



SAR·DFP

Society of Abdominal Radiology  
Disease-Focused Panels



**SAR·DFP**

Society of Abdominal Radiology  
Disease-Focused Panels

## **Gastrinomas in MEN type I**

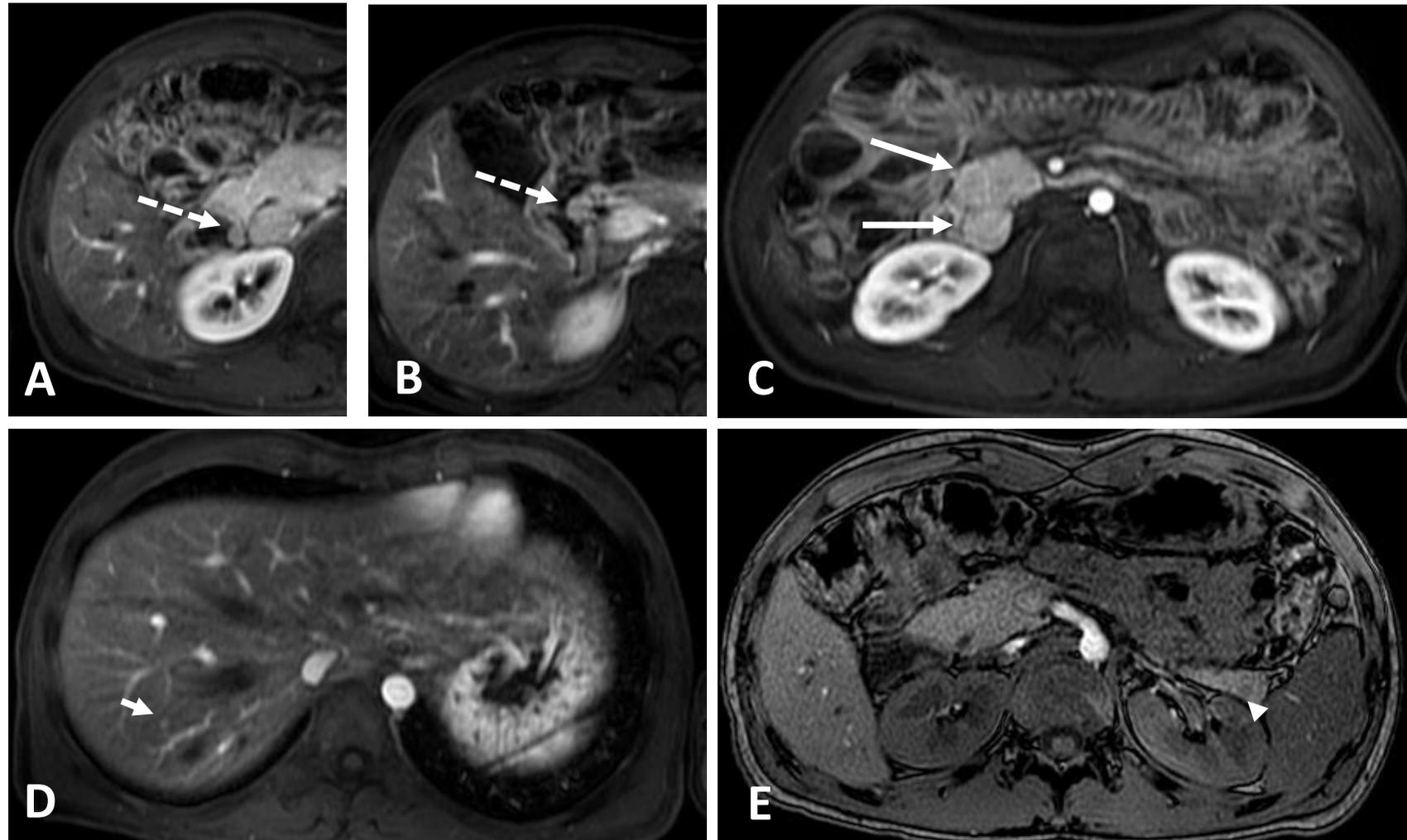
*H Dalla Pria, MD; UI Salem, MD; NA Wagner-Bartak, MD; AC  
Morani, MD*

# Case presentation

- An 18-year-old male presenting with chronic diarrhea. The patient was under surveillance due to a strong family history of multiple endocrine neoplasia type 1 (MEN-1).
- Initial abdomen MRI showed multiple enhancing polypoid lesions along the duodenal wall and subcentimeter enhancing pancreatic lesions.
- Labs: Gastrin > 20000pg/mL (Ref <100); Chromogranin A 3570 (Ref <93ng/mL)
- Clinical presentation, history and elevated serum gastrin levels were concerning for gastrinoma. <sup>68</sup>GA DOTATATE was requested for further evaluation.

