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CASE REPORT

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Frantz Tumor, a Rare Indolent Pancreatic Neoplasm Entity: A Case Report and Brief Review

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Abstract

Solid pseudopapillary neoplasm (SPN) of the pancreas, also known as Frantz tumor, is an uncommon tumor with indolent clinical features that primarily affects young women. We are presenting a 27-year-old Caucasian female with an accidental finding with pancreatic SPN who underwent complete resection of the tumor using a distal pancreatectomy and splenectomy procedure. Immunohistochemistry revealed low-grade pancreatic SPN. Despite the rare appearance, it should be considered in the differential diagnosis of a young female with a large pancreatic mass.**(International Journal of Biomedicine. 2024;14(1):175-178.)**

Keywords: Frantz tumor • solid pseudopapillary neoplasm • pancreas • distal pancreatectomy

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Abbreviation

CECT, contrast-enhanced computed tomography; **EUS**, endoscopic ultrasound; **FNA**, fine needle aspiration; **MDCT,** multiphasic multidetector-row CT; **MRI**, magnet resonance imaging; **SPN**, solid pseudopapillary neoplasm.

Introduction

Solid pseudopapillary neoplasm (SPN) of the pancreas is a low-grade malignant tumor composed of poorly cohesive epithelial cells, forming solid and pseudopapillary structures and lacking a specific line of pancreatic epithelial differentiation.(1) These entities were first described by Virginia Kneeland Frantz in 1959 as pancreatic papillary-cystic tumors.(2)

The WHO classified them as solid pseudopapillary tumors in 1996 and reclassified them as SPNs in 2010. The WHO classification describes SPNs as low-grade malignant neoplasms composed of loosely cohesive, monomorphic epithelial cells forming solid and pseudopapillary structures.(3)

Case Presentation

We are presenting a 27-year-old Caucasian female with an accidental neoplasm found from a routine check-up by a gastroenterologist. An MRI revealed a pancreatic tail mass, symptomless, adjacent to the spleen hilus (Figure 1).

Using general anesthesia, we performed laparotomy surgical treatment, and intraoperatively discovered a solid

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tumor in the distal pancreas adjacent to the spleen hilus. We performed a distal pancreatectomy with splenectomy and saved the discovered accessory spleen (Figure 2).

Postoperative recovery went well. Macro findings: encapsulated, gray-colored nodular tumor located in the distal pancreas, 6.5cm in diameter, soft consistency, which was built with stromal hyalin, myxoid vascular and hemorrhagic foci, and degenerative changes. Resection margins were microscopically free of tumor, R0 (Figure 3a).

Immunohistochemistry: SPN of the pancreas, low grade, pT3pNx (UICC 8th Edition), B-catenin (+) (Figure 3b), Vimentin (+) (Figure 3c), E-cadherin (-). The proliferation cell index measured with Ki-67 was low, about 7%. The patient has been followed up for 10 months and is in good condition.

