LESSON OF THE MONTH

A case of weak muscles

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Background: Gut involvement in inflammatory myositis is rare but causes significant morbidity and mortality.

Case report: A case of eosinophilic gastroenteritis and polymyositis occurring in the same patient is described. The interface of visceral and striated muscle involvement is discussed. The pathophysiology of eosinophilic gastroenteritis and the spectrum of gastrointestinal involvement in inflammatory myositis are also discussed.

Results: Both gastrointestinal and skeletal muscle symptoms improved with immunosuppression, suggesting a possible common underlying mechanism.

Gastrointestinal manifestations in inflammatory skeletal myopathies are known.^{1,2} There is a wide spectrum of involvement, from oesophageal dysmotility² to severe gastric outflow obstruction.³ The stomach and small intestine are the most commonly affected sites in most series. Rarer forms of gut disease, such as eosinophilic gastroenteritis in the case we describe, may also be associated with inflammatory myositis. Treatment with immunosuppressive drugs not only improved the skeletal muscle symptoms but also the gastrointestinal symptoms due to visceral

involvement. In such patients, gastrointestinal symptoms should be included in the assessment of disease activity.

CASE REPORT

A 56 year woman, originally from Pakistan, had been resident in the United Kingdom for 30 years. She was admitted to her local hospital with a two year history of generalised weakness and recurrent abdominal pains. She also had occasional nausea and vomiting, but no bowel disturbance or weight loss.

Two years before her admission she had an episode of acute pancreatitis with an amylase of 50 000 U/l. One year previously she was seen in the accident and emergency department of another hospital with chest pain and a creatinine kinase of 1510 IU/l (normal range 20-175). A diagnosis of angina was made at that time. There was no history of asthma or atopy.

On the current admission, she had a marked peripheral blood eosinophilia of $9.7 \times 10^9/l$ (normal range $0.04-0.4 \times 10^9/l$). Her haemoglobin, total white cell count, platelets, urea, electrolytes, liver function tests, C reactive protein, and erythrocyte sedimentation rate were all normal. Her creatinine kinase remained high at 990 IU/l. A full autoantibody screen, including antinuclear antibody, extractable nuclear antigens, rheumatoid factor, cytoplasmic antinuclear cytoplasmic antibodies (cANCA) and perinuclear ANCA (pANCA) were all

Figure 1 The skeletal muscle biopsy with the inflammatory infiltrate seen on the left and the marked muscle fibre damage on the right. (Haematoxylin and eosin $\times 20$.)

Figure 2 The muscle layer of the open small intestinal biopsy specimen showing a small but definite infiltrate of eosinophils (arrow). (Carbolchromatope $\times 40$.)

negative. She also had a normal chest radiograph and electrocardiograph. An examination of stools for parasites was negative.

On examination she was overweight but did not look unwell. Abdominal palpation disclosed mild tenderness in the periumbilical area, but no organomegaly. Mild wasting of the quadriceps muscles was noted. There was marked proximal muscle weakness in the arms and legs.

In view of the proximal muscle weakness and raised creatinine kinase levels, a muscle biopsy and electromyography (EMG) were arranged. The EMG showed a symmetrical, mainly proximal myopathy. The quadriceps muscle biopsy showed patchy lymphocytic infiltration, muscle fibre necrosis, variation in fibre size with fibre atrophy (fig 1), confirming a diagnosis of polymyositis. Eosinophils were not a feature.

Bowel studies were then made. A barium follow through showed an inflamed distal ileum with thickened valvulae conniventes; two strictures were also seen. An upper gastrointestinal endoscopy showed mild antral gastritis. A colonoscopy disclosed an oedematous ileocaecal valve with diffuse erythema and granularity. A gastric biopsy was normal and the colonic biopsies showed a minimal increase in eosinophils, including occasional ones infiltrating crypts. It was considered abnormal but at the time not diagnostic. A mini-laparotomy was performed to obtain a full thickness small intestinal biopsy specimen for a definitive diagnosis. At laparotomy, the two strictures noted on barium studies were identified—one was biopsied. No other macroscopic changes were seen at laparotomy. Histology of the small intestinal biopsy showed a normal mucosa but submucosal oedema and a definite though mild accompanying infiltrate of eosinophils in the submucosa and both layers of the muscularis propria (fig 2). The findings considered alongside those in the colonic mucosa and the blood eosinophilia indicated a diagnosis of eosinophilic gastroenteritis. No changes suggestive of a vasculitis were seen.

