Case: A 69-year-old man presents with a long history of abdominal pain and small bowel obstruction. Surgery was performed, which showed a torsed, dilated segment of jejunum. Gross and microscopic images are below.



Figure 1. Gross image



Figure 2. Gross image



Figure 3. H&E, (20x)



Figure 4. H&E, (40x)



Figure 5. Trichrome, histochemical stain (40x)



Figure 6. Smooth muscle actin, immunohistochemical stain (40x)



Figure 7. C-kit, immunohistochemical stain (40x)

What is your diagnosis?

- A. Chronic intestinal pseudo-obstruction
- B. Crohn's disease
- C. Diaphragm disease
- D. Fibrosing enteropathy
- E. Scleroderma

SCROLL DOWN FOR ANSWER AND DISCUSSION ...

Answer and discussion:

A. Chronic intestinal pseudo-obstruction

Chronic intestinal pseudo-obstruction (CIPO) is an unusual process whereby the intestine is obstructed for non-mechanical reasons. Normal peristalsis relies upon the smooth muscle cells and the interstitial cells of Cajal (ICC) in the bowel wall to function properly. Patients may develop dysfunction in this system due to myopathy (such as familial visceral myopathy), neuropathy (such as aganglionosis), or disorders affecting the ICC.

Myopathic forms of CIPO demonstrate fibrosis of the muscularis propria (as seen on the above trichrome special stain). This finding is sometimes present in cases caused by ICC abnormalities, which also demonstrate loss of ICC (as seen on the above c-kit immunohistochemical stain). In severe cases, this leads to dilation of the bowel, which can take the form of sacculations (as seen in the above gross photographs), which are also known as pseudodiverticula.

None of the other conditions listed cause loss of interstitial cells of Cajal. Crohn's disease can cause sacculations of the bowel wall, but this specimen does not show evidence of inflammatory bowel disease. Scleroderma can cause both sacculations and fibrosis of the bowel wall; the fibrosis is usually marked, rather than patchy, and entire layers of the muscularis propria can be obliterated.

Diaphragm disease is a rare complication of NSAID use wherein "diaphragms" form along the bowel wall. These thin membranes are composed of mucosa and submucosa and obstruct the lumen. This is the reverse of the current specimen, which features outward sacculations rather than inward constrictions.

Fibrosing enteropathy/colonopathy is a rare process typically encountered in the setting of cystic fibrosis. As a side effect of enzyme-replacement therapy, these patients develop fusiform strictures along the bowel. The right colon is most commonly involved, and the distal ileum may be affected as well. The fibrosis is primarily submucosal, and the mucosa demonstrates increased chronic inflammation. Sacculation is not a feature of this disease.

References:

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