Henoch–Schönlein purpura as a rare cause of an acute abdomen

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ABSTRACT

A 52-year-old man presented with a purpuric rash affecting his legs and hypertension. He was diagnosed with Henoch–Schönlein purpura and discharged with non-steroidal anti-inflammatory drugs. Three weeks later, he presented again with loss of appetite and vomiting before developing abdominal pain with pyrexia of 38.5°C and rigors. On examination, he was hypotensive with a distended abdomen and a national early warning score of 6. Computed tomography revealed enteropathy of Henoch–Schönlein purpura. The patient deteriorated and went for an emergency laparotomy, where 30cm of ischaemic small bowel was resected.

KEYWORDS

Gastrointestinal surgery – General surgery – Vasculitis – Henoch–Schönlein purpura

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Our case represents a rare medical cause of an acute abdomen. We hope to highlight enteropathy with ischaemic bowel as a possible complication of Henoch–Schönlein purpura (HSP) and outline the immediate surgical management required.

Case history

A 52-year-old man with a background of hypertension presented to the emergency department with a purpuric non-blanching rash on both legs. Physical examination was otherwise unremarkable and initial blood tests (including full blood count and C-reactive protein) were within normal ranges. He was diagnosed with HSP, and discharged with reassurance and non-steroidal anti-inflammatory drugs (NSAIDs). He was readmitted three weeks later with pyrexia of 38.5°C, rigors, vomiting and abdominal pain. Initial observations showed he was tachycardic and tachypnoeic, with a mildly distended but soft abdomen. There were no overt signs of peritonism. As the day progressed, the abdominal pain increased in severity. On reassessment, he had developed a distended abdomen with localised guarding on the left, suggesting localised peritonism.

Investigations

Laboratory tests showed a high white cell count ($24.5 \times 10^9/l$) and a high C-reactive protein level (364 mg/l). The patient had an acute kidney injury with urea of 11.6 mmol/l, creatinine of 219 $\mu\text{mol/l}$ and an estimated glomerular filtration rate of 28ml/min/1.73 m² (from a baseline of 87 ml/min/1.73m²). Venous blood gas testing demonstrated elevated

lactate of 4.6mmol/l. Abdominal radiography revealed dilated small bowel loops.

Urgent abdominal computed tomography (CT) showed no evidence of intestinal obstruction, pneumoperitoneum or ischaemic bowel. The imaging did demonstrate features in keeping with the rare finding of enteropathy of HSP (Fig 1)

Figure 1 Abdominal computed tomography demonstrating dilated small bowel with mural thickening, mesenteric fat stranding and free fluid along the paracolic gutters

Figure 2 Intraoperative photograph demonstrating ischaemic areas of small bowel

Figure 3 Illustration of the postoperative anatomy with a venting gastrostomy (A), 100cm of small bowel ending in an ileostomy (B), 180cm of defunctioned small bowel ending in a mucous fistula (C) and a large bowel mucous fistula (D)

and close vigilance was recommended from the radiologists as there was a risk of developing bowel ischaemia.

Differential diagnoses

This patient presented with an acute abdomen, for which the list of differential diagnoses is vast. After taking the patient's history into consideration, the likely causes were perforation secondary to NSAIDs, intra-abdominal sepsis and bowel ischaemia. All these require a similar initial assessment and management, often followed by acute surgical management to prevent mortality.

Treatment

Our patient was treated initially for sepsis with the Sepsis Six care bundle. High flow oxygen was administered, lactate was measured and blood cultures were taken. He was given intravenous antibiotics, catheterised and intravenous fluids

were administered. Hourly urine output and observations were monitored. Despite the CT failing to demonstrate any perforation, the raised inflammatory markers in conjunction with the patient's worsening symptoms and clinical signs raised the clinical concern of perforation, and he subsequently underwent an emergency laparotomy.

On induction of anaesthesia, the patient went into septic shock, managed by the anaesthetic team. Once stabilised, an emergency laparotomy was performed, where 2l of free fluid were found in the peritoneal cavity with a localised perforation in the jejunum. Multiple dusky patches of small bowel were also evident (Fig 2). The non-viable small bowel was resected and the decision was made by the surgical team to place a venting gastrostomy because of the anticipated need for prolonged gastric decompression. A venting gastrostomy was deemed safer than the alternative of multiple nasogastric tubes with their associated risks of blockage, dislodgement and migration. The patient was transferred to the critical care team with an open laparostomy to prevent the development of abdominal compartment syndrome.

A second laparotomy conducted two days later revealed further ischaemia and an additional resection of small bowel was completed. The remaining bowel anatomy is illustrated in Figure 3.

Outcome and follow-up review

The patient remained in the intensive care unit for 24 days after surgery. Small bowel biopsy samples subsequently diagnosed leucocytoclastic vasculitis, consistent with HSP enteropathy. He was discharged after 72 days in hospital with a peripherally inserted central catheter to facilitate total parenteral nutrition. He is currently awaiting surgery to restore gastrointestinal continuity and is being followed up by vasculitis specialists.

Discussion

There are many causes of an acute abdomen and most commonly these are surgical. However, there are important underlying medical causes that can present in a similar way. Surgical causes include ruptured organs, bowel obstruction and perforated viscera. The medical causes are numerous and seen less often, making the initial diagnosis challenging.

HSP is a systemic vasculitis typically affecting young males, with a peak incidence at six years of age.¹⁻⁵ The characteristic palpable purpuric rash is non-blanching and purple in colour, typically affecting the buttocks and extensor surfaces. This purpura must be present for a diagnosis of HSP to be made, along with one of the following: abdominal pain, immunoglobulin A deposition on biopsy, arthralgia or arthritis and renal involvement (proteinuria with/without haematuria).⁴

In 89% of adult cases, HSP resolves spontaneously and supportive therapy is often all that is required.² NSAIDs have a role in managing arthralgia but the gastrointestinal side effects of these medications must be considered in the presence of abdominal involvement.

Adults with HSP are more likely to experience renal complications but less likely to present with abdominal pain.^{2,4} The abdominal complications of HSP in adults are rarely seen. They may include bowel ischaemia, infarction, perforation or haemorrhage.^{3,5} One reported series from 2007 detailed three cases of HSP in adults presenting with severe abdominal pain, none of which required surgical management.⁶ Our case demonstrates the very rare complication of bowel ischaemia and perforation secondary to HSP. Close monitoring, frequent reassessment and early surgical intervention helps to reduce morbidity and improve outcome.

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