Treatment was started with prednisolone 30 mg, reducing by 5 mg each month over the next few months. Methotrexate was added as a steroid sparing agent. In view of the patient's persisting waddling gait and muscle pains, she was given two-monthly α globulin infusions. The polymyositis and also the gastrointestinal symptoms improved dramatically. At the most recent clinic review, the proximal myopathy remained unchanged and she had no gastrointestinal symptoms. The eosinophil count was suppressed to $0.6 \times 10^9/L$, with normal creatine kinase levels.

DISCUSSION

In polymyositis, involvement of smooth muscle of any part of the gastrointestinal tract is uncommon, except where polymyositis occurs alongside systemic sclerosis as an overlap syndrome. We have been unable to find any previous cases of eosinophilic gastroenteritis and polymyositis in the same patient.

Abdominal pain and eosinophilic infiltration of the bowel unassociated with polymyositis occur in the idiopathic form, known as eosinophilic gastroenteritis, or secondary to various conditions discussed below. No stool cultures from our patient grew any parasites, and no biopsy showed granulomas, invasive micro-organisms, or viral inclusions. Our patient's polymyositis and gastrointestinal symptoms all improved with immunosuppression, suggesting a common mechanism for the pathophysiology of both the smooth and striated muscle systems affected in this case.

Eosinophilic gastroenteritis itself is an uncommon disorder affecting one or more parts of the gut. It is characterised by gastrointestinal thickening of one or all layers by oedema and infiltration by eosinophils.^{4,5} Most commonly it affects the gastric antrum and proximal small intestine. The most common cause is infiltration of eosinophils as part of a general tissue reaction in a range of conditions, such as inflammatory bowel disease, collagen diseases, primary vasculitic disorders, especially Churg-Strauss vasculitis (where eosinophilia is often a prominent feature), and even lymphoma. Because eosinophilic infiltration may be part of several inflammatory gastrointestinal conditions a confident histological diagnosis can be difficult, as it was in this case, unless infiltration is florid. A peripheral blood eosinophilia is always a helpful adjunct to establishing a firm diagnosis.

Perhaps surprisingly, our patient had no evidence of eosinophilic infiltration on her skeletal muscle biopsy specimen, because idiopathic eosinophilic myopathies have been described.^{6,7} Cardiac and neurological involvement are the main causes of morbidity,⁸ but gastrointestinal involvement is rarely documented in these cases.

Gastrointestinal involvement associated with inflammatory skeletal myopathies is rare. Our unit has recently reported another case of chronic inflammatory polymyositis with visceral involvement.³ Cases of pharyngeal and oesophageal dysphagia complicated by aspiration are recognised in patients with polymyositis. In the former, symptoms are characterised by difficulty in initiating swallowing, nasal regurgitation of liquids, and dysphonia. One case of megaesophagus⁹ and another of oesophageal rupture¹⁰ have been reported. More commonly, chronic distal oesophagitis predisposes to stricture formation. Scintigraphy has been used to show delayed oesophageal and gastric emptying in patients with polymyositis or dermatomyositis.¹¹ The possible cause of such effects may be impaired smooth muscle peristaltic activity from muscle atrophy, fibrosis, lymphocytic infiltration, or secondary to neurological dysfunction.

THE LESSONS

- In patients with polymyositis, gastrointestinal symptoms should be taken seriously.
- Upper gastrointestinal endoscopy and a barium swallow may be insufficient to establish the diagnosis; a full thickness gut biopsy is needed.
- Treatment of skeletal and striated muscle disease with high dose immunosuppression may bring about improvement.
- Visceral muscle involvement can occur in association with inflammatory skeletal muscle myopathies; early detection and treatment will help prevent morbidity and mortality.
- Screening for gastrointestinal symptoms must also be carried out to assess disease activity.

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