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Case Report

Surgery Section

Pseudopapillary Tumour of the Pancreas in an Adult Female

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ABSTRACT

Pseudopapillary tumour of the pancreas is a rare tumour, seen usually in young adolescent females. The occurrence of such tumour in adult middle-aged female, as depicted in this case, is an extremely rare occurrence. This tumour is classified as a tumour of low malignant potential with rare metastasis. Surgery is the treatment of choice for such a tumour. A 50-year-old lady presented with complaints of a painless lump in abdomen, progressively increasing in size, since past five months. The patient underwent a Contrast Enhanced Computed Tomography (CECT) scan of the abdomen, which was suggestive of a pancreatic pseudopapillary tumour with compression of surrounding structures. Tumour excision with an adequate margin, distal pancreatectomy and splenectomy were done. The diagnosis was confirmed by the histopathology report. Patient had an uneventful postoperative stay and was subsequently discharged.

Keywords: Distal pancreatectomy, Surgical gastroenterology, Surgical oncology

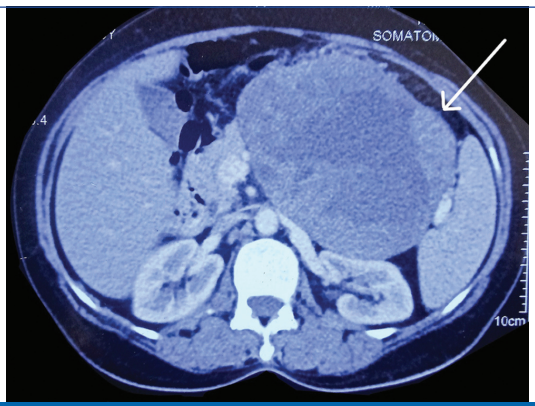
CASE REPORT

A 50-year-old female presented with a painless, progressively increasing lump in left upper abdomen for the past five months. This was associated with anorexia and weight loss since past two months. There were no complaints of pain or altered bowel and bladder habits. There was no previous history of hospital admission or any surgical or dental procedure that the patient underwent. Patient did not suffer from Hypertension, Diabetes Mellitus, Asthma or Tuberculosis. There was no history of a similar complaint in the first- or second-degree family members. General physical examination showed no pallor, icterus or lymphadenopathy. The patient was normo-tensive and the pulse rate was normal. Abdominal examination revealed a non-tender 15×10 cm lump occupying the left hypochondrium, umbilical and left lumbar region which was moving with respiration. The surface was nodular with variable consistency.

The patient's haemoglobin was 11.2 gm/dL and all other biochemical parameters were within normal limits. CECT abdomen scan revealed 13.6×11.2×19.7 cm heterogeneously enhancing lobulated lesion arising from body and tail of pancreas with necrotic areas within. The lesion was lying posterior to the body of stomach reaching up to splenic hilum and causing lateral displacement of large bowel loops. The mass was causing displacement of the splenic artery anteriorly and the splenic vein posteriorly. Inferiorly, it was reaching up to left iliac fossa with preserved fat planes with surrounding structures and with few areas of calcifications within the lesion, features suggestive of Pseudopapillary tumour of the pancreas [Table/Fig-1,2]. Serum CA19.9 and CEA levels increase significantly in patients with pancreatic malignancy but are more of prognostic value rather than diagnostic. Nevertheless, a significantly high value would suggest a malignancy, possibly of pancreatic origin in this case. The serum CA19.9 and CEA levels were normal in our patient.

Exploratory laparotomy was carried out and the tumour was seen arising from the tail of pancreas and densely adhered to the spleen. The patient underwent distal pancreatectomy and excision of mass with splenectomy [Table/Fig-3]. A left subdiaphragmatic drain was placed before closure of the abdomen. The resected specimen consisted of the tumour (20x15x10 cm), distal pancreas (6x3x2 cm), spleen (17x9x4 cm) and omentum (30x10x2 cm). Triple vaccine was administered two weeks after the surgical procedure.

