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Case Report

Pathology Section

Solid Pseudopapillary Tumour of Extrapancreatic Origin Presenting as Mesenteric Cystic Mass: A Diagnostic Dilemma

SUDIPTA CHAKRABARTI[1], SUMAN GHOSH[2], RANU SARKAR[3]

ABSTRACT

Solid Pseudopapillary Tumour (SPT) is a rare and distinctive pancreatic exocrine neoplasm. Even Rarely, such primary SPT may originate from ectopic pancreatic tissues. We are hereby presenting one such unique case, where a 50-year-old female presented with pain and a mid-abdominal lump. Radiology revealed a well-defined outline located adjacent to the tail of pancreas. The excised mass was 19×14×7cm in dimension having zones of haemorrhage, necrosis and cystic spaces filled with necrotic debris. Microscopic examination confirmed the diagnosis of SPT. SPT originating in extrapancreatic location may mimic an ovarian cystic tumours or mesenteric cysts, its proper identification is crucial.

Keywords: Extrapancreatic tumour, Intra-abdominal lump, Pancreatic exocrine tumour

CASE REPORT

A 50-year-old female presented with pain in the abdomen for three months. On clinical examination, a 10cm diameter intra-abdominal lump was noted having a smooth surface. There was no pallor, jaundice, clubbing, oedema, hepatosplenomegaly or lymphadenopathy.

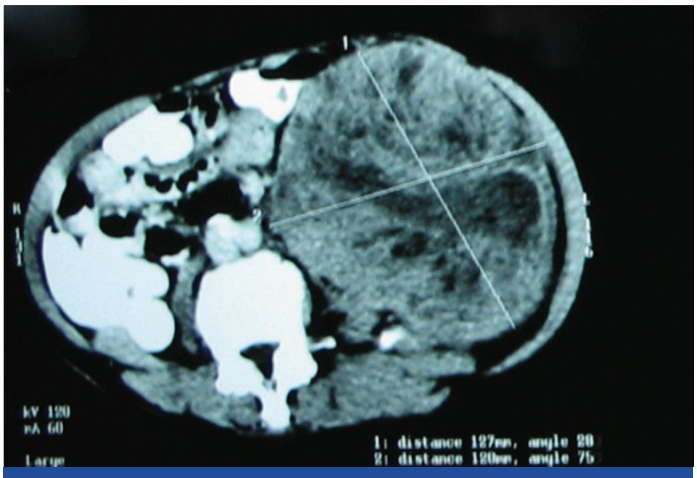
Routine hematological investigation revealed haemoglobin level of 12.3gm/dl, TLC of 10,150/cmm, with a DLC of N68 L26 M01 E05 B00. RBCs in peripheral blood smear were normochromic and normocytic. Platelet count was 186,000/cmm and ESR was 63 mm /hour. Her fasting blood glucose was 96mg/dl, urea 20.5 mg/dl and creatinine 1.1 mg/dl. All parameters of liver function test were within normal limit.

Ultrasonography (USG) detected a mass in the left mid – lower abdomen measuring 13.9x12.1x8.7cm having solid and cystic areas. Computer Tomographic (CT) scan done subsequently revealed a mixed attenuated mass having well defined outline located high up in the left lumbar region adjoining the tail of pancreas [Table/Fig-1]. There were areas of calcification. No enlarged lymph nodes or ascites was noted. Ovaries were not separately visualized. Radiological differential diagnoses were mesenteric cystic neoplasm, pancreatic cystic neoplasm and ovarian cystic neoplasm.

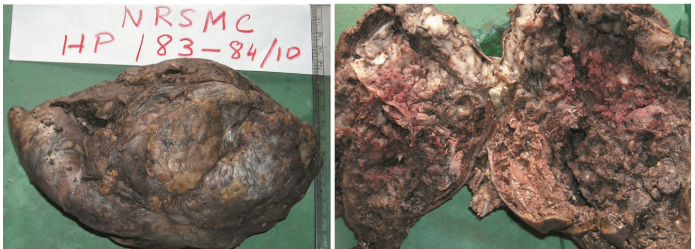
A decision of surgical excision of the mass was taken based on clinical and radiological observation. Surgical exploration revealed that the mass was located in the mesentery adjacent to the tail of pancreas but was completely separated from it. The mass was excised and sent for histopathogical examination. The resected specimen measured 19x14x7cm in dimension having lobulated surface with encapsulation. On opening, inner surface of the mass had a light brown colour having solid areas, regions of haemorrhage, necrosis, and necrotic debris in cystic spaces [Table/Fig-2]. Microscopy revealed an encapsulated cellular tumour comprising of pseudopapillae covered with layers of epithelial cells having ovoid nuclei, indistinct nucleoli and occasional mitosis. The thick fibrovascular core show mucinous change. Hyaline globules were noted. Areas of haemorrhage and cystic changes were identified [Table/Fig-3]. The final histopathological diagnosis was Solid Pseudopapillary Tumour (SPT). A two year follow up of the patient showed no evidence of local spread or metastasis.

