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Solid Pseudopapillary Tumor: A Rare Neoplasm of the Pancreas: A Case Report with Literature Review

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Abstract

Solid Pseudopapillary tumour of the pancreas (SPT) is an uncommon neoplasm of the pancreas representing about 1e3% of exocrine pancreatic neoplasm. The Histogenesis and the guidelines for its treatment remain a debatable question.We present a case of 28 years women who had epigastric. The ultrasound showed a heterogeneous, slightly echogenic and vascular ovoid mass, measuring 6 cm, situated in the pancreatic head.The patient underwent pancreaticoduodenectomy and did well post-operatively. Histological examination and immunohistochemistry staining concluded to solid peudopapillary tumor. The tumor's pathology,immunohistochemistry and treatment are discussed.SPN is a rare neoplasm that primarily affects young women. The prognosis is favorable even in the presence of distant metastasis. The majority of tumors are diagnosed through ultrasound or CT scan of the abdomen or MRI. Microscopically, the characteristic features are cellular degeneration and pseudopapillary appearance. Aggressive surgical resection is needed even in presence of local invasion, and also for recurrence as patients had a good long-term survival.

Keywords: Anatomy Pathology; Pancreas;Solide Pseudopapillary Tumour

Introduction

Solid pseudopapillary tumour of the pancreas (SPT) is an uncommon neoplasm of the pancreas representing about 1-3% of exocrine pancreatic neoplasm, usually occur in young females in the second to fourth decades of life [1], Until it was defined by the World Health Organization (WHO) in 1996 as 'solid pseudopapillary tumor' of the pancreas, this tumor was described by using various names including 'solid cystic tumor', 'papillary cystic tumor', 'papillary epithelial neoplasia', 'solid and papillary epithelial neoplasia', 'papillary epithelial tumor' and 'Frantz's tumor', 'solid and papillary tumor', 'solid-cysticpapillary epithelial neoplasm', 'benign or malignant papillary tumor of the pancreas' [2]. Histogenesis remains a debatable question: acinar, endocrine, ductal and progenitor cells have been postulated as possible beginnings of this tumor. It is frequently asymptomatic or minimally symptomatic [3].The classic CT-scann of the solid pseudopapillary tumor are a large well-encapsulated mass with varying solid and cystic components caused by hemorrhagic degeneration. Calcification and enhancing solid areas may be present at the periphery of the mass [4]. The clinical and pathological characteristics of SPT are different from pancreatic cancer. It has low malignant potential, and rarely metastasizes [1].

Observation

A 28-year-old woman with no prior medical history presented with complaints of epigastric. The pain had a sudden onset, with intermittent attacks; it was stabbing in nature and radiating to her back. It was associated with nausea and vomiting. There was no precipitating, relieving or aggravating factor. Review of systems was otherwise unremarkable. Her vital signs were stable. On physical examination, her abdomen was soft, non-tender and non-distended. There were no palpable masses, and bowel sounds were audible.The ultrasound showed a heterogeneous, slightly

echogenic and vascular ovoid mass, measuring 6 cm, situated in the pancreatic head. CT scan of the abdomen and pelvis revealed a similar appearance(Figure 1).

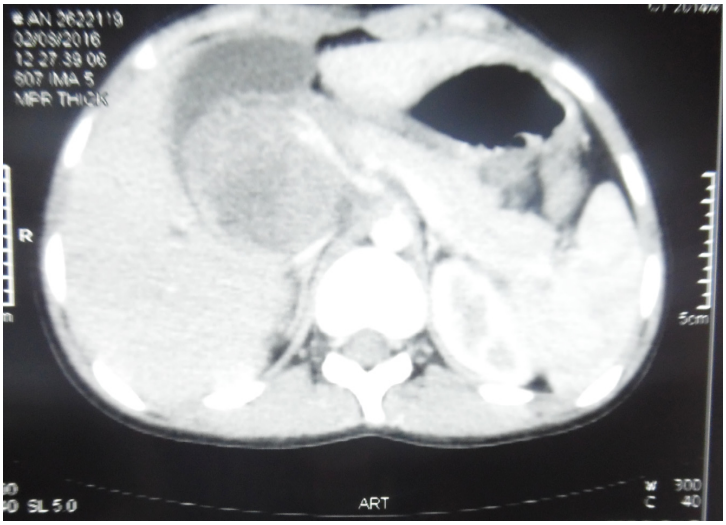

Figure 1: Rounded solid-cystic lesional process in the head of the pancreas with tissue component.

