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Gruber-Frantz tumor: a rare pancreatic neoplasm

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Mr. Editor

We present the case of a 37-year-old woman with abdominal pain in the epigastrium radiating to the right flank for a month of evolution. On physical examination, epigastric mass is palpated, firm and painless. Computed tomography (CT) showed a cystic tumor in the body and tail of the pancreas with solid areas and defined borders (12x10 cm), which displaces structures (Fig. 1-A). Endoscopic ultrasound (EUS)-guided fine needle biopsy was taken, with consistent cytology of SPT. Subsequently, a distal pancreatectomy with tumor resection and nodal dissection was performed (Fig. 1-B). Cytology reported discohesive cells, some arranged around capillaries, with small nuclei with clefts (Fig. 1-C), CK7 negative and β-catenin positive (Fig. 1-D). After 4 years of follow-up, there is no evidence of recurrence.

Discussion

Pancreatic SPT is rare (1-2% of pancreatic neoplasms), of low-degree malignancy (10-15%) and generally, in young women (25-35 years-old). It is diagnosed in asymptomatic patients as incidental finding or by nonspecific abdominal pain with/without mass effect. It is typically a large solitary lesion (6-22 centimeters) on the body and tail of the pancreas (1).

Diagnosis is by TC/MRI. In portal-venous CT, an encapsulated tumor of a solid-cystic component with intramural hemorrhage without communication to the pancreatic duct is observed. EUS is useful to characterize the lesion and taking biopsies (2). Histologically, it has monomorphic discohesive cells separated from capillaries with oval or cercariform nuclei in hyalinized myxoid material and PAS-positive grouped globules. The main positive markers are β-catenin, cyclin D-1, CTNNB1 and vimentin (3).

Treatment is surgical if the tumor is localized and circumscribed. Recurrence and 5-year survival are <10% and >96%, respectively (1-2). Metastases are usually hepatic and chemotherapy, arterial embolization (4) and radioablation (5) are described as treatment options. Its accurate diagnosis considerably improves the prognosis with respect to others pancreatic neoplasms.

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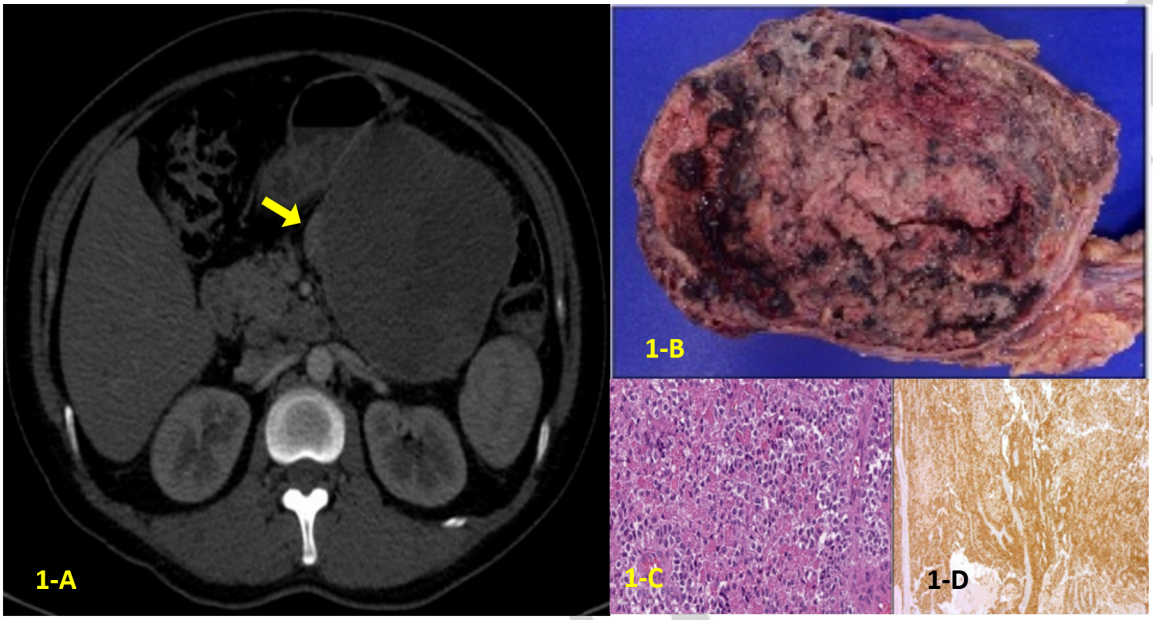

Fig. **1-A)** Axial abdominal tomography image in arterial phase demonstrates a cyst tumor in the body and tail of the pancreas with solid component (yellow *arrow*) that displaces intestinal loops, without adenopathies. **1-B)** Surgical specimen with integral capsule. **1-C)** Histopathology with cells arranged along vascular septa with small nuclei, mild anisonucleosis and nuclear clefts. **1-D)** Immunohistochemistry for β-catenin expression was positive in the nuclei.