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Solid pseudopapillary Epithelial Neoplasm Of Pancreas: A Case Series From A Tertiary Care Centre

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Abstract

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Introduction

Solid pseudopapillary neoplasm of the pancreas is an extremely rare neoplasm of the pancreas. This neoplasm is seen predominantly in the young female population with age ranging from 2 to 85 years [1]. The mean age of presentation being 28yr [2]. Patients are typically asymptomatic and may be an incidental finding or can present with abdominal mass and pain [3]. It's a primary pancreatic neoplasm and can arise from any part of the pancreas [4].

Here we present a case series reported at our institute, Sher-i-Kashmir institute of medical sciences skims Soura. These patients presented with abdominal mass. Clinical and laboratory examinations were performed. The radiological examinations of the patient were done. FNAC performed showed variable diagnostic features. The histopathological examination and IHC were confirmatory of the solid pseudopapillary neoplasm of the pancreas.

Case 1: A 40-year-old female patient incidentally detected a pancreatic head mass on USG abdomen. There was no history of pain, nausea or vomiting.

Radiology:

USG's abdomen shows a hypoechoic mass in the body of the pancreas.

Contrast-enhanced CT scan showed an exophytic mass arising from the distal body of the pancreas.

Contrast-enhanced MRI: heterogeneously enhancing solid cystic mass lesion arising from distal body and tail.

Tumour markers: were in the normal range.

Guided FNAC: diagnosed as neuroendocrine tumour

Gross examination: distal pancreatectomy with splenectomy specimen with a white well-circumscribed mass measuring 10.5 x9 cm. The lesion on sectioning had a solid appearance with very tiny cystic and haemorrhagic spaces

Case 2: A 19 -year-old female patient, presented clinically with pain abdomen and was being evaluated and diagnosed with a pancreatic head mass on the USG abdomen.

Radiology:

USG's abdomen shows a hypoechoic mass in the body of the pancreas.

CT scan and MRI were not done

Tumour markers: were in the normal range.

Guided FNAC: showed non-diagnostic smears.

Gross examination: pancreaticoduodenectomy specimen with a firm grey-white white mass in the head of the pancreas measuring 3x3cm.

Case 3:

A 33 -year-old female patient, presented clinically with pain abdomen and was accidentally diagnosed with a pancreatic head mass on the USG abdomen.

Radiology.:

USG's abdomen showed a hypoechoic mass in the body and neck of the pancreas.

CT scan showed an Isodense lesion at the junction of body and neck

Tumour markers: were in the normal range.

Guided FNAC: diagnosed as borderline papillary neoplasm

Gross examination: distal pancreatectomy with splenectomy specimen with a friable mass in the body of the pancreas measuring 1.7x1.3cm.

Case 4:

A 16 -year-old female patient, with incidentally detected pancreatic body mass on the USG abdomen.

Radiology:

USG's abdomen showed a 2x2x2 cm solid cystic mass in the body of pancreas.

CT scan showed a 2.1x2.4x2.1cm lesion at the junction of body and tail likely suggestive of solid pseudopapillary epithelial neoplasm.

Tumour markers: were in the normal range.

Guided FNAC: was not done

Gross examination: hemi pancreatectomy with splenectomy specimen with a solid cystic mass in the body of the pancreas measuring 3x2.6cm.

Case 5:

A 34-year-old female patient already a diagnosed case of solid pseudopapillary epithelial neoplasm with recurrent tumour in pancreatic bed with large metastatic secondary in right lobe of liver.

Radiology:

USG's was not done

CECT showed a large centrally necrotic cystic lesion measuring 17x13x19 cm with peripheral enhancement involving right lobe of liver.

A solid cystic lesion measuring 4.5x3.7cm in the region of the distal body and tail of pancreas, inseparable from adjacent small bowel loops .

Tumour markers: were not available.

Guided FNAC: was not done

Gross examination: right hepatectomy specimen measuring 28x24cm with a mass measuring 17x15cm, and pancreatic bed tumour mass measuring 3.5x3cm.