### **Fig 1: MRI Abdomen**

Enhancing nodules along the duodenal wall (dashed arrows in A-B), concerning for gastrinomas in the setting of elevated serum gastrin. Enlarged lymph nodes, suspicious for nodal metastases (long arrows in C). Subcentimeter arterially enhancing liver nodule (short arrow in D), that becomes isointense on the delayed postcontrast images (Not shown). Subcentimeter T1 hypointense nodule in the pancreatic tail (arrowhead in E) with associated restricted diffusion, suspicious for pancreatic neuroendocrine tumor in the setting of MEN-1.

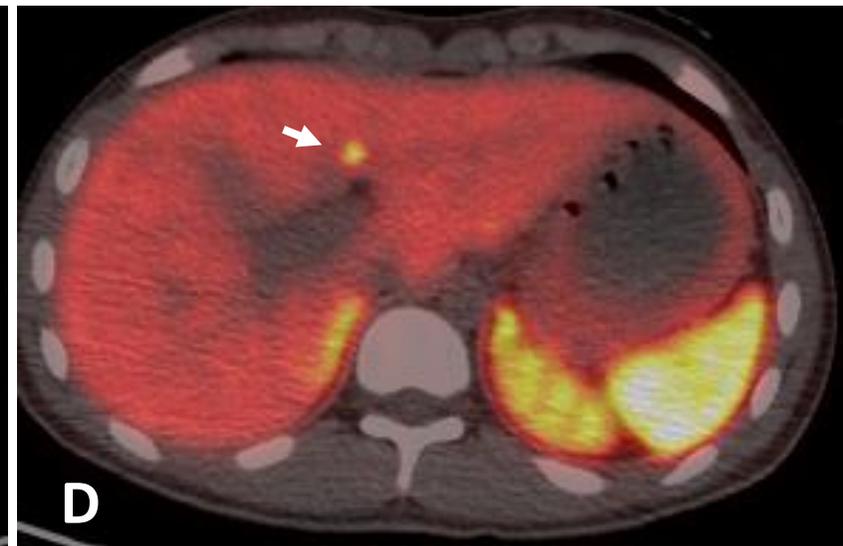
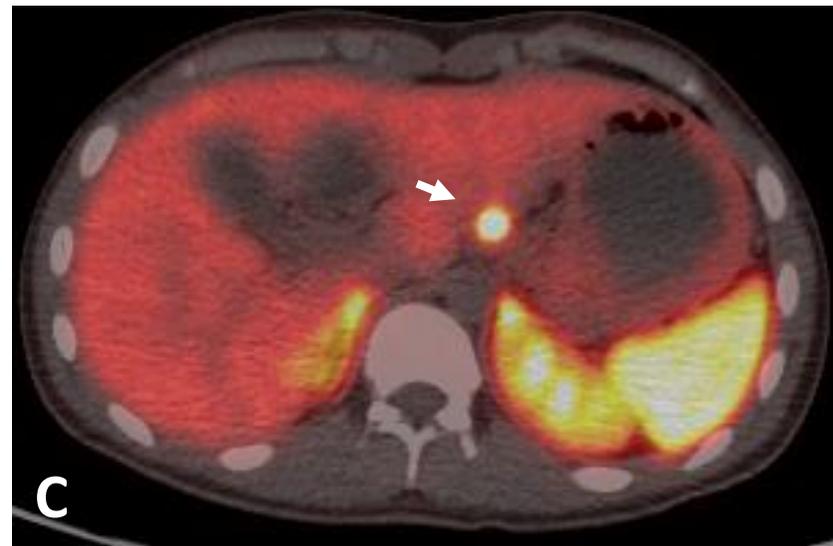
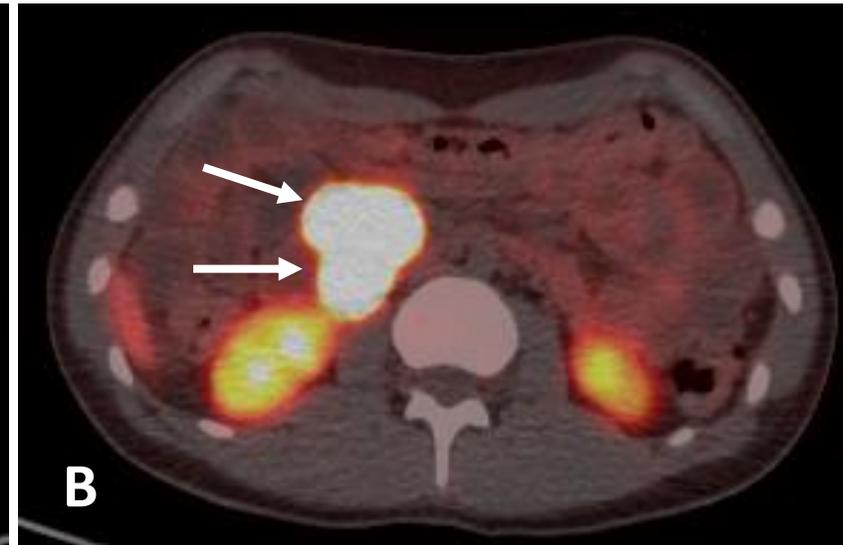