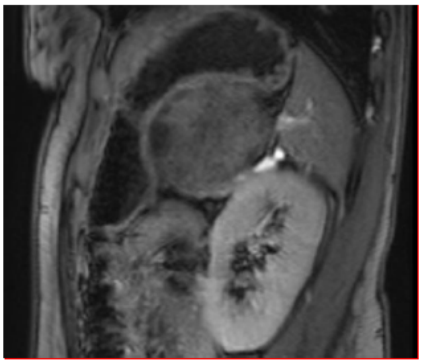

Fig.1. *MRI: A pancreatic tail mass, adjacent to the spleen hilum.*

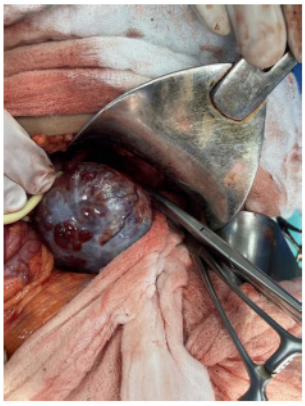

Fig.2a. *Intraoperative findings, pancreatic tail with SPN.*

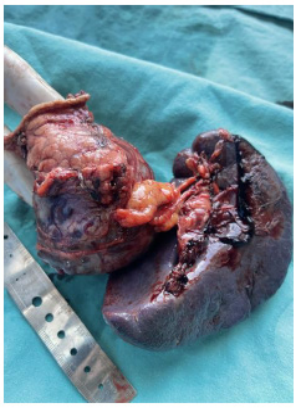

Fig.2b. *Specimen of resected distal pancreas with SPN and spleen.*

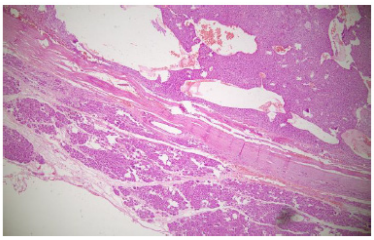

Fig.3a. *SPN and pancreas, H&E staining.*

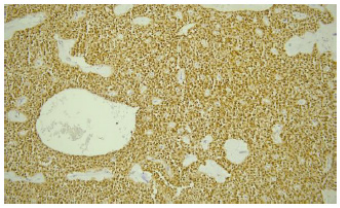

Fig.3b. *Immunohistochemistry: Beta-catenin, strong diffuse positive.*

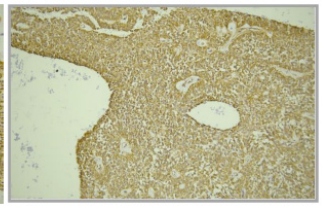

Fig.3c. *Immunohistochemistry: Vimentin positive.*

Discussion

Since the first description by Frantz as "papillary tumor of the pancreas, benign or malignant" in 1959, various names have been used to describe this rare tumor, such as a solid-cystic tumor, solid-cystic acinar tumor, papillary-cystic tumor, solid-papillary epithelial neoplasm, and Frantz tumor.[1,2] SPNs are rare, comprising approximately 0.17%-2.7% of all pancreatic tumors and only 5% of cystic neoplasms.[3]

An SPN primarily affects young women in their second and third decades. Although the hypotheses of its origin include endocrine, ductal, acinar, neurosecretory, and totipotent primordial cells, the histogenesis and pathogenesis remain unclear, which still motivates discussions.[4] Neoplasms morphologically identical to pancreatic SPNs arise in retro pancreatic tissue, ovaries, and testes.[1]

Two large retrospective reviews of 340 patients with SPN from the National Cancer Database showed that 82% of patients were female, the median age was 39 years,[5] and the mean age of the 553 SPN patients included in a review by Yu et al.[6] was 27.2 years, 88% were female. Similarly, in a study by Sun et al.[7] with a total of 118 patients, the mean age was 30.8 years, and the majority were female (n=95, 80.5%).

The exact reason for female predilection is unclear, but the literature suggests that sex hormones may be part of the pathogenesis of SPN.[8] According to studies by Lanke et al.[9] and Kurokawa et al.,[10] SPNs are considered hormone-sensitive because they express progesterone receptors, and female hormones influence the growth of SPNs. Unlike this study, Omiyale et al.[11] consider that "there is no association with functional endocrine syndromes."

The clinical symptoms are non-specific. Many patients are asymptomatic (38.1%); however, most patients are symptomatic, presenting with abdominal pain or discomfort. Other symptoms include abdominal mass, weight loss, jaundice, anorexia, fever, fatigue, abdominal discomfort, nausea, and vomiting.

