On-line Case Report

Henoch–Schonlein purpura with ischaemic bowel

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ABSTRACT

Henoch–Schonlein purpura is a vasculitis affecting small arterial vessels. Occasionally, cases are referred for a general surgical opinion due to bowel involvement in the form of abdominal pain with or without rectal bleeding. However, surgical intervention is rarely required. We describe a case of Henoch–Schonlein purpura in a young man who went on to develop ischaemic bowel requiring resection.

Keywords: Henoch–Schonlein purpura – Ischaemic bowel

An 18-year-old man was admitted with a 3-day history of ankle pain and swelling and a non-blanching purpuric rash over the dorsum of both feet. The patient also complained of wrist swelling and tenderness, as well as abdominal pain. In the preceding days, the patient had had a mild flu-like illness with a sore throat. Inflammatory markers were raised (white blood cells, 11.9; C-reactive protein, 11). A clinical diagnosis of Henoch–Schonlein purpura was made, which was further supported by an autoimmune and vasculitic screen, revealing a mildly raised IgA titre. The patient was managed supportively without the addition of immunosuppression.

The patient was re-admitted 22 days later with sudden onset of central, colicky, abdominal pain associated with anorexia, loose stools and rectal bleeding. On examination, generalised lower abdominal tenderness was present with reduced bowel sounds. The initial rash had remained predominantly affecting the lower limbs. The white cell count was found to be raised (20.5), as was the C-reactive protein (62). At laparotomy, one litre of haemorrhagic fluid was found. There was full thickness ischaemia of the small

bowel, 225 cm from the duodenojejunal flexure involving 10 cm of bowel. Patchy areas of serosal ischaemia were also found in the neighbouring segment of small bowel involving up to 50 cm of bowel. The distal small bowel was found to be normal up to the ileocaecal junction. Small bowel resection was performed, with formation of a proximal ileostomy in the right iliac fossa. The distal end was brought out at the lower end of the wound as a mucus fistula. Post-operatively, immunosuppression was achieved with methylprednisolone pulse therapy.

Histopathological analysis demonstrated acute small bowel ischaemia. Extensive mucosal ulceration with moderate-to-severe patchy infiltrate of acute and chronic inflammatory cells in the lamina propria was seen. Dilated vascular channels engorged with blood were noted in the submucosa. In addition, extensive interstitial haemorrhage involving mainly the lamina propria and submucosa was found, along with marked oedema. Haemorrhage extended into the muscle coat and mesentery. No thrombus or evidence of vasculitis was seen. The features were consistent with acute haemorrhagic infarction.

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Discussion

Henoch–Schonlein purpura is a multisystem small vessel vasculitis characterised by involvement of skin, joints, gastrointestinal tract and kidneys. Henoch–Schonlein purpura can occur at any age from infancy to adulthood although 50% of cases occur in children under 5 years of age and 75% under 10 years of age. Henoch–Schonlein purpura is uncommon in adults with few reported cases; however, adult Henoch–Schonlein purpura represents a more severe clinical syndrome with reportedly worse outcomes.¹ Adult Henoch–Schonlein purpura is characterized by a lower frequency of abdominal pain and fever, and a higher frequency of joint symptoms and renal involvement.²

Clinically, Henoch–Schonlein purpura presents with a classic tetrad of symptoms (with reported frequencies): cutaneous purpura (100%), arthralgia or arthritis (82%), abdominal pain (63%), gastrointestinal bleeding (33%). The aetiology of Henoch–Schonlein purpura is unclear. Half of the cases are associated with a preceding upper respiratory tract infection and associations have been postulated with infectious agents, particularly β -haemolytic streptococci, mononucleosis and parvovirus B19.

Gastrointestinal symptoms in Henoch–Schonlein purpura may mimic the acute abdomen. In children, the most common abdominal complication is intussusception. In adults, bowel ischaemia, infarction, intestinal perforation, fistula formation, upper gastrointestinal haemorrhage have all been described. Carmichael *et al.*³ described a case of extensive small bowel infarction with fatal outcome. Another report, in 2002,⁴ described a 41-year-old man with ischaemic small bowel presenting with an acute abdomen at the time of the initial rash. In that case also, the patient was managed conservatively; despite this, the patient's condition deteriorated after 4 days.

The natural history of Henoch–Schonlein purpura is self-limiting in most cases. In our case, however, the

presence of bowel ischaemia necessitated small bowel resection. Typically, supportive therapy and judicious hydration form the mainstay of treatment for Henoch–Schonlein purpura. Non-steroidal anti-inflammatory drugs help to alleviate joint and soft-tissue discomfort. Corticosteroids administered during the acute phase help to ameliorate the symptoms of severe abdominal pain, arthralgia. Recently, a randomised, double-blind, placebo-controlled study confirmed that prednisolone in a dose of 1 mg/kg a day for 2 weeks, then tapered for 2 weeks decreased the intensity and severity of gastrointestinal symptoms.⁵ In severe cases, steroid-sparing agents (*e.g.* cyclophosphamide, cyclosporin, azathioprine) may be used.

Conclusions

Abdominal pain in association with Henoch–Schonlein purpura is well documented in the paediatric literature. However, in adults, bowel pathology is unusual. Emerging data support the early use of immunosuppressants in cases of severe abdominal pain. Early involvement of general surgeons is advised to monitor abdominal involvement.

References

1. McCarthy R, Rosen T, Chen SH, Raimier SS. Adult Henoch–Schonlein purpura with fatal complications. *Arch Dermatol* 2001; **137**: 19–21.
2. Blanco R, Martinez-Taboada VM, Rodriguez-Valverde V, Garcia-Fuentes M, Gonzalez-Gay MA. Henoch–Schonlein purpura in adulthood and childhood: two different expressions of the same syndrome. *Arthritis Rheum* 1997; **40**: 859–64.
3. Carmichael P, Brun E, Jayawardene S, Abdulkadir A, O'Donnell PJ. A fatal case of bowel and cardiac involvement in Henoch–Schonlein purpura. *Nephrol Dial Transplant* 2002; **17**: 497–9.
4. Lawes D, Wood J. Acute abdomen in Henoch–Schonlein purpura. *J R Soc Med* 2002; **95**: 505–6.
5. Ronkainen J, Koskimies O, Ala Houhala M, Antikainen M, Merenmies J, Rjantie J *et al.* Early prednisolone therapy on Henoch–Schonlein purpura: a randomised, double-blind placebo-controlled trial. *J Pediatr* 2006; **149**: 241–7.