SOLID PSEUDOPAPILLARY TUMOUR OF THE PANCREAS: A TERTIARY CARE CENTRE EXPERIENCE

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ABSTRACT

BACKGROUND

Solid pseudopapillary tumour of the pancreas is a rare tumour of low malignant potential occurring predominantly in young females. Its incidence has been increasing due to advanced imaging modalities. As this tumour offers a good prognosis, it is important to make a proper diagnosis to offer better treatment and reduce morbidity.

MATERIALS AND METHODS

This is a prospective study for a period of 2 years (From May 2014 to April 2016). Of the 52 pancreatic specimens we received after surgery, 9 cases had a prior radiological diagnosis of solid pseudopapillary tumour of the pancreas. The clinical and histo-pathological characteristics of SPT were studied along with review of literature. Whipple resection specimens which were radiologically diagnosed as adenocarcinoma of the periampullary region were excluded.

RESULTS

Nine cases were reported radiologically as papillary neoplasm of pancreas. On histopathology, 8 of them were confirmed as solid pseudopapillary tumours of the pancreas. One was a case of serous cystadenoma and other one was pancreatic neuroendocrine tumour. One case which was suspected as pancreatic endocrine tumour radiologically was diagnosed as SPT.

CONCLUSION

SPT typically is limited to the pancreas at the time of diagnosis, and even with metastasis, an extended complete surgical excision offers good prognosis. Hence, it is important to distinguish it from other tumours of similar morphology. In this study, we discuss the process of establishing the diagnosis accurately of SPN in young patients presenting with pancreatic mass.

KEYWORDS

Solid Pseudopapillary Tumour, Frantz Tumour, Pancreatic Tumours in Females, Mass Per Abdomen.

HOW TO CITE THIS ARTICLE: Aruna L, Mekhala PM, Arasi E, et al. Solid pseudopapillary tumour of the pancreas: A tertiary care centre experience. J. Evid. Based Med. Healthc. 2016; 3(63), 3435-3438. DOI: 10.18410/jebmh/2016/739

INTRODUCTION: Solid papillary tumour of the pancreas (SPT) is also known as "Frantz tumour" which was first described by Dr. Frantz in 1959. He described this tumour as a "papillary tumour of the pancreas, benign or malignant".[1] In 1996, WHO coined its current terminology solid pseudopapillary tumour in the international histological classification of the tumours of exocrine pancreas.[2] It is known with various names like Solid and papillary epithelial neoplasm, papillary epithelial neoplasm of pancreas, solid and cystic tumour of pancreas, adenocarcinoma of pancreas of childhood.

MATERIAL & METHODS: The present study was conducted from May 2014 to April 2016 at our tertiary care centre.

Financial or Other, Competing Interest: None.
Submission 29-06-2016, Peer Review 19-07-2016,
Acceptance 30-07-2016, Published 08-08-2016.
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DOI: 10.18410/jebmh/2016/739

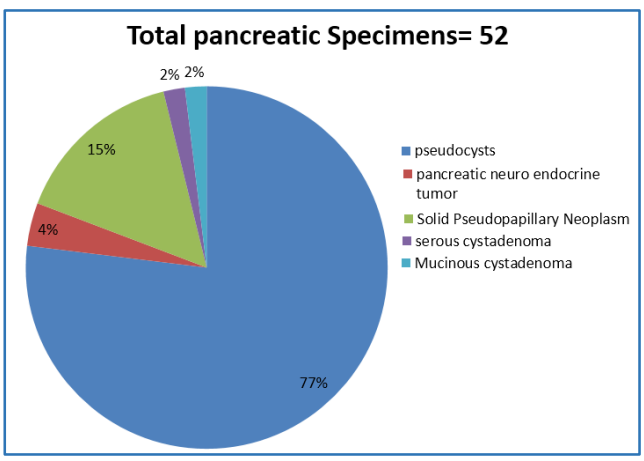
We received 52 pancreatic specimens with a preoperative suspicion of benign lesions, out of which 9 cases were suspected to be SPT radiologically. Clinical details including age, mode of presentation, radiological findings, surgery performed and pathological characteristics were analysed in all the cases. The specimens were fixed in buffered formalin, routinely processed and sections cut from paraffin blocks. Histological examination was done on Haematoxylin and Eosin (H & E) stained sections. The sections were also subjected to immunohistochemical stains. The panel of markers included were - vimentin, beta catenin, Progesterone receptor (PR), CD 10, Neuron-specific enolase (NSE), chromogranin, Ki-67 (Proliferating index). The results were correlated clinically and radiologically.