**Fig 2: <sup>68</sup>GA DOTATATE PET/CT:**

An avid focus of tracer uptake localizes to the second portion of the duodenum consistent with a primary gastrinoma (dashed arrow in A).

DOTATATE-avid lymphadenopathy is identified in the periduodenal region (long arrows in B).

Two foci of dotatate-avidity in the gastro-hepatic/cealic region and along the falciform ligament are consistent with small metastatic lymph nodes/implants (short arrows in C and D).



# Discussion

- Patient had a diagnosis of MEN-1, initially diagnosed through predictive genetic testing.
- MEN-1 is a rare inherited syndrome that affects the endocrine glands with tumors usually developing in the parathyroid glands, pituitary gland, and pancreas (and other parts of gastrointestinal tract).
- Although the patient was initially seen at the age of 6 due to a strong family history of MEN-1, he eventually was lost to follow-up. Recently, at the age of 17, the patient came back presenting with chronic diarrhea, concerning for MEN-1 gastrinoma, with imaging findings supporting the clinical diagnosis.

# Discussion

- Initial MRI showed enhancing nodules along the duodenal wall, and metastatic lymphadenopathy was confirmed by corresponding 68Ga Somatostatin analogue (DOTATATE)-avidity. EUS revealed multiple submucosal duodenal nodules (gastrinomas) and lymphadenopathy compatible with metastatic gastrinoma.
- Subcentimeter pancreatic nodules did not show DOTATATE-avidity nor EUS abnormality, but continued to show enhancement on follow-up CT. Pituitary adenoma/hyperprolactinemia were identified on MRI (not shown)
- Ga-DOTATATE PET/CT is very helpful for the work-up of MEN1 providing a comprehensive picture of MEN1-related lesions and prognostication in these cases

# References

- Rockall, A. G., & Reznick, R. H. (2007). Imaging of neuroendocrine tumours (CT/MR/US). *Best practice & research Clinical endocrinology & metabolism*, 21(1), 43-68.
- Sato, Y., Hashimoto, S., Mizuno, K. I., Takeuchi, M., & Terai, S. (2016). Management of gastric and duodenal neuroendocrine tumors. *World journal of gastroenterology*, 22(30), 6817.
- Ruszniewski, P., Amouyal, P., Amouyal, G., Grangé, J. D., Mignon, M., Bouché, O., & Bernades, P. (1995). Localization of gastrinomas by endoscopic ultrasonography in patients with Zollinger-Ellison syndrome. *Surgery*, 117(6), 629-635.
- Tiwari, R., Singh, A. K., Somwaru, A. S., Menias, C. O., Prasad, S. R., & Katabathina, V. S. (2019). Radiologist's primer on imaging of common hereditary cancer syndromes. *Radiographics*, 39(3), 759-778.
- Brandi, M. L., Gagel, R. F., Angeli, A., Bilezikian, J. P., Beck-Peccoz, P., Bordi, C., ... & Marx, S. J. (2001). Consensus: guidelines for diagnosis and therapy of MEN type 1 and type 2. *The Journal of Clinical Endocrinology & Metabolism*, 86(12), 5658-5671.
- Lastoria S, Marciello F, Faggiano A, Aloj L, Caracò C, Aurilio M, D'Ambrosio L, Di Gennaro F, Ramundo V, Camera L, De Luca L, Fonti R, Napolitano V, Colao A. Role of (68)Ga-DOTATATE PET/CT in patients with multiple endocrine neoplasia type 1 (MEN1). *Endocrine*. 2016 Jun;52(3):488-94. doi: 10.1007/s12020-015-0702-y. Epub 2015 Aug 5.