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A rare case report of Solid Pseudopapillary Tumor of the pancreas with portal hypertension



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ABSTRACT

INTRODUCTION: Solid Pseudopapillary Tumor of the pancreas (SPT) is a rare pancreatic tumor and represents 1–3% of all pancreatic tumors. It usually presents in young females with abdominal pain, nausea, vomiting and abdominal fullness. The first case report was documented in 1959 and since then multiple case reports have been documented on the various surgical approaches for SPT. However, there are not many reported cases where surgery has been performed on SPT with portal hypertension.

PRESENTATION OF CASE: In our case report, a 19 year old girl presented with a mass in the left side of the abdomen with associated dragging pain. Ultrasound Abdomen and CT (computed tomography) confirmed an SPT with portal hypertension, with the lesion involving the body and tail of pancreas.

DISCUSSION: Although few reports are available on SPT with portal hypertension, ours is the first report on a benign SPT with sinistral portal hypertension treated with a distal pancreatectomy. The presence of portal hypertension made the excision of the tumor and delineation of the vessels very difficult. However, when great care is taken while handling the dilated vessels, dissection can be completed with minimal blood loss.

CONCLUSION: Meticulous surgical technique along with accurate identification of vasculature will aid in the resection. Although some SPTs behave aggressively, most of them are benign and patients with SPT have an excellent prognosis.

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1. Introduction

Solid Pseudopapillary Tumor of the pancreas (SPT) is a rare pancreatic tumor that usually presents in young females. Multiple studies have demonstrated the course and nature of this tumor. Various surgical approaches for SPT have been documented. However, not much has been written about the scope for surgery in an SPT with Portal Hypertension. We describe the possibility for resection of tumor and discuss the outcome in a 19 year old who presented with SPT with Portal Hypertension.

2. Presentation of case

A 19-year-old girl was admitted with complaints of fullness in the left side of the abdomen for the past month. She also complained of a dragging pain in the abdomen for the past week. There was no

history of vomiting, constipation, fever or yellowish discoloration of eyes. Patient was otherwise asymptomatic.

On general examination vitals were stable, patient was well-built and well-nourished and general condition was fair.

On examination of the abdomen, a mass was palpable in the left hypochondrium extending into the epigastric region. The mass was 15 × 18 cm in size, non-tender and moving well with respiration. All borders except the superior border were well made out. No other mass was palpable in the abdomen.

Ultrasound abdomen showed moderate splenomegaly with a heterogeneous lesion involving the body and tail of the pancreas. CECT (Contrast enhanced Computed Tomography) (Fig. 1) abdomen revealed a huge heterogeneous mass lesion replacing the body and tail of pancreas with extension mass effect indicative of a Solid Pseudopapillary Tumor of the pancreas with left sided portal hypertension. Gastric varices were made out on upper GI (Gastrointestinal) endoscopy. The cause of the left sided portal hypertension was due to the compression caused by the large pancreatic tumor on the splenic vein leading to splenic vein thrombosis. Reconstruction of the vessels was done to study the vascular anatomy.

After a preanaesthetic work up, patient was taken up for surgery. Patient underwent an exploratory laparotomy and a 16 × 10 cm lesion involving the body and tail of pancreas (Fig. 2) was noted

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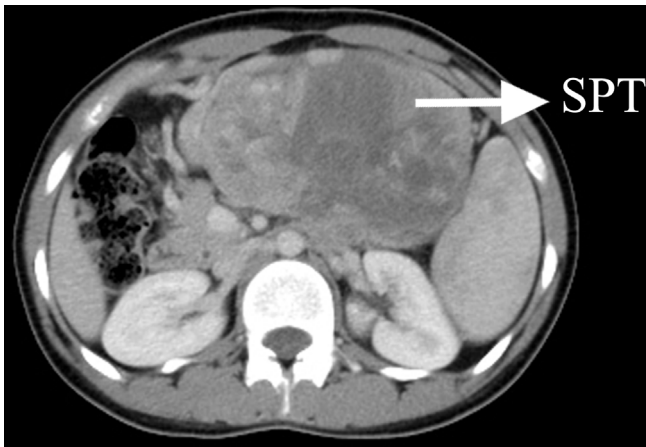


Fig. 1. CECT Abdomen showing a heterogeneous mass lesion replacing the neck, body and tail of pancreas.

along with dilated and tortuous gastric and splenic vasculature (Fig. 3). Excision of the tumor was done which required a distal pan-

createctomy and this was combined with a splenectomy (Fig. 4). Spleen preserving pancreatotomy was not attempted in view of the varices, as the splenectomy also acts as a treatment modality for the varices. Post-operative vaccinations and antibiotics were given. Oral feeds were started on day 3 and sutures removed on day 13. Final histopathology report confirmed an SPT and tumor board discussion reassured that no chemotherapy was needed. Patient was reviewed after 1 month, 3 months and 1 year and continues to do well. Upper GI endoscopy and ultrasound of the abdomen were done at the 3 month follow up which showed no gastric varices or signs of portal hypertension.