RESULTS: Out of the 52 pancreatic specimens we received, 40 cases [77%] were pancreatic pseudocysts. 8 cases [15%] were of SPT. 2 cases were of serous cystadenoma and mucinous cystadenoma [4%]. Remaining 2 cases were of pancreatic neuroendocrine tumour [4%]. Among the 8 cases, one was a male [M: F=1:7]. All the patients were in the age group of 11-48 years.

Among 6 cases - 3 cases each [37.5%] presented with mass per abdomen and pain abdomen respectively, while 2 cases [30%] presented with both mass per abdomen and associated pain. Location of the tumour in all the patients was variable. In 2 cases each, the tumour was located in the head, body of the pancreas and tail of the pancreas respectively. In 1 case, the tumour involved both head and body of the pancreas. In another patient, tumour was located in body and tail. Radiologically, CT scan revealed both solid and cystic components and a diagnosis of SPT was offered in all the cases. In 1 case, a diagnosis of pancreatic endocrine tumour was made. The surgery performed for tumours located in head of the pancreas was Whipple's resection. For tumours in the body, excision of the tumour was performed. For tumours located in the tail, distal pancreatectomy and splenectomy was done. In one case, differential diagnosis of gastrointestinal stromal tumour (GIST) was made intraoperatively as the patient presented with mass per abdomen and CT scan revealed a large mass compressing duodenum.

On gross examination, the tumours were well encapsulated in all the cases. Size of the tumour varied from 3 cm to 15 cm. Both solid and cystic areas were present. 3 specimens showed foci of haemorrhage. Microscopically, the tumour tissue was arranged in papillary pattern with thin vascular core. At few areas they were arranged in solid pattern with cystic spaces. Individual cells were having moderate amounts of eosinophilic cytoplasm. Nucleus was round to oval, vesicular with nuclear grooving and inconspicuous nucleoli in some. Foci of haemorrhage and necrosis were also present. In 1 case, stroma showed hyalinisation.

Of the 9 cases radiologically diagnosed as SPT, one case on histopathology was confirmed as serous cystadenoma of the pancreas and other as pancreatic neuroendocrine tumour which showed a strong positivity for chromogranin and negative for vimentin, beta catenin and PR receptor. The case radiologically diagnosed as pancreatic endocrine tumour turned out to be SPT. The sections were subjected to a panel of immunohistochemical markers which included beta catenin, vimentin, CD 10, Progesterone receptor (PR), Chromogranin, Neuron specific Enolase and Ki-67. Most of the cases showed strong nuclear positivity for PR receptor, vimentin showed cytoplasmic positivity, while beta catenin showed positivity for both. CD 10 and NSE positivity was seen in 2 cases. All the 8 cases were negative for chromogranin. Of the 8 cases, one showed Ki-67 index of 20%, rest of the cases showed very low mitotic activity.



Sl. No	Age/Sex	Presenting Symptoms	Location of the Tumour	Surgery Performed
1	11/f	Abdominal Pain	Head of the Pancreas	Whipple's Resection
2	26/f	Abdominal Pain	Head & body of the Pancreas	Whipple's Resection
3	21/f 11/f	Mass Per Abdomen	Tail of the Pancreas	Distal Pancreatectomy+ Splenectomy
4	48/m	Mass Per Abdomen & Pain	Tail of the Pancreas	Distal Pancreatectomy+ Splenectomy
5	18/f	Abdominal Pain	Head of the Pancreas	Whipple's Resection
6	19/f	Mass Per Abdomen	Body and Tail of the Pancreas	Excision of the Tumour
7	26/f	Mass Per Abdomen	Body of the Pancreas	Excision of the Tumour
8	35/f	Mass Per Abdomen & Pain	Body of the Pancreas	Excision of the Tumour

