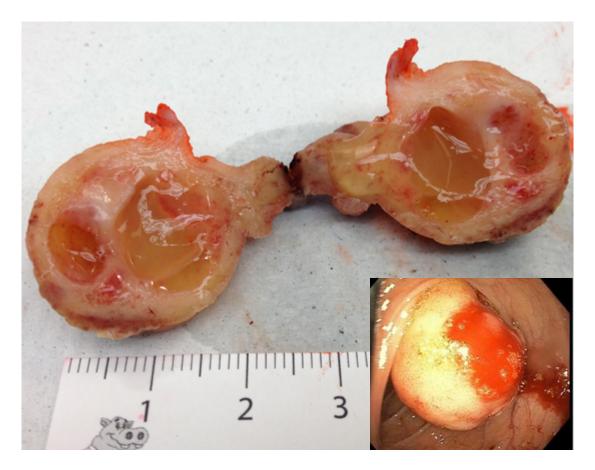
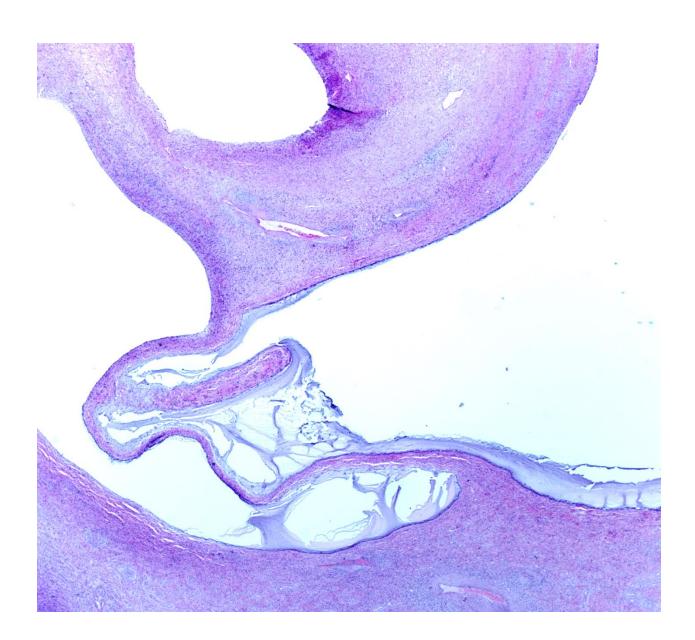
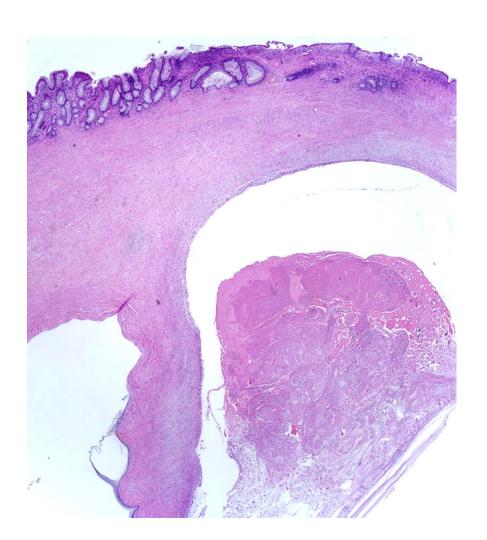
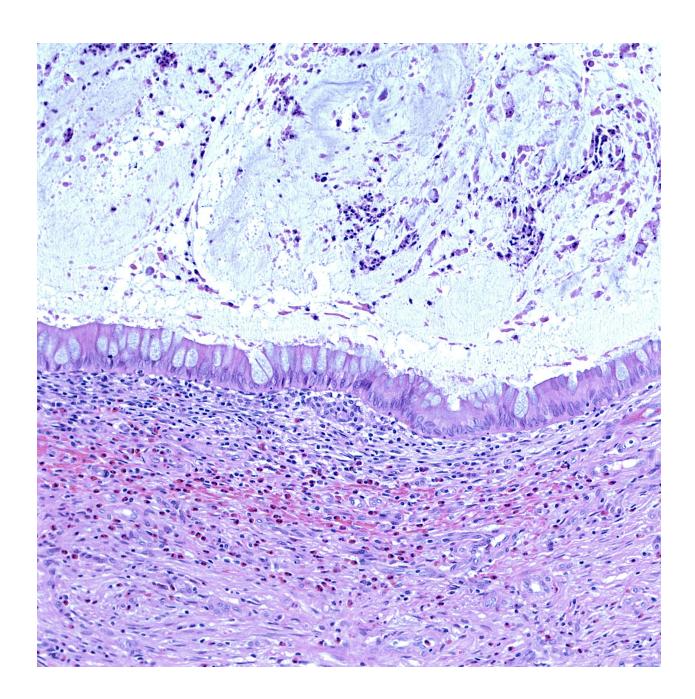
Case: A 69 year-old man with a history of gastrointestinal polyps presents with occult blood in the stool and iron deficiency anemia. He reports no weight loss, melena or hematochezia. Colonoscopy shows a 3.5 cm, pedunculated polyp in the splenic flexure oozing blood. Gross and microscopic pictures of the polyp are shown below.

