

## Solid pseudopapillary neoplasm of the pancreas :A rare presentation :A study of two cases.



### Medical Science

KEYWORDS :

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#### Introduction :

Solid pseudopapillary neoplasm of pancreas (SPN) is a rare entity. It is almost exclusively seen in females and occurs in the second or third decades of life<sup>1</sup>. Due to the paucity of the number of cases seen, the natural history of the disease is not fully understood.

There has been a steady increase in the number of diagnosed cases of SPN recently, with more than two-thirds of the total cases described in the last 10 years. This study was undertaken to examine the clinico-pathological characteristics of the disease and to evaluate the outcome of surgical intervention in a tertiary referral cancer centre.

#### CASE REPORT 1:

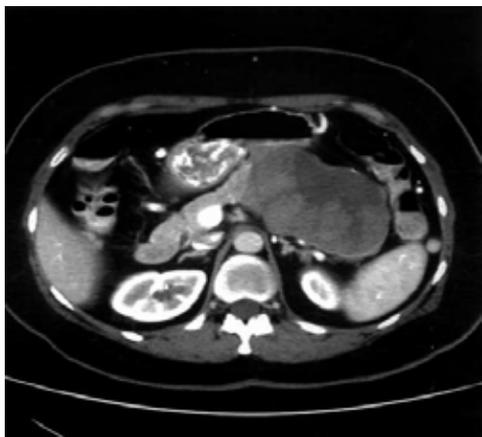
A 18 year old female presented with history of acute abdominal distension with no signs of intestinal obstruction.

Clinical examination revealed massive ascites with a mass felt in the left hypochondrium after ascitic tap.

USG revealed left retroperitoneal mass and guided aspiration cytology showed round cells.

#### Investigations:

CECT showed large intra peritoneal heterogenous mass lesion with few cystic with solid component with gross ascites.



#### MANAGEMENT:

- Therapeutic ascitic tapping to relieve respiratory distress, but Fast re-accumulation of peritoneal fluid noted.
- Diagnostic laparoscopy revealed haemorrhagic fluid with large cystic mass arising from tail of pancreas.
- Resection of the tumor done.

Post operative stay was uneventful.

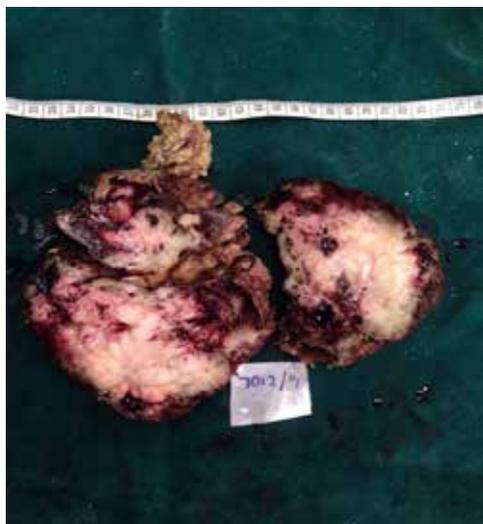
#### Intra op pictures:



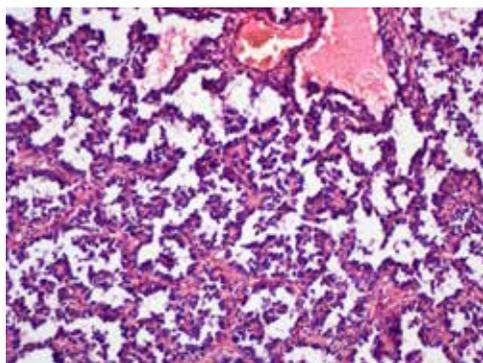
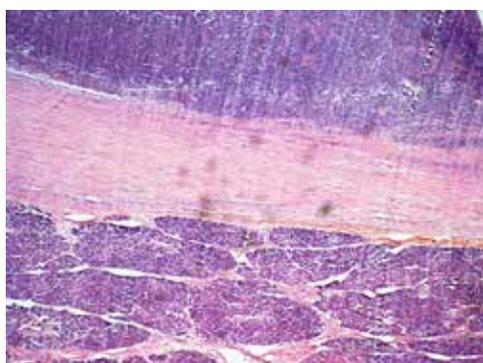
#### Macroscopic picture:

- Large grey mass of size 16 x 14 x 6 cm with multiple cystic spaces and areas of hemorrhagic fluid. borders well defined showing no infiltration.





