CASE REPORT

Fulminant ischaemic colitis with atypical clinical features complicating sickle cell disease

A Karim, S Ahmed, L J Rossoff, R Siddiqui, A Fuchs, A S Multz

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.\(^1\)\(^2\) Virtually every organ system in the body is at risk for ischaemic injury, which results in the characteristic multisystem manifestations of the disease.\(^1\)\(^2\) 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

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.)
Ischaemic colitis and sickle cell disease

Oxygen. Physical examination was remarkable for tenderness over the sternum, lower back, and lower extremities and presence of an arteriovenous graft on right forearm. Laboratory evaluation revealed a white cell count of 14.9 × 10⁹/L, haemoglobin of 100 g/L, absolute reticulocyte count of 0.217 × 10⁹/L (reference range 0.018–0.158 × 10⁹/L), and platelet count of 273 × 10⁹/L. Serum chemistry showed potassium of 5.3 mmol/l, bicarbonate of 29 mmol/l, blood urea nitrogen of 18.56 mmol/l, and creatinine of 954.7 µmol/l. Liver enzymes were normal and chest radiography revealed a minimal infiltration in the left lower lobe. He was hospitalised and intravenous ceftriaxone and azithromycin were initiated. Hydromorphone was administered for pain control and haemodialysis resulted in normalisation of his blood pressure. Blood cultures were negative and the infection on the chest radiograph resolved within 48 hours. He failed to improve and became febrile to 38.3°C. His haemoglobin concentration decreased to 80 g/l and white cell count increased to 26 × 10⁹/L. Abdominal examination revealed decreased bowel sounds without guarding or rigidity and faecal haemoccult test was negative. He became increasingly lethargic, unresponsive, and required mechanical ventilation and transfer to the intensive care unit. His blood pressure remained stable. Arterial blood gas analysis on 50% fractional inspiratory oxygen revealed a pH of 7.31, carbon dioxide pressure of 3.33 kPa (25 mm Hg) (reference range 4.67–6.00 kPa (35–45 mm Hg)), oxygen pressure 23.5 kPa (176 mm Hg) (10.7–13.3 kPa (80–100 mm Hg)), bicarbonate of 13 mmol/l, and oxygen saturation 99.5%. Serum chemistry demonstrated an anion gap of 30 and lactate level of 11.8 mmol/l. Computed tomography of the head was unrevealing. The computed tomogram of abdomen revealed circumferential inflammation and thickening of the colon with diffuse colonic dilatation consistent with colitis. No free air or perirectal fluid or abscesses were identified. Stool examination was negative for Clostridium difficile toxin. Antibiotic coverage was broadened. A diagnostic peritoneal lavage did not reveal faeculent material and demonstrated a white cell count of 595/µl and erythrocytes of 8268/µl. Sigmoidoscopy was performed and demonstrated areas of necrosis and haemorrhage at the rectosigmoid junction consistent with ischaemic colitis. Emergency total colectomy and partial small bowel resection was performed. The patient continued to deteriorate and became hypotensive, and developed a severe coagulopathy and died.

Pathological examination of the resected bowel revealed multiple erosions and ulcers with gangrenous lesions with extensive transmural necrosis consistent with colitis. No free air or perirectal fluid or abscesses were identified. Stool examination was negative for Clostridium difficile toxin. Antibiotic coverage was broadened. A diagnostic peritoneal lavage did not reveal faeculent material and demonstrated a white cell count of 595/µl and erythrocytes of 8268/µl. Sigmoidoscopy was performed and demonstrated areas of necrosis and haemorrhage at the rectosigmoid junction consistent with ischaemic colitis. Emergency total colectomy and partial small bowel resection was performed. The patient continued to deteriorate and became hypotensive, and developed a severe coagulopathy and died.

DISCUSSION

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 illness was raised by unexplained metabolic acidosis in the context of diminished bowel sounds and was subsequently supported by imaging studies and endoscopy. The paucity of abdominal findings may be explained to some extent by the masking effect of narcotic administration.

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 polymerisation 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, leucocytes, 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, especially if there is a recent sickle cell vaso-occlusive crisis and may be difficult to distinguish from acute abdominal disease processes, such as acute cholecystitis, hepatic infarction, splenic sequestration, and appendicitis. Severe ischaemic colitis may manifest atypically with minimal abdominal findings.

Narco tic administration to the patient with acute surgical disease process may mask the progression of signs and symptoms delaying surgical evaluation and appropriate treatment. Clinicians should have a low threshold of suspicion for ischaemic bowel in patients with unexplained metabolic acidosis.

Learning points

- Sickle cell pain crisis can be complicated by fatal ischaemic colitis and it should be considered in the differential diagnosis of abdominal pain.
- Abdominal pain frequently accompanies sickle cell vaso-occlusive crisis and may be difficult to distinguish from acute abdominal disease processes, such as acute cholecystitis, hepatic infarction, splenic sequestration, and appendicitis.
- Severe ischaemic colitis may manifest atypically with minimal abdominal findings.
- Narcotic administration to the patient with acute surgical disease process may mask the progression of signs and symptoms delaying surgical evaluation and appropriate treatment.
- Clinicians should have a low threshold of suspicion for ischaemic bowel in patients with unexplained metabolic acidosis.
unexplained metabolic acidosis should raise the suspicion of an ischaemic bowel injury and prompt surgical evaluation and treatment.

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REFERENCES

IMAGES IN MEDICINE

A bleeding aortic graft enteral fistula diagnosed by push enteroscopy

A 72 year old man with a previous history of an emergency abdominal aortic aneurysm repair presented with rigors and recurrent episodes of melena. Both oesophagogastroduodenoscopy and colonoscopy were unremarkable. Computed tomography with contrast failed to show evidence of a leak. He subsequently underwent a small bowel push enteroscopy which clearly demonstrated erosion of the Dacron graft through the distal duodenal wall with active bleeding. Emergency surgery was undertaken and the fistula successfully repaired.

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