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CASE REPORT | ESOPHAGUS

Massive Esophageal Variceal Bleeding as a Rare Complication of Sickle Cell Anemia

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Abstract

A 24-year-old man with sickle cell anemia presented with fatigue, dark stool, and coffee ground emesis. He was found to have large esophageal varices and experienced massive variceal hemorrhage in the hospital. The varices were caused by diffuse splanchnic venous thrombosis, and his only risk factor for hypercoagulability was sickle cell anemia. Splanchnic venous thrombosis due to sickle cell anemia is exceedingly rare.

Introduction

Sickle cell anemia is a known hypercoagulable state.[1] Thromboses in sickle cell anemia usually consist of deep venous thrombosis or pulmonary embolism. Thrombosis of the splanchnic venous system (i.e., mesenteric, splenic, portal, and suprahepatic veins), however, is very rarely seen, with only 2 cases reported in the literature.[2,3]

Case Report

A 24-year-old African American man with a history of sickle cell anemia presented with complaints of fatigue, dyspnea on exertion, dark stools, and coffee ground emesis. His hemoglobin was 5.8 g/dL. He was transfused 2 units of packed red blood cells and an esophagogastroduodenoscopy (EGD) showed large esophageal varices throughout the esophagus with stigmata of recent bleeding (red wale signs; Figure 1). No gastric varices, gastric ulcers, or duodenal ulcers were noted. During the EGD, he developed uncontrolled variceal hemorrhage that was not amenable to banding. Ethanolamine injection was unsuccessful at achieving hemostasis, so a Sengstaken-Blakemore tube was inserted to attempt an urgent transjugular intrahepatic portosystemic shunt (TIPS) procedure. Abdominal computed tomography (CT) later noted complete thrombosis of the splenic vein, non-occlusive thrombus of the superior mesenteric vein extending into smaller mesenteric branches, and complete thrombosis of the left portal vein along with incomplete thrombosis of the right portal vein and cavernous transformation (Figure 2). This eliminated TIPS as an option.

EGDs were performed the following 4 days with attempts to deflate the Blakemore tube. However, persistent variceal bleeding was noted each time. Attempts to control the bleeding with ethanolamine and band ligation were unsuccessful and the Blakemore tube was re-inserted after each attempt. On hospital day 8, after consultation with transplant surgery, hepatology, and interventional radiology, successful embolization of the gastroesophageal variceal collaterals originating from the left gastric artery was performed. The following day, EGD showed small decompressed (<5 mm) esophageal varices with scarring and ulcerations in the esophagus, but no active bleeding. The Blakemore tube was removed and did not need to be replaced for the remainder of the hospitalization. He required no more blood transfusions or EGDs and was discharged to home in stable condition on hospital day 23. In total, he required 6 EGDs and was transfused 11 units packed red blood cells.

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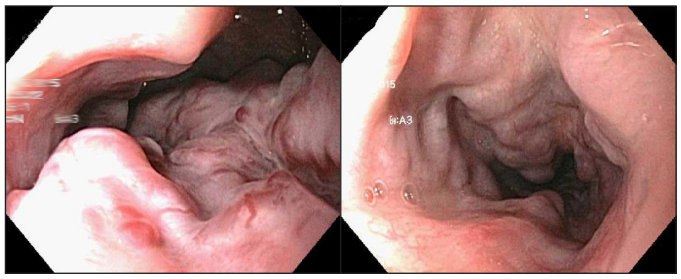

Figure 1. Endoscopic views from initial EGD showing large varices throughout the entire esophagus prior to intraprocedural hemorrhage.

Since this initial presentation, our patient has been admitted with gastrointestinal bleeding due to esophageal variceal hemorrhage 3 times. He required intubation and placement of a Sengstaken-Blakemore tube two of those times. Given his demonstrated high risk for re-bleeding, we have been unable to treat him with any form of anticoagulation. Current efforts are focused on potential surgical options, including dual liver–small bowel transplant or modified Sugiura procedure.

