

## CASE IMAGE

# A rare pancreatic neoplasm in a 40-year-old male patient

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**Key Clinical Message**

The differential diagnoses of solid pseudopapillary neoplasm of the pancreas include cystic pancreatic neuroendocrine tumor, acinar cell carcinoma, and pancreatoblastoma.

**Abstract**

Solid pseudopapillary neoplasm (SPN) is a low-grade malignant pancreatic tumor which accounts for 0.9%–2.7% of all exocrine pancreatic neoplasms. It predominantly affects young females (90%) and less frequently occurs in male patients. Its prognosis after surgical resection remains excellent. Herein, we report a case of SPN in a male patient.

**KEYWORDS**

abdominal mass, cephalic duodenopancreatectomy, immunohistochemistry, pancreas, pathology, solid pseudopapillary tumor, surgery

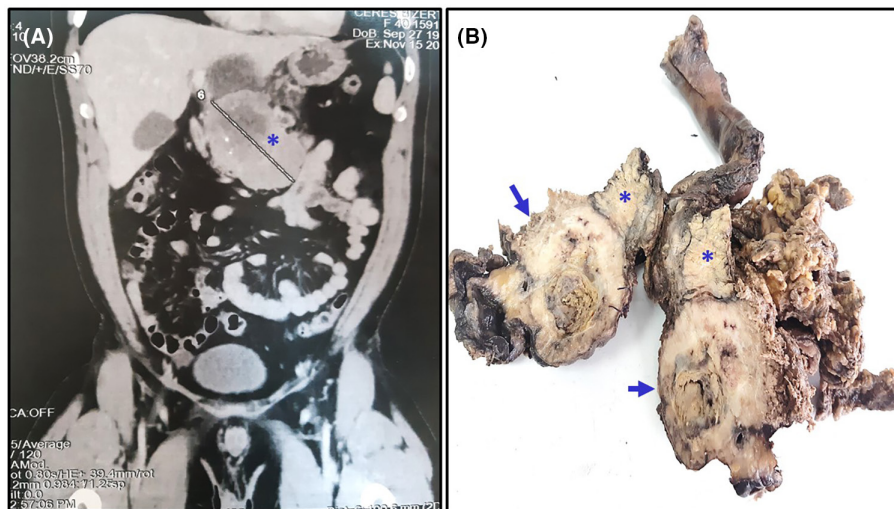
## 1 | CASE IMAGE

A 40-year-old male patient who had no significant past medical history, complained of epigastric and abdominal pain that had been present for the previous 2 months. Physical examination and laboratory tests were normal. Abdominal CT scan revealed a well-defined heterogeneous mass of the pancreas with focal calcifications (Figure 1A). The patient underwent cephalic duodenopancreatectomy. Grossly, the tumor was well-demarcated with a fibrous pseudocapsule and measured 9 × 7 × 3 cm. On cut section, it was solid with focal areas of necrosis, hemorrhage, and pseudocystic spaces (Figure 1B). The wall of the neoplasm contained calcifications and it was well-separated from the adjacent pancreatic parenchyma that it did not invade (Figure 1B). Microscopically, the tumor was encapsulated and composed of poorly cohesive tumor cells forming solid and pseudopapillary structures (Figure 2A,B). In the solid areas, the tumor cells were arranged in sheets and nests, separated by small vessels (Figure 2C). In areas where

