Pneumatosis intestinalis in a patient with polymyositis

Sirs,

The patient is a 38-year-old patient with a 4-year history of polymyositis, with proximal weakness of the limbs, markedly increased serum levels of CPK, and typical electromyographic and muscular pathological findings. His serological tests disclosed positive fluorescent anti-nuclear antibodies, which were negative for anti-Jo1, RNP, and scleroderma 70 and anti-centromeres. At this time the patient responded to treatment with corticosteroids (prednisone 1 mg/kg) and high dose intravenous immunoglobulins (IVIG) (0.4 mg/kg for 5 days, once a month, for 12 months).

In January 1997, while being treated with low dose prednisone and azathioprine, he first complained of abdominal pain and distension, early satiety, constipation and a 3 kg weight loss. Gastroscopy revealed severe esophagitis and a very dilated stomach. Barium meal of the upper gastrointestinal (GI) tract confirmed a dilated stomach. Isotopic scanning of the esophagus and stomach with Technecium 99 phitate demonstrated very delayed emptying. Colonoscopy showed megacolon. The patient was treated with proton-pump inhibitors and cisapride and was recommended to eat small and multiple meals.

On March 1999, an exacerbation of his polymyositis was noted with recurrent proximal muscular weakness and an increase in serum CPK. The prednisone dosage was increased to 40 mg/d. During the following weeks, his muscular strength improved but he developed severe colicky abdominal pain, early satiety and constipation. He lost 10 kg in 5 weeks. On examination, he looked ill and in severe pain. The abdomen was very distended and tender on palpation.

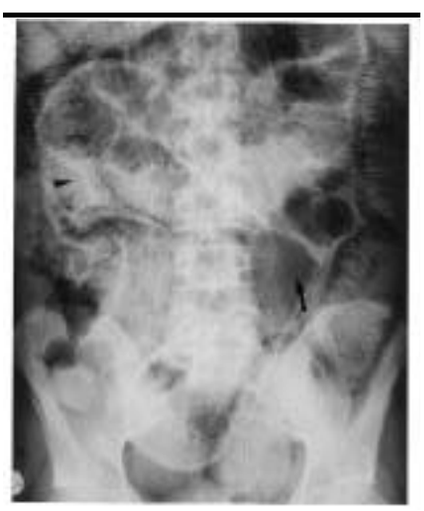

Fig. 1. Plain abdominal radiograph demonstrating pneumatosis intestinalis (thin arrow) and free abdominal air (thick arrow).

The plain abdominal film and CT scan (Fig. 1) performed at the same time showed free air in the abdominal cavity and within the wall of all visualized bowel loops of the small and large intestines. The patient was diagnosed as having pneumatosis intestinalis of the small and large intestines with free abdominal air. He underwent explorative laparotomy which confirmed the diagnosis of extensive pneumatosis intestinalis excluding a perforated viscera or ischemic bowel.

The patient was subsequently treated with high dose IVIG, total parenteral nutrition, metronidazole (1.5 gr/d) (after performing a Xylose test which disclosed significant bacterial overgrowth),daily injections of octreotide 50 g and cisapride (30 mg/d). After 10 days, a dramatic improvement was noticed with resolution of the abdominal pain and distension and weight gain. CT scan of the abdomen 2 months later revealed complete resolution of the free air and significant decrease in the pneumatosis.

Pneumatosis intestinalis is defined as subserosal or submucosal infiltration of the intestinal wall with multiple gaseous cysts. Several mechanisms for the appearance of gas in the mucosa have been proposed including breaks in the integrity of the gastric mucosa, ischemic bowel with subsequent bacterial overgrowth, and increased intraluminal pressure in the bowel as well as tracking down from leaking pulmonary emphysematous bullae (1). Pneumatosis intestinalis has been associated with several conditions such as duodenal ulcers, sigmoidoscopy, kidney and bone marrow transplantation - mainly attributed to corticosteroid therapy- ischemic bowel disease, chronic obstructive lung diseases and connective tissue disease (1). Pneumatosis intestinalis may be asymptomatic, but in most cases it leads to abdominal pain, distension, constipation and/or diarrhea (2). The diagnosis can be established on plain abdominal radiography or CT (2, 3).

Within the connective tissue diseases, systemic sclerosis is the most frequent cause of PI, where it is attributed to hypoxemia, hypomotility of the bowel with consequent bacterial overgrowth and increased intraluminal pressure (4). Pneumatosis has also been reported in systemic lupus erythematosus (5), mixed connective tissue disorders, polyarteritis nodosa, rheumatoid arthritis and temporal arteritis (2). In these cases, corticosteroid therapy as well as vasculitic involvement of the bowel has been suggested as a trigger of PI.

Pneumatosis intestinalis has been seldom described in poly/dermatomyositis (6-9). It seems to be more frequent in children (8, 9) and has been primarily attributed to ischemic vasculitic bowel involvement,

In our patient, the combination of corticosteroid therapy with a severe hypomotility disorder of the gastrointestinal tract probably induced the PI. Muscular involvement of the digestive system in PM is generally subclinical,although case reports of megaesophagus (10) and a study which demonstrated an impaired motility of the esophagus and stomach in 13 patients with PM (11) suggest that it may be more frequent than previously suspected.

The management of PI depends on the underlying disease. Treatment generally includes the use of nasogastric decompression, antibiotic therapy and induction of high concentration of oxygen by delivering oxygen by mask or using hyperbaric oxygen (1). If underlying GI hypomotility is present, treatment with cisapride or octreotide may further improve the clinical symptoms (1).

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