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A Large Solid Pseudopapillary Tumour of the Body and Tail of Pancreas: A Rare Entity Treated with Combined Modality

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Authors' contributions

This work was carried out in collaboration with all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Solid Pseudopapillary Tumour (SPPT) of the pancreas are rare tumours, accounting for only 1-2% of all pancreatic neoplasms. Increased rate of detection of this neoplasm in the last three decades is probably due to the increased awareness and the increased provision of imaging. Though surgery is the mainstay of its treatment, single agent chemotherapy with Inj. Gemcitabine is considered to be of choice for the adjuvant therapy in case of large tumours with high risk features.

We report here a case of a huge Solid Pseudopapillary Tumour (SPPT) of the Body and Tail of Pancreas [Stage pT2N0M0] in a 23 year old lady presenting with vague pain abdomen and vomiting. At first, it was suspected by Magnetic Resonance Cholangio-Pancreatography (MRCP) and finally it was diagnosed by histopathological examination of specimen obtained by surgery. Patient is now being treated with single agent adjuvant chemotherapy (Gemcitabine) and is completely asymptomatic.

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Keywords: Pseudopapillary; SPPT; SPT; pancreatic tumour; solid tumour; carcinoma pancreas.

1. INTRODUCTION

Solid pseudopapillary tumour (SPPT) of the pancreas represents less than 1-2% of all exocrine pancreatic tumours [1,2]. More than 90% SPPTs occur in young women, with a median age of 20–30 years [2–4]. Less than 10% of cases involve men and usually they are about a decade older than the affected women [5,6]. In the past three decades, an apparent growth in the incidence of this tumour has been noted probably due to improved imaging techniques [7,8]. We report here a case of SPPT in a 23 year old female.

2. CASE REPORT

A 23 year old lady of average built (Body Mass Index - 29), euglycaemic, normotensive without any other comorbidities, with no addiction history and no significant family history presented to us in July 2017 with a non-specific pain upper abdomen for the past four months, and intermittent episodes of vomiting for the last two weeks. Her vitals as well as blood parameters including Liver and Renal Function Tests were unremarkable with Serum amylase-109 U/L & Serum Lipase-152 U/L. Abdominal palpation revealed no significant abnormality except mild tenderness in left flank and umbilical region. A Contrast Enhanced Computed Tomography (CECT) scan of Whole Abdomen (10/07/17) suggested sequela of acute pancreatitis - a pseudocyst along with bilateral minimal pleural effusion with adjacent basal atelectasis. Just after three days on 13/07/17 another Magnetic Resonance Cholangio-Pancreatography (MRCP) was done which revealed a large heterogenous Space Occupying Lesion (SOL) measuring 110 mm (antero-posterior) X 158 mm (medio-lateral) X 150 mm (supero-inferior) located postero-infero-lateral to the stomach and spleen overlapping the pancreas, (Fig. 1 & Fig. 2).