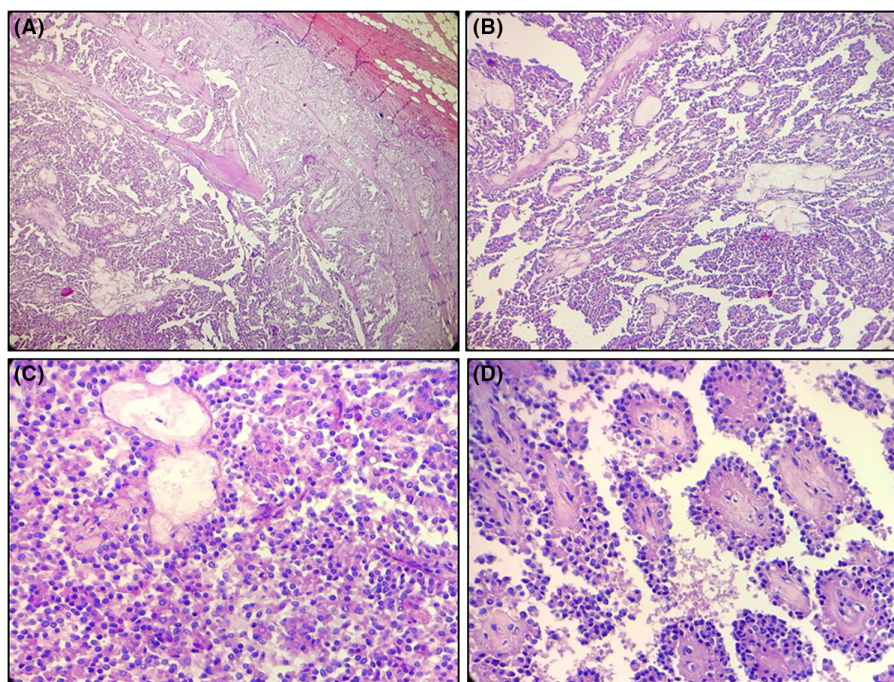
early degenerative changes were more apparent, the tumor cells displayed pseudopapillae with hyalinized fibrovascular cores lined by poorly cohesive tumor cells (Figure 2D). The neoplastic cells were small and monomorphic. Their cytoplasm was eosinophilic or clear. The nuclei were round to oval and focally showed grooves. Mitotic figures were absent. We focally noted the presence of foamy histiocytes (Figure 3A), cholesterol clefts, foreign body giant cells, and calcifications (Figure 3B). There was neither perineural infiltration nor vascular invasion. The peripancreatic lymph nodes were not metastatic. Immunohistochemically, the tumor cells showed positive immunostaining for  $\beta$ -catenin, vimentin (Figure 3C), CD56, CD10, CD99, and progesterone receptors. However, they were negative for chromogranin A (Figure 3D) and cytokeratin. Based on histological and immunohistochemical findings, the diagnosis of SPN was established. Postoperative course was complicated by hemorrhagic shock. Consequently, the patient was transfused. At present, the patient is still being followed up. No adjuvant therapy was administered.

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**FIGURE 1** (A) Abdominal computed tomography scan showing a  $9 \times 7 \times 3$  cm heterogeneous mass of the pancreas with focal calcifications (blue asterisk). (B) Macroscopic examination of the cephalic duodenopancreatectomy specimen revealing a well-demarcated tumor (blue arrow). On cut section, the tumor is solid with focal areas of necrosis, hemorrhage, and pseudocystic spaces. It is well-separated from the adjacent pancreatic parenchyma (blue asterisk) that it does not invade.

