Solid Pseudopapillary Neoplasm of Pancreas – A Case Series from Bangalore, India

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ABSTRACT

BACKGROUND

Solid pseudopapillary neoplasm (SPN) of pancreas is a rare epithelial tumour of low malignant potential. SPN accounts for less than 1 to 2 % of exocrine pancreatic tumours. It mainly affects women between the second and third decade of life, and its management is not well defined. The aim of this study was to report clinicopathological characteristics of SPN and its outcome.

METHODS

A retrospective study was conducted in a tertiary care centre from January 2015 to December 2019. All patients who were diagnosed and treated as SPN of pancreas in our institute were retrospectively reviewed. A data of the characteristics of these patients was developed, including age, gender, size, location of tumour, treatment, histopathological and immunohistochemical features.

RESULTS

Six patients were diagnosed as having SPN of pancreas, during the 5-year period. All 6 patients were female. Youngest age of occurrence was 15 years. Maximum age was 41 years. Average age was 25 years. All patients were symptomatic and the most common symptom was dull aching upper abdominal pain. Contrast enhanced computed tomography (CECT) was done for all patients. 3 patients had typical features of SPN. Endoscopic ultrasound (EUS) was done for 4 patients and EUS fine needle aspiration cytology (FNAC) was done for 3 patients. Patients were provided with procedure details and informed consent was taken. All patients were subjected to surgical treatment. Out of six patients, two underwent laparoscopic spleen preserving distal pancreatectomy, two patients underwent classical Whipple's procedure and two patients had undergone median pancreatectomy.

CONCLUSIONS

SPN are rare neoplasms, typically affecting young females without clear histogenesis and with a malignant potential. Appearance from imaging studies can be adequate to guide surgical resection without pre-operative pathological assessment. But in unclear cases, EUS-FNAC with immunohistochemistry helps in establishing a pre-operative diagnosis. Surgical resection should be offered when feasible. Prognosis of SPN of the pancreas is good due to its favourable biological features, even in the presence of distal metastasis.

KEYWORDS

Solid Pseudopapillary Neoplasm (SPN)

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BACKGROUND

The first description of the disease was published by Frantz in 1959. These tumours were subsequently reclassified as solid pseudopapillary tumours (SPTs) by the World Health Organization (WHO) in 1996, and then as solid pseudopapillary neoplasms (SPNS) in 2010.[1] They are classically presented as a solitary, large, well-circumscribed lesion, which can have a completely cystic, mixed cystic and solid, or a purely solid appearance on abdominal imaging.

SPN rarely metastasizes and even in disseminated cases, surgical treatment is still an option with a favourable outcome. Due to its unusual behaviour and rareness, SPN of the pancreas is often associated with diagnostic and therapeutic challenges. Hence, the correct identification and treatment of these fewer common neoplasms of the pancreas is becoming increasingly important. An increasing incidence has been recorded during the last 15 years, which most likely is due to an increased use of computed tomography (CT) scans and magnetic resonance imaging (MRI), since many tumours are slow growing and found incidentally.[2]

The aim of this study is to report clinicopathological characteristics of SPN and its outcome**.**

METHODS

A retrospective study was conducted in a tertiary care center, Bangalore Medical College & Research Institute, Bangalore, India from 2015 to 2019. All patients who were diagnosed and treated as SPEN in our institute were retrospectively reviewed**.** This is a case series study. Data on clinical features, intra operative findings, pathological reports were retrieved from patient records. Patients were examined according to standardized procedures. All patients had a pre-operative triple-phase CT scan and if needed supplementary MRI / EUS. Follow-up was done with clinical examination, blood investigations and ultrasound abdomen once in 6 months.

RESULTS

During the 5-year period, 6 patients were diagnosed with SPN. All 6 patients were female. Youngest age of occurrence was 15 years. Maximum age was 41 years. Average age was 25 years. All patients were symptomatic and the common symptom was dull aching upper abdominal pain. Two patients had tumour in tail of pancreas, two in the head, and the other two in head and neck.

