

Nodular Regenerative Hyperplasia-Induced Peristomal Variceal Bleeding: A Rare Thiopurine Side Effect

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Introduction

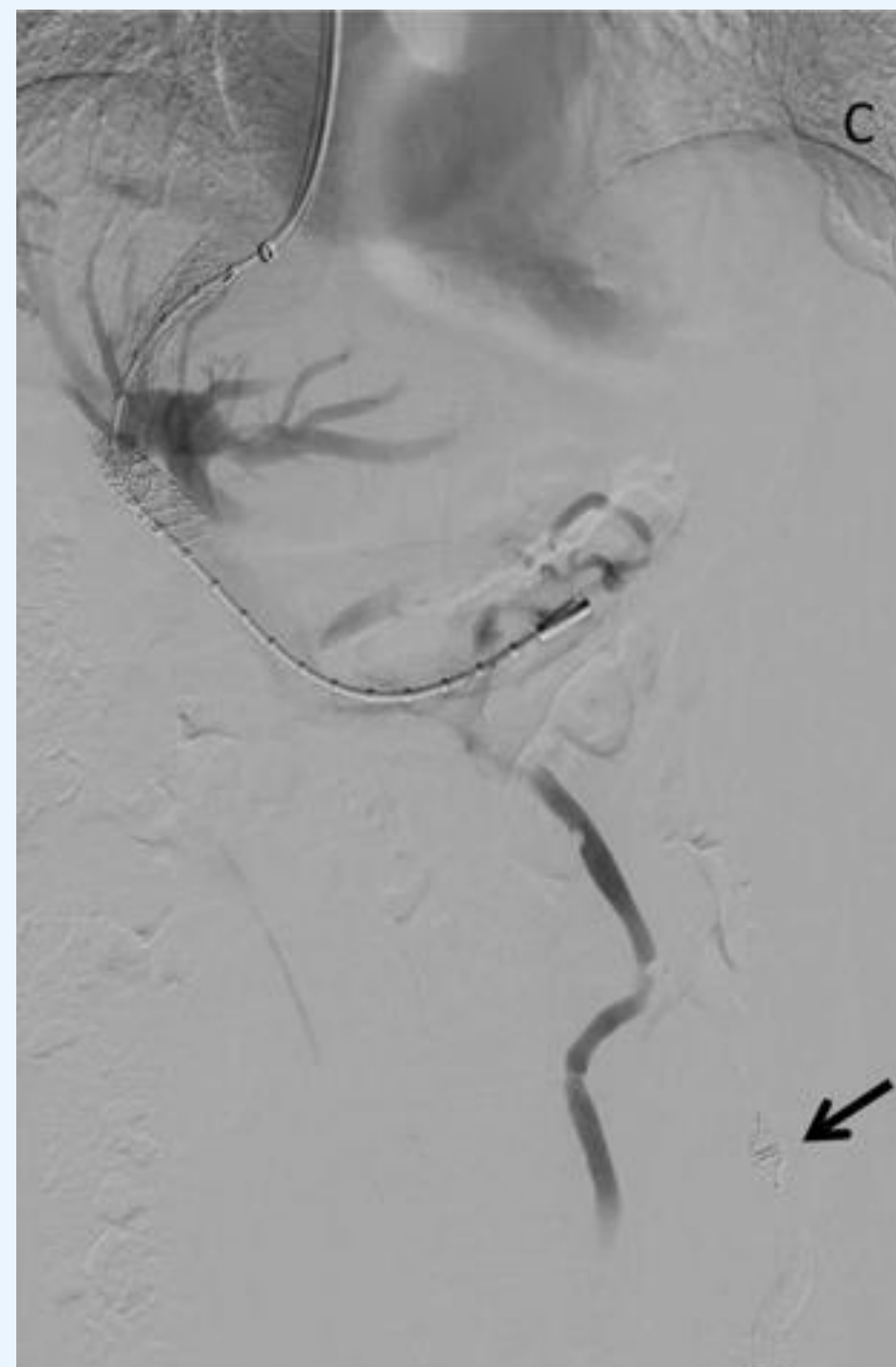
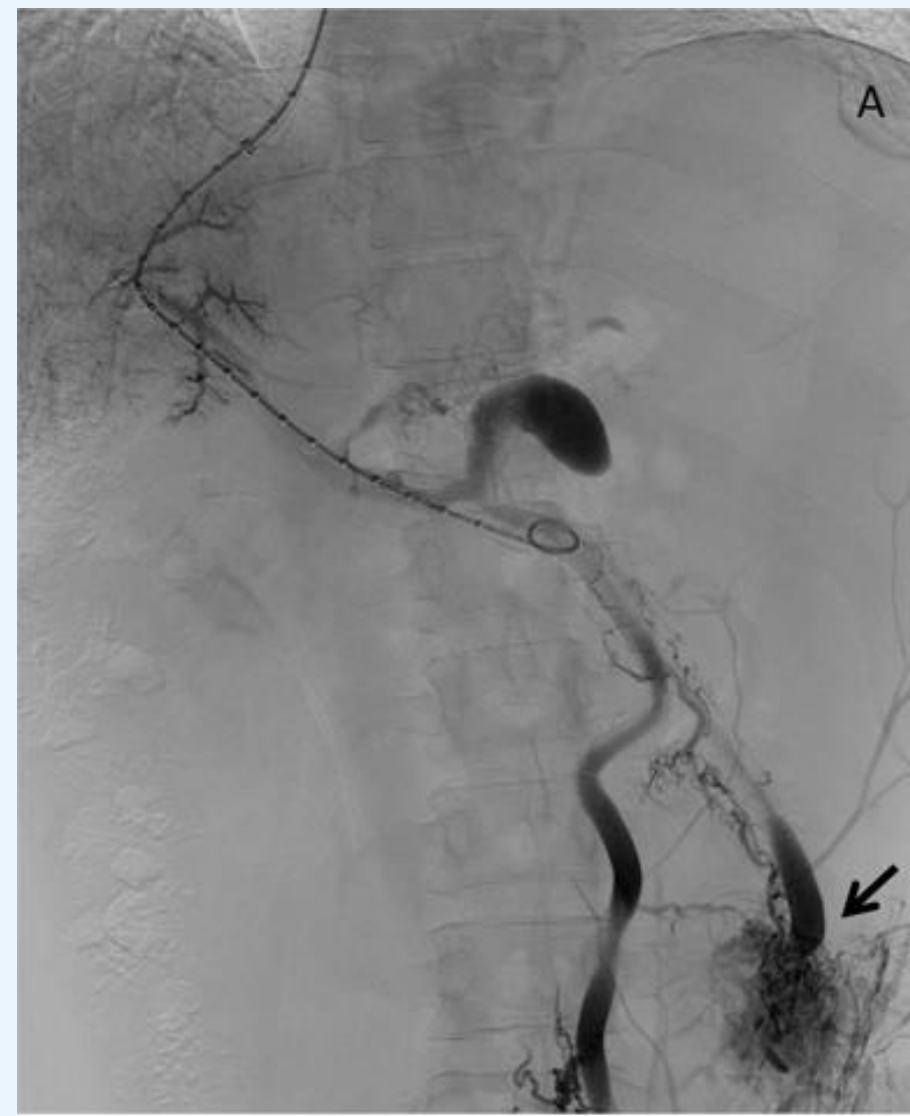
Azathioprine (AZA) and 6-Mercaptopurine (6-MP) are agents used to treat Inflammatory Bowel Disease (IBD). One of the rare side-effects described in cases throughout the literature is the development of Nodular Regenerative Hyperplasia (NRH) of the liver. NRH can lead to the development of non-cirrhotic portal hypertension (NCPH) which can then progress to ascites and varices. We present a patient who developed NRH from long-standing 6MP leading to peristomal variceal bleeding.