The patient underwent pancreaticoduodenectomy and did well post-operatively. Histological examination demonstrated a solid and vascular pattern with pseudopapillary cores. The tumor was found to be confined to the pancreas, with involvement of the intrapancreatic duct. There was no margin involvement. The histology also revealed perineural and probable angiolymphatic invasion tumor cells with papillary architecture(Figure 2).

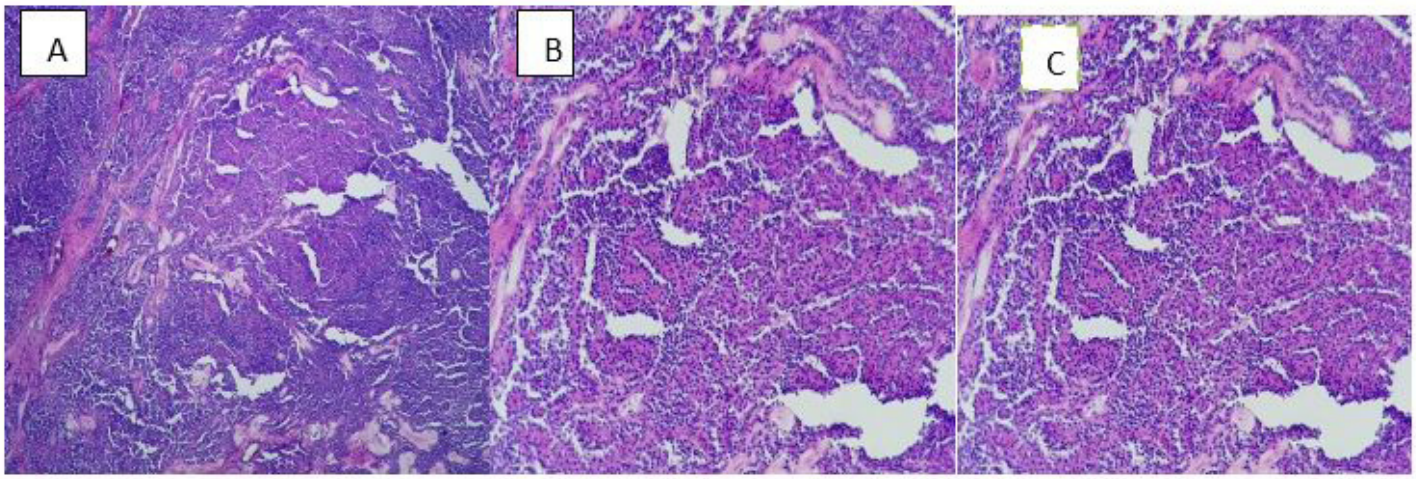

Figure 2:(A)+(B):solid and vascular pattern with pseudopapillary cores (HE x4); **(C):** Thin vascular structures covered by cuboidal cells with no cellular atypia (HE x40)**(B).**

An immunohistochemical analysis revealed that the tumor cells were positive for beta-catenin, vimentin, NSE, CD56 and progesterone receptor. These findings were consistent with solid pseudopapillary neoplasm (Figure 3).Given the concern of neurovascular involvement, the patient was enrolled into an imaging surveillance program.She has been asymptomatic, not requiring any adjunctive therapy.