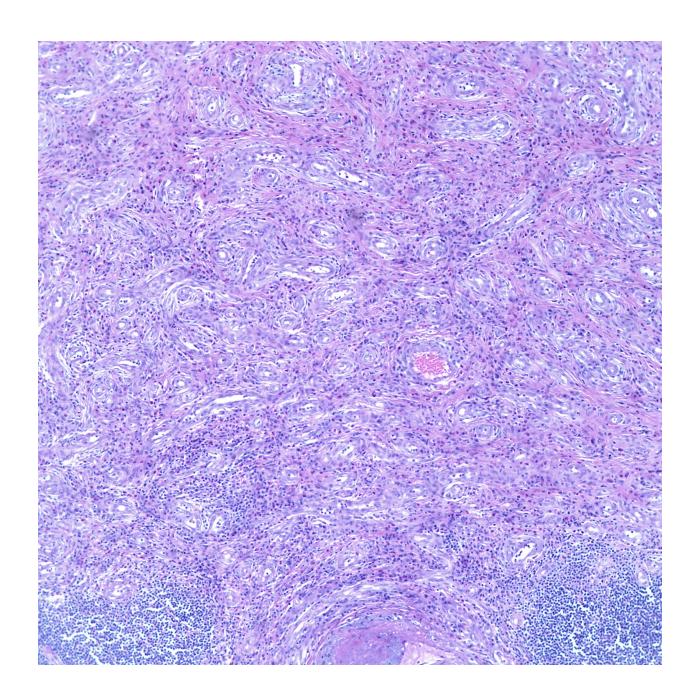


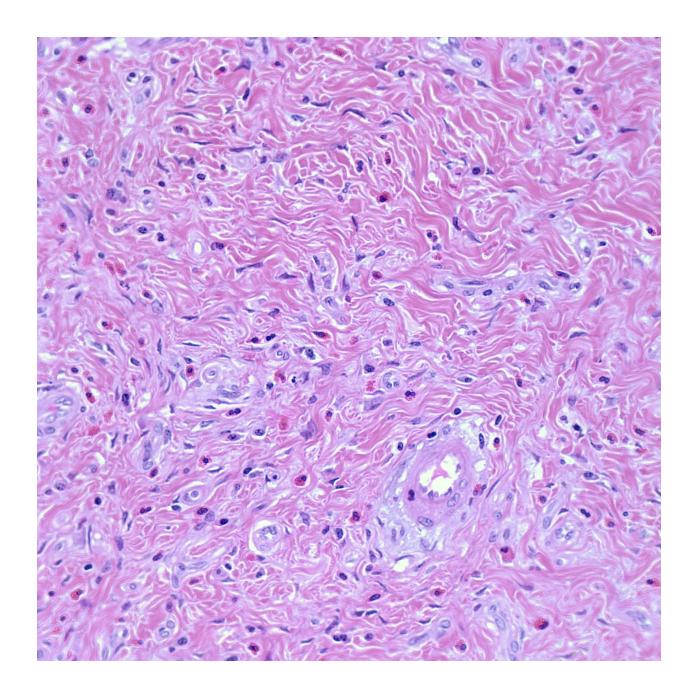
**Figure 1. Polypectomy specimen.** Polyp bisected showing cystic spaces filled with mucinous material. **Inset: Colonoscopy.** Hemorrhagic polyp in splenic flexure.











# What is your diagnosis?

- A. Colitis cystica profunda
- B. Isolated juvenile polyp
- C. Inflammatory fibroid polyp containing cystically dilated glands
- D. Inflammatory myoglandular polyp
- E. Mucinous adenocarcinoma

#### Answer and Discussion:

C. Inflammatory fibroid polyp containing cystically dilated glands

This case is that of an inflammatory fibroid polyp (IFP) containing cystically dilated glands. The gross photo shows a polypoid lesion containing cystic spaces with mucinous material. Microscopic examination shows a submucosal lesion composed of cystically dilated glands filled with mucin in a background of fibrovascular proliferation with abundant eosinophils and focal sclerotic changes. The glandular epithelium showed no atypia.

IFP is a rare benign lesion of unknown etiology which usually forms a solitary mass in the luminal gastrointestinal tract. IFP was first described by Vanek in 1949, as "submucosal granuloma with eosinophilic infiltration". Only 4% of the at least 1,000 cases of IFP reported in the literature have been described in the colon and rectum. The histological and immunohistochemical characteristics of IFP have been studied mostly in gastric polyps 4,6,11,12,13 as 70% of the reported IFPs have been found in the stomach.