Case 6:

A 30 -year-old female patient, with incidentally detected pancreatic body mass on the USG abdomen.

Radiology:

USG, CECT, MRCP were suggestive of a solid cystic lesion in the body of pancreas measuring 2x2x2cm

Tumour markers: were not done in this patient

Guided FNAC: was not done

Gross examination: distal pancreatectomy specimen with a solid cystic mass in the body of the pancreas measuring 2.5x1.8cm.

Case 7:

A 24 -year-old female patient, complaining of abdominal pain with radiologically detected pancreatic body mass.

Radiology:

CT scan showed a 4x4cm lesion in the body of pancreas suggestive of solid pseudopapillary epithelial neoplasm.

Tumour markers: were normal

Guided FNAC: was not done

Gross examination: distal pancreatectomy with splenectomy specimen with a solid cystic mass in the body of the pancreas measuring 3.6x3x3cm.

Case 8:

A 30-year-old female patient complaining of epigastric pain since 4 years, was diagnosed distal pancreatic mass on USG.

Radiology:

USG showed a large solid mass arising from distal pancreas.

CE-CT was suggestive of solid pseudopapillary epithelial neoplasm

Tumour markers: were not available.

Guided FNAC: was not done

Gross examination: distal pancreatectomy with splenectomy specimen with a friable mass in the tail of the pancreas measuring 11x9.5x8cm.

Case 9:

A 33-year-old female patient complaining of pain with accidentally detected mass radiologically.

Radiology .:

CT showed an Isodense lesion in the body

Tumour markers: were not available.

Guided FNAC: Showed features of borderline papillary neoplasm with cystic changes.

Gross examination: distal pancreatectomy with splenectomy specimen with a friable mass in the body of the pancreas measuring 1.7x1.3x1cm.

Histopathology: on histopathological examination, all the cases revealed a well-circumscribed tumour with tumour cells arranged in a pseudopapillary pattern, minimal pleomorphism, fine chromatin, intranuclear grooves and moderate eosinophilic cytoplasm.

Discussion: solid pseudopapillary neoplasm of the pancreas presents usually with abdominal pain followed by abdominal mass, loss of appetite [5]. Approximately 30% of patients are asymptomatic and mass is detected on routine examination or on imaging procedures [6]. In our study 5 cases presented with pain abdomen and 3 cases were detected incidentally on imaging and 1 case was already a diagnosed case of Solid pseudopapillary epithelial neoplasm of the pancreas with recurrence into the pancreatic bed and metastasis to liver.

In our study all the 9 patients were females as is seen in other studies where female preponderance is seen. In a study conducted by choi et al, female to male ratio was seen to be 5:1.9 [7]. Imaging studies have been a very important diagnostic tool in detecting these tumours as a result the diagnosis of Solid pseudopapillary epithelial neoplasm of the pancreas has increased. These lesions are shown to have well circumscription, hypoechoic, heterogeneous solid cystic masses [8]. In our study, 5cases were detected by Ultrasonography, 2 cases were detected by CT scan and 1 case of recurrence was followed by

CECT. FNAC has not been helpful in our study in the diagnosis of these lesions.

As in other studies, IHC staining for Beta-catenin has been important in diagnosing these lesions [9]. In our study, all 8 cases were strongly positive for Beta-catenin.

The mainstay of treatment is complete surgical excision of the mass even if it is locally invasive or shows distant metastasis. The most common procedures being distal pancreatectomy pancreatoduodenectomy [10]. In our study 7 cases were operated by distal pancreatectomy with splenectomy and 1 case was operated pancreaticoduodenectomy and 1 case of recurrence with hepatic metastasis was operated by right hepatectomy with pancreatic tumour bed mass excision.

The most common site of the tumour in studies is pancreatic tail and head of pancreas followed by body and tail [1]. In our study 4 cases presented with mass in the body of the pancreas and 2 case with mass in the head of the pancreas, 1 case in the tail of pancreas, 1 case at the junction of body and neck and 1 case with metastasis into liver with recurrence in pancreatic bed.