DISCUSSION

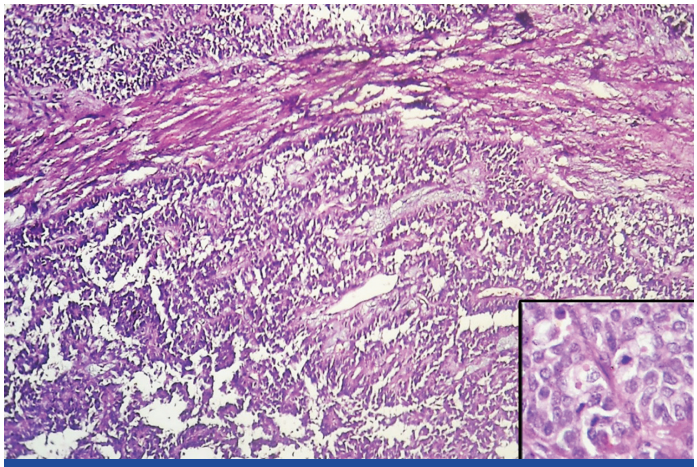
Solid Pseudopapillary Tumour (SPT) is quite a distinctive and rare pancreatic exocrine tumour that usually originates in the body or tail of pancreas and has a low malignant potential. Only about 718 cases were reported till 2005 [1]. It was first described by Frantz [2]. Even rarer is the occurrence of primary SPT in mesocolon, omentum, retroperitoneum, gastrointestinal tract and ovary where it presumed to develop from the ectopic pancreatic tissue [3]. Such extrapancreatic tumour poses diagnostic problems when it occurs in elderly persons. Uniqueness of the present case is that



[Table/Fig-1]: Computer Tomographic (CT) scan showing a mass having well defined outline and mixed attenuation near the tail of pancreas.



[Table/Fig-2]: The resected mass measured 19x14x7cm (left image) and the inner surface was light brown in colour having solid areas, zones of haemorrhage and cystic spaces filled with necrotic debris (right image).



[Table/Fig-3]: The tumour comprising of pseudopapillae covered with layers of epithelial cells thick fibrovascular cores showing mucinous changes (H&E, x100). The inset showing epithelial cells having ovoid nuclei, indistinct nucleoli, scattered mitosis and occasional hyaline globules (H&E, x400).

SPT has exceptionally originated outside the pancreas resulting in a wide clinical and radiological differential diagnosis including an ovarian cyst.

Different nomenclatures imparted to this tumour include solid and cystic neoplasm, Frantz's tumour and 'solid pseudopapillary tumour' of the pancreas by the World Health Organization in the year 2000. Histopathological criteria of malignant SPT are presence of angio-invasion, perineural invasion or deep invasion into the surrounding pancreatic parenchyma [4].

Extra pancreatic site of origin of SPT is rarely described in the literature. A search for cases using PubMed since 1990 revealed only 13 instances of extrapancreatic SPT till 2013 [3]. The extrapancreatic origin of SPT is explained by the presence of ectopic pancreatic tissue having no structural connection to the normal pancreas [5,6]. Recently, Guo et al., has reported one case each of an extrapancreatic and a metastatic SPT in 2016 [7]. Only a few cases of SPT originating from the retroperitoneum have been so far reported in the literature till 2016 [3,7].

Intra-abdominal cystic or solid and cystic masses can generate profound diagnostic confusion if origin could not be discerned by clinical or radiological parameters. Kurtz et al., in a review of reported cases of mesenteric and retroperitoneal cysts in the English literature noted that correct pre-operative diagnosis was achieved in only 30 out of 122 patients (about 25%) because of diverse clinical presentation [8]. Mete et al., reported diagnostic confusion in a case of primary pancreatic SPT with multiple metastasis in omentum [9], peritoneum and both ovaries, which was ultimately resolved by histopathological study. In another case of pancreatic SPT, radiological differential diagnosis offered was mesenteric desmoid tumour or papillary cyst adenoma of the pancreas which was similarly resolute by histopathological evaluation [10].

Pseudocyst constitutes 80% of cystic lesions of pancreas, while primary cystic neoplasm like serous cystadenoma, mucinous cystic tumour and SPT constitute the rest. In histopathological study, diagnosis of these lesions is mostly done by morphology in routine H&E stain rather than by immunohistochemistry [11].

Nishihara et al., compared the histological features of three metastasizing and 19 nonmetastasizing solid pseudopapillary neoplasms [12], and found that venous invasion, degree of nuclear atypia, mitotic count and prominence of necrobiotic cell were associated with malignancy. However as metastasis even can occur in absence of these histopathological features, a morphologically benign SPT must be categorized as lesions of uncertain malignant potential.

A complete surgical excision of these tumours would prevent recurrence in more than 95% of cases, emphasizing its excellent prognosis. A two year follow up of the present case was unremarkable.

CONCLUSION

Solid Pseudopapillary Tumour (SPT) occurring in an elderly woman in an extrapancreatic location is exceptional. SPT originating in extrapancreatic location may mimic ovarian cystic tumours and mesenteric cyst. Precise histological diagnosis is essential for correct management.

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PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of Pathology, ESI PGIMSR, Manicktala, Kolkata, West Bengal, India.
2. Assistant Professor, Department of Pathology, NRS Medical College and Hospital, Kolkata, West Bengal, India.
3. Professor, Department of Pathology, NRS Medical College and Hospital, Kolkata, West Bengal, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Sudipta Chakrabarti,
Associate Professor, Department of Pathology, ESI PGIMSR, Manicktala,
54 Bagmari Road Kolkata-7000054, West Bengal, India.
E-mail : sudiptach@gmail.com

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