Image Quiz

Multiple Colonic Submucosal Tumors in a Patient with Chronic Lung Allograft Dysfunction

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A 62-year-old man presented for a colonoscopy for positive fecal occult blood test. He had a past medical history of lung transplantation for idiopathic pulmonary fibrosis complicated by chronic lung allograft dysfunction in treatment with high dose of immunosuppressive drugs and steroids. The endoscopy revealed three polyps of the sigmoid colon (about 1 cm in size) and multiple submucosal lesions of varying sizes (1-3 cm) in the ascending and transverse colon, covered by normal mucosa [Figure 1a and b]. The polyps have been resected by snare-polypectomy, and histology showed a completely removed tubular adenoma with mild dysplasia. Biopsy specimens of submucosal tumors were inconclusive, revealing mild inflammation.

QUESTION

1. What is the diagnosis?

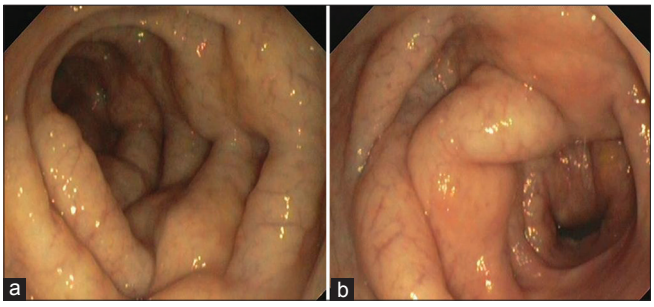

Figure 1: (a, b) Endoscopic view of multiple colonic submucosal tumors

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ANSWER

A computed tomography (CT) scan of the abdomen was performed that showed the presence of extensive gas collections within the gross bowel wall [Figure 2a and b]. Gastrointestinal perforation was excluded and no sign of pneumoperitoneum was found. On these findings, a diagnosis of pneumatosis cystoides intestinalis (PCI) was made. Because of total absence of symptoms, no treatment has been recommended.

PCI is an uncommon condition characterized by the presence of gas-filled cysts localized inside the intestinal wall.[1] Its pathogenesis is not completely understood. It is considered to result from primary mucosal damage due to varying causes, either benign (chronic pulmonary disease, infectious diseases, connective tissue diseases, graft vs. host disease, glucocorticoids, and immunosuppressive drugs) or life threatening (necrotizing enterocolitis, intestinal ischemia, sigmoid volvolus, mesenteric vascular disease).[1,2] However, most of the PCI are asymptomatic, incidentally discovered, with a benign course, and do not require any treatment. The diagnosis is generally suspected by endoscopy and confirmed by cross-sectional imaging, especially CT scan that can differentiate benign from severe conditions.[3] Sometimes histological examination of endoscopic biopsy specimens can confirm the diagnosis. The endoscopic differential diagnosis of more common diseases can be difficult. Treatment options include oxygen therapy and/or antibiotics. Surgical intervention is only required in rare cases of perforation and peritonitis.[1,2]

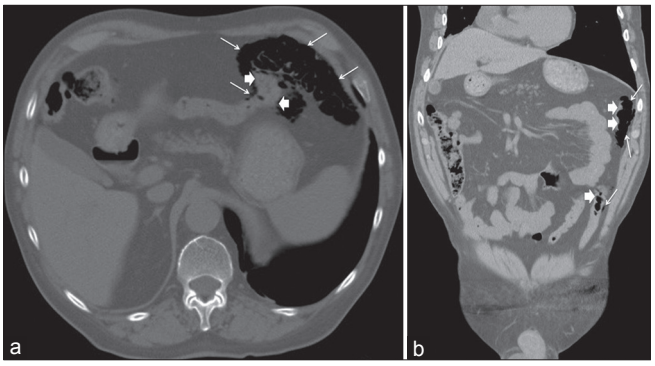

Figure 2: (a, b) Computed tomography scan demonstrating gross bowel loops (short arrows) with extensive intramural gas (long arrows)

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