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Learning Objectives

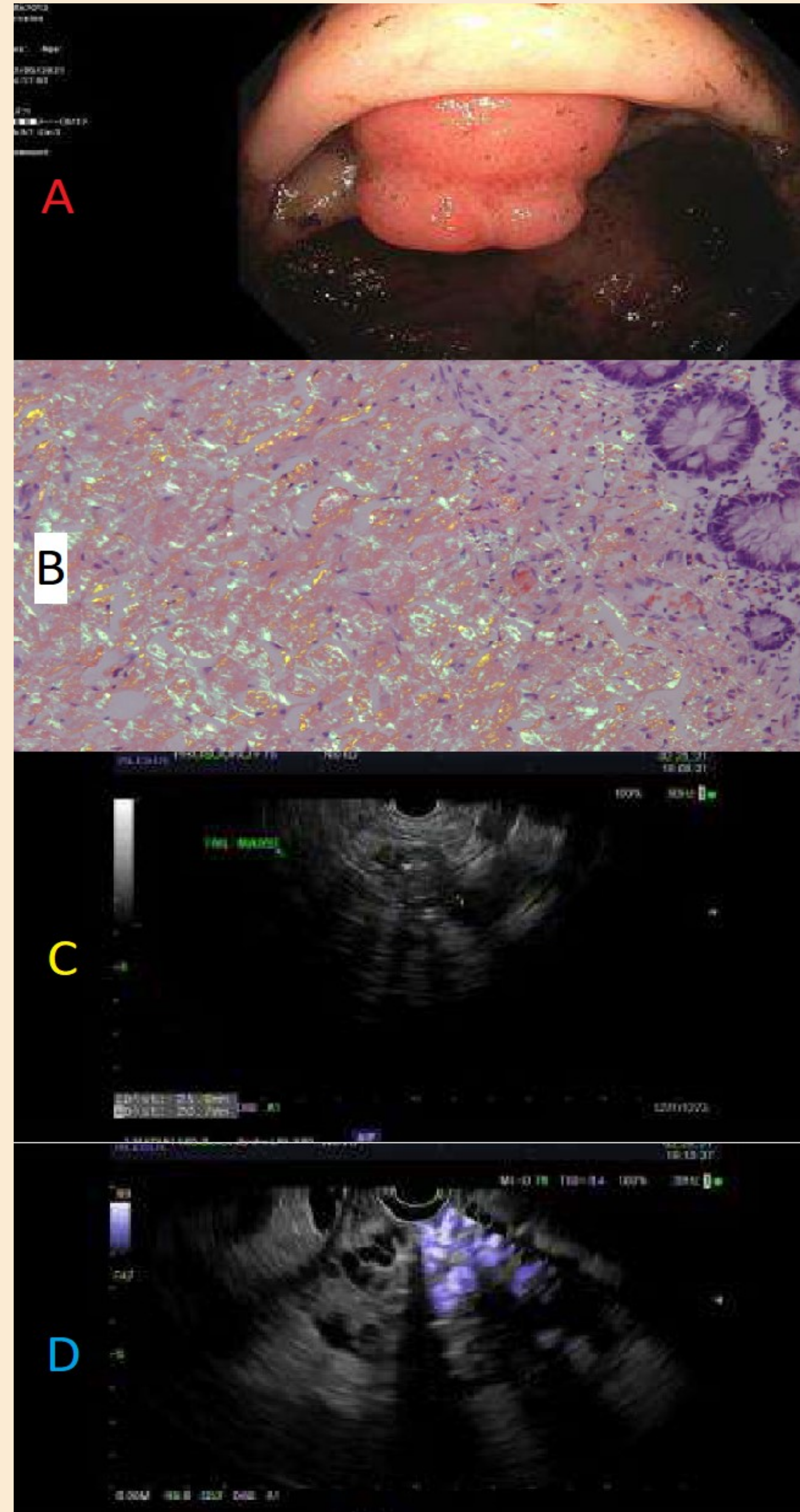
- Pancreatic Neuroendocrine tumors (pNETs) are endocrine tumors arising from the pancreas and are one of the most common NETs.
- Splenic vein thrombosis with resulting sinistral hypertension is a rare complication of pNETs causing isolated gastric varices and less commonly portal hypertensive colopathy if portal vein becomes involved.
- In this case, we present a 78-year-old male with intestinal amyloidosis complicated by pancreatic neuroendocrine tumor (NET), splenic vein occlusion with gastrosplenic shunt formation and gastrointestinal hemorrhage secondary to diffuse colonic angioectasias.

Case Background Information

- A 78-year-old male with history of ADPKD, CAD, and HTN was initially found to have colonic mass on screening colonoscopy. Biopsies at that time illustrated Congo red staining positivity for light chain (AL) amyloidosis. He was maintained on daratumumab in the outpatient setting and remained complication free for nearly two decades.
- Following a cardiac catheterization with stent placement, patient was started on dual antiplatelet therapy and had resulting hematochezia requiring admission. Colonoscopy showed friable mucosa with ulceration from rectum to cecum as well as ulcerated polypoid lesions.
- The following year patient was found to have pancreatic tail mass on CT and subsequently underwent EUS with FNA revealing pNET.
- Clinical course was further complicated by splenic vein thrombosis, large gastrosplenic shunt formation and type II isolated bleeding varices.
- He ultimately underwent coil embolization with IR and was started on lanreotide injections for pNET.

Physical Exam / Lab Values

Physical Exam				
General: NAD	141	107	53	INR: 1.3
Eyes: anicteric conjunctivae	4.5	14	1.8	AST: 11
CVS: RRR, normal S1/S2				ALT: 14
Resp: CTAB		10.3		TBili 0.7
GI: soft, NT	12.5		129	ALP 129
Rectal: +Melena		35		



Endoscopic and Pathology Findings

- A. Large amyloidoma extruding from terminal ileum.
- B. Cecum histopathology: Positive Congo red stain with apple-green birefringence under polarized light.
- C. EUS showing 25mm x 21mm hypoechoic lesion in pancreatic tail with few calcifications.
- D. Hypoechoic pancreatic tail lesion (pNET) with intervening fundal varices preventing biopsy.

Current Admission Hospital Course

- The patient now presented with melanic stools of two days duration.
- EGD redemonstrated nonbleeding type I isolated gastric varices and colonoscopy revealed numerous actively bleeding colonic angioectasias treated with APC ablation.
- Due to persistent hematochezia, CT venogram was performed which demonstrated pneumoperitoneum concerning for perforated viscus.
- He was seen by colorectal surgery, underwent total colectomy with ileostomy and was eventually discharged to rehab in stable condition.

Take Home Points

- Splenic vein thrombosis is a rare complication of pNETs which can cause isolated gastric varices and is recognized as an important cause of upper GI bleeding.
- This case highlights a rare complication of pNET whereby a splenic vein occlusion and large gastrosplenic shunt eventually involved the portal vein causing true right sided portal hypertension with development of portal hypertensive colopathy characterized by bleeding angioectasias.

References

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