sigmoid colectomy for diverticular disease, presented to the Emergency Department with symptomatic anemia in the setting of maroon stools, found to have a hemoglobin level of 6.2 g/dL. Following resuscitation, the patient underwent esophagogastroduodenoscopy (EGD) and colonoscopy, which failed to identify a culprit lesion. In the ensuing days, the patient continued to have maroon stools and daily blood transfusion requirements. Further evaluation with capsule endoscopy and double-balloon enteroscopy were inconclusive. Tagged RBC scan and CT angiography were likewise negative, though CT angiogram did note prominent venous collaterals associated with the IMV and LGV, concerning for ectopic varices (Figure 1). An interdisciplinary meeting between Gastroenterology, Hepatology, Hepatobiliary Surgery, and Interventional Radiology was held to discuss potential options. Transjugular intrahepatic portosystemic shunt was contraindicated due to the degree of liver decompensation. Given the lack of other identifiable lesions, it was felt reasonable to attempt direct embolization of the varicocities between her IMV and LGV. Pre-embolization venogram did demonstrate a large shunt with extensive varices between the IMV and LGV (Figure 2). Amplatzer plugs and embolization coils were successfully deployed into 3 branches of the distal IMV (Figure 3). The patient was monitored in the hospital for a few days with stability of her hemoglobin, and she was discharged home. The patient followed up 1 month post-hospitalization with no recurrence of bleeding. Case Highlights: This case illustrates the difficulty that may be encountered in the identification and treatment of ectopic varices. In this case, the unusual location of her varices is postulated to have been related to her post-surgical state. A multi-disciplinary approach was needed in order to properly manage and treat this patient.
7.5 and lactate of 5. Her Hb, PTT and factor VIII levels improved with 2 units of packed red blood cells and recombinant factor VII. Her high-volume hematochezia persisted along with ongoing coagulopathy with transfusion requirements. The risks and benefits of proceeding with endoscopic evaluation were discussed and ultimately pursued. Colonoscopy revealed a large subepithelial hematoma near the hepatic flexure that involved two-thirds of the circumference of the wall and extended proximally at least 10 cm (Image), without evidence of active bleeding. Cross-sectional imaging ruled out transmural involvement or bowel necrosis. She was started on aminocaproic acid and rituximab, but remained in the hospital for over one month receiving recombinant factor VII for ongoing intermittent bleeding. Intestinal intramural hematomas are rare but account for approximately 5% of GI bleeds in patients with hemophilia. They can also be seen in patients on anticoagulation, with vasculitis, or following blunt trauma, and are thought to be due to rupture of a terminal artery arising from the mesentery which dissect the muscularis mucosa. In addition to GI bleed, intramural hematomas can present with mass-like or obstructive symptoms. It is important to keep intestinal intramural hematomas on the differential diagnosis in patients with the risk factors outlined above, and it is essential to be able to recognize them endoscopically and on cross-sectional imaging.

2010

A Rare Case of Bleeding Duodenal Arteriovenous Malformations in Association with Amyloid Enteropathy

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Amyloidosis is a rare disease caused by accumulation of abnormal amyloid fibrils within the extracellular matrix leading to structural and functional disruption of the involved tissue. Gastrointestinal (GI) involvement is seen in primary, secondary and dialysis related amyloidosis. AVM’s (arteriovenous malformations) in association with amyloidosis are a rare cause of GI bleed. We report an unusual case of duodenal AVM’s associated with gastrointestinal amyloidosis (AL), presenting as an acute upper GI bleed. An 82-year old Caucasian male with history of coronary artery disease and atrial fibrillation treated with anticoagulation, presented with a complaint of melena. Physical examination was significant for pallor. Laboratory investigation revealed iron deficiency anemia with a hemoglobin of 7.9 g/dL. Subsequent RGD showed thickened small bowel polypoidal mucosal folds with multiple AVM’s in the second and third portions of the duodenum, few of which were bleeding actively. APC (Argon Plasma Coagulation) therapy of the bleeding AVM’s was performed with resultant satisfactory hemostasis. Mucosal biopsies of the thickened duodenal folds were also performed. Histology revealed amyloidosis involving the muscularis mucosa and vessels, which displayed an apple green birefringence under polarized light microscopy using Congo Red stain (Figure 1).

Further laboratory investigation revealed a negative urine immunofixation test for monoclonal free and light chains. Mass spectroscopy revealed AL type amyloidosis, however the workup for plasma cell dyscrasias (multiple myeloma/Waldenstrom macroglobulinemia) was negative. GI tract involvement occurs more often with AA amyloidosis rather than with the AL type. Patients with amyloid enteropathy can present with GI bleeding, the etiology of which is usually secondary to ischemia, mucosal ulcers, and submucosal hematomas. Rarely AVM’s have been found to be associated with amyloidosis. Published data has shown such association as with gastric and pulmonary AVM’s. This case demonstrates a rare presentation of GI bleed in the setting of amyloid enteropathy secondary to duodenal AVM’s. This is the first reported case of bleeding duodenal AVM’s with amyloidosis. Hence it is our recommendation, that the presence of AVM’s with thickened or polypoidal small bowel folds should prompt the endoscopist to rule out amyloidosis.

2010 Figure 1. (a, arrow) Thickened polypoidal mucosal folds (b, arrow) Multiple AVM’s visualized in the duodenum (c, arrow) Actively bleeding AVM (d) Duodenal wall H&E stain showed homogenous eosinophilic deposits of muscularis mucosa and vessels, suggestive of amyloid enteropathy; (e, arrow). Positive Congo red staining of amyloid deposits in the vessel (f) Apple green birefringence (arrow) of Congo red stain under fluorescence polarized microscopy confirmed amyloid rich deposition.