In our study, Lymphovascular invasion was noted in the case of hepatic metastasis however no lymph node metastasis, perineural invasion was noted. 9-15% of patients treated with surgical excision show local recurrence or metastasis [10]. However, in our cases, no metastasis or recurrence was seen on follow-up so far but 1 case which was an already operated case of pancreatic resection had presented with metastasis to liver and pancreatic bed recurrence.

No association was seen with tumour markers or laboratory results in relation to SPN in our cases.

Conclusion: SPN is a rare neoplasm of the pancreas. The tumour is seen predominantly in the female population. Imaging techniques are important in the preoperative diagnosis of these lesions. IHC staining for Beta-catenin is a very useful marker in the diagnosis of these lesions. The mainstay of treatment is complete surgical excision but few cases may show recurrence of tumour. Follow up of patient with radiology is must.

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Fig. 1: Gross photograph of distal pancreatectomy with splenectomy specimen with a well circumscribed mass in the body of the pancreas.



Fig. 2: Microphotograph showing well circumscribed tumour

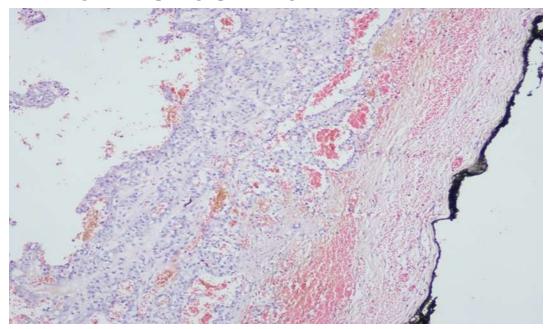


Fig. 3: Microphotograph showing tumour cells arranged in a pseudopapillary pattern, minimal pleomorphism.

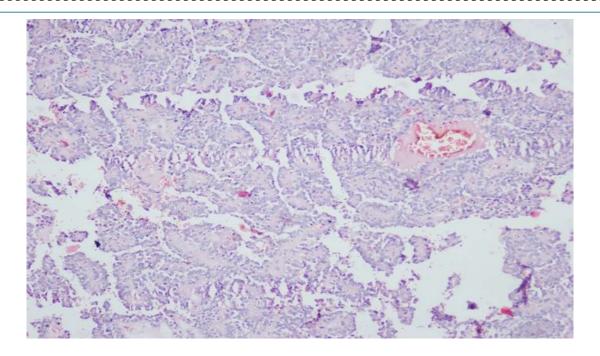
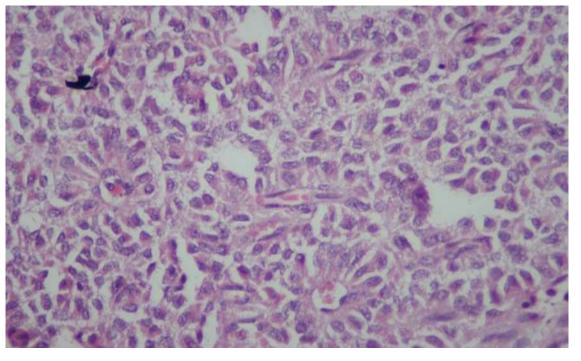


Fig. 4: Microphotograph showing tumour cells arranged in a pseudopapillary pattern, minimal pleomorphism, fine chromatin, intranuclear grooves and moderate eosinophilic cytoplasm



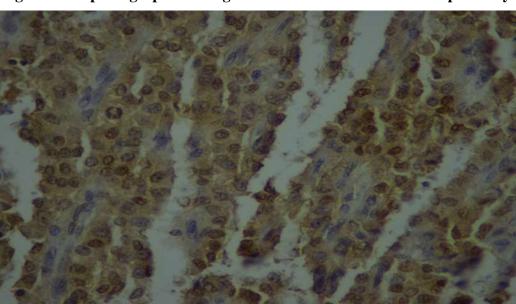


Fig. 5: Microphotograph showing tumour cells with Beta-catenin positivity.

Fig. 6: Microphotograph showing tumour cells with Synaptophysin positivity.

