CASE REPORT

Spontaneous Rupture of Solid Pseudopapillary Tumor of Pancreas

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ABSTRACT

Solid pseudo papillary neoplasms are rare, potentially malignant tumors of the pancreas which commonly present with abdominal pain or incidentally picked on imaging. Acute presentation of these tumors is quite rare. Here we report a case of SPN in a 13 yr old girl who presented with feautures of acute abdomen with hypotension. She had sudden onset of abdominal pain associated with giddiness, evaluation revealed hemoperitoneum and she received 2 units of red blood cell transfusion. Contrast computed tomography of the abdomen was suggestive of a solid lesion in the lesser sac close to the spleen. She was operated and a distal pancreatectomy plus splenectomy was done. Final histopatholgy was suggestive of solid pseudopapillary tumor with rupture.

INTRODUCTION

Solid Pseudopapillary Neoplasms also known as Frantz Tumors are rare tumors of the pancreas,first described by Frantz in 1959 [1]. In 1996, World Health Organization classified these tumors as Solid Pseudopapillary Tumors (SPT's) [2] accounting for nearly 2% of resected pancreatic neoplasms. These tumors are most commonly seen in young girls in their second decade of life, they are usually well circumscribed and most often seen in the distal pancreas [3]. These lesions are commonly picked up when the patient presents with symptoms like upper abdominal pain, fullness in the abdomen. Presentation of these lesions with rupture is quite rare and the cause for rupture is found to be either spontaneous or following trauma, with both causes being reported with similar incidence.

CASE REPORT

Informed consent from the patients family and an Institutional ethics committee approval was obtained to report this rare presentation. Our patient is a well preserved 13 year old girl, who presented to another hospital with features of sudden onset upper abdominal pain associated with giddiness. She was initially treated elsewhere, received blood transfusion (for a haemoglobin level of 6g %), symptomatic management and cross sectional imaging (Contrast Enhanced CT). She was referred to us for further management. Patient was stable when she was first seen by us. General examination was unremarkable, abdominal examination revealed a swelling in the epigastric and left hypochondrial regions associated with tenderness. CT images were reviewed and she was planned for surgery in view of features of a ruptured pancreatic neoplasm **(Figure 1a,b,c)**.

Laparotomy was performed with a roof top incision. Altered blood was noted in the peritoneal cavity. There was a large mass in the lesser sac protruding through the gastro-colic omentum. Lesion was extending from the supra renal region till the upper pole of spleen with adhesions to the splenic capsule, necessitating splenectomy. A distal pancreatico-splenectomy was completed within 90 mins, with a blood loss approximately 200 ml. Patient was discharged on the 7th postoperative day after an uneventful course **(Figure 2)**. Final histopathology was a solid pseudo papillary tumor of the tail of pancreas with no evidence of atypical features, associated with rupture. Immunohisto chemical profile was done and tested for vimentin, neuron specific enolase, chromogrannin –A, pan cytokeratin, CD10, EMA, Synaptophysisn and the result was positive for Vimentin and Neuron Specific Enolase **(Figure 3)** which were in favour of the diagnosis of Solid Pseudo papillary Tumor. She has been under follow up for the past one and a half year and no disease has been identified clinic radiologically.

DISCUSSION

SPN's are known to behave indolent and frequently present with vague abdominal symptoms. In a large review,

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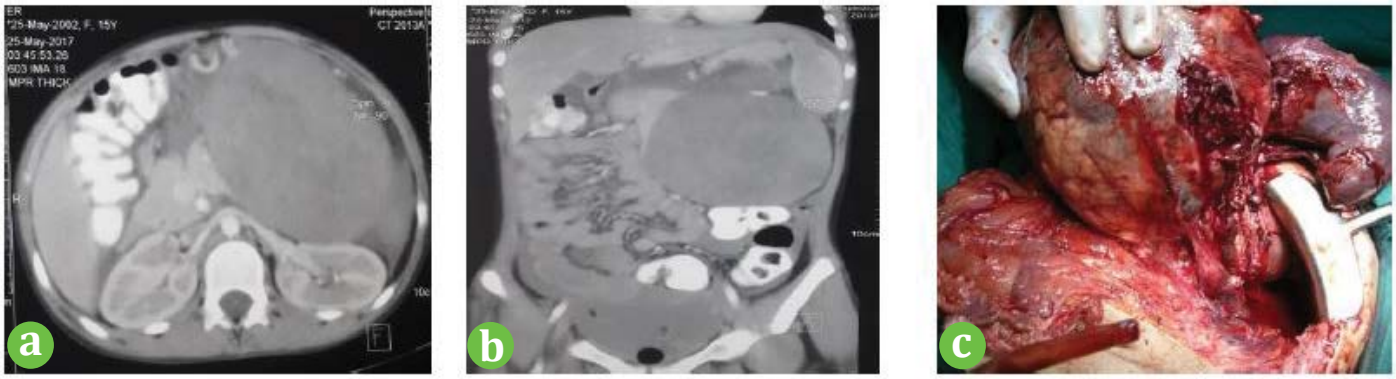

Figure 1. (a, b). CECT images showing a large heterogenous mass lesion in the distal pancreas. **(b).** Operative image showing rupture.