Most colonic polyps are pedunculated (68%) and tend to occur proximally.<sup>3</sup> Although all age groups are affected, IFP typically presents in the 5<sup>th</sup> to 7<sup>th</sup> decade of life.<sup>2</sup> Clinical presentation varies depending on anatomic site. Abdominal pain and hematochezia are the most common symptoms of colonic IFPs.<sup>3</sup>

The etiology of IFPs is enigmatic. Proposed mechanisms range from an inflammatory process in response to injury to a hamartomatous or neoplastic process.<sup>1,6</sup> A study by Lasota et al. showed consistent expression and common mutational activation of PDGFRA in small intestinal IFPs, which had also been found in their gastric counterparts, suggesting that these lesions should be considered PDGFRA-driven benign neoplasms.<sup>1</sup>

Microscopic examination of IFP reveals numerous small vessels and fibroblasts in association with an inflammatory infiltrate consisting predominantly of eosinophils. The fibroblasts are typically arranged concentrically around the small vessels in an "onion-skin"-like fashion.<sup>4</sup> At least four distinct histologic patterns have been described, namely classical fibrovascular, nodular, sclerotic and edematous.<sup>4</sup>

This case is unique in that it presented as a polyp in the splenic flexure of the colon and showed cystically dilated glands containing mucin, a pattern which has not been described in the literature thus far, to the best of our knowledge.

A colonic IFP with cystically-dilated glands must be differentiated from other entities presenting with mucin-containing, cystic spaces such as colitis cystica profunda (CCP), isolated juvenile polyp (IJP), inflammatory myoglandular polyp (IMGP), and, most importantly, mucinous adenocarcinoma. CCP, which consists of mucin-filled cysts beneath the muscularis mucosa, can also present as a polypoid lesion. <sup>14,15</sup> IJP is the most common gastrointestinal polyp in children, but it is not an uncommon entity in adults. <sup>16</sup> IMGP is a colorectal polyp displaying cystically dilated glands which can look grossly like IFP. <sup>17,18</sup> However, the exuberant fibrovascular proliferation with predominantly eosinophilic inflammatory infiltrate, which characterizes IFP is not seen in these entities. In mucinous adenocarcinoma, the pools of mucin dissect through the stroma and are partially lined or filled by malignant epithelium. <sup>19</sup>

IFPs may be removed endoscopically. Recurrence of these benign entities is rare.<sup>3</sup>

Choice A is incorrect: Colitis cystica profunda consists of mucin-filled cysts beneath the muscularis mucosa and can also present as a polypoid lesion; however, there is no associated fibrovascular proliferation or eosinophilic inflammatory infiltrate.

Choice B is incorrect: Isolated juvenile polyp is the most common gastrointestinal polyp in children, but it is not an uncommon entity in adults. Microscopically, there is granulation tissue and ulceration covering abundant cystically dilated glands filled with mucus in an edematous and inflamed stroma.

Choice D is incorrect: Inflammatory myoglandular polyp is a colorectal polyp displaying cystically dilated glands which can look grossly like an inflammatory fibroid polyp; however, the exuberant fibrovascular proliferation with predominantly eosinophilic inflammatory infiltrate, which characterizes inflammatory fibroid polyps, is not seen in these entities. Inflammatory granulation tissue in the lamina propria mucosae, and proliferation of smooth muscle are also seen in inflammatory myoglandular polyps.

Choice E is incorrect: In mucinous adenocarcinoma, the pools of mucin dissect through the stroma and are partially lined or filled by malignant epithelium.

