SAR·DFP

Society *of* Abdominal Radiology
Disease-Focused Panels

Gastroenteropancreatic & Genitourinary Neuroendocrine tumors

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Pancreatic NET

Presented By:

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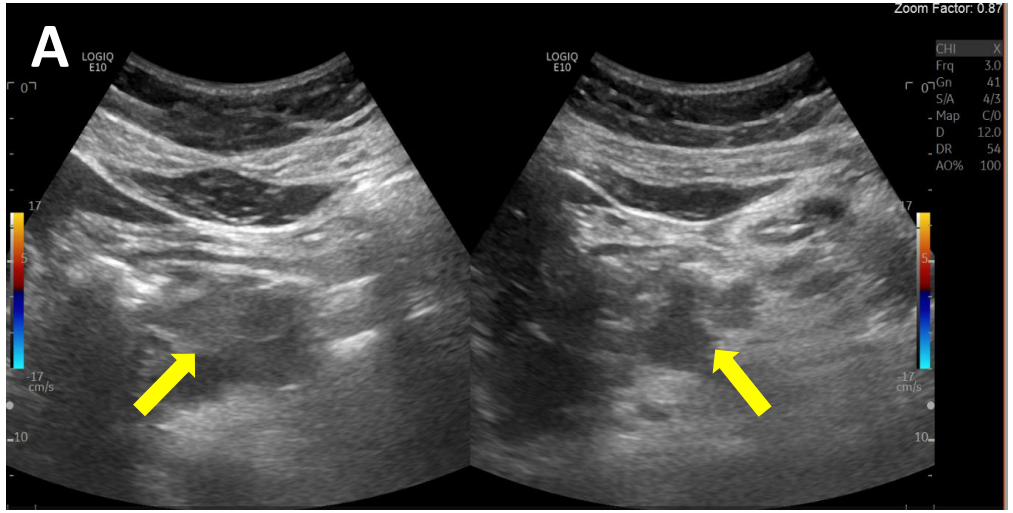
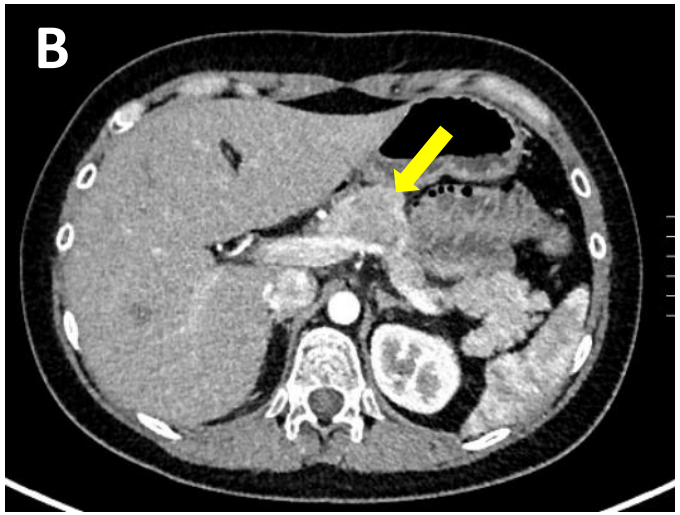
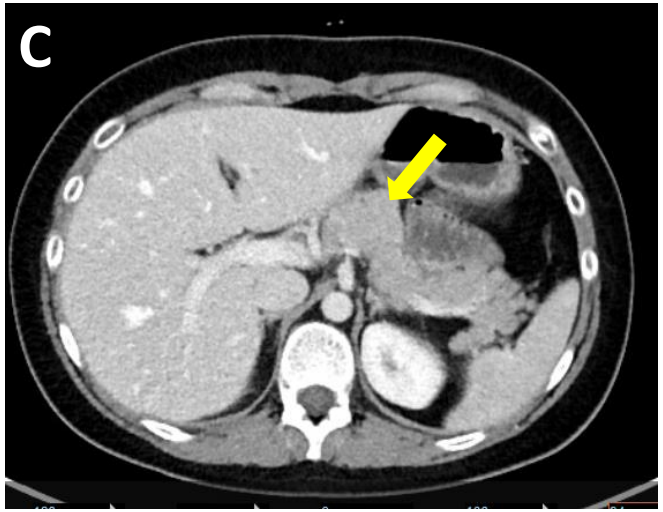
37 year-old woman with PMH of FAP syndrome presented to the ED with upper abdominal pain and mildly elevated lipase



