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Solid pseudopapillary neoplasm of the pancreas: Report of a rare case and review of the literature on an enigmatic entity

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Abstract
Solid pseudopapillary neoplasm pancreas SPN is a rare primary neoplasm of the pancreas that typically affects young women, SPN show nonspecific clinical presentation with vague radiologic features and are often histologically benign.
We here report a 21-year-old Libyan girl presented with gradually progressing, episodic, abdominal pain accompanied by anorexia and significant weight loss (11 kg in 2 months), without any significant laboratory findings. On CT scan a heterogenous mass was found at the distal pancreas. The patient underwent distal pancreatectomy with spleen preserving operation with the presumptive diagnosis of Solid pseudopapillary neoplasm of the pancreas (SPN). The tumor was well-circumscribed, encapsulated, 17x12 cm in dimensions, 2.5 kg weight, the patient was not given any adjuvant therapy and shows no sign of disease after eight months follow-up
Surgical decision should be built depend on information gained collectively from preoperative CT examination, intraoperative findings of tumor location, capsule integrity, and invaded surrounding tissues, immunohistochemical staining which lead to relatively clear diagnosis of solid pseudopapillary neoplasm favoring more tendency for curable radical resection even in large swellings or capsular invasion, as conclusion it is important to differentiate this tumor from other pancreatic neoplasms because this neoplasm is amenable to cure after complete surgical resection, unlike malignant tumors of the pancreas.
We aim to review the current literatures regarding the diagnosis, management, and outcomes of patients with solid pseudopapillary neoplasms of the pancreas.

Keywords: Solid pseudopapillary neoplasm, SPN, Pancreas, Tumor

Introduction
Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare neoplasm, which represents 0.2-2.7% of pancreatic cancers [1-3]. The name of this entity dates back to 1959 when Virginia Frantz first described a “papillary cystic tumor of the pancreas” in the Armed Forces Institute of Pathology (AFIP) band on tumors of the pancreas. The patient was a 2-year-old boy who died during an attempted pancreatic-duodenectomy [4]. In 1970 Hamoudi *et al.* described the ultrastructural features of the tumor, which led to its acceptance as a separate clinic-pathological entity [5]. Until its inclusion in the World Health organization (WHO) classification of pancreatic tumors in 1996 as ‘‘solid pseudo- papillary tumor’’ of the pancreas [6], this entity has been described by different names in the literature such as “papillary epithelial neoplasm of pancreas”, “solid and cystic tumor of the pancreas”, “adenocarcinoma of pancreas of childhood”, “papillary-cystic tumor” and “solid and papillary epithelial neoplasm” [7], all reflecting histogenesis and biology of this lesion as well. In the current WHO classification [8], SPN is defined as a low-grade malignant neo- plasm of the exocrine pancreas. The term SPN gained wide acceptance and is currently the most frequently used name for this entity [9].
To date, around 700 cases have been reported [1], more than two-thirds of them in the last 10 years [10, 11]. This probably reflects the increasing awareness of the clinicopathologic and radiographic features of SPN and the uniformity of the nomenclature used for SPN in the last years. However, the etiology and the differentiation status of SPN remained challenging and still enigmatic [9]. Herein, we present the clinical, histopathological, immune-histochemical and therapeutic characteristics of one SPN case with a review of the literature

Case report

A 21-year-old Libyan girl presented with gradually progressing, episodic, epigastric pain radiating to the left hypochondrium with recent accentuation. Pain was accompanied by anorexia and significant weight loss (11 kg in 2 months). There was no relationship to food or bowel movements. General and systemic examination revealed no abnormality, abdominal examination reveals gross distention involve most of left abdomen regions maximally left hypochondrium accompanied by mild, dull left sided-tenderness, Hematologic and metabolic parameters, including tumor markers were within normal limits.