Postoperative period was uneventful. The abdominal drain was removed on postoperative day four when the output was reduced

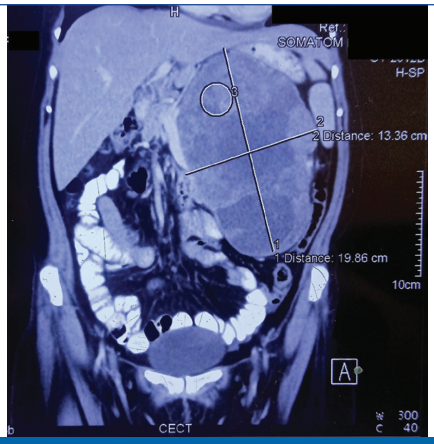


[Table/Fig-1]: CECT abdomen axial section showing tumour (white arrow) adherent to the spleen and compressing the left kidney.

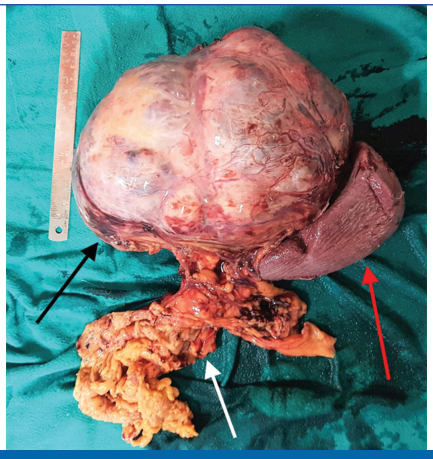
to 30 mL and, subsequently she was discharged on the 5th postoperative day. The patient was fully tolerating oral meals and the bowel movements had returned. Histopathological examination report described the origin of tumour from the tail of pancreas. The gross cut section showed partly solid, partly cystic and partly haemorrhagic regions within. The resected margins were clear of the tumour cells by 2 cm. An R0 resection was achieved. Microscopic examination showed the tumour to be composed of solid nests of cells arranged as papillae with few cells showing nuclear groove. Few hyaline globules were also identified. Focal necrosis and areas of haemorrhage were also present [Table/Fig-4]. Hilum of spleen was free from tumour and spleen showed no tumour deposits; only focal areas of congestion were present in the omentum and spleen. Immunohistochemistry panel done at our centre showed positivity for CD10, CD56 and vimentin [Table/Fig-5,6]. Patient has been on regular follow-up since discharge from the hospital and is doing well. A CECT abdomen is planned after six months of the surgery. An ultrasonography of the abdomen and pelvis was done at the four-month visit and shows no abnormality or recurrence.

DISCUSSION

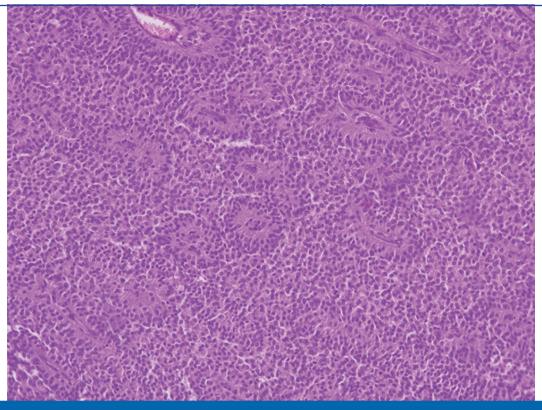
Solid Pseudopancreatic Tumour (SPT) is a relatively rare neoplasm contributing only 0.2-2.7% to exocrine pancreatic malignancies and approximately 9% of all cystic pancreatic malignancies [1,2]. It was first reported by Frantz in 1959; therefore, it is also called as Frantz tumour [3]. It has various names like papillary epithelial neoplasm of



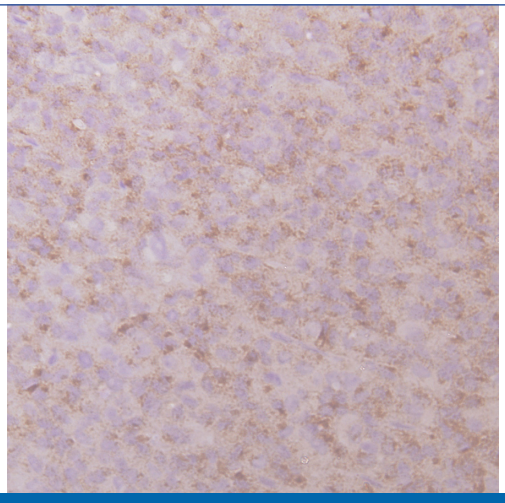
[Table/Fig-2]: CECT abdomen coronal section showing the tumour location and dimensions.