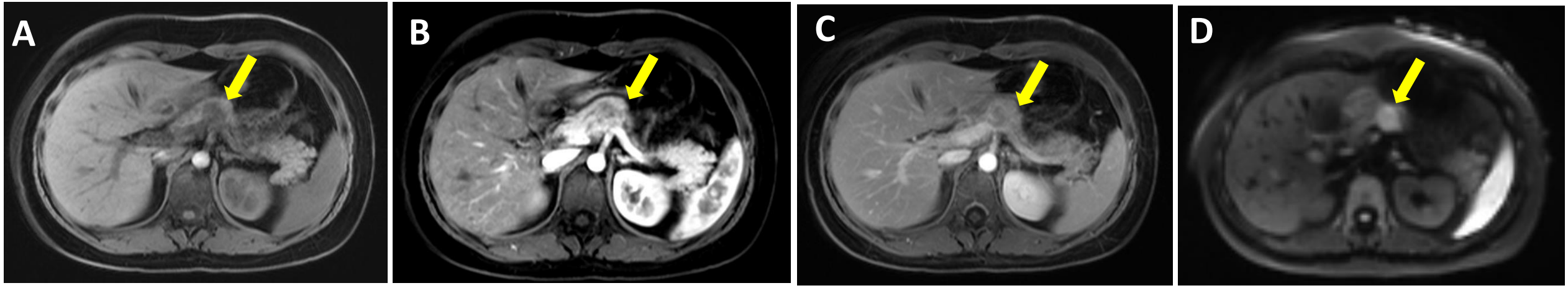




Initial ultrasound of upper abdomen demonstrates an irregular hypoechoic area in the pancreatic neck region (A).

Same day axial contrast-enhanced CT of the upper abdomen demonstrates heterogenous enhancement in the pancreatic neck region on the arterial phase (B), which appears less conspicuous with similar density to the adjacent normal pancreas on the portal venous phase (C). Given the pattern of enhancement, this finding was suspicious for a focal mass.

37 year-old woman with PMH of FAP presented to the ED with upper abdominal pain and mildly elevated lipase



Follow-up MRI of abdomen demonstrates a hypointense lesion in the pancreas head on pre-contrast T1 weighted image (A), which shows brisk progressive enhancement on arterial (B) and subsequent delayed images (C). The lesion shows restricted diffusion on DWI (D).
Given the reduced conspicuity relative to the background pancreatic enhancement due to the hypervascularity of the lesion, the pre-contrast and associated restricted diffusion images help with identifying the lesion and its local extension.

