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Case Report

Pathology Section

Malignant Solid Pseudopapillary Neoplasm of Pancreas- Case Report with Review of Literature

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ABSTRACT

Solid Pseudopapillary Neoplasm (SPN) is a rare tumour with excellent prognosis seen predominantly in females. Patients usually remain asymptomatic or have vague symptoms like abdominal discomfort, pain, vomiting arising due to compression effect. Only a few cases with aggressive nature have been reported. Here we report a case of 25-years-old female who presented with abdominal mass of 15x15 cm arising from the pancreas. Following surgical resection, microscopic examination of mass revealed tumour composed of atypical cells arranged in sheets and pseudopapillae, showing high mitosis, necrotic foci, capsular and vascular invasion which is unusual form of presentation. Informed consent of patient was taken prior to procedure.

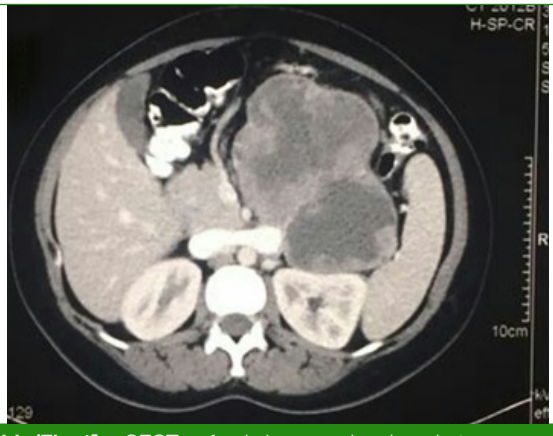
Keywords: Pancreas, Solid pseudopapillary neoplasm, Spleen

CASE REPORT

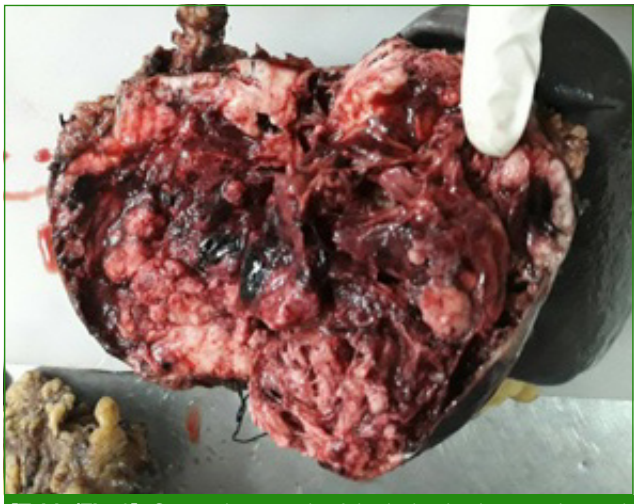
A 25-years-old female patient presented to the Surgery Department of our hospital with complaints of abdominal pain and bilious vomiting since six months. No history of weight loss, diarrhoea, constipation was noted. On per abdominal examination, a vague mass about 15x15 cm in the upper abdomen moving with respiration was palpable. CECT scan was performed which revealed a large well defined heterogeneously enhancing lesion in the left upper abdomen with areas of necrosis, superiorly displacing the body and head of the pancreas, abutting part of the colon, stomach left kidney, spleen , duodenum, duodenojejanal junction, proximal jejunum with ill defined planes arising from the same [Table/Fig-1]. Following consent of patient, surgery was performed. Per-operatively, a mass of size 15x15 cm was seen in the lesser sac arising from body and head of the pancreas. The mass was resected along with the body, tail and part of pancreatic head. Spleen and part of the transverse colon with omentum were also resected. The specimens were received at our department for histopathological examination.

Gross examination revealed an encapsulated solid mass of size 15x13x10 cm arising from the body of the pancreas. Cut section of the mass showed solid, variegated lesion with large areas of haemorrhage, necrosis and focal cystic changes [Table/Fig-2]. On microscopy the tumour was composed of sheets of cells with focal pseudopapillae formation [Table/Fig-3a]. The individual cells showed high N:C ratio oval to polygonal nuclei and prominent nucleoli showing moderate pleomorphism, high mitosis (2-3/ hpf), [Table/Fig-3b] with large areas of necrosis and haemorrhage. Prominent vessels were noted with tumour cells cuffing around the vessels. Focally, foamy macrophages were also seen. Areas of capsular and vascular invasion were also noted [Table/Fig-3c]. Sections from spleen, colon and mesentry were unremarkable.

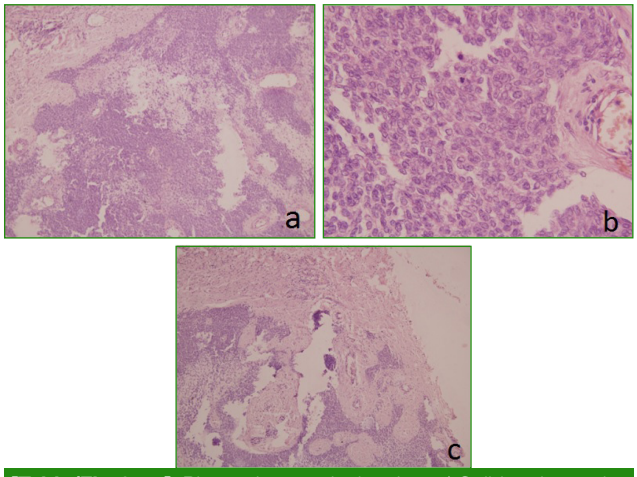
Immunohistochemistry was performed which showed strong positivity for vimentin and focal positivity for beta catenin. A final diagnosis of malignant solid pseudopaillary neoplasm was made. The patient was discharged following surgery and is receiving chemotherapy. After four months of surgery the patient remains free of malignancy.