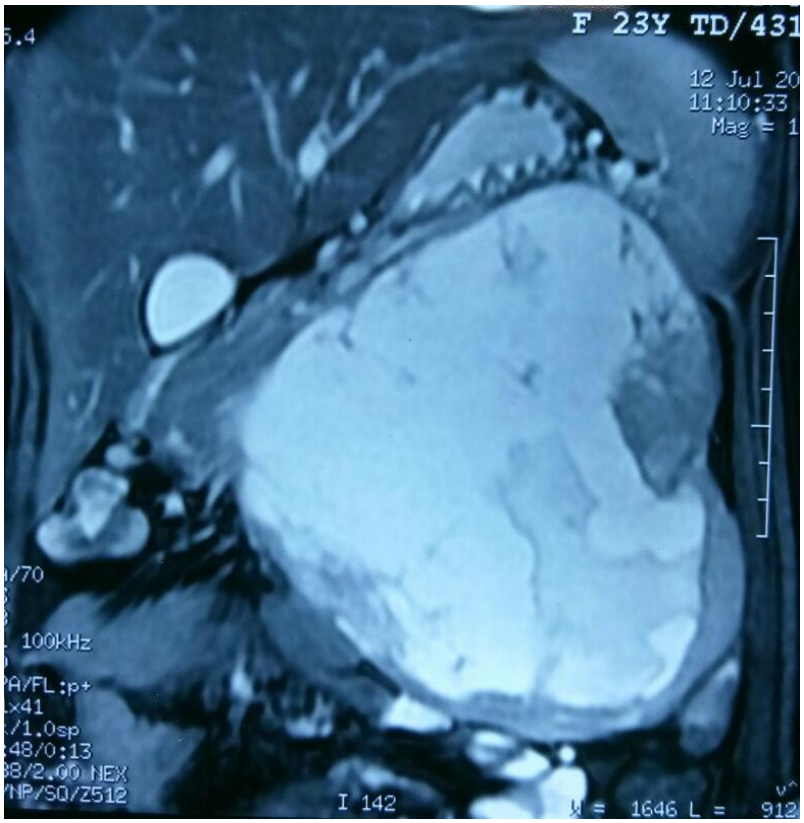

Fig. 1. MRI showing the tumour in coronal view

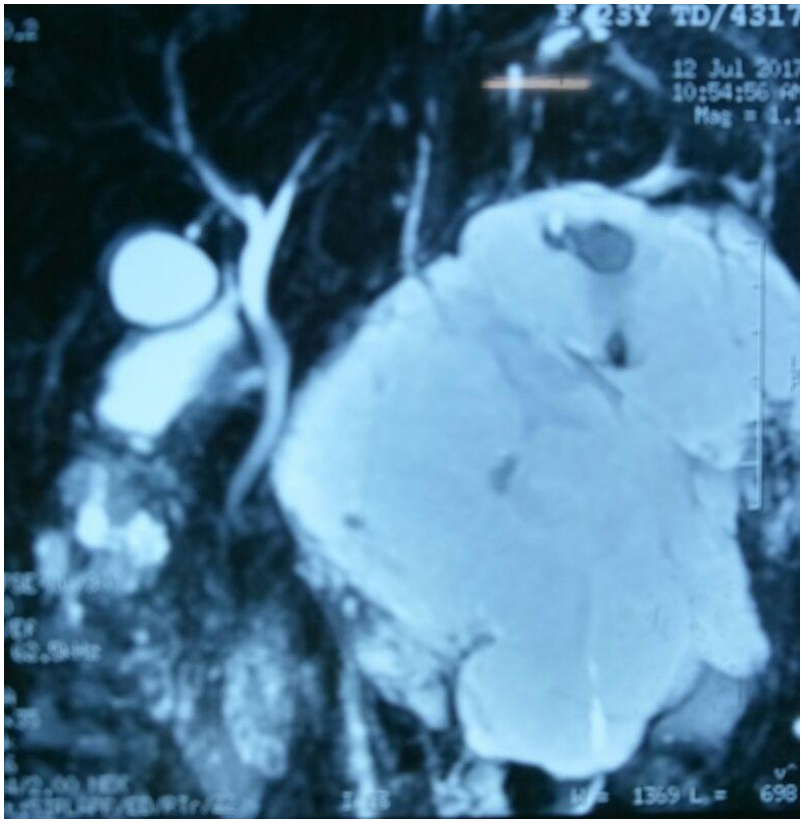

Fig. 2. MRCP showing the tumour in coronal view

She underwent exploratory laparotomy which was subsequently converted to a distal pancreatectomy (body and tail) and splenectomy on the 20th July, 2017. Histopathological examination report (22/07/17) opined that the tumour involving body and tail of the pancreas was composed of sheets and small nests of uniform round or oval cells. Pseudopapillae covered by several layers of epithelial cells were present. The fibrovascular cores of some of the pseudopapillae showed myxoid changes. Areas of haemorrhage and cystic degeneration were seen. High risk features like, invasion of capsule and lymphovascular invasion were also present. The resection margins were however free and adherent spleen was not involved. Hilar lymph nodes also showed reactive hyperplasia. Presence of both papillary areas and solid areas (Fig. 3 & Fig. 4) confirmed the diagnosis of Solid Pseudopapillary Tumour (SPPT).

TNM staging was pT2N0M0. The patient was discussed in the Multidisciplinary Tumour Board and considering the pre-mentioned risk factors, is now being treated with single agent adjuvant chemotherapy with Inj. Gemcitabine (1000 mg/ sq. m BSA, IV D1, D8 & D15 q 28 days). She is completely asymptomatic after the fourth cycle administered on 01/11/17. A triple Phase CECT scan of whole abdomen (04/11/17) also showed no residual disease.

