

Ischemic colitis in a known sickle cell disease patient

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Abstract

Patients affected with sickle cell disease (SCD) suffer from recurrent vascular occlusions that lead to ischemia and distal tissue infarction in multiple organs. These vascular occlusions manifest as sickle cell pain crises. Abdominal pain is common in sickle cell pain crises. It is attributed to vasoocclusion or infarction in the mesenteric vasculature. Progression to significant ischemic damage of the intestine is rare in sickle cell disease. The objective was to show the rare case of ischemic colitis in SCD patient. The patient's images (plain abdominal x rays and barium enema images) and case file were reviewed and summarized. The subject matter of ischemic colitis in a known SCD was reviewed in the literature. The index case was discussed and compared with literature. We report an 18-year-old known SCD patient diagnosed since childhood but not regular on medication and follow up. The patient presented to SCD clinic of Aminu Kano Teaching Hospital (AKTH), Kano with 2-year history of recurrent blood stained and watery stool associated with abdominal pain and low-grade fever. There was history of easy fatigability and multiple blood transfusions in the past. Other systemic reviews were unremarkable. This man is the only sickle cell anemic child among 9 other siblings of the family, and on presentation, a second-year student of a junior secondary school. Examination revealed a stunted young man, mild pale with a tinge of jaundice. Abdominal examination revealed hepatomegaly 4 cm below the costal margin. The spleen was not enlarged. Other systemic reviews were unremarkable. Latest hemoglobin was 5.7 g/dL. Renal and the liver function tests were normal. Stool microscopy did not isolate ova, cyst or protozoa. Barium enema was then requested and showed loss of haustration and cobblestone appearance involving the entire colon. Fine mucosal granulations were also noted in the rectum. Diagnosis of colitis most probably ischemic was made. The patient was commenced on analgesics and antibiotics including azithromycin and metronidazole and made significant improvement.

Ischemic colitis in SCD may still be encountered in practice despite its rarity.

Introduction

Sickle cell disease (SCD) is an autosomal recessive inherited hemoglobinopathy in which hemoglobin S aggregates into large polymers under low oxygen tension, leading to erythrocyte distortion and impaired deformability.¹⁻⁴ It most often affects people of African, Middle Eastern, Mediterranean and Asian descent. In Nigeria, sickle cell anemia occurs with a prevalence of 2%^{1,5} and sickle cell hemoglobin disease occurs in 0.7% of the population.¹ Affected patients suffer from recurrent vascular occlusions that lead to ischemia and distal tissue infarction in multiple organs. These vascular occlusions manifest as sickle cell pain crises. Abdominal pain is common in sickle cell pain crises, with an incidence of 30% to 57% in patients presenting with pain.⁶ It is attributed to vasoocclusion or infarcts in the mesenteric vasculature. Progression to significant ischemic damage of the intestine despite adequate analgesia and fluids is rare⁷ in sickle cell disease. This is likely because the colon has an abundant collateral blood supply and low oxygen extraction. To the best of the author's knowledge, only few cases have been described in the available literature.² Hence, the need to present this case of ischemic colitis (IC) in an 18-year-old sickle cell disease patient.

Case Report

MM is an 18-year-old known SCD patient diagnosed since childhood using hemoglobin electrophoreses at alkaline Ph, but not regular on medication and follow up. The patient presented to SCD clinic of Aminu Kano Teaching Hospital (AKTH), Kano with 2-year history of recurrent blood stained and watery stool associated with abdominal pain and low-grade fever. There was history of easy fatigability and multiple blood transfusions in the past. Other systemic reviews were unremarkable. This man is the only sickle cell anemic child among 9 other siblings of the family, and is presently a second-year student of a junior secondary school. His steady state PCV was 27%. Examination revealed a stunted young man, mildly pale with a tinge of jaundice. Abdominal examination revealed hepatomegaly 4 cm below the costal margin. The spleen was not clinically palpable. Other systemic reviews were unremarkable.