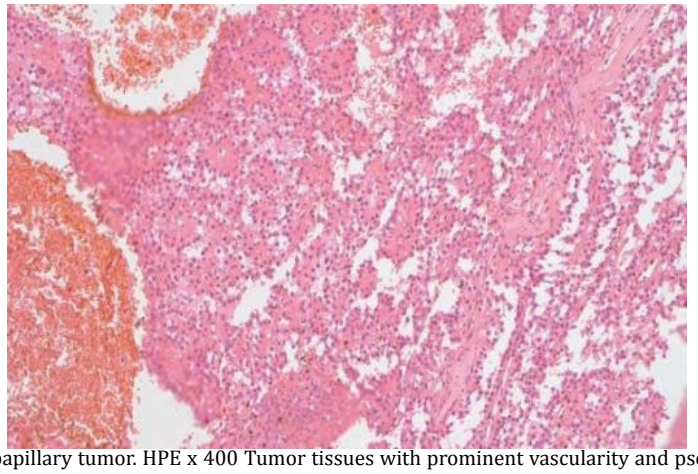

Figure 2. Features suggestive pseudopapillary tumor. HPE x 400 Tumor tissues with prominent vascularity and pseudo papillae with no nuclear atypical features.

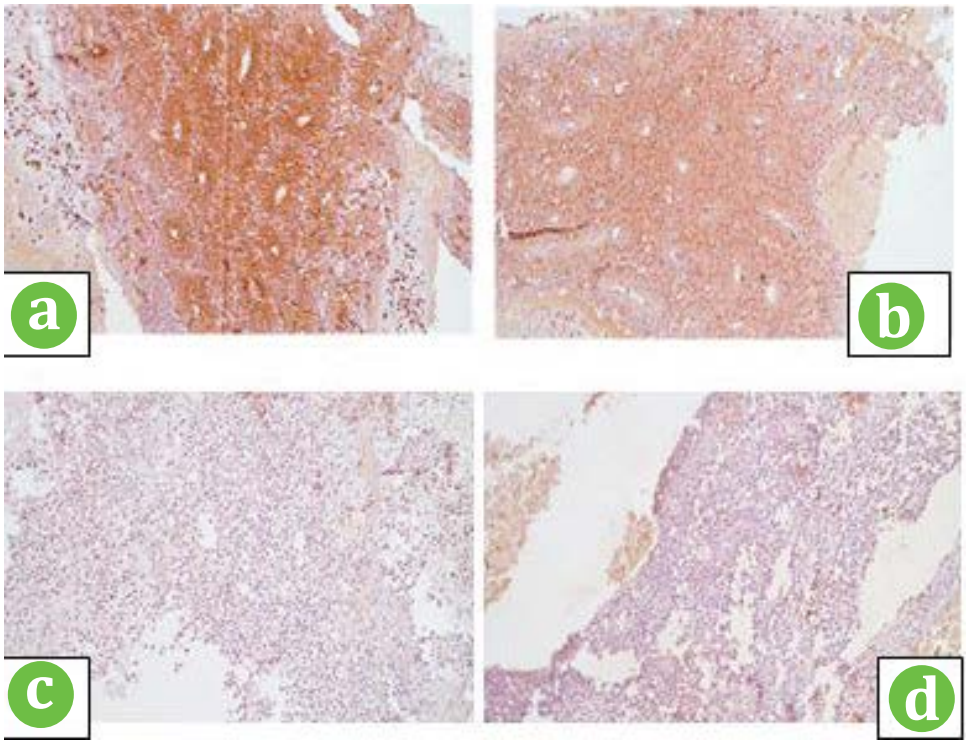

Figure 3. Immuno Histochemistry. **(a).** vimentin – positive, **(b).** neuron specific enolase- focal positive, **(c).** chromogrannin a – negative, **(d).** PAN cytokeratin – negative.

these tumors were found to present most commonly with abdominal pain (47%) followed by swelling (35%) [3]. In this case there was an aggressive presentation of the tumor in the form of spontaneous rupture. 17 cases of SPT rupture have been reported in literature so far with 10 cases of traumatic rupture and 7 cases of spontaneous

rupture [4, 5, 6, 7, 8, 9, 10, 11, 12, 13, 14, 15, 16, 17, 18]. Spontaneous rupture is a very rare complication, reported less than 1% [14]. SPN's usually show significant degenerative component, hence the rupture may be explained by sudden increase in the intracavitary pressure due to a bleed, infection or necrosis. It is known that any ruptured neoplasm can potentially lead to implantation of the tumor cells in the peritoneal cavity and portends a poor prognosis in most of the intra abdominal malignancies, however this does not preclude surgical intervention in the case of SPN [19]. As most of these lesions are benign/ borderline malignancies a complete extirpation must be attempted due to the favourable biology, even though a risk of recurrence exists [20]. Patients with metastatic disease are still expected to have a long survival [21] and resection of metastatic disease is advocatedwhen feasible.

CONCLUSION

Our patient was followed up for one year till date and there is no evidence of disease clinico-radiologically.

Conflict of Interest

The authors have no conflicts of interests to declare.

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