Table 1: Clinical Details of the Patients

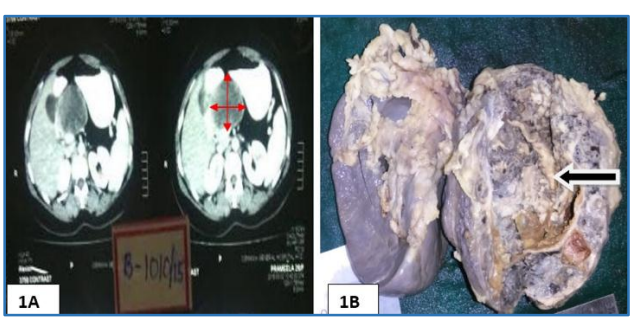


Fig. 1A: CT scan Imaging Depicting Tumour in the Head and Body of the Pancreas.
Fig. 1B: Distal pancreatectomy+ splenectomy Performed For Tumour Located in the Tail of the Pancreas

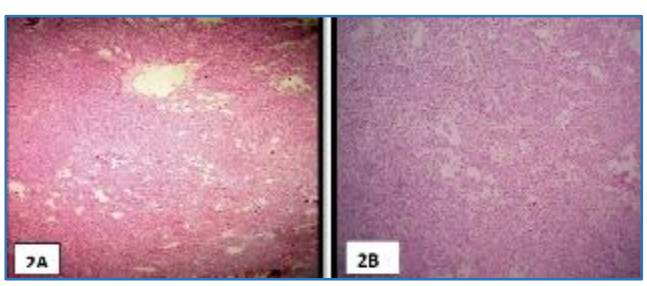


Fig. 2A: Scanner View of the Tumour.
2B: Low Power Magnification Showing Papillary Pattern

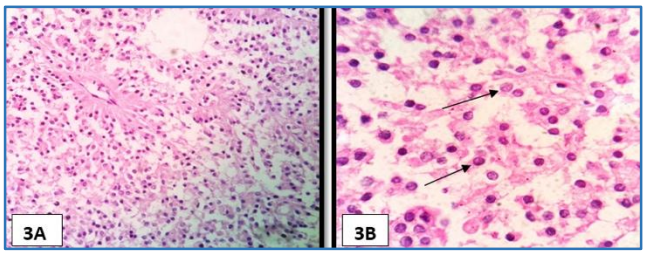


Fig. 3A: 40x Magnification of the Tumour
Fig. 3B: Tumour Cells Showing Grooving of the Nucleus

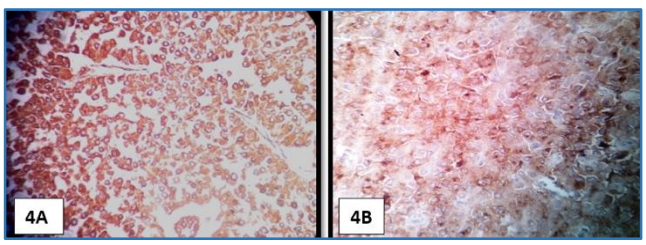


Fig. 4A: IHC - Beta Catenin Positive
Fig. 4B: Vimentin - Positive

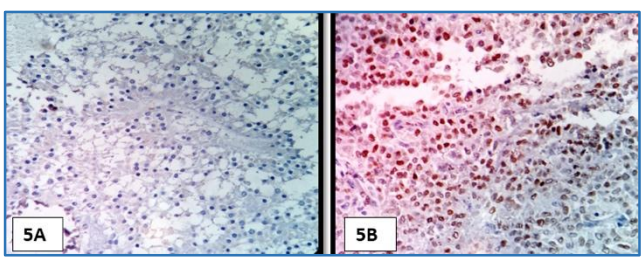


Fig. 5A: Chromogranin Negative
Fig. 5B: Progesterone Receptor - Nuclear Positivity

