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Case Report

Endoscopic Ultrasound in Solid Pseudopapillary Tumor of the Pancreas: About two Clinical Cases

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Abstract

Background: Solid pseudopapillary tumor of the pancreas (SPPT) or Frantz's tumor is a rare exocrine pancreatic tumor, it generally occurs among females within the second or third decade of life. The clinical symptomatology is non-specific, frequently presenting as a slowly growing abdominal mass without any biological abnormalities. The imaging usually shows a well-limited mass with little vascularity. Endoscopic ultrasound (EUS) with fine needle aspiration (FNA) helps the diagnosis. The treatment of these tumors is surgical, and the prognosis is excellent after complete resection. We report two new cases of Frantz tumor, detailing the clinicopathological features of this rare neoplasm. ***Case presentation:*** Case 1: A 19-year-old woman presented with middle upper abdominal pain. Computed tomography (CT) scan showed a solitary encapsulated mass in the pancreatic body. Endoscopic ultrasound showed a regular, well-defined, heterogenous lesion, measuring 50 × 46 cm, in the pancreatic body. endoscopic ultrasound fine-needle aspiration was then performed with cytopathological analysis compatible with SPPT. Body computed tomography confirmed the absence of metastases and she underwent a central pancreatectomy. 12 months after the diagnosis, she remains asymptomatic, continuing regular follow-up. Case 2: A 22-year-old woman presented with epigastric pain and weight loss. CT scan revealed a single 5 cm well defined mass in the pancreatic head. EUS showed a well-defined heterogenous hypoechoic encapsulated mass in the pancreatic head, measuring 64 × 44 mm; EUS-FNA was performed and cytopathological analysis was suggestive of SPPT. She underwent conventional cephalic duodenopancreatectomy. She presented septic postoperative complications, with a favorable outcome after medical and surgical therapy. she remains asymptomatic, after 6 months follow-up. ***Conclusion:*** Solid pseudo papillary pancreatic tumor is a rare exocrine low-grade neoplasm. EUS is a sensitive tool in the diagnosis of SPPT, identifying and characterizing the pathologic lesions, and allowing EUS-FNA with cytomorphological recognition, making it an invaluable tool for establishing diagnosis, helping clinical management and surgical planning.

Keywords: Solid pseudopapillary tumor, Frantz's tumor, pancreatic tumor, Endoscopic ultrasound, Pancreatectomy.

INTRODUCTION

Solid pseudopapillary tumor of the pancreas (SPPT) or Frantz's tumor is a rare exocrine pancreatic tumor, it generally occurs among females within the second or third decade of life. The clinical symptomatology is non-specific, frequently presenting as a slowly growing abdominal mass without any biological abnormalities. The imaging usually shows a well-limited mass with little vascularity. Endoscopic ultrasound (EUS) with fine needle aspiration (FNA) helps the diagnosis. The treatment of these tumors is surgical, and the prognosis is excellent after complete resection. We report two new cases of Frantz tumor, detailing the clinicopathological features of this rare neoplasm.

CASE PRESENTATION

Case 1

A 19-year-old female, with no relevant past medical or surgical history, was admitted to our hospital

with a 3-month history of constant upper abdominal pain radiating to the back, without weight loss nor any other symptoms. No anomalies were found on physical examination.

Computed tomography (CT) scan identified a roughly rounded homogeneous well defined encapsulated mass in the pancreatic body measuring 54 * 57 mm with moderate enhancement after contrast injection, pushing forward the gastric antrum without invasion.

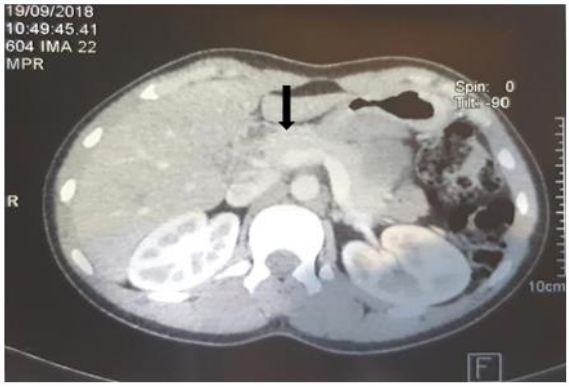
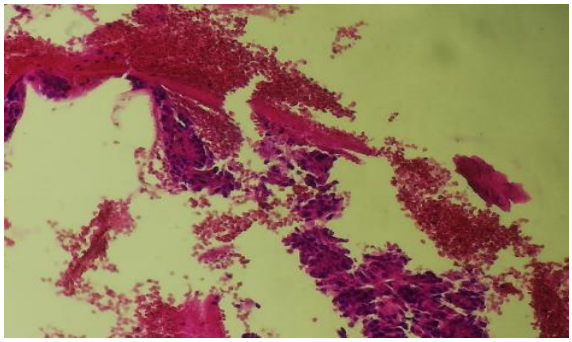


