formation of pigmented bile stones that are rich in bilirubin. Another suggested mechanism is recurrent transfusions for anemia which puts these patients at risk of hepatitis from a viral infection and iron overload. Also, intra sinusoidal sickling and hypoxia lead to hepatocellular damage. Clinically patients present with acute abdominal pain and jaundice. Labs show elevated liver enzymes due to intrahepatic cholestasis, choledocholithiasis, or cholecystitis.

2. Case Presentation

A 27-year-old man, a diagnosed case of Sickle cell Disease and multiple admissions for sickle cell crises about 3-4 crises/year, initially presented with left lateral chest pain without cough or shortness of breath. However, this episode is different from his usual sickle crisis in which he usually gets extremity pain and burning He was vitally stable on presentation. The chest and abdomen examination were unremarkable. On presentation, he was found to have a hemoglobin of 5.7 g/dL which dropped to 4.2 g/dL within 2 days of admission with CBC showing a hemolytic picture with a simultaneous elevation of LDH, total bilirubin of 4.8 with indirect bilirubin more than direct bilirubin; and an elevated retic count. The patient was transfused 2 units of packed RBC's; however, his liver function tests started trending up from initial values of ALT of 37 IU/L and AST of 43 IU/L to ALT of 1996 IU/L and AST of 1236 IU/L within a course of five days of hospital stay (Table 1).

There were no documented episodes of hypotension during hospitalization. Serology for hepatitis B, hepatitis C, HIV, and autoimmune liver disorders were all negative (Table 2).

Table 1. Laboratory Data L, Low; H, High

Labs	Reference range	Day 1	Day 5	Day 10 (Discharge)
Hemoglobin (Hb)	12.9-16.7 g/dl	5.7 (L)	7.1 (L)	9.8 (L)
Alanine Transaminase (ALT)	21-72 U/L	87 (H)	1987 (H)	604 (H)
Aspartate Transaminase (AST)	17-59 U/L	150 (H)	1450 (H)	108 (H)
Lactate Dehydrogenase (LDH)	313-618 IU/L	1323 (H)	2785 (H)	1006 (H)
Total Bilirubin	0.2-1.3 mg/dl	14.2 (H)	10.2 (H)	3.2 (H)
Indirect Bilirubin	0.2-0.8 mg/dl	11.2 (H)	8 (H)	2.4 (H)

Table 2. Other investigations for liver injury

Lab	Reference range	Results
Herpes Simplex Virus PCR	Not detected	Not detected
Cytomegalovirus PCV	<2.3 log IU/ml	<2.3
Epstein-Barr Virus (EBV) IgM	<36 U/ml	<36
Hepatitis A virus IgM	Negative	Negative
Hepatitis C antibody	Negative	Negative
Hepatitis B surface antigen	Negative	Negative
Ceruloplasmin level	20-35 mg/dl	15
Iron level	49-181 ug/dl	148
Antinuclear antibody (ANA)	Negative	Negative
Anti Smooth Muscle Antibody	<20 U	<20
Anti Mitochondrial Antibody	Negative	Negative

There was no history of the use of any hepatotoxic drugs. Abdominal ultrasonography was unremarkable for any liver, spleen, pancreas, or kidney pathology. Based on all of these findings acute ischemic hepatitis due to hepatic sequestration crisis was confirmed as the likely diagnosis. The patient received an exchange transfusion after which his liver enzymes started to trend down and his clinical condition also markedly improved and was subsequently discharged. Currently, he is doing well and continues to follow at the sickle cell clinic.

3. Discussion

Hepatic manifestations of Sickle Cell Disease widely vary in pathogenesis and clinical severity. It could range from mild cholestasis to acute hepatic failure and even cirrhosis. Mild abnormalities in liver function test could occur in the majority of patients however explicit picture of acute hepatic injury is fairly uncommon.

Given the clinical presentation of our patient, he showed signs of ischemia with falling hemoglobin, elevated lactate, and indirect bilirubin levels, all of which indicate a sequestration crisis. In sickle cell disease sequestration crisis most commonly occurs in the spleen and very few cases occur in the liver. His hepatitis panels were negative, and ultrasound showed no signs of obstructive disease. Hence it was concluded that he was suffering from an acute liver sequestration crisis leading to ischemia and hypoxia. Clinically it could merely be lab abnormality or present with intense right quadrant abdominal pain with tender hepatomegaly and rarely acute hepatic failure with encephalopathy and coagulation disorder [6]. Hence it is important for clinicians to distinguish the self-limiting cholestasis abating with mere hydration and analgesics to potential liver failure for which prompt recognition and exchange transfusion are considered the only effective therapeutic options [7]. This case reiterates the need for early detection of liver involvement in sickle cell disease hence preventing potentially fatal liver failure.

4. Conclusion

Hepatic sequestration crisis leading to acute hepatic failure in sickle cell disease in an uncommon phenomenon as most of the cases of sequestration crisis occur in the spleen. Early recognition and prompt management of this condition with exchange transfusion could be lifesaving. We present a case of a 27-year-old man who had no liver involvement, present with acute liver injury due to hepatic sequestration crisis. Immediate diagnosis and exchange transfusion lead to significant improvement in his lab abnormalities as well as his clinical condition.

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None.

Statement of Competing Interests

The authors have no competing interests.

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