CASE REPORT

Fulminant ischaemic colitis with atypical clinical features complicating sickle cell disease

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Clinically significant ischaemic bowel injury is an exceedingly rare complication of sickle cell disease. It manifests as acute surgical abdomen and may respond to conservative treatment. An unusual fatal case of ischaemic colitis with minimal abdominal findings in a young male during a sickle cell vaso-occlusive pain crisis is described. This case demonstrates that an acute surgical abdomen should be considered in such patients who fail to respond to conservative management as untreated this condition may be fatal.

Chronic haemolytic anaemia and episodic vaso-occlusive pain characterise sickle cell disease. Virtually every organ system in the body is at risk for ischaemic injury, which results in the characteristic multisystem manifestations of the disease. We describe a case of fatal ischaemic colitis with minimal abdominal findings complicating a sickle cell pain crisis.

CASE REPORT

A 35 year old African-American male with a history of sickle cell anaemia (haemoglobin SS), hypertension, and end stage renal disease and a prior splenectomy and cholecystectomy presented with an acute painful episode involving chest, abdomen, lower back, and extremities. The episode was typical of his previous painful crises and he denied fever, dyspnoea, cough, or diarrhoea. His current medications were enalapril, metoprolol, calcium acetate, and erythropoietin. There was no history of substance abuse. On admission through the emergency department his blood pressure was 186/112 mm Hg, pulse 82/min and regular, respiratory rate 18/min, temperature 37.2°C, and oxygen saturation 99% on 2 litres of oxygen.

Figure 1 (A) Colonic mucosa with inflammation and necrosis (× 40) and (B) at higher magnification (× 100). (C) Submucosal thrombosed blood vessels and vascular congestion with red blood cells sickling (× 100) and (D) at higher magnification (× 600). (All stained with haematoxylin and eosin.)
Abdominal pain is a common component of sickle cell pain crisis. It is thought to result from microvascular occlusion, with resulting ischaemia of the mesentery and abdominal viscera. Usually no precipitating cause is identified and symptoms resolve with conservative measures. Clinically significant ischaemic bowel injury is rare during sickle cell crisis. Ischaemic colitis typically manifests as sudden onset abdominal pain, distension, and the detection of maroon blood in stool. The diagnosis relies on a combination of clinical, radiographic, and endoscopic findings. “Thumb printing” on barium enema, attributed to submucosal oedema and haemorrhage, is the classic radiological finding. However, endoscopic examination with its higher sensitivity and potential for tissue biopsy has largely supplanted barium studies. Angiography is often non-diagnostic as it cannot reliably image the smaller vessels involved in the vascular injury leading to ischaemic colitis. Treatment is determined by the severity of ischaemia and varies from conservative medical treatment to surgery. Increasing abdominal tenderness, rising temperature, metabolic acidosis, and ileus indicate bowel infarction necessitating emergent laparotomy and bowel resection.

To our knowledge, there is only one previous report of ischaemic colitis in adults complicating sickle cell disease. In contrast to the previous report, our case manifested only minimal abdominal findings. The suspicion of acute surgical disease process may mask the progression of ischaemic bowel in patients with unexplained metabolic acidosis.

Ischaemic colitis in sickle cell pain crisis is likely to be the consequence of vascular occlusion and impairment of blood flow in the microcirculation which is responsible for other manifestations of sickle cell disease. The polymaturation of haemoglobin S at low oxygen tension results in a distorted erythrocyte with decreased deformability leading to vascular occlusion and injury. Furthermore, the complex interaction between endothelium, plasma factors, leukocytes, and rigid sickled red cells play an important part in vaso-occlusion. The diffuse ischaemic involvement of large bowel in our case supports small rather than large vessel occlusion. Although the colon with its a relatively low blood flow is considered susceptible to ischaemia, diffuse involvement during sickle cell vaso-occlusive crisis is rare. This may partially be explained by its abundant collateral blood supply and low degree of oxygen extraction. The bowel can tolerate up to a 75% reduction of mesenteric blood flow for 12 hours with no ischaemic changes. However, a low perfusion pressure may significantly increase the likelihood of vascular obstruction and ischaemic injury. Although the specific trigger of the ischaemic colitis in our patient is unknown, it is plausible that predisposing factors may have included end stage renal disease, administration of erythropoietin, a relatively high level of haemoglobin, and a vascular anatomy with poorly developed collaterals.

In summary, although ischaemic injury of the colon is rare during a sickle cell crisis it should be considered in the differential diagnosis of abdominal pain. Unfortunately, generalised pain, and the use of analgesics and sedatives may mask the abdominal findings. Progressive clinical deterioration and
unexplained metabolic acidosis should raise the suspicion of an ischaemic bowel injury and prompt surgical evaluation and treatment.

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REFERENCES


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A bleeding aortic graft enteric fistula diagnosed by push enteroscopy

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