Computerized tomography Figure 1. Imaging revealed a well-circumscribed, enhancing, rounded, well defined. partly cystic and partly solid mass measuring 17x12 cm in the tail of the pancreas very close to posterior wall of stomach and extended to occupying all areas below left hemidiaphragm, supero-anterior to the left kidney with no clear demarcation from left adrenal gland and spleen; (Arrow heads), a proposed endoscopic ultrasound and fine needle aspiration cytology of the mass was declined by the patient.

The patient was operated on with the presumptive diagnosis of SPN or cystadenoma/ cystadenocarcinoma of the pancreas, as well as possibilities of retroperitoneal and left adrenal masses not excluded due to large size of the mass as well as proximity to left upper abdomen organs in preoperative imaging.

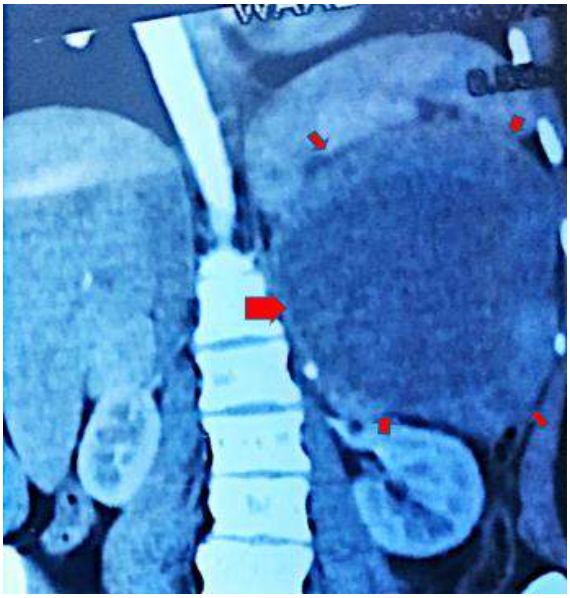


Fig 1: Pre-operative imaging

Per-operatively, there was a large mass at the tail of the pancreas adherent to the stomach, the peritoneal cavity, liver and spleen were normal (There was no definite infiltration into the surrounding tissues) Figure 2 (A& B).

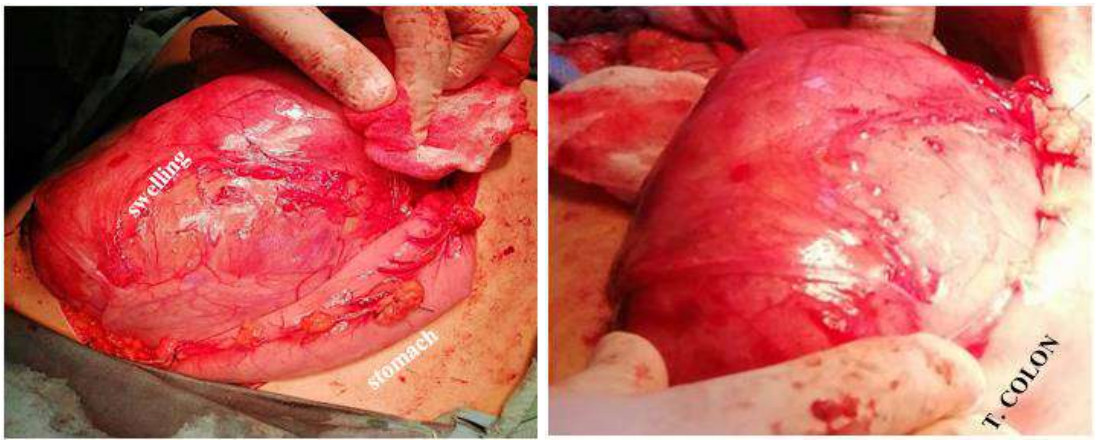


Fig 2 A, B: Intra-operative mass excision from posterior gastric wall

The patient underwent a spleen-preserving distal pancreatectomy, made an uneventful post-operative recovery. Since the disease was localized and the excision was complete, the patient was not given adjuvant treatment, and shall be on regular follow-up. The resected mass was 17x12 cm in dimensions, 2.5 kg weight, well circumscribed, and solid brown on color. Fig. 3 (A& B)