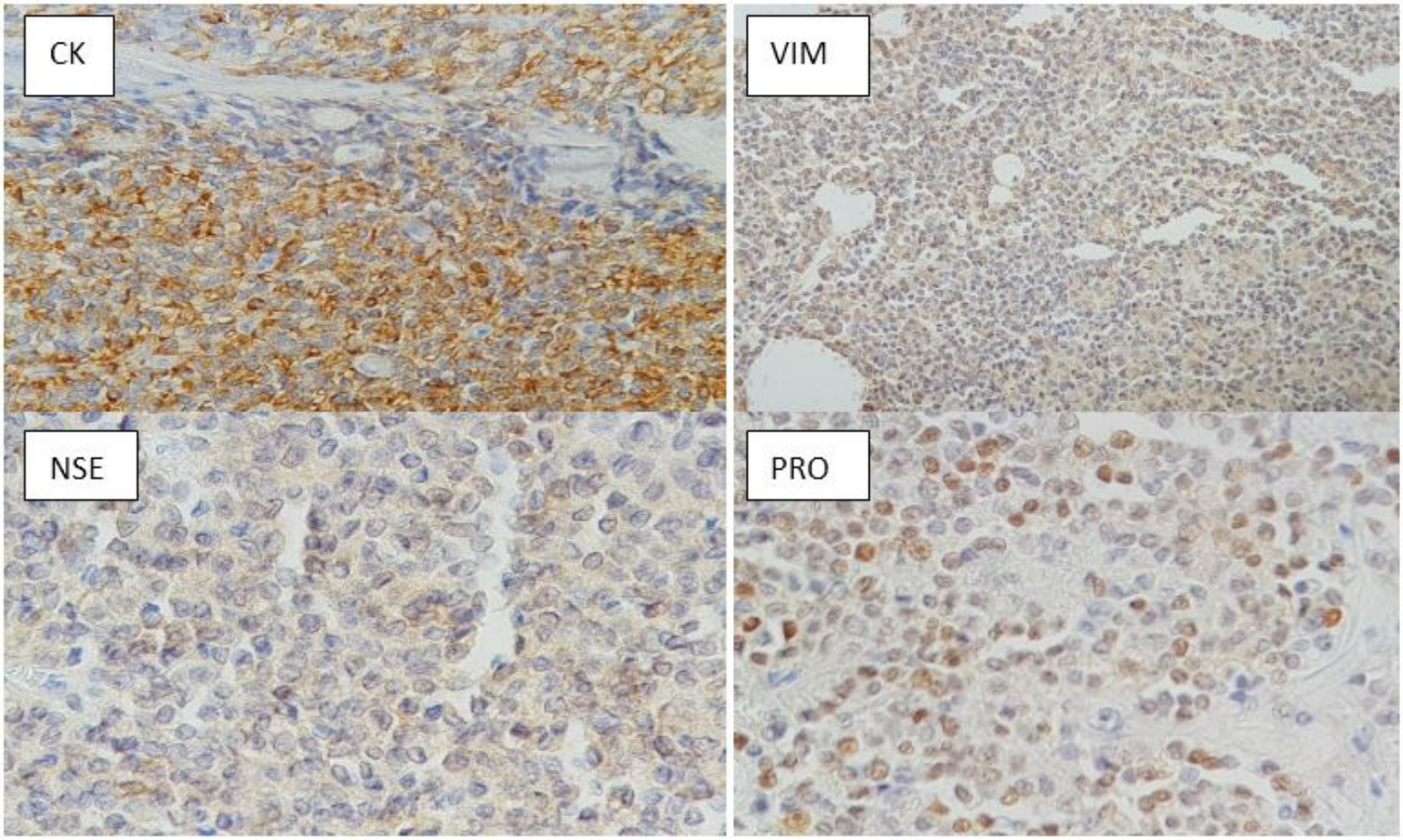

Figure 3: Tumor cells typically show strong immunoreactivity for Vimentin, Ck, NSE and progesterone anti-bodies.

