Steroids in villous atrophy: opening a can of worms?

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Abstract

A 60 year old man presented to the emergency department with septic shock and respiratory distress. He noted significant weight loss over the last 6 months with associated postprandial bloating. Endoscopy demonstrated partial villous atrophy. He had been on a gluten free diet, received empirical treatment for giardiasis and was receiving treatment with high dose systemic steroids for retroperitoneal fibrosis. This case discusses histology findings, differentials and reason for this acute presentation.

Clinical presentation

A 60-year-old man who immigrated to England from Bangladesh presented in extremis with septic shock and respiratory distress. He had lost 25 kg over 6 months and described postprandial bloating without diarrhoea. Endoscopy performed 5 months earlier demonstrated partial villous atrophy and CT was consistent with retroperitoneal fibrosis. Coeliac serology, strongyloides antibodies and stool cultures were negative; he had received empirical treatment with tinidazole for giardiasis and was put on a trial of a gluten free diet. His past medical history included type 2 diabetes mellitus, hypertension, and a clinical diagnosis of protein losing enteropathy secondary to lymphangiectasia. He was receiving treatment with high dose systemic corticosteroids (2 mg/kg) for retroperitoneal fibrosis and persistent villous atrophy that had not responded to the gluten free diet.

On examination the patient was febrile, hypotensive and tachycardic. Empiric treatment was initiated with intravenous hydrocortisone to cover for adrenal insufficiency. Examination noted marked cachexia and T12 dermatome shingles infection. Blood tests were consistent with disseminated intravascular coagulation,

severe anaemia (Hb 3.5 g/dl) and leucocytosis (white blood cells 21.1×10^9 , neutrophils 20.4×10^9 /l, eosinophils 0.1×10^9 /l). The HIV and anti-HTLV1 antibody test were negative.

A repeat oesophago-gastro-duodenoscopy was performed.

Questions

- What does the histology show?
- What is the best diagnostic test to identify the underlying pathology?
- What was the cause for his sudden deterioration?

Answers

Histology shows *Strongyloides stercoralis*. This intestinal nematode is endemic in tropical areas. Adult worms are found in the duodenum/jejunum but larvae may penetrate the bowel wall and migrate to the lung. Chronic infection may be asymptomatic or present with vague abdominal discomfort, bloating and loose stools. Heavy infestation may cause bowel wall thickening, diarrhoea, malabsorption, asthma and pulmonary haemorrhage. Eosinophilia is found in 50% of cases; however, in this patient it was only present during the acute admission. Stool microscopy often reveals larvae or eggs but may be negative in a third of cases.¹ Endoscopy may show villous atrophy, lymphocytic infiltration, eggs and larvae in the submucosa. The gold standard test is an *S stercoralis* ELISA. This has a sensitivity of 80–100%; however this is much reduced in immunocompromised patients.²

Immunosuppression with corticosteroids is an important cause of *S stercoralis* hyperinfection. This is characterised by increased worm burden resulting from impairment of host cellular immunity. Parasitic penetration of the gut predisposes to Gram-negative sepsis from enteric bacteria. Death of intraluminal

Figure 1 H&E stain of gut mucosa

Figure 2 H&E stain of gut mucosa

and intramural larvae leads to production of inflammatory and vasoactive mediators, resulting in disseminated intravascular coagulation.

The case highlights the need to exclude and treat strongyloidiasis as a cause of villous atrophy prior to starting immunosuppressants as this can lead to life-threatening hyperinfection. It should be noted that villous atrophy is not pathognomonic for coeliac disease. Villous atrophy may occur as the result of infections, immune deficiency and other autoimmune conditions such as inflammatory bowel disease.³ A diagnosis of coeliac disease is suggested by the presence of villous atrophy in addition to >25 intraepithelial lymphocytes per 100 enterocytes.⁴ Finally, the HLA-DQ phenotype may be used as a means of excluding coeliac disease in patients with equivocal small bowel histological findings.⁵

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