Fig-1: CT scan of the abdomen showed a well-defined pancreatic tumor

EUS found a well-defined round heterogeneous mass in the pancreatic body, measuring 50 * 46mm. A fine needle aspiration (FNA) with a 22-gauge needle was performed obtaining a representative tissue sample without complications. The cytopathological analysis of two slides and one cell block showed a tumoral proliferation with a papillary and micropapillary architecture bordered by slightly atypical cell with rounded monomorphic nucleus, fine chromatin and smooth nuclear contours evoking a solid pseudopapillary pancreatic tumor, Immunohistochemical staining showed positive expression of synaptophysin, CD10, β-catenin, Vimentin, Progesterone Receptor.



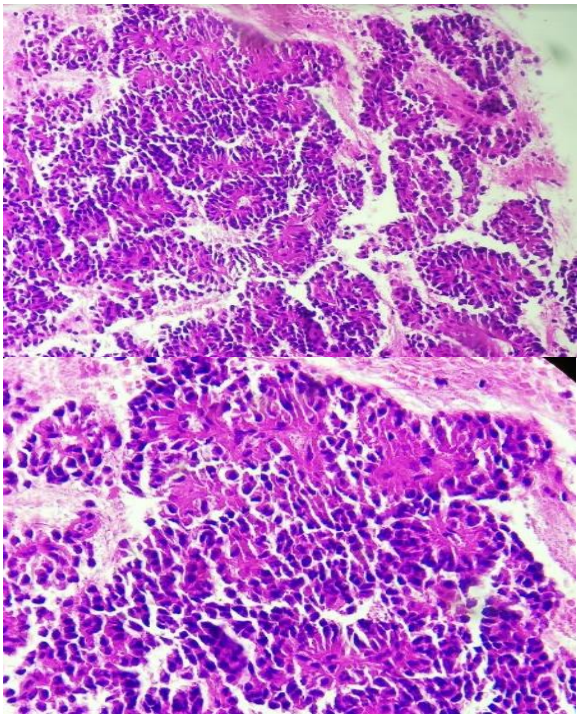


Fig-2: Histology reveals features of a solid pseudopapillary tumor

All biological parameters were within normal range: no pancreatic insufficiency or elevated pancreatic enzymes, no abnormal liver function or cholestasis. Tumor markers carcinoembryonic antigen (CEA), carbohydrate antigen (CA19-9) were all within normal range.

The patient underwent a central pancreatectomy, with R0 resection. Histopathological examination of the surgical specimen showed an encapsulated tumor proliferation with a compact and pseudopapillary architecture, monomorphic rounded tumor cells, moderately abundant eosinophilic cytoplasm oval nucleus with fine chromatin and no mitosis, compatible with a pseudopapillary and cystic tumor of the pancreas. No additional therapy was administered.

The follow-up was 12 months without either any evidence of recurrence during this period of neither tumor recurrence nor pancreatic insufficiency.

Case 2

A 22-year-old female, with no relevant past medical or surgical history, presented to our department with abdominal pain localized to the epigastric region and a weight loss (3kg in 2 months). Physical examination was normal.

CT scan identified a well-defined mass located in the pancreatic head measuring 45 * 50mm, heterogeneous and hypodense with peripheral enhancement. Magnetic resonance imaging (MRI) scan

showed a well-circumscribed encapsulated tumor in the pancreatic head measuring 44 * 57 mm, evoking an SPPT, with a mass effect on common bile duct without dilatation or invasion, and no secondary liver location.