#### References

- 1. Lasota, J., Wang, Z. F., Sobin, L. H., & Miettinen, M. (2009). Gain-of-function PDGFRA mutations, earlier reported in gastrointestinal stromal tumors, are common in small intestinal inflammatory fibroid polyps. A study of 60 cases. *Modern Pathology*, 22(8), 1049-1056.
- 2. Wysocki, A. P., Taylor, G., & Windsor, J. A. (2007). Inflammatory fibroid polyps of the duodenum: a review of the literature. *Digestive surgery*, *24*(3), 162-168.
- 3. Ihimoyan, A., Chelimilla, H., & Balar, B. (2012). An inflammatory fibroid polyp in the hepatic flexure of the colon treated with argon plasma coagulation, endoscopic clipping and polypectomy. *Case reports in gastroenterology*, *6*(1), 63-68.
- 4. Kim, Y. I., & Kim, W. H. (1988). Inflammatory fibroid polyps of gastrointestinal tract. Evolution of histologic patterns. *American journal of clinical pathology*, 89(6), 721-727.
- 5. Vaněk, J. (1949). Gastric submucosal granuloma with eosinophilic infiltration. *The American journal of pathology*, 25(3), 397.
- 6. Johnstone, J. M., & Morson, B. C. (1978). Inflammatory fibroid polyp of the gastrointesthal tract. *Histopathology*, *2*(5), 349-361.
- 7. Helwig EB, Ranier A. Inflammatory fibroid polyps of the stomach. *Surg Gynecol Obstet* 1953;96:335–367.
- 8. Nakamura, S. I., Kino, I., & Akagi, T. (1992). Inflammatory Myoglandular Polyps of the Colon and Rectum A Clinicopathological Study of 32 Pedunculated Polyps, Distinct From Other Types of Polyps. *The American journal of surgical pathology*, 16(8), 772-779.
- 9. Park, Y. B., Cheung, D. Y., Kim, J. I., Park, S. H., Cho, S. H., Han, J. Y., ... & Choi, K. Y. (2006). A large inflammatory fibroid polyp in the sigmoid colon treated by endoscopic resection. *Internal medicine* (*Tokyo, Japan*), 46(19), 1647-1649.
- 10. Kim, B. C., Cheon, J. H., Lee, S. K., Kim, T. I., Kim, H., & Kim, W. H. (2008). Needle knife-assisted endoscopic polypectomy for a large inflammatory fibroid colon polyp by making its stalk into an omega shape using an endoloop. *Yonsei medical journal*, 49(4), 680-686.
- 11. Trillo, A. A., & Rowden, G. (1991). The histogenesis of inflammatory fibroid polyps of the gastrointestinal tract. *Histopathology*, *19*(5), 431-436.
- 12. Kolodziejczyk, P., Yao, T., & Tsuneyoshi, M. (1993). Inflammatory fibroid polyp of the stomach: a special reference to an immunohistochemical profile of 42 cases. *The American journal of surgical pathology*, *17*(11), 1159-1168.

- 13. Matsushita, M., Hajiro, K., Okazaki, K., & Takakuwa, H. (1997). Gastric inflammatory fibroid polyps: endoscopic ultrasonographic analysis in comparison with the histology. *Gastrointestinal endoscopy*, *46*(1), 53-57.
- 14. Herman, A. H., & Nabseth, D. C. (1973). Colitis cystica profunda: localized, segmental, and diffuse. *Archives of Surgery*, 106(3), 337.
- 15. Jung, M. E., Cho, G. M., Byun, S. W., Do, K. R., Kim, H. K., Cho, Y. S., ... & Kim, S. S. (2010). Colitis cystica profunda presenting with a mucus pool within the stalk of a pedunculated colon polyp. *Endoscopy*, 42(S 02), E114-E115.
- 16. Odze, R. D., & Goldblum, J. R. (Eds.). (2009). *Surgical pathology of the GI tract, liver, biliary tract, and pancreas*. Elsevier Health Sciences.
- 17. Hirasaki, S., Okuda, M., Kudo, K., Suzuki, S., & Shirakawa, A. (2008). Inflammatory myoglandular polyp causing hematochezia. *World journal of gastroenterology: WJG*, *14*(34), 5353.
- 18. Meniconi, R. L., Caronna, R., Benedetti, M., Fanello, G., Ciardi, A., Schiratti, M., ... & Chirletti, P. (2010). Inflammatory myoglandular polyp of the cecum: case report and review of literature. *BMC gastroenterology*, 10(1), 10.
- 19. Bosman, F. T., Carneiro, F., Hruban, R. H., & Theise, N. D. (2010). *WHO classification of tumours of the digestive system* (No. Ed. 4). World Health Organization.

### **Authors:**

Diana M. Morlote, M.D., 1,2 diana.morlote@gmail.com

Brett Neustater M.D., bneustater@gastrohealth.com

Antonio E. Martinez M.D.<sup>2,3</sup> anmartinez@gastrohealth.com

## **Corresponding author:**

Diana M. Morlote, M.D.
Arkadi M. Rywlin, M.D. Department of Pathology
Mount Sinai Medical Center
4300 Alton Road
Miami Beach, Florida 33140
P: 305-674-2277

F: 305-674-2999

<sup>&</sup>lt;sup>1</sup>Arkadi M. Rywlin, M.D. Department of Pathology, Mount Sinai Medical Center. 4300 Alton Road, Miami Beach, Florida 33154

<sup>&</sup>lt;sup>2</sup>Florida International University Herbert Wertheim College of Medicine. 1240 S.W. 108 AVE, Path, University Park, Miami, FL 33174

<sup>&</sup>lt;sup>3</sup> Gastro Health. 9500 South Dadeland Blvd., Suite 802, Miami, FL 33156

e-mail: diana.morlote@msmc.com

The authors declare that they have no significant relationships with, or financial interest in, any commercial companies pertaining to this article.