Polymyositis associated with ulcerative colitis

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Abstract
An elderly woman with chronic ulcerative colitis who developed proximal muscle weakness, increased serum creatine phosphokinase activity, and histological and electromyographic abnormalities characteristic of polymyositis is described. Treatment with corticosteroids and 5-acyclysalic acid was followed by a remission in bowel symptoms, improvement in muscle power, and reversal of electromyographic changes. An autoimmune link between the two disorders seems likely.

Extraintestinal manifestations of inflammatory bowel disease are known to affect several organ systems. Although 'granulomatous' myositis is mentioned occasionally as a rare complication of inflammatory bowel disease, most reports on ulcerative colitis do not mention any type of myositis as a complication or association of the disorder. We have been able to find only five cases of myositis of different types complicating ulcerative colitis, of which only two are published in English. Because of its rarity, we document the case of an elderly woman who developed polymyositis as a complication of long standing ulcerative colitis.

Case report
A 78 year old woman was admitted to hospital for investigation and management of diarrhoea and muscle weakness. She had been having recurrent episodes of diarrhoea associated with the passage of blood and mucus, low grade fever, and a feeling of exhaustion for the past 15 years. Each episode lasted for 7 to 10 days, followed by a symptom free period of 2-3 months. She had never visited hospital and was treated symptomatically by general practitioners with anti-diarrhoeal drugs. During the past 15 days, she had started having arthralgias that affected multiple joints and appreciable proximal muscle weakness of both lower and upper limbs. The patient was prescribed indomethacin (25 mg thrice daily) for arthralgia by her treating physician. She had lost about 4 kg of weight over the previous 2 weeks.

Physical examination at the time of hospital admission showed moderate pallor and mild dehydration. The patient’s pulse rate was 96 bpm and the volume was slightly decreased. Her blood pressure was 100/60 mm Hg. The small joints of the hands were tender to pressure and movement. Abdominal examination showed mild tenderness in the hypogastrum, and the patient’s spleen was palpable 2 cm below the left costal margin and was firm in consistency. Neurological examination showed weakness of the proximal muscles of the lower and upper limbs and muscles of the trunk and flexors of the neck (grade 3/5). The superficial and deep reflexes were normal and her plantars were flexor. Ocular fundi were normal.

Laboratory investigations showed a haemoglobin concentration of 78 g/l and a haematocrit of 23%. The patient’s total leukocyte count was 8600/mm³ (8.6×10⁹/l), with neutrophils 85%, lymphocytes 13%, and eosinophils 2%. The erythrocyte sedimentation rate was 67 mm in the first hour (Westergren) and reticulocyte count 5%. A coagulogram was normal. Platelets were 110000/mm³ (110×10⁹/l). Serum Na was 135 mmol/l and K 3.3 mmol/l. Blood urea was 28 mg/dl (9.9 mmol/l) and serum creatinine 1.0 mg/dl (88.4 mmol/l). Serum bilirubin was 0.8 mg/dl (13.6 mmol/l); SGOT was 13 U/l, SGPT was 7 U/l and alkaline phosphatase was 10 KAU. The total serum protein was 74 g/l with an albumin concentration of 36 g/l, globulin 38 g/l, and an albumin/globulin ratio of 0.95. The patient’s urine was negative for protein and sugar, and microscopy showed 10–12 pus cells/high power field. Urine culture showed growth of Pseudomonas aeruginosa. Stools showed the presence of red cells (+ + + +) but no ova or cysts, and culture grew Candida albicans. Upper gastrointestinal endoscopy showed features of oesophagitis and erosive gastritis. A barium enema showed extensive ulceration in the entire large bowel (Figs 1 and 2) and pseudopolyp formation (Fig 3). Colonoscopy confirmed the presence of multiple ulcerations and pseudopolyps. Rectal biopsy specimen showed infiltration of the lamina propria by inflammatory cells severe mucodepletion, and cryptitis (Fig 4). No granuloma formation or vasculitis was noted. A biopsy specimen from the left quadriceps muscle showed focal intermyseal infiltration by lymphocytes and occasional phagocytosis of degenera-
tive muscle fibres (Fig 5). No evidence of regenerative activity, vasculitis, or granuloma was seen. Electromyography of the right deltoid and right quadriceps muscles showed normal insertional activity. No spontaneous activity was observed. Motor unit potentials were small in amplitude and duration. The interference pattern was full, and 30–40% polyphasia was noted (Fig 6). Myopathic motor unit potentials and significant polyphasia were consistent with the diagnosis of polymyositis.

Nerve conduction studies were normal. The patient’s creatine phosphokinase activity was increased five fold (1010 U/l). Rheumatoid factor was positive, and antinuclear antibodies were negative.