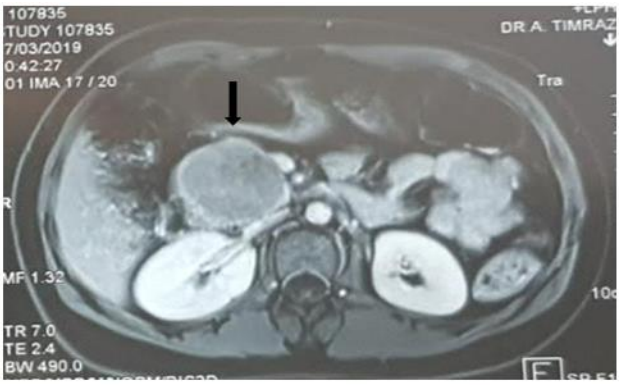


Fig-3: MRI showed a well-defined pancreatic tumor

EUS showed a regular well-defined heterogeneous hypoechoic encapsulated tumor in the pancreatic head measuring 64* 44mm, containing microcalcifications.

Two samples of FNA were taken from the suspected lesion and they were then subjected to immunohistochemistry and evaluated by a pathologist: the microscopic and immunohistochemical profile was that of a Frantz tumor. All biological parameters were within normal range.

The patient underwent a cephalic duodenopancreatectomy with R0 resection. In the Postoperative course, the patient presented a sepsis; the investigations found a high account of white blood cells and an elevated C-reactive protein, the imaging showed a collection repressing the pancreas in favor of a perihepatic abscess which was treated by surgical abscess drainage and received intravenous antibiotic therapy with a favorable outcome.

No additional therapy was administered. After 6 months of follow-up neither signs of tumor recurrence neither endocrine nor exocrine pancreatic insufficiency are present.

DISCUSSION

SPPT was described for the first time by Dr. Frantz in 1959 [1]. Also referred to as papillary epithelial neoplasm, papillary cystic neoplasm, solid-papillary neoplasm, solid-cystic neoplasm, and low-grade papillary neoplasm of the pancreas [2] accounting for less than 1%–2% of all exocrine pancreatic tumors [3]. It mainly affects young women ranging from 20 to 30 years [4]. Our patients fit into this group.

The pathogenesis of the tumor is unclear, the cell origin of the SPPT were hypothesized to be from multipotent primordial cells, whereas others investigators suggest an extra pancreatic origin, from genital ridge angle-related cells [5].

Most common sites of the tumor are at the tail of the pancreas (35.9%) and the head -as presented in our first patient- (34%) then the body –as presented in our second patient-(14.8%), the neck (1.01%) and the uncinate process of the pancreas (0.43%). The tumor can be located in multiple pancreatic sites (13.35%), or have a extra pancreatic localization (1.01%). mass, with a mean diameter of 6cm [4].

The Clinical symptoms are not specific and are represented by vague abdominal pain, sometimes associated with nausea or a slowly enlarging upper abdominal mass or weight loss [6, 7], although many are incidental findings. Both our patients presented upper abdominal pain with no significant weight loss.

The pancreatic enzymes are within normal range with non-endocrine imbalance [8]. The value of tumor markers is limited in the diagnosis of SPPT In a Chinese study, alpha fetoprotein, CEA, CA19-9, CA125, and CA242 were found to be slightly increased in 11 out 553 cases of SPPT [9]. In our case, no biological anomalies were found.

The imaging can show a mixed solid and cystic pancreatic lesion. The CT scans show a heterogenous mass, usually found to be a vascular or hypo vascular from arterial phase of CT, often with peripheral contrast enhancement corresponding to the fibrous pseudo capsule [4, 10]. Unenhanced CT shows calcifications in one third of the tumors [11].

A study about MRI features of small solid pseudopapillary tumors showed that these tumors can have completely (82%) or partially (18%) well defined margin, occur in the pancreatic tail, be round, have low or very low signal intensity on fat-saturated unenhanced T1- weighted and have high or very high signal intensity on fat-saturated T2-weighted images and have early heterogeneous and progressive enhancement [12].

EUS is more accurate as a diagnostic modality for pancreatic tumor, especially for tumors less than three centimeters [13].

The typical EUS appearance of SPPT is a well demarcated, hypoechoic, solid appearing mass. SPPT can also appear as either a mixed solid and cystic lesion or a purely cystic lesion, due to hemorrhagic necrosis and irregular calcifications occasionally present in up to 20 % of cases [14]. In our cases, EUS showed well defined heterogeneous hypoechoic mass.