CECT in 5 patients had typical features of SPN. One patient had suspicion of neuro endocrine tumours (NET) which is the most common differential diagnosis of SPN and the other one patient had suspicion of mucinous cystadenoma in CT. In CT, one patient had tumour with external compression in portal vein confluence.

Sl. No.	Age / Sex	Location of Tumour	Surgery
1	19 / F	Tail of pancreas	Laparoscopic distal pancreatectomy
2	41 / F	Tail of pancreas	Laparoscopic distal pancreatectomy
3	15 / F	Head of pancreas	Whipple's procedure
4	25 / F	Head of pancreas	Whipple's procedure
5	20 / F	Head & neck of pancreas	Median pancreatectomy
6	32 / F	Head & neck of pancreas	Median pancreatectomy

Table 1. Tumour Location and Type of Surgery

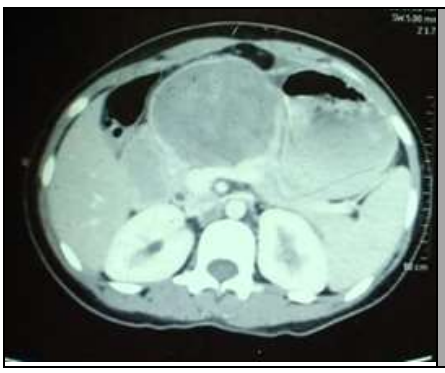

Figure 1. CT-Scan: Well Defined Altered Density Lesion at Head of Pancreas

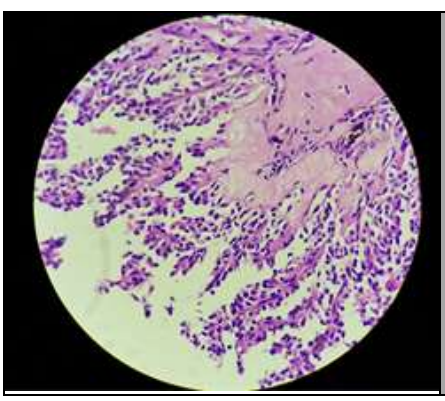

Figure 2. Tumour Cells Arranged in Pseudopapillary Pattern & Stroma Reveal Hyalinisation

EUS was done for 4 patients and EUS FNAC was done for 3 patients. One was diagnostic of SPN and other patient's EUS FNAC was? neuro endocrine tumour and the third one was non diagnostic aspirate.

All patients were subjected to surgical treatment. Out of six patients, two underwent laparoscopic spleen preserving distal pancreatectomy, two patients underwent classical Whipple's procedure and two patients had median pancreatectomy. Two distal pancreatectomy patients developed grade A pancreatic leak, which was managed conservatively and one among them had post-operative bleeding, which was treated conservatively with blood transfusion. This patient's post-operative histopathology (HPE) report showed capsular and lymphovascular invasion. Margins and lymph nodes were negative for tumour.

One patient who underwent median pancreatectomy with pancreatico gastrostomy, had hematemesis on post-operative day one. Emergency laparotomy showed spurter at pancreatico gastrostomy anastomosis site, which was ligated. Other patient who underwent median pancreatectomy had multiple peri pancreatic and peri choledochal lymph node. Patients who underwent Whipple's procedure had 24 and 32 lymph nodes harvested respectively. All were negative for malignancy. None of the patients received neo adjuvant or adjuvant therapy. All

patients were in follow up once in six months without disease recurrence.

DISCUSSION

SPN is a very rare type of pancreatic neoplasm with a low rate of progression to malignancy. The symptoms of SPN are usually non-specific, with abdominal pain being the most common, accounting for approximately 37 % of the cases. Other signs and symptoms such as abdominal fullness, jaundice, nausea, vomiting, anorexia, and weight loss may also be present. Approximately one-third of the patients are asymptomatic.[3]