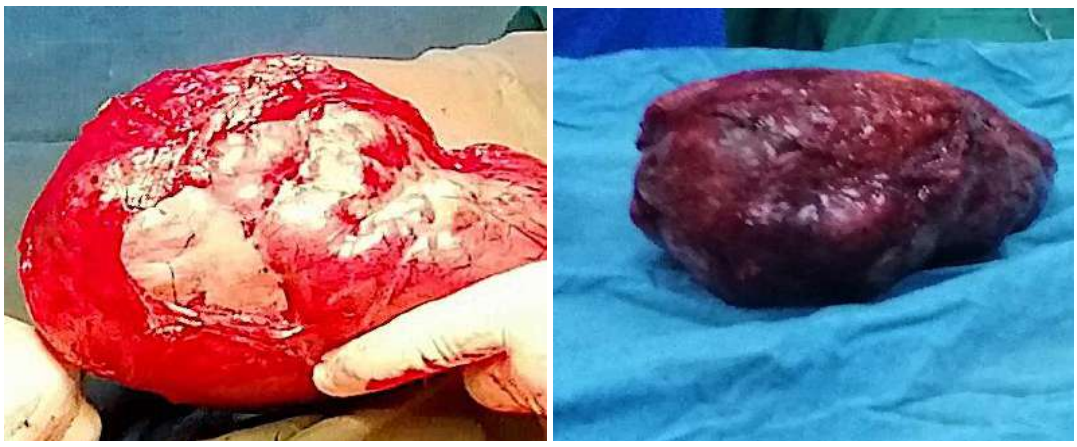
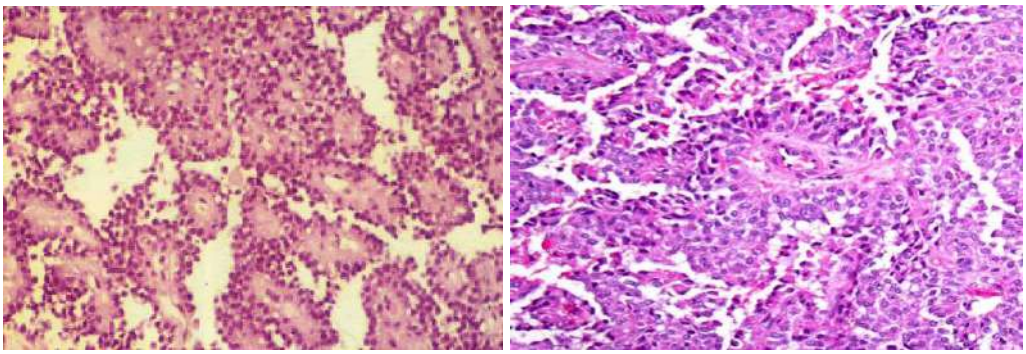


Fig 3 A, B: Resected mass

Microscopically, there were solid and cystic areas with pseudorosetting and pseudopapillae formed by polygonal cells with eosinophilic cytoplasm, vesicular nuclei with grooves foamy histiocytes, nuclear groove which is characteristic finding of SPN, as well as minimal atypia and occasional mitosis**,** neither capsular nor vascular invasion, (Fig.4 A&B)



A **B**

Fig 4 A, B: Minimal atypia and occasional mitosis**,** neither capsular nor vascular invasion

In addition, Immunohistochemistry evaluation was performed: The tumor cells showed strong cytoplasmic positivity for beta-catenin, vimentin, CD 10 and negative for (Ki 67 (Fig.5 A,B, C&D).