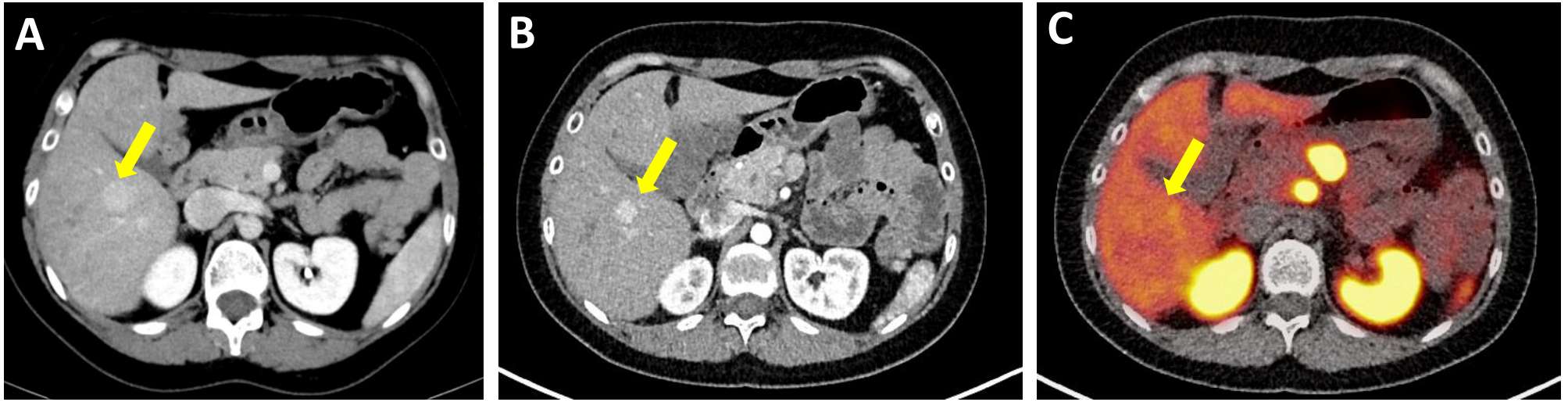
6 months Prior

Time of Presentation to ED

Time of Presentation to ED

Patient had history of multiple bouts of acute pancreatitis in the past. A contrast-enhanced CT 6 months prior shows a cystic lesion in the pancreas head, which was thought to be likely a pseudocyst and sequela of prior pancreatitis (A). The enhancing lesion appears to have developed in the interval at a similar location (B). ^{68}Ga-DOTATATE PET/CT demonstrates focal radiotracer uptake to the new lesion (C) which is highly suspicious for a neuroendocrine tumor.

6 months Prior | Time of Presentation to ED | Time of Presentation to ED



An enhancing liver lesion was noted on the CT from 6 months prior (A) and diagnostic CT at the time of presentation to the ED (B). Given the stability, the lesion was thought to be likely representing a focal nodular hyperplasia (FNH). However, an enhancing metastatic lesion could not be excluded. The lesion demonstrated no focal ^{68}Ga-DOTATATE uptake on the PET/CT images, confirming that the lesion is likely benign.

Diagnosis: Neuroendocrine Tumor of Pancreas in FAP

- Although initial considerations included focal pancreatitis or sequela of prior necrotizing pancreatitis, the degree of arterial enhancement on CT and MRI, as well as focal uptake on ^{68}Ga-DOTATATE PET/CT was diagnostic for NET.

- The lesion is more conspicuous on pre-contrast fat saturated T1 images, and becomes less distinguishable from normal adjacent pancreas on post contrast sequences. Marked restricted diffusion also adds value to diagnosis of focal NET in the pancreas.

- EUS and biopsy was performed subsequently which confirmed the diagnosis of NET.

Discussion:

Familial Adenomatous Polyposis (FAP) Syndrome:

- Autosomal dominant entity characterized by adenomas throughout the colon and a variety of common signs, such as upper GI tract polyps, osteomas, dental anomalies, congenital hypertrophy of the retinal pigment epithelium, thyroid, brain and periampullary carcinomas, and desmoid tumors.
- Caused by mutations in the APC gene with estimated prevalence of over 1/10,000.
- Near 100% risk of developing colorectal carcinoma by the 4th decade of life.
- Estimated relative risk for PDAC is 4.5 times higher than for the general population.
- Rare cases of NET have been reported in patients with FAP, however unclear whether this is a co-existence or genetic association.

Discussion:

- The initial clinical presentation of this patient was more likely to be another episode of acute pancreatitis, but the follow-up imaging and biopsy confirmed the diagnosis of NET at a similar location of previously noted pancreatic pseudocyst.
- NET is a less commonly known cause of acute pancreatitis. Majority of previously reported cases have been nonfunctioning tumors and the pancreatitis tended to be mild. Moreover, majority of the tumors were diagnosed in advanced stages, hindering curative treatment.
- In this case, no definite solid enhancing lesion was seen on the previous images. Interval development of NET at this similar location raises the possibility of initial focal pancreatitis in the setting of gradually developing NET in this case.
- 68Ga-DOTATATE PET was helpful in distinguishing benign focal pancreatitis from the tumor and confirming the benign characteristics of liver FNH and excluding hepatic metastasis.

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