Routinely, three imaging modalities were used to evaluate pancreatic lesions: trans-abdominal ultrasound (US), CT scanning, and magnetic resonance imaging. Trans-abdominal US, while having the advantage of being readily available and inexpensive, is operator-dependent, and is limited in its ability to visualize the entire pancreas. Furthermore, the sensitivity of US for characterization of pancreatic cystic processes is limited by significant bowel gas. Radiologically CECT and MRI both are almost equally diagnostic, MRI is slightly better than CT in identifying capsule, haemorrhage and cystic degeneration while CT is advantageous in its ability to detect central calcifications. On CT scan these tumours are well encapsulated, hypodense with various solid cystic components.[4] Typical SPN on CT has surrounding capsule with demarcation between solid and cystic components and hypoattenuation during pancreatic phase. On MRI typically, a large, well-defined, encapsulated lesion with heterogeneous high or low signal intensity on T 1 - weighted, heterogeneous high signal intensity on T2 weighted, and early peripheral heterogeneous enhancement with progressive fill-in is found on gadolinium-enhanced dynamic MRI. These features help differentiate this rare tumour from other pancreatic neoplasms.[5] Atypical imaging features include invasion into surrounding pancreatic parenchyma, adjacent organs and vascular invasion are considered to be relevant with their aggressive behaviours, which may cause the tumours to be prone to recur.

The diagnosis of SPN with EUS has been increasing in popularity. Patients who do not have a clear diagnosis of an SPN using only image studies may benefit from EUS.[6] Pancreatic cystic neoplasms, thought to be pre-malignant or malignant, make the use of EUS, with or without fine needle aspiration, an attractive option in work up. Aspirated cystic fluid should be evaluated for biochemical and cytological analysis. The biochemical tests which should be routinely ordered are amylase and CEA level. But it may not be accepted by others because of the uncertainty in diagnosis and the possible tumour spread. The F-18 fluoro deoxy glucose (FDG) uptake of SPN on positron emission tomography (PET) is not well studied. FDG uptake of SPN on PET is related to proliferative index, tumour cellularity or histological malignancy. SPN having a greater proportion of solid component has more FDG uptake than with haemorrhagic or cystic component. Kim et al.[7] reported that PET CT can lead to high false positivity in SPEN but useful in detecting hepatic metastasis. Studies showed that tumour markers such as carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA19-9) were usually within normal ranges in patients with this disease. Thus, routine tumour markers are of no value to predict malignant SPN.[8]

On gross, these tumours are well encapsulated. On microscopic examination there is solid cystic mass with center of haemorrhagic and necrotic material and peripherally by solid tissue. Pseudopapillary formation, foamy histiocytes, nuclear grooves are characteristic finding of SPN. On immunohistochemistry (IHC), they are characteristically positive for vimentin, CD 10, AACT, atypical; antibody test (AAT), β-catenin, neuron-specific enolase (NSE), Syn, and progesterone receptors (PRs) all of which are very useful in differentiating them from endocrine pancreatic tumour cells.[9] In our study all the patients were stained strongly for vimentin, CD 10, β-catenin.

The origin of solid pseudopapillary tumours still remains unclear. These neoplasms have been suggested to have a ductal epithelial, neuroendocrine, multipotent primordial cell, or even an extra-pancreatic genital ridge angle-related cell origin.[10]

Surgical resection is the treatment of choice for SPN and organ preservation is advocated if feasible[11] SPN exhibits benign or low-grade malignancy and is usually surrounded by a pseudo capsule. Incidence of lymph node metastasis is very rare in SPN and hence routine lymphadenectomy is not indicated.[12] According to the location of the tumour, distal pancreatectomy with or without splenectomy, pancreaticoduodenectomy (Whipple operation), median pancreatectomy or enucleation can be performed. In our series two patients underwent laparoscopic distal pancreatectomy, two patients underwent median pancreatectomy and in other two Whipple's procedure was done. Metastasectomy of the liver is advocated at the time of primary resection or even for the recurrences when feasible.[13] The role of chemotherapy and radiation in SPN is not clear. On histology, feature of malignant tumour is angioinvasion, perineural invasion, and deep pancreatic tissue invasion.[14] High-risk features of tumour recurrence are larger tumours > 5 cm, lymphovascular invasion, lymph node metastasis, synchronous metastasis and positive margin.[15] The overall five - year survival rate of patients with SPN is about 95 %.[16]

CONCLUSIONS

SPN are rare neoplasms, typically affecting young females without clear histogenesis and with malignant potential. Appearance from imaging studies can be adequate to guide surgical resection without pre-operative pathological assessment. But in unclear cases, EUS-FNAC with immunohistochemistry helps in establishing a pre-operative diagnosis. Surgical resection should be offered when feasible. Prognosis of SPN of the pancreas is good due to its favourable biological features, even in the presence of distal metastasis.