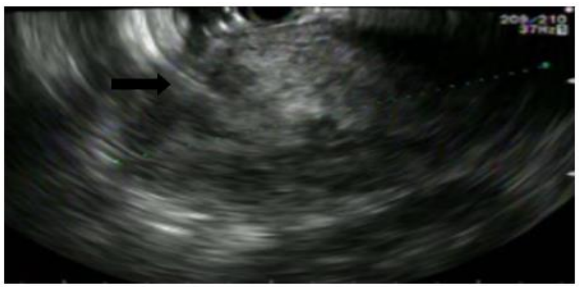


Fig-4: EUS showing an SPPT adjacent to the gastric wall [15]

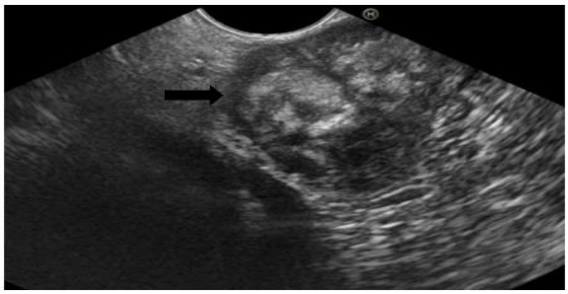


Fig-5: SPPT containing calcification components with an inhomogeneous echo pattern [16].

EUS-FNA improved the preoperative diagnostic yield of SPPT, as showed in an international multicenter case series [17]. The diagnostic yield of CT 23.5% and EUS 41.2 %, CT and EUS combined had a diagnostic yield of 52.9 %. The addition of EUS-FNA increased the diagnostic yield to 82.4 %.

EUS-guided FNA of pancreatic masses is becoming the standard for obtaining cytological diagnosis; it also avoids the risk of cutaneous or peritoneal contamination, unlike CT/US-guided FNA [18].

Both of our patients had an accurate preoperative diagnosis of SPPT by EUS-FNA, confirmed by histological study of postoperative specimens.

Some authors advocate preoperative fine-needle biopsy (FNB) which helps the diagnostic in 75% of cases [14]. Other authors may not accept this because of the uncertainty in diagnosis and possible tumor cell spread.

The SPPT has a malignant potential, a tumor size ≥ 5 cm was associated with an increased risk of high-grade malignancy [19].

The SPPT is usually operable at presentation unless it shows local vascular invasion or distant metastases, usually to the liver and peritoneum [4].

Macroscopically the SPPT is encapsulated tumor with solid and cystic compositions, and areas of hemorrhages and central necrosis [4]. The main character of the tumor is the presence of both solid and pseudopapillary patterns, which give rise to the designation of solid-pseudopapillary pancreatic tumor [20, 21].

Microscopically, the tumor exhibits branching papillae with myxoid stroma surrounded by monomorphic neoplastic cells [22], similar to what was found in the specimen from our case.

The neoplastic cells are like neuroendocrine tumor cells, thus Immunohistochemically it is important to use special stains vimentin, CD10, and beta-catenin to differentiate these tumors, chromogranin A has been regarded as typically negative in SPPT [4, 23].

Surgical procedure is the main treatment for pancreatic SPPT, and the type of operations depends on the location of the tumor. Accurate pre-operative diagnosis enables minimal surgery to preserve the pancreas. Distal pancreatectomy and pancreaticoduodenectomy (Whipple or Longmire) followed by local resection and enucleation are the most common operations. Resection of only midportion of the pancreas leaving the head and the tail of pancreas can be done for tumors at the body of pancreas mass [4].

Both patients had complete surgical resections; none of them required additional therapy. There has been no evidence of recurrence with a mean period follow up of 9 months after surgical resection.

In general, the prognosis of SPPT is good, with a 5-year survival of 95% following complete surgical resection [4]. SPPT is regarded as benign tumor with low malignant potential and less than 5% risk of local recurrence [24]. In rare cases metastases may occasionally develop with a latency of several years [25].

CONCLUSION

Solid pseudo papillary pancreatic tumor is a rare exocrine low-grade neoplasm that generally occurs in young women. The imaging investigations help the diagnosis. EUS is a sensitive tool in the diagnosis of SPPT, identifying and characterizing the pathologic lesions, then allowing EUS-FNA with cytomorphological recognition, making it an invaluable tool to diagnose SPPT with typical cytomorphologic and immunohistochemical features prior to treatment. SPPT has a favorable prognosis provided that the treatment is a complete surgical resection. The clinicopathological features of SPPT in our population are similar to those described in literature.

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