EDITOR'S QUIZ: GI SNAPSHOT

Jejunal perforation and central retinal vein occlusion in a 55-year-old European man

CLINICAL PRESENTATION

A 53-year-old white European male builder presented emergently with haematemesis and melaena. Four months previously, he underwent an exploratory laparotomy, small bowel resection and anastomosis for a spontaneous jejunal perforation. His immediate postoperative course was uneventful. One month prior to this admission he lost the sight in his right eye because of a central retinal vein occlusion (CRVO): blood cultures, vasculitis and thrombophilia screens were negative and an echocardiogram and carotid Dopplers unremarkable. There was no significant family history. For the treatment of his CRVO, he was taking 15 mg of prednisolone daily and 150 mg of aspirin. He consumed 14 units of alcohol per week but did not smoke cigarettes.

Figure 1 Macroscopic appearance of the small bowel.

Figure 2 H&E x10 of small bowel mucosa.

Figure 3 H&E x40 of small bowel mucosa.

Initial laboratory tests showed haemoglobin 103 g/L (normal range 130–180), mean corpuscular volume 97.6 fL (82–98) and lymphopenia $0.80 \times 10^9/L$ (1.1–3.5). Tissue transglutaminase antibodies were 1.2 U/mL (0–10.0) with normal IgA levels and antiendomysial antibodies were negative.

Oesophagogastroduodenoscopy demonstrated a duodenal ulcer with a bleeding vessel that was clipped and injected with epinephrine. Four days later, however, he developed peritonitis that required an emergency laparotomy and small bowel resection (see figures 1–3).

QUESTION

What was the diagnosis?

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ANSWER

The small bowel macroscopically was thickened and oedematous with multiple perforations and abundant serosal exudate. The mucosa showed blunting of the intestinal villi with intraepithelial lymphocytosis (see figures 4 and 5): the immunophenotype was characteristic for type 1 refractory coeliac disease. A population of neoplastic T lymphocytes with a clonal cytotoxic immunophenotype were seen to invade the lamina propria indicating enteropathy-associated T cell lymphoma.

Figure 4 H&E x10 showing areas of mucosa ulceration and wall of the perforation site with florid acute inflammation.

Figure 5 H&E x20 showing diffuse infiltration by a population of blasts with abundant apoptotic debris.

About one-third of patients diagnosed with small bowel lymphoma have evidence of proximal villous atrophy.¹ Our patient was seronegative but carried the coeliac risk-serotype HLA-DQ2. Seronegative coeliac disease is rare among patients with coeliac disease but is a common cause of seronegative villous atrophy and is associated with high mortality.^{2,3} Enteropathy-associated T cell lymphoma is a rare form of extranodal T cell lymphoma that is usually, but not always, associated with untreated or undiagnosed coeliac disease.⁴ Patients are typically diagnosed in the sixth or seventh decade of life and are more often male. The jejunum is the most commonly affected part of the GI tract and the diagnosis is frequently made because of stricturing or perforation.⁴ The prognosis is poor: the estimated overall and treatment failure-free survival at 5 years is 20% and 4%, respectively.⁵

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