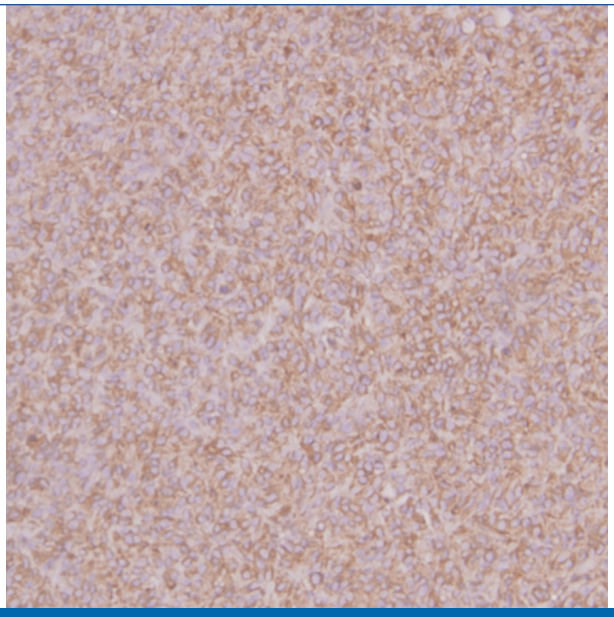
[Table/Fig-3]: Resected gross specimen of tumour (black arrow) with distal pancreatectomy (white arrow) and spleen (red arrow).



[Table/Fig-4]: Photomicrograph Haematoxylin and Eosin (200x): Showing pseudopapillae with central vascular core lined by discohesive cells showing moderate amount of eosinophilic cytoplasm and prominent nuclear grooving.



[Table/Fig-5]: Immunohistochemistry for CD 10 (400x): Tumour cells stain positively.



[Table/Fig-6]: Immunohistochemistry for Vimentin (200x): Tumour cells stain positively.

pancreas, papillary cystic neoplasm, Hamoudi tumour and currently, solid pseudopapillary tumour (WHO nomenclature) [4].

According to some studies, the tumour occurs approximately 6 to 10 times more in the female gender as compared to males [1,5,6]. This tumour most commonly occurs in young females in 2nd and 3rd decade or the 3rd and 4th decades of life [7]. An extensive review of 2744 patient of SPT, reported in literature between 1961 and 2012, showed that the average age of presentation is 28.5 years [8]. The patient described herein was 50-year-old, which is not a common age for the occurrence of this tumour.

The most frequent presenting complaints with which a patient of SPT would report to the clinician are pain and mass in the abdomen. The third most common presentation is an asymptomatic patient [1,8]. Discomfort, vomiting, nausea, post trauma, fever and jaundice; in the order of occurrence, are the less frequent symptoms that a patient may present with. Less than 1% of the patients present with anorexia, weight loss, pseudocyst or other symptoms [1]. Rare presentations include post blunt trauma abdomen. In one such reported case, the patient's CT scan report was suggestive of either pancreatic pseudocyst or duodenal haematoma and the diagnosis was eventually confirmed after one year [9]. Our patient presented to the outpatient department with the complaints of a painless, rapidly increasing lump in the abdomen associated with anorexia and weight loss (5 kg over the last two months). This might be attributed to the mass effect on the gastrointestinal tract of the rapidly growing tumour. However, these tumours may even remain indolent and not increase in size for as long as three years [10]. In contrast, our patient presented with a very large mass in the abdomen, which had rapidly increased in size over the last five months. Different studies have shown the mean tumour size in the patient of SPT to be 8.6 cm [8], 6 cm [6], 6.5 cm [3] and 6.08 cm [1], respectively. However, the tumour described in this report had a maximum diameter of 20 cm.