**Microscopy:**  
**Tumor cells with sheets, clusters and pseudopapillary pattern with increased mitosis and necrosis.**



Histopathological pictures : description explained above.

**CASE REPORT 2:**

A 32 year old female patient was admitted on 07-05-2015 with complaints of:

Pain abdomen since one year, insidious in onset, dull aching type, radiating to the back and increases on doing household work.

There was no history of jaundice vomiting, haemetemesis, melena, No history of loss of weight and appetite .

Patient was diabetic since one year on treatment.

**Past history:**

Patient had underwent caesarean section 3 times and underwent surgery for incisional hernia 4 years ago .

- General physical examination patients vitals were stable and no significant pathology noted on GPE.
- on examination:

**Inspection:**

Shape of the abdomen was distended and there were previous surgery scars which were healed by primary intention and there was no mass seen.

On palpation also no mass palpable and there was no organomegaly.

Other systems examination was normal.

**Blood investigations**

HB was 13.8 gm% ,LFT was normal and other investigations were normal.Upper GI Endoscopy : Normal study .

**Usg abdomen and pelvis :** A large heterogenous mass noted in the head of the pancreas of 10×6 cms

- ? Carcinoma head of pancreas
- ? GIST

**CT abdomen and pelvis:**

A well defined circumscribed mass lesion in the head of the pancreas with cystic and solid components with no fat stranding suggestive of Carcinoma head of Pancreas.

**TREATMENT:**

- Patient Underwent Whipples procedure.
- On table tumor was arising from head of the pancreas and second part of duodenum and was hard in consistency and the tumor content was necrotic material.



**Histopathology:**

**Gross:**

**GROSS-SPPT**





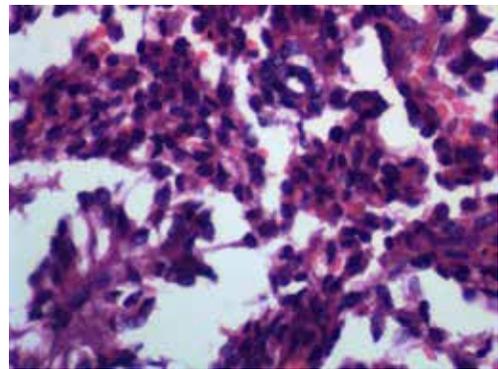
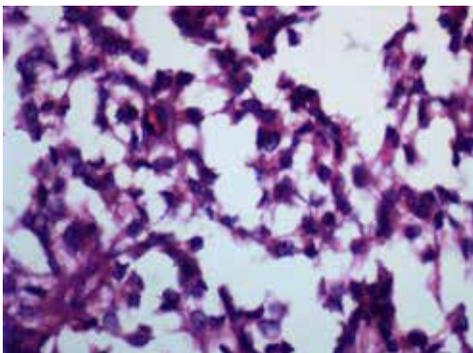
**GROSS-SPPT**



The tumor may occur anywhere in the pancreas and presents macroscopically as a round, deceptively well demarcated lesion, measuring 2 to 17 cm in diameter (average 8 cm). Sectioning demonstrates a solid mass with pseudocystic areas, and hemorrhage is common.

**SPPT-MICRO:**

Micro- solid portions contain sheets, cords, and nests of uniform, rather small, and fairly round cells; this organoid appearance can mimic a neuroendocrine tumor. The cytoplasm is eosinophilic or vacuolated, containing clustered large, PAS-positive hyaline globules. Nuclei appear round to oval and have finely dispersed chromatin and inconspicuous nucleoli. Some nuclei contain a groove or indentation. Mitoses are rare. Solid portions characterized by rich, delicate vascular network. Cells farthest from the vessels undergo degeneration, causing the remaining cells around the vessels to form pseudorosette or pseudopapillary patterns. The cystic zones result from more extensive degenerative changes, hemorrhage, cholesterol granulomas, aggregates of foamy histiocytes.