Discussion

Due to a number of alterations in the clotting cascade, patients with sickle cell disease are known to be hypercoagulable.[1,4,5] Nearly all thrombotic events related to sickle cell disease involve deep venous thrombosis or pulmonary embolism.[4,6,7] Thrombosis of the splanchnic venous system caused by sickle cell disease is exceedingly rare. Very few cases of splanchnic venous thrombosis attributed to sickle cell disease have been reported.[2,3] The most commonly recognized risk factors for splanchnic venous thrombosis include myeloproliferative disorders/neoplasms, liver cirrhosis, abdominal malignancy, pregnancy, paroxysmal nocturnal hemoglobinuria, and inherited thrombophilias (deficiencies of anti-thrombin, protein C, and protein S).[8,9]

Our patient had no suggestion of abdominal or myeloproliferative malignancy, and did not have cirrhosis. Paroxysmal nocturnal hemoglobinuria, protein C and S deficiency, factor V Leiden mutation, anti-thrombin III deficiency, and antiphospholipid antibody syndrome were ruled out with appropriate laboratory testing. Consequently, sickle cell anemia wass his only identified risk factor for hypercoagulability and was determined to be the cause of our patient's diffuse splanchnic thrombosis, large esophageal varices, and massive gastrointestinal hemorrhage.

Disclosures

Author contributions: M. Malamood performed the literature search, wrote the manuscript, and is the article guarantor. G. Bernstein, Z. Malik, and M. Mathur edited the manuscript.

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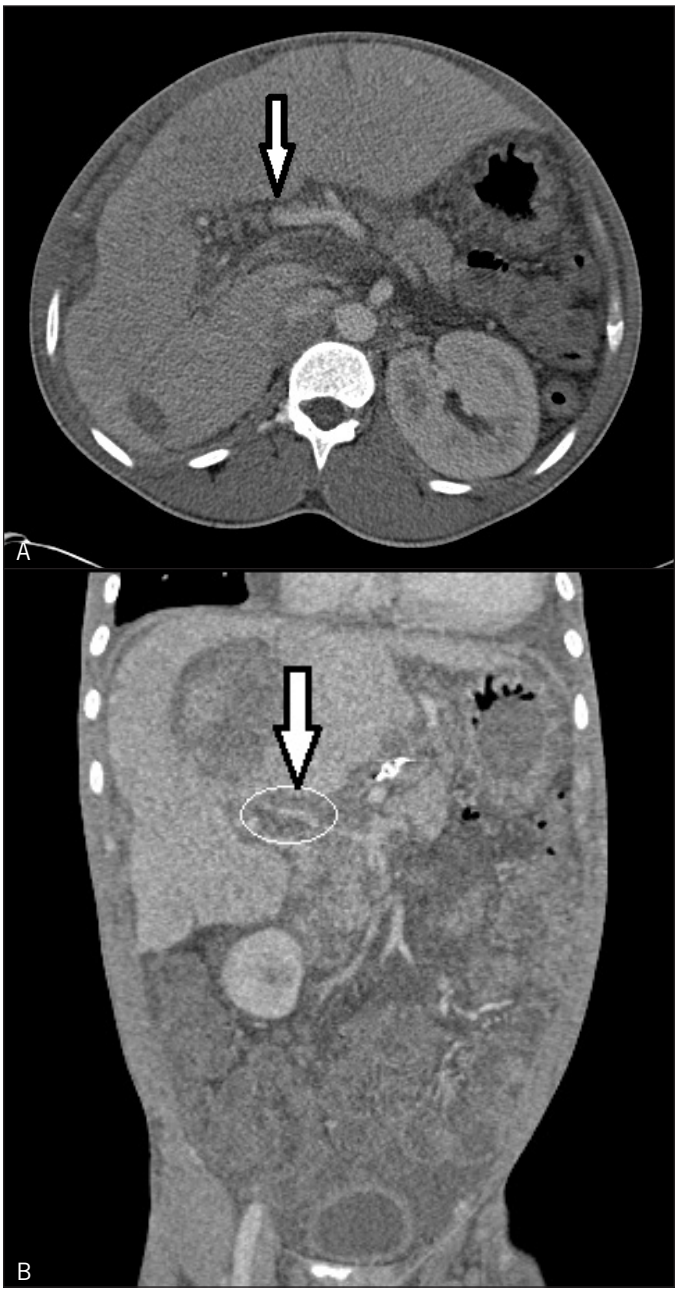

Figure 2. (A) Axial CT with IV contrast showing near complete occlusion of the main portal vein with cavernous transformation (arrow). (B) Coronal CT with IV contrast showing the confluence of the splenic and superior mesenteric veins into the main portal vein (arrow) with only a sliver of contrast passing through (circled).

Informed consent was obtained for this case report.

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