Case

A 12-year-old male with fistulizing and stricturing Crohn's disease, was being treated with 6-MP monotherapy from ages 14-18 and then restarted at age 27. At age 29, he underwent a partial colectomy with colostomy creation. Several weeks after his colectomy, he began to see blood in his ostomy bag. The source of this blood was determined to be from peristomal varices.

Abdominal CT revealed lower esophageal varices and a nodular appearing liver. He underwent a liver biopsy which revealed NRH. Given his peristomal varices he was diagnosed with NCPH. The patient underwent a successful TIPS procedure with resolution of the varices and has not had any further bleeding.

Images



A – Digital subtraction angiographic (DSA) image shows catheter-directed venography of a dilated branch of the inferior mesenteric vein (IMV) that supplies a network of peristomal varices (black arrow).

B - After successful creation of a transjugular intrahepatic portosystemic shunt (TIPS) with a covered stent (white arrow), a DSA image shows a plug and foam (black arrow) in the dilated IMV branch with decreased blood flow into the peristomal varices.

C- Final DSA image of completion IMV venography shows absent blood flow into the dilated IMV branch and peristomal varices.

Discussion/Conclusion

While variceal bleeding due to NCPH from NRH caused by thiopurines has been documented in the literature, a case of peristomal variceal bleeding from thiopurines has yet to be described. NRH is a poorly understood condition that is characterized by diffuse transformation of normal liver parenchyma into small regenerative nodules with little to no fibrosis. Increased liver nodularity can cause NCPH and subsequently formation of varices throughout the GI tract.

Patients with IBD on thiopurines have a cumulative incidence of NRH of 0.6% and 1.28% at 5 and 10 years, respectively¹. NRH has a variable clinical presentation- from asymptomatic to severe complications of NCPH--and patients often do not present until late in the disease course. Several risk factors associated with development of NRH are male sex, stricturing and/or fistulizing disease, and history of a bowel resection. While NRH and its pursuing complications from thiopurines remains a rare entity, clinicians should be cognizant of this potential complication--particularly in those with fistulizing/stricturing disease and/or a surgical resection with an ostomy who present with gastrointestinal bleeding.

References:

1. Musumba CO. The Association between Nodular Regenerative Hyperplasia, Inflammatory bowel disease and Thiopurine therapy. *Aliment Pharmacol Ther.* 2013; 1025-1037.



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