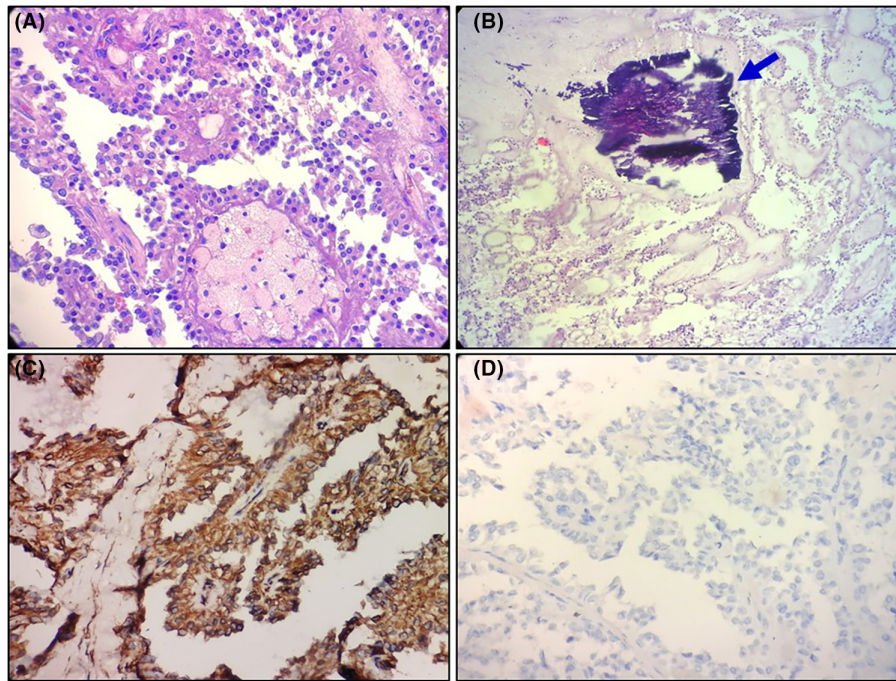


**FIGURE 2** (A) Low-power view of the tumor showing an encapsulated tumor proliferation made of cystic and solid areas. The tumor consists of peripheral solid areas and central pseudopapillary structures. (hematoxylin and eosin, magnification  $\times 40$ ). (B) Low-power view of the tumor showing areas of degenerative changes, in which the tumor cells displayed pseudopapillary formation with abundant microvasculature (hematoxylin and eosin, magnification  $\times 40$ ). (C) Sheets of uniform polygonal epithelioid cells with round or oval nuclei and eosinophilic cytoplasm. Mitotic figures and atypia were not found (hematoxylin and eosin, magnification  $\times 400$ ). (D) The tumor shows pseudo papillae with hyalinized fibrovascular cores lined by poorly cohesive-appearing, cytologically bland tumor cells (hematoxylin and eosin, magnification  $\times 400$ ).

## 2 | DISCUSSION

Solid pseudopapillary neoplasm is a rare pancreatic neoplasm with an uncertain etiopathogenesis.<sup>1,2</sup> Its clinical

presentation is not specific. The definitive diagnosis of SPN relies on histopathological examination coupled with an immunohistochemical study.<sup>1,2</sup> Histologically, SPN is composed of poorly cohesive epithelial cells



**FIGURE 3** (A) Within the tumor proliferation we noted the presence of clusters of foamy histiocytes (hematoxylin and eosin, magnification  $\times 400$ ). (B) Focally, we noted the presence of basophilic calcifications within the tumor proliferation (blue arrow) (hematoxylin and eosin, magnification  $\times 100$ ). (C) The tumor cells showed intense and diffuse positive immunostaining for vimentin (Immunohistochemistry, magnification  $\times 400$ ). (D) The tumor cells showed negative immunostaining for chromogranin A (Immunohistochemistry, magnification  $\times 400$ ).

forming solid and pseudopapillary structures that lack a specific line of pancreatic epithelial differentiation.<sup>2</sup> Resection of the tumor typically offers a favorable prognosis. Even when the tumors are found in the pancreas' head, laparoscopic resections of solid pseudopapillary tumors can be carried out securely and with sufficient excision margins. In the majority of cases, complete resection of SPN is feasible with a minimal recurrence incidence. To prevent tumor rupture, only qualified surgeons should conduct laparoscopic excision of SPN.<sup>3</sup> Postsurgical monitoring is necessary as these tumors can still have a malignant tendency and recurrence risk even after being surgically removed.<sup>3</sup> In conclusion, SPN should be suspected in young males presenting with pancreatic cystic masses. It is crucial for the pathologist to be aware of the tumor's key radiological, histopathologic, and immunohistochemical characteristics in order to differentiate it from other more aggressive pancreatic neoplasms.

#### AUTHOR CONTRIBUTIONS

**Dr. Faten Limaïem** prepared, organized, wrote, and edited all aspects of the manuscript. She performed the gross and microscopic pathologic evaluation of the pathology specimen. **Dr. Mohamed Hajri** participated in the conception and design of the study, the acquisition of data, analysis and interpretation of the data, the drafting of the

article, and revising it critically for important intellectual content and final approval of the manuscript before its submission.

#### ACKNOWLEDGMENT

None.

#### FUNDING INFORMATION

We did not receive funding for this article.

#### CONFLICT OF INTEREST STATEMENT

The authors have no conflict of interest to declare.

#### DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

#### ETHICS STATEMENT

All procedures performed were in accordance with the ethical standards. The examination was made in accordance with the approved principles.

#### CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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**How to cite this article:** Limaiem F, Hajri M. A rare pancreatic neoplasm in a 40-year-old male patient. *Clin Case Rep*. 2023;11:e7387. doi:[10.1002/ccr3.7387](https://doi.org/10.1002/ccr3.7387)