DISCUSSION: SPT is considered a rare tumour of low grade malignancy potential.[2] It is most commonly seen in young females between age group 10-25 yrs. SPT is an unique tumour with no clear origin as it lacks clear evidence of ductal, acinar or endocrine differentiation.[3] The most favoured theory is that SPT originates from multipotent primordial cells while others suggest an extrapancreatic origin, from genital ridge angle-related cells.[4],[5],[6] Its incidence has been increasing lately because of improved imaging modalities available and histopathological correlation. In TB Patil et al study, over 10 years, 14 cases were reported. Rao et al reported 8 cases over a 3-year period. In the present study, we have 8 cases over a period of 2 years. Female preponderance [F: M 7: 1] was observed in the present study in concordance with Martin RC, Klimstra DS et al[3],[7],[8]

Female preponderance is probably due to proximity of primordial pancreatic cells to the ovarian ridge during development.[9] SPT can occur in male patients and it has an indolent behaviour.[10] Of the 8 cases, one was a 48-year-old male. SPT with malignant behaviour seems more prevalent in the paediatric population.[7] In present study, age ranged from 11 years-48 years. Youngest patient was a female and oldest was a male.

In a large retrospective review of 718 SPT cases, mean age was 21.97 years.[11] Patients usually present with intermittent abdominal pain in epigastrium and/or palpable abdominal mass.[12],[13] In the present study, most of the patients presented with mass per abdomen followed by pain abdomen. Maha Arafah et al have reported hypoglycaemia as a presenting symptom which has been attributed to the presence of multiple foci of hyperplastic islets cells,[9] Imaging studies of SPT show a well-circumscribed, heterogeneous, solid and cystic mass. Calcification may be seen. Trivedi et al and Sommer et al have suggested that CT scan and Endoscopic ultrasonography are more sensitive and specific and have shown more accuracy in diagnosing SPT.[14],[15] CT scan was done in all our cases. In our study, tumour was located mostly in the body of pancreas followed by head and tail respectively. Tumours can arise from an extrapancreatic location and a case of SPT arising from heterotopic pancreatic tissue located in the mesocolon has been reported.[16-17]

The tumour was well encapsulated in all the cases and the gross and microscopic features was in concordance with the literature available. Aggregates of foamy histiocytes, cholesterol clefts and cytoplasmic vacuolisation can also be seen.[5] None of our cases showed infiltration into the adjacent parenchyma. Immunohistochemically, most SPTs were immunoreactive for Progesterone receptor [PR], vimentin (vim), beta catenin in concordance with other studies. CD10 showed membrane positivity in few cases. Occasionally, neuron specific enolase and synaptophysin can be positive.[8] Tumour is nonreactive for S-100, CA 19.9 and chromogranin. The main differential diagnosis of SPT includes pancreatic neuroendocrine tumour, acinar cell carcinoma, papillary mucinous carcinoma and intraductal papillary mucinous tumour (IPMT). Pancreatic neuroendocrine tumour has a distinct clinical feature like hypoglycaemia. Macroscopically, it is mostly solid and microscopically pseudopapillary structures usually are not seen.

In most patients, the tumour has an indolent clinical course. Local invasion, recurrence (5-7%) or metastasis to peritoneum and liver can occur.[3],[16],[18] Malignant transformation could occur in 15% of adults and 13% of children with the risk rising in males and advancing age.[19],[20] All the cases in our study were followed up for 6 months to 1 year and none of the cases presented with recurrence or metastasis.

CONCLUSION: SPT is not an uncommon tumour now with improved diagnostic modalities and awareness. It is important to diagnose SPT when a young female presents with mass per abdomen or pain abdomen. CT scan helps in providing an accurate diagnosis. Endoscopy guided FNAC may be attempted which helps in aiding diagnosis. In spite of it being termed as a tumour of low malignant potential, surgical resection offers good prognosis.

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