SPT is a tumour of low malignant potential; however, metastasis have been reported in 15% of cases with the liver being the most common site followed by regional nodes and peritoneum [11]. Malignant SPTs have been described by features of angio- or perineural invasion or extension into the normal pancreas on histopathology [4]. Local or distant metastasis or vascular invasion is not a contraindication for surgical resection [12]. The goal of surgical resection should be R0 resection and an en bloc excision of the affected adjacent organs [13]. It is advocated that metastasis is to be treated with debulking procedures and liver metastasis was treated with enucleations or lobectomies [1]. The tumour commonly arises from the tail of pancreas (35.9%) followed by the head (34%) [1]. Extremely rare though, the tumour may be present at sites other than the pancreas. According to Wu H et al.,

who discussed the first case of mesenteric SPT in their article, only 16 cases of extra-pancreatic SPT have been reported till 2017 [14]. These tumours have been found in the retroperitoneum (4 cases), omentum (2 cases), ovary (6 cases), mesocolon (1 case), mesentery (1 case), stomach (1 case) and duodenum (1 case) [15].

The clinical presentation of extra-pancreatic SPT is similar to pancreatic SPT. In a case of retroperitoneal SPT localized at the site of left adrenal gland, CT scan depicted a 6 cm solid encapsulated mass with low attenuation and again, having solid and cystic contents. The patient underwent a laparoscopic left adrenalectomy and the diagnosis was established by the histopathological examination report. The report also revealed the presence of ectopic pancreatic tissue within the tumour [15]. It is hypothesized that entrapment of some of the genital ridge omnipotent cells occurs within the pancreatic anlage during development, while the other omnipotent cells follow the normal ovarian descent. SPT may then originate from anywhere along this route. Therefore, SPT may be influenced by the female hormones [16]. SPT typically shows a peripheral enhancement, of similar Hounsfield units as that of the pancreas, in arterial and venous phases on CECT. This feature helps to rule out pancreatic adenocarcinomas (hypo-attenuation on venous phase) and pancreatic neuroendocrine tumours (enhancement on arterial phase). CECT scan shows hypoattenuation on unenhanced images and remain the same during pancreatic and portal venous phase. Calcifications are commonly seen [17]. Other malignancies which must be ruled out include pancreatic adenocarcinomas, pancreatic neuroendocrine tumours, serous cystadenoma, intraductal papillary mucinous neoplasm and acinar carcinoma.

Preoperative diagnosis can be established by endoscopic ultrasound guided fine needle aspiration [12]. Ultrasonography is non-specific for establishing a diagnosis and therefore, computed tomography and magnetic resonance imaging are commonly used. These tumours can be small (≤ 3 cm) and large (> 3 cm). Large SPTs are heterogeneous, consisting of both, solid and cystic component. MRI can diagnose haemorrhage in the lesions. Tumour is hypointense on T1 and T2 images. Small lesions are homogeneous [18]. Histopathological examination is diagnostic of the tumour; which shows solid sheets of neoplastic cells with variable areas of discohesive tumour cells surrounding delicate blood vessels creating a pseudopapillary appearance. Tumour cells consistently express vimentin, CD 10, CD 56, progesterone receptors, neuron specific enolase, alfa-1-antitrypsin and alfa-1-antichymotrypsin. β-catenin and CD10 show membrane staining in SPT cells. Synaptophysin is strongly positive for neuroendocrine tumours of the pancreas but is not expressed in SPT [19]. Surgery is the main stay of treatment and is associated with excellent prognosis with approximately 95% of 5-year survival rate [20].

CONCLUSION

Solid Pseudopapillary Tumour is a rare pancreatic tumour, especially in middle aged females. Numerous aspects of the case discussed in this report cause it to deviate from the classical features of SPTs. These include age of 50 years, complaints of a large abdominal mass with anorexia and weight loss and a short history of a rapidly growing mass reaching to very large size (20 cm). Radiological investigations can aid in diagnosis preoperatively, but confirmation is by histopathology only. Complete surgical excision is the main stay of treatment for such tumours.

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