Latest hemoglobin was 5.7g/dL at pres-

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entation. Renal and the liver function tests were normal. Stool microscopy did not isolate ova, cyst or protozoa. Barium enema was then requested and showed loss of haustration and cobblestone appearance involving the entire colon. Fine mucosal granulations were also noted in the rectum. Diagnosis of colitis most probably ischemic was made. The patient was commenced on analgesics and antibiotics including azithromycin and metronidazole as an outpatient treatment and made some improvement (Figures 1-3).

Discussion and Conclusions

Ischemic colitis (IC) is relatively rare in patients with SCD.⁷ This is in part due to the low degree of oxygen extracted by the bowel (15-20% of oxygen delivered) and rapid flow due to arteriovenous shunting in the bowel wall. Even though uncommon, ischemic injury of the colon should be considered in those patients who develop severe intestinal colic, rectal bleeding, or signs of peritonitis.^{7,8} This patient presented with both abdominal pain, blood stained stool and fever. The clinical presentation however varies, depending on the severity and extent of the disease. None of the symptoms and signs is specific.

Given that the presentation of ischemic colitis is not specific and is highly variable, diagnosis and management is clinically challenging.⁸ Diagnosis requires a high



Figure 1. Plain abdominal radiograph showing loss of haustral markings in the outlined transverse colon.



Figure 2. Barium enema showing loss of haustrations involving the sigmoid, descending, transverse and ascending colon.



Figure 3. Barium enema showing cobblestone appearance of the involving mainly the transverse colon.

index of clinical suspicion. The chronology of symptoms and the clinical situations upon which these symptoms appear must be taken into account. In the present case, symptoms started 2 years prior to presentation. Any part of the colon may be affected but the left colon is the predominant location in approximately 75% of patients.⁸ Similarly, the colon was predominantly involved in this patient. Splenic flexure is involved in approximately one-quarter of patients and isolated right colon ischemia (IRCI) in about 10% of cases. The splenic flexure, transverse colon and descending colon were also involved in this patient.

Plain abdominal radiography can reveal nonspecific findings such as thumb printing, air-filled loops, colonic aperistalsis and mural thickening in up to 21% of patients.⁸ Barium enema may suggest colon ischemia in up to 75% of patients with thumb printing being the most common finding. Findings are however nonspecific.⁸

Sonography is a sensitive technique for the early detection of changes in the colon wall resulting from ischemia. Location and length of the involved colonic segment, colon wall thickening and abnormal echogenicity of the pericolic fat and peritoneal fluid are some of the findings on sonography.

Computed tomography (CT) scan may demonstrate bowel wall thickening, thumb printing, and pericolic stranding, with or without peritoneal fluid. If there is total vascular occlusion without reperfusion (infarction), the colonic wall remains thin and unenhancing, associated with dilatation of

the lumen. Even though CT scan was not done in this case, but it may demonstrate a thrombus in the corresponding mesenteric vessel. Pneumatosis and/or gas in the mesenteric veins are ominous signs when associated with bowel wall thickening and are due to bowel infarction. Pneumatosis coli or pneumatosis intestinalis can be diagnosed by demonstrating air bubbles in the colonic or intestinal wall.⁸

Treatment of ischemic colitis depends on acuteness and severity of presentation. Most cases are transient and resolve spontaneously. Very mild cases can be managed on an outpatient basis with liquid diet, close observation and antibiotics. Empiric broad-spectrum antibiotics are given to cover aerobic and anaerobic bacteria and minimize bacterial translocation and sepsis which has been shown to occur with the loss of mucosal integrity.⁸ About 20% of patients with acute IC will require surgery.⁸ At laparotomy, the diagnosis is confirmed and all affected bowel resected. A colectomy is followed by colostomy or ileostomy. Despite these interventions, several case reports describe significant complications, including bowel perforation and shock resulting in death.⁹

The present patient had been placed on conservative treatment including liberal fluid intake, analgesic and antibiotics and made remarkable improvement and is currently on follow up.

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