3. DISCUSSION

In retrospect, a series of three cases of SPPT along with pathological descriptions was published by Frantz in 1959 [9]. Hamoudi et al. [10] first characterised the electron microscopic features of pancreatic SPPTs. This is why SPPTs are also referred as Frantz tumours or Hamoudi-Frantz tumors.

To the best of our knowledge, T. Papavramidis and S. Papavramidis in 2005 published the largest case series of SPPT comprising 718 cases [2]. According to Papavramidis' series, the diagnosis was made at a mean age of 22 years with a female preponderance of 10:1 [2]. However, Mean age of diagnosis in males (37 years) is a decade later than that in the females (25 years) [11]. There have been very few reported cases from India, specifically from Eastern India.

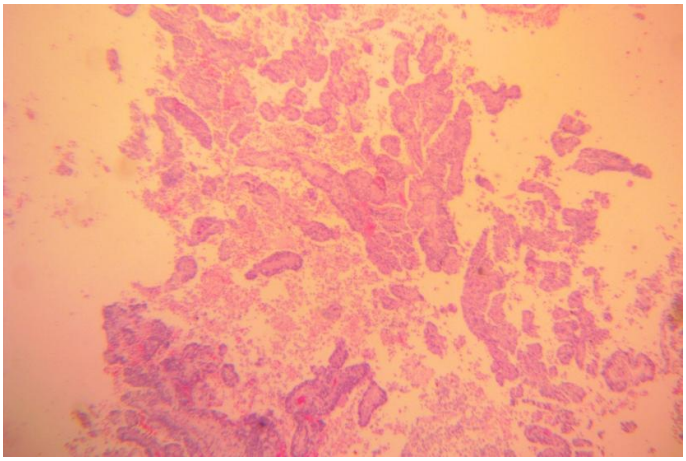

Fig. 3. Papillary areas in scanner view (4x X 10; Haematoxylin and Eosin)

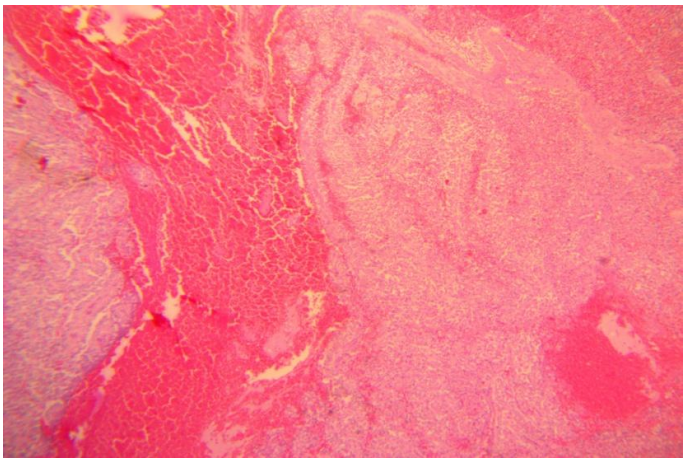

Fig. 4. Solid area in scanner view (4x X 10; Haematoxylin and Eosin)

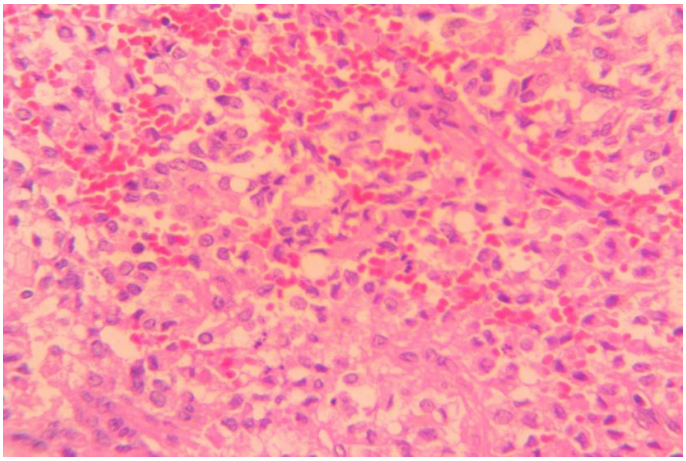

Fig. 5. Sheets and nests of uniform round or oval tumor cells in high power view (40x X 10; Haematoxylin and Eosin)

