



## Case Report

## Solid pseudopapillary tumour of pancreas

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## ABSTRACT

**Introduction:** Solid Pseudopapillary tumor (SPT) is a tumor with low malignant potential and comprises around 1-2% of all tumors occurring in Pancreas. Pseudocyst of Pancreas, Microcystic adenoma, neuroendocrine tumor and mucinous cystic neoplasm are considered as challenging differentials of SPT. High suspicion followed by accurate diagnosis is key to management because SPT is known for its good prognosis and successive cure rate on surgical resection.

**Case Presentation:** 21-year-old female patient presented with complaints of abdominal pain and dyspnoea. Imaging studies revealed a retroperitoneal tumor. Surgical resection carried out. Gross examination of the specimen showed circumscribed lobular brown mass weighing around 468 gm with a measurement of 15x12x10 cm. On sectioning, the tumor had a variegated appearance containing multiple cysts varying from 1 - 3 cm in size. Microscopic examination revealed an encapsulated cellular neoplasm with individual tumor cells arranged in solid, pseudopapillae and cystic patterns. Tumor cells were round to oval with no distinct nucleoli. Mitotic figures were very few in number. Hyaline globules were noted. Mucinous changes were seen focally. Occasional areas exhibited clear cell change of tumor cells. Cystic areas were filled with hemorrhage. Vascular areas were unremarkable. Normal Pancreatic parenchyma was noted at the tumor edge.

**Conclusion:** Morphology of SPTs are varied ranging from solid to cystic appearance. No definitive morphology is identified to differentiate benign from malignant SPT. SPT are known to mimic a wide range of pancreatic and retroperitoneal lesions that may pose a challenge to diagnostic pathologists as seen in this case.

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## 1. Introduction

SPTs are rare epithelial tumors occurring in Pancreas. Cell of origin is not known. About 2-3% of tumors occurring in pancreas are SPTs. It is known to occur predominantly in females and very low malignant potential.<sup>1</sup> WHO has classified it as borderline malignant neoplasm.<sup>2</sup> Age of occurrence is young age especially 2<sup>nd</sup> decade. The tumor seems to have predilection for African American and Asian women. Since, the tumor has a good prognosis and rarely metastasize, surgical treatment is standardised option. Many

of the tumors are silent and found incidentally. Rising incidence of SPTs is attributed to increasing use of CT and MRI scans.<sup>3</sup> Lichtenstein first reported and described pseudopapillary in the tail of pancreas in 1959.<sup>4</sup> Soon, Frantz described pathology of SPT thoroughly.<sup>5</sup> Several synonyms were used to describe the tumor until WHO named it as solid pseudopapillary tumor in 1996.<sup>2</sup> Many SPTs have been reported as case reports previously. SPTs share radiological and immunohistochemical features with many tumors like neuroendocrine neoplasms, mucinous cystic neoplasms, pseudocyst of pancreas, acinic cell tumors and pancreatoblastomas.<sup>6</sup> Hence, it is important document

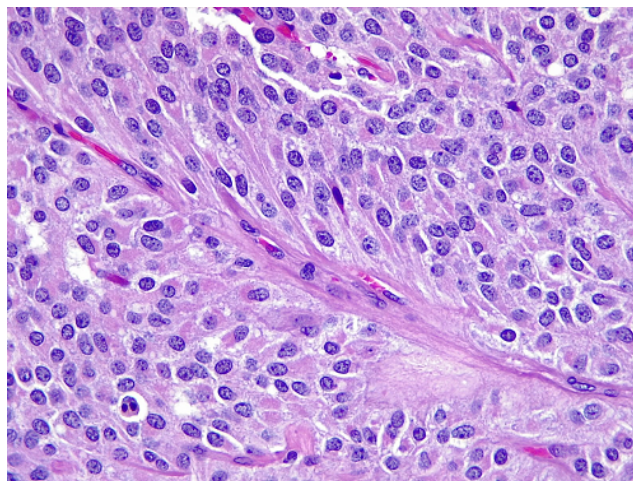
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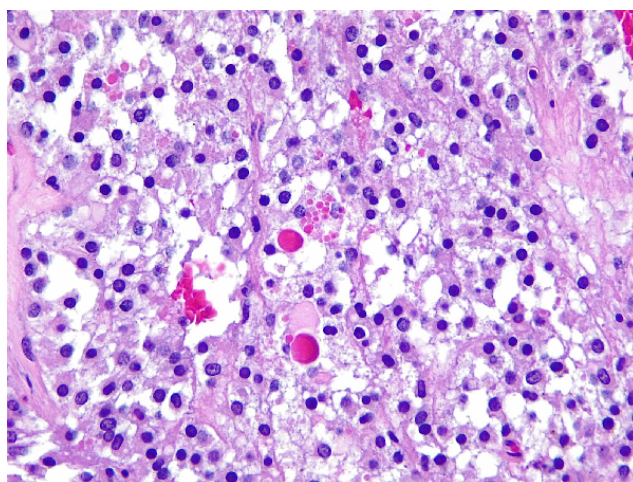
SPTs along with its characteristic features. We present a case of SPT occurring in a 21-year-old female with its pathological features.

