

Fibrosing colonopathy in a man with cystic fibrosis on pancreatic enzyme supplements



Fig: Discrete colonic ulcers surrounded by normal mucosa

A 29-year-old man with cystic fibrosis, on pancreatic enzyme supplement for about two years, presented with abdominal pain and watery diarrhea for a week. His medications included ten capsules daily of *Pancrease MT-16* (each containing lipase 16,000 U, amylase 48,000 U, protease 48,000 U; Ortho-Mcneil, New Jersey, USA). Abdominal examination was unremarkable. Stool cultures were negative for common pathogens. Colonoscopy showed scattered, discrete, well-circumscribed ulcers throughout the colon, each 1 cm in size (Fig) and surrounded by normal colonic mucosa. Histology showed focal ulceration and acute inflammation at the border of the ulcers with prominent fibroblastic proliferation at the base. Acid-fast stain and Grocott methanamine silver stain for fungi were negative. Immunostains were negative for *Cytomegalovirus* and *Herpes Simplex virus*. The patient improved with supportive care.

Fibrosing colonopathy occurs in patients with cystic fibrosis taking pancreatic enzyme supplements. Patients

may present with abdominal pain, diarrhea, hematochezia or refractory distal intestinal obstruction syndrome. Radiologic studies may show strictures in the colon and small bowel. Numerous hypotheses for the mechanism of injury have been postulated, including direct toxicity of the enteric coating of the supplements,^{1,2} colonic obstruction and intramural enzymatic digestion, immune-mediated injury, and predisposition of the large intestine due to increased permeability.

We believe that the ulcers in our patient were due to a direct effect of the capsules. It is possible that the enteric-coated capsules did not dissolve in the small intestine and because of slow transit adhered to the colonic mucosa, resulting in direct injury. It is not possible to say whether the injury was secondary to the Eudragit® coating (metha-acrylic acid copolymer) or to the enzymes themselves. This acute injury has the potential to lead to fibrosing colonopathy and stricture due to enzymatic effect on deeper layers.

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