Table 1 Complementary treatment modalities for SPPT

Neoadjuvant	Adjuvant
Gemcitabine 1000 mg/m.sq, day 1,8,15	Gemcitabine
Gemcitabine 800 mg/m.sq + Cisplatin 30 mg/m.sq weekly	Gemcitabine+Capecitabine
5FU+EBRT	5FU+Cisplatin
	Cisplatin+Etoposide+Doxorubicin
	EBRT

5FU (5-fluorouracil); EBRT (external beam radiation therapy)

Though the majority of the reports originated from Europe, Japan, and North America [2], there is no recognised racial predilection for SPPTs.

The reported expression of progesterone receptors in some cases [12] and the strong female predilection points to an association between female sex hormones and pathogenesis but the exact cell of origin is still undetermined [13].

The predominant localisation of tumor is the body and tail of the pancreas, followed by the head and the neck.The most common extrapancreatic sites were the mesocolon and ovary, sometimes even in the testicle. Song et al. [14] reported one case located at site surrounded by the left kidney, spleen, and pancreatic tail which invaded the left kidney, caused the symptom of back pain and hematuria, and finally was resected with both tumor and kidney.

Less than 10% of patients are symptomatic at presentation [15], with the commonest symptom being vague abdominal pain [16]. As these lesions enlarge, symptoms from mass effect, such as vomiting and early satiety due to gastric outlet obstruction arises. Jaundice is not a common feature [2]. SPPT may also be detected incidentally on imaging. Advanced tumours may have a palpable mass detectable. Our patient presented with a vague abdominal pain for a period of four months and vomiting for two weeks.

The diagnosis is usually made on cross-sectional imaging. Encapsulated, well-defined mass with central areas of calcification, necrosis, hemorrhage, and/or cystic degeneration are major radiological features [17]. In both the arterial and venous phases, there is usually peripheral enhancement with similar Hounsfield unit density as the nearby pancreatic parenchyma [17]. The diagnosis can usually be made on multiphase contrast enhanced CT with an estimated 60% overall accuracy [18], but some authorities also advocate magnetic resonance imaging because it may be more able to delineate tissue characteristics [19].

After the diagnosis, young patients with good performance status should be managed operatively. Preoperative histologic confirmation is usually reserved for patients who have high operative risk or who require complex resections. The lesion is comprised of small ovoid or polygonal cells with small central nuclei and abundant cytoplasm [20].

Patients with resectable lesions who are candidates for operation should be treated by en bloc resection with clear margins, since this provides the best chance for a cure. and there are many reports of long-term survival after complete resection in excess of five years [13, 15,16,21,22].

There is no good comparative data evaluating the role of systemic chemotherapy. Therefore, the decision to administer systemic therapy is usually an individualised one and depends on presence of high risk features i.e. involvement of other abdominal structures, residual disease, lymphovascular invasion, perineural invasion, invasion of capsule and presence of poorly differentiated areas. Some advocate systemic therapy when poor prognosticators [23-27] or metastatic disease [26-28] is present. Role of EBRT (external beam radiation therapy) is controversial. Complementary treatment modalities as described by Bochis et al. [29] are depicted in Table 1

4. CONCLUSION

Though rare, Solid Pseudopapillary Tumour (SPPT) of Pancreas is either asymptomatic at presentation or presents with vague abdominal pain in majority. If operable, surgery forms the mainstay of curative treatment. In cases of huge mass, residual disease or other high risk features adjuvant chemotherapy can be administered with single agent Gemcitabine. It can be administered

as neo-adjuvant chemotherapy also in cases of upfront inoperability. Prognosis is good even in large tumours, if resectable. However, high risk features or residual disease, if present can be encountered with adjuvant therapy to proceed towards a favourable outcome. We report this case not only because of its rarity but also because it is a challenging clinical problem and often requires an MRI and appropriate pathologic characters for exact categorisation.

CONSENT

All authors declare that written informed consent was obtained from the patient (or other approved parties) for publication of this paper and accompanying images.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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