LESSON OF THE MONTH

Polymyositis as a cause of total gut failure

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Background: Gastrointestinal manifestations are seen in systemic sclerosis and mixed connective tissue disorders but are rare in pure polymyositis.

Case report: A 44 year old woman with polymyositis who developed total gut failure requiring treatment with total parenteral nutrition is described.

Results: The patient's polymyositis is now fully controlled biochemically, but her gastrointestinal symptoms persist.

Gastrointestinal manifestations are well recognised in patients with systemic sclerosis and mixed connective tissue disorders. In pure polymyositis they are rare, with the exception of pharyngeal dysphagia.^{1,2} The case of a 44 year old woman with polymyositis who developed total gut failure requiring treatment with total parenteral nutrition is described here.

CASE REPORT

In 1994 a 44 year old Afro-Caribbean woman presented with a five month history of exertional dyspnoea, cough, and proximal muscle weakness. Salient features on examination were power of 3/5 proximally and 4/5 distally. Her chest was clear and there was no rash. Table 1 shows the relevant serological findings. An EMG and deltoid muscle biopsy were characteristic of polymyositis. A chest x ray examination and computed tomography of the abdomen and pelvis showed no evidence of malignancy. Treatment was started with oral prednisolone 60 mg daily; muscle strength improved and the creatine kinase (CK) level fell.

She subsequently developed nausea and abdominal distention, an axial x ray examination disclosed dilated loops of small bowel, and an obstruction was diagnosed. At laparotomy dilated loops predominantly of the small bowel were seen but no obstructing lesion was located. A few adhesions following a hysterectomy were divided.

On follow up her polymyositis was inadequately controlled (CK 452–3000 IU/l). Treatment included oral prednisolone 15–30 mg daily and methotrexate 10–20 mg/week. Azathioprine was substituted for methotrexate owing to a lack of

Figure 1 (A) A barium follow-through showing a dilated oesophagus with markedly dilated loops of the small bowel; (B) abdominal computed tomography showing dilated, fluid filled loops of the small and large bowel.

Table 1 Serological findings

Variable	Value
Creatine kinase (IU/l)	17700
LDH (IU/l)	627
AST (IU/l)	455
ANA	1/400
Anti-Jo antibody	+ve
dsDNA, ANCA, LKM, smooth muscle, parietal cell, thyromicrosomal, thyroglobulin antibodies	–ve

LDH, lactate dehydrogenase; AST, serum aspartate aminotransferase; ANA, antinuclear antibodies; ANCA, antineutrophil cytoplasmic antibodies; LKM, liver/kidney microsome antibodies.

Abbreviations: BMI, body mass index; CK, creatine kinase; GI, gastrointestinal; TPN, total parenteral nutrition

clinical response. She was admitted to hospital several times with nausea and vomiting, each settling with conservative management. Symptoms were initially attributed to methotrexate but persisted even when the drug was discontinued.

By 1998 she was housebound with severe malabsorption, her body mass index (BMI) was 15, albumin 26 g/l, haemoglobin 72 g/l, and potassium 2.7 mmol/l, so a referral was made to a gastroenterologist. Investigations included upper gastrointestinal (GI) endoscopy: Barrett's oesophagus, duodenal biopsies were normal. A barium follow-through showed a dilated oesophagus with markedly dilated loops of the small bowel and prolonged transit time (fig 1A). Abdominal computed tomography showed dilated, fluid filled loops of the small and large bowel (fig 1B). Colonoscopy failed twice as adequate preparation was precluded by intestinal dysmotility. A barium enema showed multiple redundant loops of atonic bowel. A hydrogen breath test was compatible with bacterial overgrowth. A pancrealauryl test confirmed malabsorption.

Parenteral hydrocortisone, intramuscular methotrexate, cyclical antibiotics, prokinetic agents, and enteral nutrition were started. Her BMI fell to 14, haemoglobin to 51 g/l, and albumin to 25 g/l. Ultimately, treatment was started with total parenteral nutrition (TPN) and iron, and she was trained to manage TPN at home.

Twenty four months later she is physically independent, her BMI is 24, and haemoglobin, CK, and albumin are normal, but she has developed diabetes mellitus. She tolerates small quantities of food orally but remains dependent on TPN. Current treatment includes TPN, prednisolone 12.5 mg daily, methotrexate 7.5 mg subcutaneously weekly, oestraderm patches, calciferol 500 µg daily, and lansoprazole 30 mg daily.

DISCUSSION

Our patient had no evidence of an overlap syndrome, yet developed profound GI manifestations with an atonic bowel, bacterial overgrowth, and malnutrition, only rectified by TPN. A full thickness intestinal biopsy specimen was not obtained

owing to her poor clinical state, so it remains a possibility that her GI disease was a chance association with polymyositis. There have, however, been two other case reports of pseudo-obstruction in patients with pure polymyositis.^{3,4} There are no reported data on the effect of treatment of polymyositis on GI symptoms. Although our patient's polymyositis is now fully controlled biochemically, her GI symptoms persist after minimal oral intake.

THE LESSONS

- Detection of GI involvement in connective tissue disorders requires a high degree of awareness by the doctor as malnutrition often develops insidiously.
- GI involvement can usually be rectified by enteral supplementation, but a small minority of patients need parenteral nutrition, and a multidisciplinary approach is required.

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