COURSE AND MANAGEMENT
The patient was put on parenteral nutrition, oral prednisolone (40 mg once daily), and 5-acetylsalicylic acid (400 mg thrice daily). She also received 20 mg of prednisolone daily by enema and 3 units of blood. In addition, she was given ciprofloxacin (500 mg twice daily) for 7 days for urinary tract infection and ketoconazole (200 mg twice daily) for candidiasis. Her ulcerative colitis went into remission after 4 weeks and a significant improvement in her muscle power (grade 4/5) became noticeable after 12 weeks of therapy. Repeat study of the electromyographic pattern 3 months later showed a distinct improvement and this was accompanied by a significant decrease in the serum creatinine phosphokinase activity. The enlarged spleen regressed in size and became impalpable.

Discussion
Skeletal muscle involvement associated with inflammatory bowel disease was first reported in 1970 by Spiro, who illustrated granuloma formation in striated muscle in a patient with Crohn’s disease without giving any further details. In a subsequent study, Tydd and Dyer performed muscle biopsies in 15 patients with Crohn’s disease in a search for granulomata. None of these patients, however, had any clinical evidence of myopathy, and the muscle histology was normal in all of them. In 1976, Menard et al reported granulomatous myositis and myopathy in a middle aged man with Crohn’s disease. Two more cases were reported subsequently, one by Gilliam et al, who described an unusual vasculitic myositis in a young man with Crohn’s disease, and another by Hall et al, who reported a non-specific localised myositis without histological evidence of muscle granulomata or vasculitis in a 32 year old woman with long standing Crohn’s disease.

The earliest documentation of myositis com-
Figure 6: Electromyograph of the right deltoid muscle showing myopathic motor unit potentials, polyphasia (above) and normal interference pattern (below).

Comparison of three reported cases of myositis in ulcerative colitis

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<tr>
<td>Age at onset of myositis (y)</td>
<td>57</td>
<td>42</td>
<td>78</td>
</tr>
<tr>
<td>Sex (M/F)</td>
<td>F</td>
<td>M</td>
<td>F</td>
</tr>
<tr>
<td>Duration of colitis prior to onset of muscle weakness (y)</td>
<td>11</td>
<td>12</td>
<td>15</td>
</tr>
<tr>
<td>Drugs before onset of myositis</td>
<td>Steroids</td>
<td>Steroids and sulfasalazine</td>
<td>Absent</td>
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<tr>
<td>Splenomegaly</td>
<td>Massive, regressed after treatment of myositis</td>
<td>No steroids or sulfasalazine</td>
<td>Mild, regressed after treatment of myositis</td>
</tr>
<tr>
<td>Creatine phosphokinase activity</td>
<td>301 U/l</td>
<td>262 U/l</td>
<td>1010 U/l</td>
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<tr>
<td>Antinuclear factor</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Rheumatoid factor</td>
<td>+</td>
<td>Not mentioned</td>
<td>+</td>
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Plicating ulcerative colitis came from Oshitani et al, who described dermatomyositis and mononeuritis multiplex in ulcerative colitis and Basedow's disease. Four more cases, excluding ours, have subsequently been reported to date, of which only two are reported in English. 

Bhigee et al recorded the case of a 44 year old man who presented with ulcerative colitis of 12 years' duration and muscle weakness of 3 years. He underwent colectomy for his bowel disease but despite surgery, the leg weakness progressed resulting in wasting of the quadriceps muscles.

Kaneoka et al described polymyositis in a 63 year old woman who suffered from ulcerative colitis and developed liver cirrhosis secondary to post-transfusion hepatitis. After a relatively symptom free course of colitis for several years, she developed severe proximal muscle weakness and generalised atrophy. She was observed to have an enlarged spleen, which regressed after administration of steroids for polymyositis.

In both cases mentioned above and the one observed by us, myositis occurred after long standing ulcerative colitis (Table). Serum creatinine phosphokinase activity was raised and there was no evidence of vasculitis or granulomata on histology. In the absence of a history of sulfasalazine ingestion before the onset of myositis in two cases, one reported by Kaneoka et al and the second observed by us, the role of this drug in the pathogenesis of muscle disease is doubtful. In addition, it is evident from the case described by Bhigee et al that presence of active colitis is not essential for the progression of the muscle disease. The disease entity seems to affect both sexes and all ages since the condition has also been observed in a child. 

Regression of splenomegaly after steroid therapy in two patients (Table) and the presence of autoantibodies in the present patient as well as that described by Kaneoka et al suggest a possible autoimmune link between ulcerative colitis and polymyositis.

In the somewhat unrelated case of a 19 year old man with ulcerative colitis and necrotising cerebral vasculitis observed by Nelson et al, a biceps muscle biopsy specimen showed 'a few necrotic myofibres and mild perivascular inflammation'. Though not commented on by the authors, the presence of myositis with or without vasculitis remains a distinct possibility in their patient. Recognition of myositis as an extraintestinal manifestation of ulcerative colitis seems to be important for future documentation of this association.