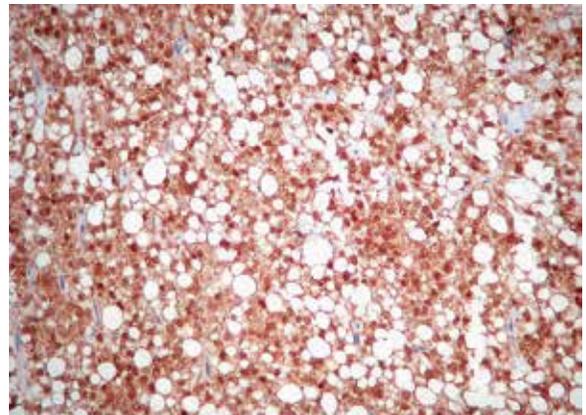


**SPPT-IHC**

Positive for  $\alpha$ 1-antitrypsin, NSE, CD56, CD10,

$\beta$ -catenin, Galectin3, Claudin2&5, Progesterone nuclear receptor, vimentin, and Some faintly express synaptophysin and cytokeratin. Positive staining for chromogranin or pancreatic enzymes, however, is never seen.

**IHC-  $\beta$ -catenin- nuclear positivity**



**DIFFERENTIAL DIAGNOSIS:**

SPPT histologically similar to those in the pancreas have been rarely described outside the pancreas, particularly in the ovary.

D/D of SPPT is primarily versus Pancreatic neuroendocrine tumors and Acinar cell carcinomas.

**Discussion:**

SPN is predominantly encountered in young, female patients (first three decades of life), but has also been reported in males and in children<sup>2</sup>.

These tumours may be discovered by chance during diagnostic imaging procedures or may be suspected in the presence of an asymptomatic palpable mass in young women. Depending on the tumour position (head, body or tail of the pancreas), the differential diagnosis includes adrenal mass, pancreatic endocrine tumour, liver cyst or tumour, or a pseudocyst<sup>3</sup>.

CT scan, ultrasonography (US) and endosonography (EUS) have been used with variable success in diagnosing SPN. CT scan and EUS are more sensitive and specific and have shown more accuracy in diagnosing SPN. Magnetic resonance imaging (MRI) can be diagnostic. Typically, a large, well-defined, encapsulated lesion with heterogeneous high or low signal intensity on T1-weighted, heterogeneous high signal intensity on T2-weighted, and early peripheral heterogeneous enhancement with progressive fill-in is found on gadolinium-enhanced dynamic MRI. These features help differentiate this rare tumour from other pancre-

atic neoplasms<sup>5</sup>. In our series, we relied on CT imaging for the pre-operative work-up.

FNAC has been used for the preoperative cytological diagnosis of SPN. The cytology specimen is usually highly cellular and is characterized by the presence of epithelioid cells that present singly or in aggregates containing fibrovascular cores. No evidence of pleomorphism or mitotic activity is seen in the cells. The most conclusive criterion for identification of SPN is the pseudopapillary arrangement with bland appearing tumour cells. EUS-guided FNAC has been reported, and this can help in correctly diagnosing SPN pre-operatively<sup>4,5</sup>. The histogenesis of these tumours is unknown but they possibly originate from the primordial cells and lack definite endocrine and exocrine differentiation.

On gross examination, SPN is a well encapsulated tumor. On cut section it shows solid and cystic areas with necrotic and haemorrhagic patches. Some tumours also demonstrate firm, fibrotic regions within the tumour. On microscopy, there are solid areas composed of polygonal epithelioid cells with intervening stroma. There is evidence of cellular degeneration. Aggregates of foamy histiocytes, cholesterol clefts and cytoplasmic vacuolization can be seen<sup>6</sup>. Despite being an encapsulated tumour, the microscopic interface between tumour and adjacent normal pancreas does show an infiltrative growth pattern.