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REFERENCES

[1] Dai G, Huang L, Du Y, et al. Solid pseudopapillary neoplasms of the pancreas: clinical analysis of 45 cases. Int J Clin Exp Pathol 2015;8(9):11400-11406.
[2] Law JK, Ahmed A, Singh VK, et al. A systematic review of solid-pseudopapillary neoplasms: are these rare lesions. Pancreas 2014;43(3):331-337.
[3] Yu PF, Hu ZH, Wang XB, et al. Solid pseudo papillary tumor of the pancreas: a review of 553 cases in Chinese literature. World J Gastroenterol 2010;16(10):1209-1214.
[4] Park MJ, Lee JH, Kim JK, et al. Multidetector CT imaging features of solid pseudopapillary tumours of the pancreas in male patients: distinctive imaging features with female patients. Br J Radiol 2014;87(1035):20130513.
[5] Ventriglia A, Manfredi R, Mehrabi S, et al. MRI features of solid pseudopapillary neoplasm of the pancreas. Abdom Imaging 2014;39(6):1213-1220.
[6] Law K, Stoita A, Wever W, et al. Endoscopic ultrasound-guided fine needle aspiration improves the pre-operative diagnostic yield of solid-pseudopapillary neoplasm of the pancreas: an international multicenter case series (with video). Surg Endosc 2014;28(9):2592-2598.
[7] Kim YI, Kim SK, Paeng JC, et al. Comparison of F-18-FDG PET/CT findings between pancreatic solid pseudopapillary tumor and pancreatic ductal adenocarcinoma. Eur J Radiol 2014;83(1):231-235.
[8] Yang F, Fu DL, Jin C, et al. Clinical experiences of solid pseudopapillary tumors of the pancreas in China. J Gastroenterol Hepatol 2008;23(12):1847-1851.
[9] Geers C, Moulin P, Gigot JF, et al. Solid and pseudopapillary tumor of the pancreas--review and new insights into pathogenesis. Am J Surg Pathol 2006;30(10):1243-1249.
[10] Eder F, Schulz HU, Röcken C, et al. Solid-pseudopapillary tumor of the pancreatic tail. World J Gastroenterol 2005;11(26):4117-4119.
[11] Romics L Jr, Oláh A, Belágyi T, et al. Solid pseudopapillary neoplasm of the pancreas--proposed algorithms for diagnosis and surgical treatment. Langenbecks Arch Surg 2010;395(6):747-755
[12] Tipton SG, Smyrk TC, Sarr MG, et al. Malignant potential of solid pseudopapillary neoplasm of the pancreas. Br J Surg 2006;93(6):733-737.
[13] Lam KY, Lo CY, Fan ST. Pancreatic solid-cystic-papillary tumor: clinicopathologic features in eight patients from Hong Kong and review of the literature. World J Surg 1999;23(10):1045-1050.
[14] Zhang H, Liang TB, Wang WL, et al. Diagnosis and treatment of solid pseudo papillary tumor of the pancreas. Hepatobiliary Pancreas Dis Int 2006;5(3):454-458.
[15] Gao H, Gao Y, Yin L, et al. risk factors of the recurrences of pancreatic solid pseudopapillary tumors: a systematic review and meta-analysis. J Cancer 2018;9(11):1905-1914.
[16] Papavramidis T, Papavramidis S. Solid pseudopapillary tumors of the pancreas: review of 718 patients reported in English literature. J Am Coll Surg 2005;200(6):965-972.