Discussion

SPT represents an enigmatic tumour from which the origin, prognosis and natural history are unknown[1]. SPN is very rare; in fact, they only constitute about 5% of cystic pancreatic tumors and about 1 to 2% of exocrine pancreatic neoplasms[2]. The incidence rate of SPT of the pancreas is 0.2e2.7% of exocrine pancreatic neoplasm[5]. SPT of the pancreas are common in young women especially aged from 20 to 40 years). It is seen more frequently in femaleswith male-female ratio of 1:9[1]. The origin of solid pseudopapillary tumors is unclear. Many investigators favor the theory that SPTs originate from multipotent primordial cells, whereas others suggest an extrapancreatic origin, from genital ridge angle-related cells [6]. Unlike the aggressive malignant behavior shown by pancreatic carcinomas, these tumors are often benign and have indolent course. Pancreatic body and tail are the most common sites of presentation. Malignant potential is seen only in 10-15% of all cases [7]. They may present as an abdominal mass, abdominal pain or, rarely, with jaundice [3]. There are no pathognomonic features on blood investigations and tumor markers are usually unremarkable [8]. The majority of tumors are diagnosed through ultrasound or CT scan of the abdomen, but MRI also defines the hypervascular, well-encapsulated, round tumors with mixed cystic and solid components. Echo-endosonography may provide FNA biopsy with the possibility of pre-operative pathologic diagnosis[9].

Grossly, it appears as a large and encapsulated mass, generally well demarcated from the remaining pancreas. These present as well-circumscribed tumor masses usually well demarcated from the pancreas, with large spongy areas of hemorrhage on its cut surface alternating with both solid and cystic degenerative components[7]. In fact, invasion of the adjacent organs, such as the spleen or the duodenal wall, is rare[2]. Preoperative diagnosis with imaging alone

is technically challenging due to difficulties differentiating this tumor from other pancreatic conditions such as a pancreatic cyst or other histological variants such as adenocarcinoma, cystadenoma, cystadenocarcinoma, islet cell tumor, neuroendocrine tumor, or teratoma [7]. Preoperative histologic confirmation is not necessary and is usually reserved for patients who have high operative risk orwho require complex resections (borderline resectable tumor, vascular resection required, and/or nodal disease outside of the resection margins).

The morphological appearance of SPT varies from solid to cystic components with cellular degenerative changes [10]. The lesion is comprised of small ovoid or polygonal cells with small central nuclei and abundant cytoplasm. Histologic markers of poor prognosis include a high mitotic rate, spindling of tumor cells, anaplastic giant cells, capsular invasion, and lympho-vascular involvement[11].These tumors are typically positive for vimentin, alpha1 antitrypsin, alpha1antichymotrypsin, and neuron-specific enolase. Nguyen,et al. showed 100% sensitivity and specificity of beta catenin in these tumors while positivity for synaptophysin was seen in 36% and for chromogranin in 15%[7]. The proliferation of these tumors may be affected by sex hormones as 80% of these tumors have progesterone receptor positivity [12]. Lesions are also known to progress during pregnancy.The positive rate is variable for estrogen receptor. IHC staining of Ki-67 can help determine malignant potential for these tumors [7].

Once the diagnosis of SPN is made, surgery is the first choice of treatment. SPN is usually surrounded by a pseudocapsule and exhibits benign or low-grade malignancy. Conservative resection with preservation of as much pancreatic tissue as possible is the treatment of choice. According to the location of the tumor, distal pancreatectomy with or without splenectomy, pylorous preserving pancreato-duodenectomy, Whipple operationor enucleation can be performed [2]. Resection of distant metastasesshould be done at the time of primary resection. Resection should also be tried for recurrences[13]. The common sites of local invasion are liver, portal/splenic vein/superior mesenteric vein, spleen, diaphragmatic muscle, omentum or peritoneum, duodenum, stomach, colon, or left kidney [14]. 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 the treatment of SPN is also unclear. In some studies, adjuvant chemotherapy and radiotherapy are reported in some unresectable cases with good results. Neoadjuvant chemotherapy or chemoradiotherapy is also reported to have been successful in a few cases[2]. The overall five-year survival rate of patients with SPN is about 95% [15]. Death ascribed directly to the tumor is rare [16].

Conclusion

SPT of the pancreas is a rare neoplasm with low malignant potential find primarily in young women. It's a distinct histological and biologic disease entity with an indolent clinical course. Surgery is the primary therapeutic modality. The use of adjuvant radiotherapy and chemotherapy is individualized depending upon the risk for local recurrence or metastasis. long term survival is excellent even after recurrence if surgical resection is performed.

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