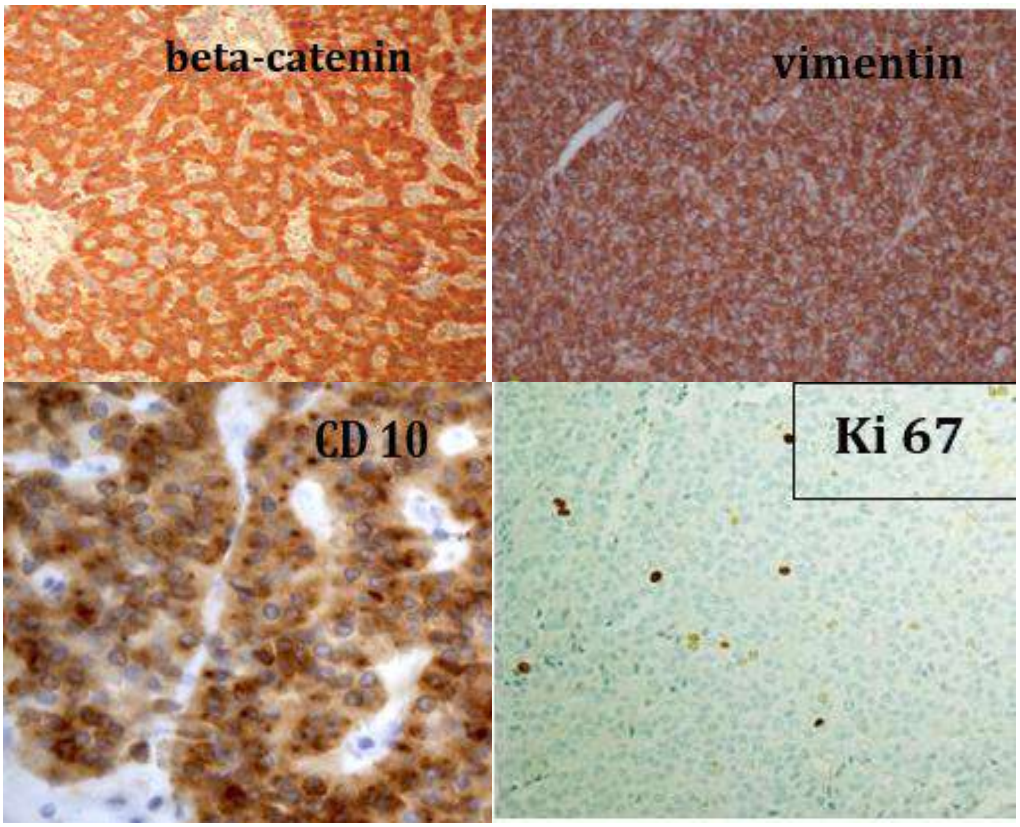


Fig 5 A, B, C D: Immunohistochemistry positive (beta-catenin, vimentin, CD 10) negative for (Ki 67)

Discussion and literature review

This case report on solid pseudopapillary neoplasm of the pancreas (SPN) highlights the enigmatic histogenesis of this neoplasm and the need to distinguish it from more aggressive pancreatic tumors The spectrum of cystic and solid and cystic neoplasms of the pancreas is wide and encompasses at least 14 different tumor types [12], however, the most commonly encountered cystic neoplasms of the pancreas may be classified into five categories: serous microcystic adenoma, mucinous cystic neoplasms (Cystadenoma or cystadenocarcinoma), intraductal papillary mucinous neoplasm cystic neuroendocrine neoplasms and solid pseudo- papillary neoplasm of the pancreas (SPN). Among these uncommon pancreatic tumors, SPN represents an exceedingly rare entity [13]

SPT is a very rare entity that was first described by Frantz in 1959. SPT has been categorized as a borderline tumor of the pancreas by WHO, 1996. An SPT often presents as imprecise nonspecific symptoms. The most common symptoms are mild abdominal pain or discomfort. Patients can also present with fullness associated with nausea and early satiety, which is secondary to a mass effect. Approximately 15% of patients are asymptomatic [14].