Our patient was asymptomatic and we incidentally found SPN through imaging. SPN can occur in all parts of the pancreas. Most tumors (59%) were in the tail.[12] Outside the pancreas, they can occur in the retroperitoneum, liver, stomach, mesentery, duodenum, omentum, ovary, or lung. SPN can also be found in regional lymph nodes, the portal vein, the colon, the spleen, and blood vessels.[9]

The diagnosis may be difficult because of indolent clinical features. Studies have shown that tumor markers such as carbohydrate antigen 19-9 (CA19-9) and carcinoembryonic

antigen (CEA) were usually within normal ranges in patients with this disease.[13]

SPT is often diagnosed during complementary imaging investigations, such as ultrasound or CT scan of the abdomen, usually showing a well-encapsulated complex mass with solid and cystic components. MRI is better than CT in detecting the cystic or solid components of the tumor.[6]

Typically, SPNs are large, well-encapsulated masses that demonstrate variable degrees of internal hemorrhage and cystic degeneration and are often associated with calcifications. When these features are encountered in a young female patient, this neoplasm should be a strong diagnostic consideration with multiphasic multidetector-row CT (MDCT).[14]

Hanada et al.[15] compared pathologic and image findings at cystic component, with MRI: specificity was 80%, accuracy 68%. On delayed phase contrast-enhanced CT (CECT), pathologically aggressive SPNs may show greater enhancement than non-aggressive SPNs.[16]

It is recommended to request an MRI, besides the abdomen CT, of all patients suspected of SPN of the pancreas to avoid possible diagnostic mistakes. [17] MRI shows a well-defined mass with heterogeneous signal intensity on T1- and T2- T2-weighted images indicative of the tumor's variably solid and cystic nature.[18] EUS-FNA significantly increased the pre-operative diagnostic yield of SPN to 82.4%.[19]

Radical surgical resection is established as the standard treatment protocol for the disease; it is also recommended to perform metastasectomy, vascular resections, and/or resections of other compromised organs to ensure therapeutic success in 95% of the cases.[17,20,21] Different surgical procedures are available depending on the tumor localization, such as the Whipple procedure, central pancreatectomy, distal pancreatectomy with or without splenectomy, enucleation, etc. The outcomes are excellent if complete resection is achieved. Similarly, with an open approach, considerations should be made for a minimally invasive approach in patients with SPN.[4,8]

In appropriate indications, the enucleation of SPNs can be considered a safe and effective surgical procedure for pediatric patients.[22] The need for lymphadenectomy has been discussed due to the description of ganglion metastases in approximately 15% of cases.[23]

There were no significant clinical factors, such as age, sex, tumor size, tumor location, elevated carcinoembryonic antigen levels, and elevated carbohydrate antigen 19-9 levels, suggesting malignant potential.[21]

Patients with SPN who undergo resection have an excellent survival at 5 years, from 95%-97.7%.[6,7,24]) A reported 10-year disease-specific survival rate of 96%.[25]

SPNs have an excellent prognosis with minimal recurrence after resection. There have been reports from 1.8%-4.5% of patients with evidence of recurrence at the last follow-up. [7,12]

Adjuvant therapy is used only in a small number of patients because of the high resectability of SPN. The role of chemotherapy or chemoradiotherapy in treating SPN is also unclear. In some studies, adjuvant or neoadjuvant chemotherapy, radiotherapy, or chemoradiotherapy are reported in some unresectable cases with good results. [26,27]

In conclusion, pancreatic SPN is a rare entity that primarily affects young women. With indolent clinical features affecting young age, this distinctive neoplasm creates diagnostic difficulties. Surgery with R0 resection is the curative treatment of choice. Mandatory follow-up for early local recurrence or distant metastasis diagnosis is important. We should consider an asymptomatic young female who presents with a large pancreatic mass, accidentally revealed by diagnostic tools, as a possibly pancreatic SPN patient.

Competing Interests

The authors declare that they have no competing interests.

Ethical Considerations

Publication of the report was approved by the Ethics Committee at the National Institute of Public Health (Prishtina, Kosovo). Informed written consent was obtained from the patient to publish this case report and any accompanying medical images.

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