3. Discussion

SPT represents 1–3% of all pancreatic tumors [1]. It usually presents in young females with abdominal pain, nausea, vomiting, and abdominal fullness [2]. The first case report was published in 1959 by Frantz [3] and since then multiple case reports and studies have been documented on both laparoscopic and open surgical options for SPT. However, not many case reports have been documented where surgery has been performed on cases with sinistral portal hypertension. One case report by Wani et al. describes a case

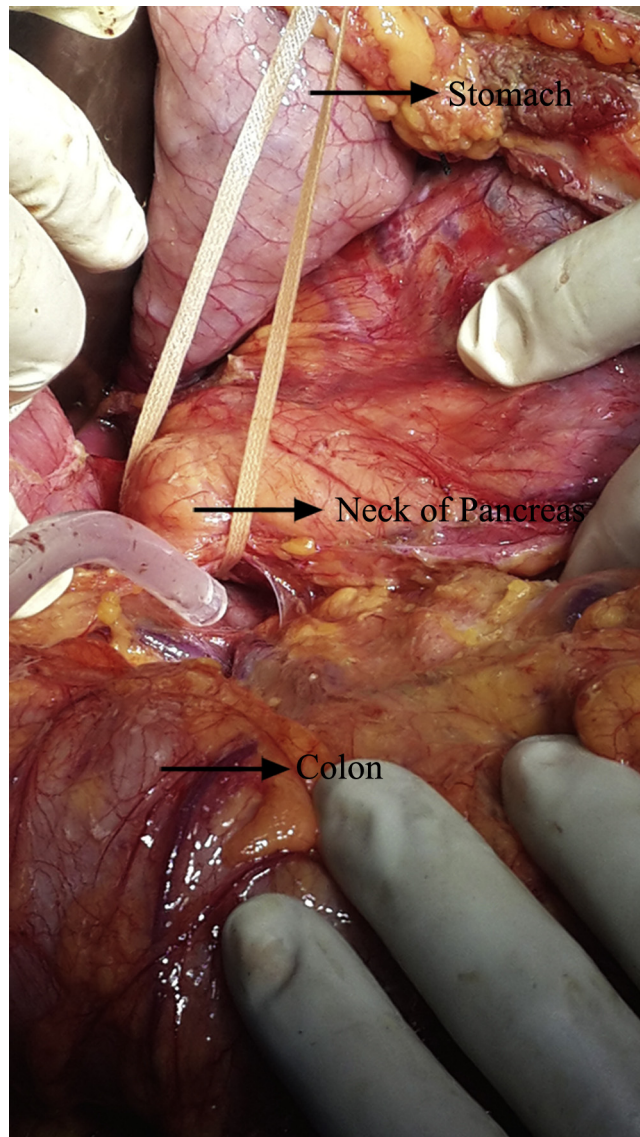


Fig. 2. Picture illustrating the plane of dissection with umbilical tape hooked around the neck of pancreas.

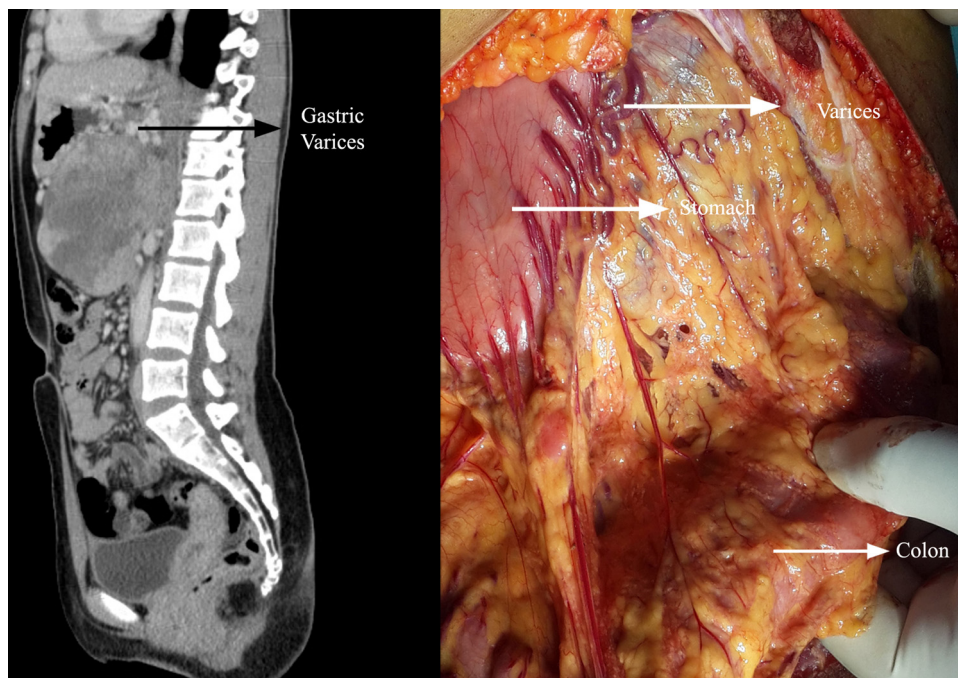


Fig. 3. These pictures depict the gastric varices due to left sided portal hypertension, which was due to the compression caused by the large pancreatic tumor on the splenic vein leading to splenic vein thrombosis.