[Table/Fig-1]: CECT of abdomen showing heterogeneously enhancing lesion in the left upper abdomen with areas of necrosis.



[Table/Fig-2]: Gross photograph of the lesion having a variegated appearance with areas of haemorrhage, necrosis and cystic change.



[Table/Fig-3a-c]: Photomicrograph showing- a) Solid and pseudo-papillary areas (H&E,100X); b) Atypical mitosis (H&E,400X); c) Vascular invasion (H&E,400X).

DISCUSSION

SPN accounts for 0.2-2.7% of all tumours arising from exocrine pancreas. However, due to improved imaging techniques the incidence has been increased to 5%. Most cases of SPN arise from body or tail of pancreas and are usually large at the time of presentation [1].

SPN is seen in young female population. Described for the first time by Dr. Frantz in 1959, hence named Frantz tumour, the neoplasm has been called by different names such as solid pseudopapillary tumour, solid cystic tumour, papillary cystic tumour, adenocarcinoma of pancreas in childhood. It was finally named solid pseudopaillary neoplasm of the pancreas by WHO in 2010 and categorized as low grade malignant neoplasm [2]. The female- male ratio of SPN is 5:1.9 [3].

Most patients remain asymptomatic with tumour detected incidentally on radiological examination. Other patients present with vague symptoms like abdominal pain, increased abdominal girth, nausea and vomiting due to mass effect over adjacent organs. Our patient was a 25-year-old female who presented with vague abdominal discomfort. Male patients usually present much later (mean age 31 years). No association of SPN with any medical condition has been found till date [4].

Typical presentation of the tumour is solitary mass which is well demarcated arising from the pancreas, body and tail being the most common site. The size of the tumour is usually large, but cases of variable size have been reported in literature. The tumour is encapsulated with large areas of haemorrhage and necrosis with cystic changes. On microscopy the tumour cells are arranged in discohesive sheets with prominent blood vessels in the stroma imparting the pseudopapillary appearance. Foamy histiocytes and foreign body giant cells have been reported in SPN [5]. In conventional cases mitosis is rare with less than 1% Ki67 index. The present case presented with classical histology but had a high mitotic count coupled with capsular and vascular invasion.

IHC may be helpful in reaching the accurate diagnosis. However, no IHC marker is specific of SPN. The tumour cells generally show positivity for vimentin, beta catenin, CD10, CD56 while focally for synaptophysin. Many of the cases show positivity for progesterone receptor while negative for estrogen receptors. CK may be positive in 30% to 70% of the cases. Almost all cases of SPN exhibit mutation in exon3 of CTNNB1 gene which codes for beta catenin. As a result of the mutation there is abnormal localization of beta catenin in the nucleus hence positive for beta catenin on IHC [6].

The prognosis remains excellent with cure rate between 85 to 90% after complete excision of the tumour. Lymph node metastasis is rare. Around 15% cases demonstrate metastases to liver and peritoneum [7].

The features favouring aggressive clinical course remains blurred. In 2000 WHO stated that features such as angioinvasion, perineural invasion and invasion into the surrounding soft tissue were more in favour of malignant behaviour. These features are non specific and inconsistent with the outcome of the patient [8]. In a case study by Mat Zin AA et al., a 14 years old girl with typical histological features of SPN with perineural invasion showed no evidence of residual tumour [9]. Similarly, Sperti C et al., reported a case with features of angioinvasion, moderate cellular atypia and mitosis with multiple metastatic deposits in liver. The patient was however disease free following multiple chemotherapeutic regimen [10].

Tang LH et al., in his case series described two cases with fatal outcome, both cases demonstrating diffuse pattern of growth with less fibrovascular stroma, extensive necrosis and high mitotic count upto 70/50 hpf, pleomorphism, high nuclear cytoplasmic ratio. One of the cases showed multinucleated giant cells as well. Also high Ki 67 index, 30-40% was noted in both the cases. Both the cases died early at 6 months and 16 months respectively. Hence, they concluded that a mitosis of

more than 15/50 hpf is associated with poor prognosis [11].

Recently Reindl BA et al., reported case of SPN in a 17-year-old girl with tumour cells showing mitotic count up to 30 per 50 hpf, nuclear pleomorphism, extensive necrosis. The patient developed recurrence following surgical resection, received multiple chemotherapeutic regimens and died eventually after 11 months of initial diagnosis [2].

CONCLUSION

SPN is a rare tumour with excellent prognosis, seen predominantly in young female population. Few SPN cases show aggressive features which are associated with fatal outcome. Criteria for malignancy remains uncertain however features such as high mitosis, extensive necrosis and pleomorphism as demonstrated in our case and previous case reports may be useful in recognition of cases with poor prognosis. Despite advancement in imaging technique the diagnosis of SPN remains challenging even more so for aggressive SPN.

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