## 2. Case Presentation

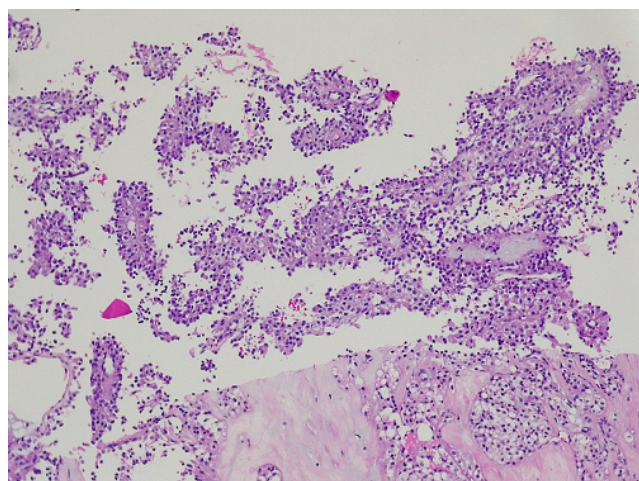
21-year-old patient presented with vague abdominal pain and difficulty in breathing. Physical examination showed a large hypochondriac mass. Radiological examination revealed a retroperitoneal mass. Trucut biopsy and immunohistochemical analysis indicated probable malignant tumor of renal origin. Resection of the retroperitoneal mass was performed. Operation findings showed that the tumor was seen in the lesser sac (right hypochondrium) and was seen to be attached to pancreas and lesser curvature of stomach mesocolon. Tumor was separated from stomach mesocolon and excised along with cuff of pancreas. Specimen consisted of a circumscribed multilobated brownish mass weighing 468 grams and measuring 15x12x10cm. Cut surface showed a variegated appearance including multiple cysts measuring 1 to 3cm in diameter. Microscopically, showed a cellular encapsulated tumour composed of solid, pseudopapillae and cystic areas. The pseudopapillae were covered by several layers of epithelial cells (Figure 1). The nuclei were ovoid with indistinct nucleoli and few mitosis (Figure 2). Hyaline globules (Figure 3) and collections of foamy cells were seen. The thick fibrovascular cores showed prominent mucinous changes (Figure 4). Tumour clear cells were prominent. Extensive areas of haemorrhage and blood clots were seen. The blood vessels and lymphatics were unremarkable. Normal pancreatic tissue was seen at the edges of the tumour.



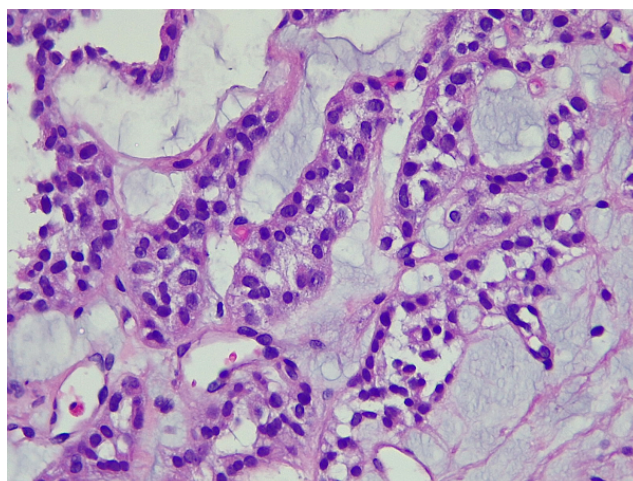
**Fig. 2:** 40X H&E Pseudopapillary pattern



**Fig. 3:** 40X H&E Hyaline globules



**Fig. 1:** 10X H&E Pseudopapillary pattern



**Fig. 4:** 40X H&E Mucinous areas

### 3. Discussion

The most common location of the tumor is the tail of the pancreas, followed by the head, the body and the neck, lastly. Morphology of SPTs are widely varied ranging from solid to cystic appearance. No definitive morphology is identified to differentiate benign from malignant SPT. SPT are known to mimic a wide range of pancreatic and retroperitoneal lesions that may pose a challenge to diagnostic pathologists as seen in this case.

Grossly, SPTs are usually large and well encapsulated, with a variable amount of solid and cystic areas along with hemorrhagic and necrosis. The cystic cavities in SPTs are considered as pseudo cysts as they do not contain an epithelial lining. The cystic spaces are degeneration and necrosis. Microscopically, tumor typically has pseudopapillae with hyalinized or no fibrovascular cores lined by several layers of bland epithelial cells with clear to eosinophilic cytoplasm. Variable amount of mucinous changes can be seen within the core.<sup>7,8</sup> The frequently seen hyaline globules as seen in this case are known to be PAS + diastase resistant. Pseudopapillary pattern is exhibited because of tumor cells degrading and moving away from the blood vessels that look like rosettes.<sup>9</sup> Current case exhibit cells arranged in the form of sheets and cords along with areas of microcysts and hyaline degeneration in the stroma that are specific to SPTs. Tumor cells, if infiltrated into the stroma, did not cause any reaction. The cystic areas often contained necrotic debris, clusters of foamy macrophages, giant cells and blood. Stromal degeneration is often noticed in the form of myxoid change and calcification. Mucinous/myxoid areas in the stroma were noticed as a sign of degeneration in this case.

A variant of SPT known as clear cell variant has been reported previously. This variant has tumor cells with a clear cytoplasm that is attributed to distended endoplasmic reticulum and mitochondria. This clear cell variant of SPT must be distinguished from other tumors with a clear cell morphology that includes clear cell variant of pancreatic endocrine tumor, ectopic adrenal tumors and metastatic renal cell carcinoma and may require immunohistochemical help.<sup>10</sup> Present case showed some tumour cells with clear cell change. Unique immunostaining patterns for beta-catenin (nuclear and cytoplasmic staining) and cyclinD1(over expression) are other molecular markers that can be used to distinguish SPTs from other similar lesions of the retroperitoneum.

### 4. Source of Funding

None.

### 5. Conflict of Interest

None.

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