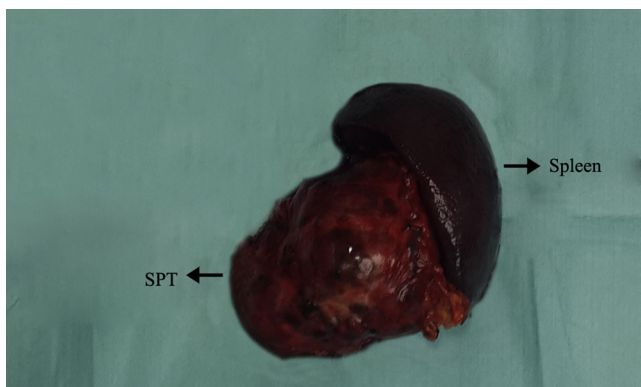


Fig. 4. Specimen of the pancreatic tumor with the spleen.

of malignant SPT treated by exploratory laparotomy and excision with splenectomy [4]. Ours is the first report on a benign SPT with sinistral hypertension treated with distal pancreatectomy.

The presence of portal hypertension, which in this case was secondary to splenic vein thrombosis due to compression by the large pancreatic tumor, makes the excision of the tumor and delineation of the vessels difficult. However with careful preoperative workup, vascular reconstruction, utmost care while handling the dilated vessels, dissection can be completed with minimal blood loss.

SPTs have a typical gross and histologic appearance. They are composed of discohesive polygonal cells that surround delicate blood vessels and form a solid mass, with frequent cystic degeneration and intracystic hemorrhage. On microscopy eosinophilic globules, foam cells and cholesterol clefts are usually present [5]. Immunohistologic analysis usually shows positivity for chromogranin and synaptophysin [6].

Typical laboratory tests include a normal serum amylase and tumor markers [1]. CT is the main investigatory modality with an additional MRI if needed for further delineation [7]. CT will pick up the location of tumor, presence of portal hypertension,

involvement of vessels, tumor calcification and exact relation to surrounding structures. 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 [8]. An MRI was not used in our centre. Depending on the location of the tumor, an ultrasound or endosonography may also be useful. The use of a biopsy is controversial as CT guided biopsy of the tumor is needed for confirmation of the diagnosis and to rule out malignancy as the treatment for SPT only consists of an enucleation which would be insufficient in the case of a malignancy. However, the accuracy of the biopsy is limited [1,2].

As SPT is considered to be a tumor of low-grade malignant potential. Complete surgical excision is the treatment of choice. Thus surgery should always be attempted irrespective of the extent of resection.

A majority of the cases of SPT are cured with radical excision, however there are reports stating that aggressive behavior of the tumor also exists. In a study by Reddy et al., 11% of the patients presented with metastasis; either lymph nodal metastasis or metastasis to the liver. Local metastasis, with involvement of the superior mesenteric artery or the duodenum has been noted [6].

A recent series from Memorial Sloan-Kettering Cancer Center recommends complete surgical excision with even metastatectomy if required. These authors conclude that long-term survival improves with complete surgical resection of primary with metastectomy for synchronous or meta-chronous lesions. Other investigators also reported long-term survival as many as ten years [9].

A local recurrence rate of 6.2% is reported in cases treated by radical surgical excision [10].

Few authors have reported an increased rate of resectability after chemotherapy [11]. Chemotherapeutic agents like gemcitabine, cisplatin, etoposide, vincristine have all been documented to reduce the size of tumor making it favorable for surgery [12–14]. There is a case report which has shown a survival benefit with radio-

therapy in a locally advanced lesion involving the portal vein [15]. However, most authors agree that aggressive surgical resection is the best modality of treatment for achieving curative results and a better long-term survival [16,17,18].

4. Conclusion

In conclusion, SPT is a rare neoplasm, usually presenting with vague symptoms like abdominal pain and a mass. A high index of clinical suspicion along with investigatory modalities can help make an accurate diagnosis. Though several reports of SPT with portal hypertension have been published, this is the first time to our knowledge, that a detailed surgical approach has been discussed. This would motivate surgeons to operate upon a case of SPT with portal hypertension safely. Formal resection may be performed in similar situations and the presence of portal hypertension need not be a contraindication for surgery. Meticulous surgical techniques along with accurate identification of vasculature will aid in the resection. Although some SPTs behave aggressively, most of them are benign and patients with SPT have an excellent prognosis after surgical excision.

Methods: This case report has been reported in line with the CARE criteria [19].

Consent: Appropriate informed consent has been obtained.

Conflicts of interest

Dr. Asha, Dr. Saravanan, Dr. Dilip and Dr. Narayanan have no conflicts of interest.

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Ethical approval

This study does not include a research study.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Dr. Asha—data collection, writing the paper, data analysis.
 Dr. Saravanan—study concept and design, writing the paper.
 Dr. Dilip—data collection, data analysis.
 Dr. Narayanan—study concept and design, data interpretation.

Guarantor

Dr. Asha Reddy.

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