Diagnosis with imaging alone is technically challenging in small tumors and in those without cystic component. Other major pancreatic cystic neoplasms. The presence of SPN is highly suggested when certain pathognomonic features are identified on

CT scan: well-defined, encapsulated mass with cystic and solid component, areas of central calcification, necrosis or hemorrhage. Tumors are encapsulated and usually well demarcated. Tumor capsule as well as the solid part enhance after intravenous contrast administration to a degree similar to normal pancreatic tissue during both arterial and venous phases. Calcification found approximately in one third of cases, usually peripheral and less common at the central part of the mass as in our presented case.

Sometimes the exact diagnosis of SPN is not simple. Bektas *et al*. reported a SPN case that the final histological specimen was initially assessed differently by two departments of pathology: one classified the tumor initially as endocrine, the other as a solid pseudopapillary lesion. In our case the guided biopsy diagnosis shows the same diagnosis two department of pathology [15].

The pathologic diagnosis of SPN is made primarily based on the distinct solid and cystic arrangement and typical pseudopapillary characteristics under the microscope [16]. On the cut surface, a variegated manifestation is seen with variable arrangement of solid hemorrhagic and cystic- necrotic parts. The microscopic features of SPN are solid areas which alternate with a pseudopapillary pattern composed of a fibrovascular stalk surrounded by several layers of epithelial cells [17].

Histopathologic criteria of malignancy are not well established but size >5 cm, nuclear atypia, vascular, perineural and invasion of surrounding structures and high proliferation rate may predict aggressive behavior [16].

In our presented case, tumor cells showed similar histopathology features without any criteria of malignancy potential.

Immunohistochemically, SPN cells were typically positive for vimentin, a1-antitrypsin, anti-chymotrypsin, epithelial markers (CK and EMA), CEA, alfa fetoprotein, neuron-specific enolase and progestron receptor. [18, 19]. In presented case the immunohistochemical showed strong positivity for beta-catenin, vimentin, CD 10, and negative for Ki 67.

In our presented case the SPN in pancreatic tail as in most of literatures, but it can be detected in any part of pancreas, Bektas reported a case of SPN in a young woman presented with unspecific complaints in the upper abdomen [15]. They detected a mass in the area of the pancreatic head in the other case presented by Hu and colleagues reported a 19-year old female patient with huge mass in distal of pancreas pushed the stomach. They undertook the patient distal pancreatectomy and splenectomy [13].

Our patient had elective operation distal pancreatectomy with preserving of spleen and not gives rise to a significant deficit of normal pancreatic function.

In recent years, with the advance and maturity of laparoscopic techniques, the unique ad- vantages of minimally invasive could develop the new avenue of SPN treatment.

Sokolov treated 2 SPN children by the application of laparoscopic surgery and achieved good treatment effects [20] The 2 children followed up 6 months and 2 years, respectively, and no tumor recurrence and metastasis were observed.

Petrosyan also believed that, for SPN were located in the body and tail of the pancreas, the surgery strategy of laparoscopic distal pancreatectomy was feasible and safe [21].

However, Fais [22] considered that laparoscopic biopsy or resection of the tumor could result in intra-abdominal spreading of tumor cells due to injected gas.

In most patients, surgical therapy is curative and neither chemotherapy nor radiotherapy should be combined. In the few cases where surgery is not feasible, radiotherapy can be applied since these tumors seem to be radiosensitive [23]. Some study had shown role of gemcitabine and radiotherapy to downstage the tumor [24, 25] Our limitation in this case report is the short time of postoperative fallow up, which was 8 months, it seems if it would be longer the consequence of treatment could be more reliable.

The overall prognosis of SPN of the pancreas is good because of their favorable biologic manifestations. Proper preoperative diagnosis is required since these patients may be definitively cured with sufficient surgical resection.

Conclusion

In conclusion, SPN is a rare pancreatic neoplasm of unclear histogenesis that typically affects young females without significant symptoms. Appearance on imaging is fairly characteristic and may suggest diagnosis, Complete surgical resection of the tumor is the only effective treatment option. SPN should be considered in the differential diagnosis of any solid and partly cystic pancreatic or upper abdominal mass, particularly in young females.

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The authors declare that there is no conflict of interests.

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