Immunohistochemical studies have shown that SPN is reactive for vimentin, antitrypsin, cytokeratin, S-100 protein and neuron-specific enolase. Flow cytometry shows aneuploidy. C-Ha-as oncogene presumably is linked to the development of the tumour. No pathologic factor is of proven prognostic importance<sup>7</sup>. Laboratory values are not contributory, although a few cases do show raised levels of CA19-9.

SPN is considered to be a tumour of low-grade malignant potential. The logical conclusion is that complete surgical excision is the best option for patients who have SPN. Thus surgery should always be attempted in a suspected case of SPN even if it implies that major resections (like pancreaticoduodenectomy along with adjacent organ resection) have to be performed<sup>7</sup>.

A local recurrence rate of 6.2% is reported in cases treated by radical surgical excision, and hepatic or Krukenberg-type distant metastases develop in 5.6% of cases<sup>4</sup>. Few authors have reported an increased rate of resectability after chemotherapy<sup>8</sup>. There are few case reports which have shown a survival benefit with radiotherapy. However, most authors agree that aggressive surgical resection is the best modality of treatment for achieving curative results and a better long-term survival.

A recent series from Memorial Sloan-Kettering Cancer Center, New York, USA, recommends complete surgical excision with even metastatectomy if required<sup>5</sup>. These authors conclude that long-term survival improves with complete surgical resection of primary with metastectomy for synchronous or meta-chronous lesions.

### Conclusions

A high index of clinical suspicion is necessary to suspect and diagnose SPN. This diagnosis should be borne in mind when young female patients present with a pancreatic mass. CT scan and EUS are valuable pointers to the pre-operative diagnosis. FNAC appears to be of value in the specific diagnosis of SPN. Surgical excision offers the best chance for cure and should always be attempted irrespective of the magnitude of resection involved. Patients with SPN have an excellent prognosis after surgical excision.

## REFERENCE

- Compagno J, Oertel JE, Kemzar M. Solid and papillary epithelial neoplasm of the pancreas, probably of small-duct origin: a clinico-pathological study of 52 cases (Abstract) *Lab Invest.* 1979;40:248-9. | 2. Jagannath P, Bhansali MS, Murthy SK, Mohandas KM, Swaroop VS, DeSouza LJ. Solid and cystic papillary neoplasm of pancreas-a report of seven cases. *Indian J Gastroenterol.* 1994;13:112-14. | 3. Mao C, Guvendi M, Domenico DR, Kim K, Thomford NR, Howard JM. Papillary cystic and solid tumors of the pancreas: a pancreatic embryonic tumor? Studies of three cases and cumulative review of the world's literature. *Surgery.* 1995;118:821-8. | 4. Martin RC, Klimstra DS, Brennan MF, Conlon KC. Solid-pseudopapillary tumor of the pancreas: a surgical enigma? *Ann Surg Oncol.* 2002;9:35-40. | 5. Canzonieri V, Berretta M, Buonadonna A, Libra M, Vasquez E, Barbagallo E, et al. Solid pseudopapillary tumour of the pancreas. *Lancet Oncol.* 2003;4:255-6. | 6. Trivedi N, Sharma U, Das PM, Mittal MK, Talib VH. FNAC of papillary and solid epithelial neoplasm of pancreas. A case report. *Indian J Pathol Microbiol.* 1999;42:369-72. | 7. Stommer P, Kraus J, Stolte M, Giedl J. Solid and cystic pancreatic tumors. Clinical, histochemical, and electron microscopic features in ten cases. *Cancer.* 1991;67:1635-41. | 8. Cantisani V, Mortele KJ, Levy A, Glickman JN, Ricci P, Passariello R, et al. MR imaging features of solid pseudopapillary tumor of the pancreas in adult and pediatric patients. *AJR Am J Roentgenol.* 2003;181:395-401. |