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Intestinal Pseudo-obstruction: Initial Manifestation of Systemic Lupus Erythematosus

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A 20-year-old man was hospitalized for 2 months after the onset of vomiting, cramping abdominal pain and dysuria. He had been diagnosed as IgA nephropathy 4 months ago. His mother was recently diagnosed as systemic lupus erythematosus (SLE). Physical examination revealed hypoactive bowel sounds and distended abdomen with diffuse tenderness. There was no rash on the face and body. The laboratory investigations showed hemoglobin 11.2 g/dL, leukocyte 10,500/μL, platelet 160,000/μL and albumin 3.2 g/dL. Antinuclear antibody (titer of 1:320 with speckled pattern), anti-Sm and ribonucleotide protein antibodies were positive. The 24 hour urine protein was 1,100 mg/day. Abdominal X-ray demonstrated multiple air-fluid levels of small bowel (Fig. 1). Abdominal CT showed fluid-filled dilatation of small bowel, bilateral hydronephrosis and thickened bladder wall. There was no abnormality on esophagogastroduodenoscopy and colonoscopy. Anorectal manometry showed decreased anal sphincter pressure and intact anaorectal inhibitory reflex.

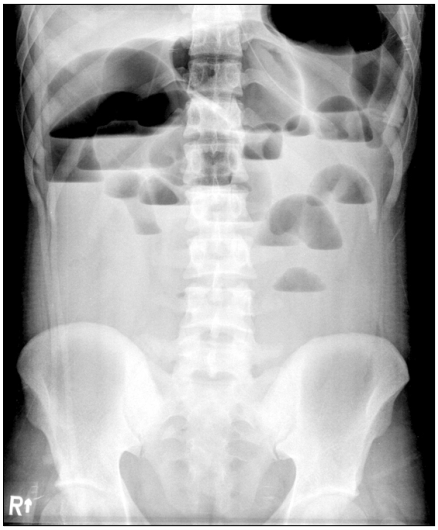

Figure 1. Simple abdominal erect view. Simple abdominal erect film shows extensive dilatation of the small bowel and multiple air-fluid levels.

He was treated with empirical prednisolone 30 mg/day for 3 days but abdominal pain was worsened and plain X-ray showed aggravation of ileus. To find out the underlying cause of intestinal pseudo-obstruction (IPO), subtotal colectomy was performed. Grossly there was no mass, ulcer or mechanical obstruction. Histopathology demonstrated diffuse muscular degeneration with little vasculitis suggestive of SLE (Fig. 2). Steroid and azathiopurine were started after surgery and he became well.

After IPO in SLE was first described in 1993,[1] few cases have been reported thereafter.[2-4] As in this case, IPO may appear as the initial presentation of SLE[2] and be associated with interstitial cystitis.[3-5] The pathophysiology of IPO in SLE is dysmotility of the intestinal muscle, caused by neuropathy, vasculitis or primary myopathy.[3-5] As IPO usually respond to high-dose ste-

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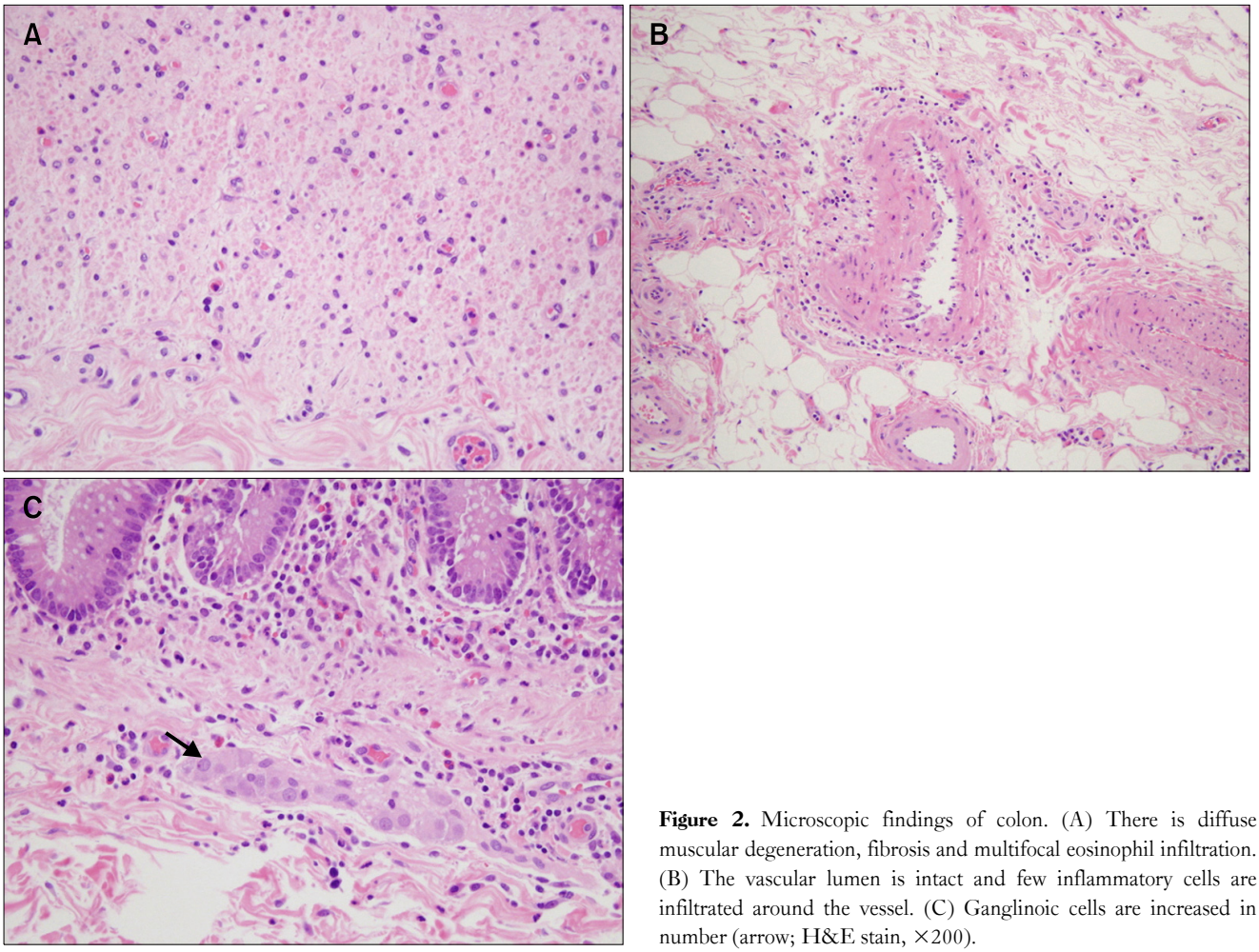

Figure 2. Microscopic findings of colon. (A) There is diffuse muscular degeneration, fibrosis and multifocal eosinophil infiltration. (B) The vascular lumen is intact and few inflammatory cells are infiltrated around the vessel. (C) Ganglinoic cells are increased in number (arrow; H&E stain, ×200).

roid and additional immunosuppressant, early diagnosis and timely initiation of treatment are important to avoid unnecessary surgery.[6]

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