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A Multicenterstudyof Solid-Pseudo Papillary Tumor of the Pancreas

Ravi Krishnappa[a], Vijay Yadav[b], Tarun Batra[c]

Author's Affiliation: [a]Assistant Professor, Department of Surgical Oncology, JSS Medical College, Mysore, Karnataka 570015, India. [b]Consultant, Oncosurgeon, Manipal Hospital, Jaipur, Rajasthan 302013, India. [c]Assistant Professor, Department of Surgical Oncology, BHU University, Varanasi, Uttar Pradesh 221005, India.

Corresponding Author: Ravi Krishnappa, Assistant professor, Department of Surgical Oncology, JSS Medical College, Mysore, Karnataka 570015, India.
E-mail: ravisashkrish@gmail.com

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Abstract

We report a retrospective study of Solid Pseudo Papillary Tumor (SPPT) of the pancreas, a rare entity, which is a low grade malignant abdominal tumor at manyinstitute; this is relatively frequent in young women, with a discussion about its presentations and management.

In our study we report 20 Solid pseudo papillary tumors of pancreas over a period of 10 year. 15 out of 20patients in our study were female with the median age at diagnosis was 27.25 years (range 11-58 years). Abdominal pain and lump were the most common presenting complaints. Median size was 10 cm (range 3-17 cm) Distal Pancreatectomy(11 pts) and Pancreatico-duodenectomy (6pts) were the most common surgeries performed. 2 patients were inoperable and in one patient wide excision were done.

Diagnosis of Solid pseudo papillary tumors of pancreas should be considered in young women presenting with a large solid cystic pancreatic mass and the treatment of choice consists of surgical resection. The outcome after surgical resection is excellent with long term survival of 90%.

Keywords: Solid Pseudo Papillary; Rare Tumor; Multi Centric; Pancreas.

Introduction

Solid pseudopapillary tumor of the pancreas is a rare condition of which only about 450 cases have been reported in literature. This tumor was first described in 1959. It is known as FRANTZ tumor, named after the author who first described it, also as solid cystic tumor [1]; papillary epithelial neoplasia; solid and papillary epithelial neoplasia; and papillary epithelial tumor. The origin of solid-pseudopapillary tumor has not ever been clarified. It is argued that it does originate either from ductal epithelium [2], Acinar cells [3], or endocrine cells [4]. Another hypothesis is that this tumor arises from pluripotent embryonic cells of the pancreas or from the ridge ovarian analogue related cells, which were attached to the pancreatic tissue during early embryogenesis [5].

Solid pseudo papillary tumor of the pancreas has a tendency to predominantly affect young women [6,7,8]. This tumor rarely affects men. It is characterized by a long asymptomatic course and nonspecific symptoms. Therefore, it is not uncommon that solidpseudopapillary tumor is detected only when it has grown to a remarkable size (8-10cm) [7]. One feature of this tumor is its low malignant potential. Although the liver is found to be the mostly affected site by metastases, these are only rarely seen [5,7,9].

Objective

To evaluate the Presentation, outcome and management of Solid-Pseudo Papillary Tumor of the Pancreas

Material and Methods

This is a retrospective study to assess different presentation, diagnostic methods and management of this rare disease in India. We reviewed the patient's files with pathology report of pseudopapillary tumor of pancreas that were operatedby Department of Surgical Oncology at Hospital, Jaipur, Rajasthan, Indi, JSS Medical College, Mysore, Karnataka and from

the BHU university, Varanasi, Uttar PradeshBetween the year 2000 to 2010. We collected and recorded these data: age, sex, signs and symptoms, past medical history, physical examinations, pre-operational diagnostic measures, laboratory data, per-operational findings, macroscopic and microscopic features of tumors, post-operation and follow up of the patients. In the follow up period, the patients were visited in planned intervals (after 2 weeks, 3 months, 6 months, and then every 6 months) by the operating surgeon from 2000 to 2010. The group consisted of 15 females and 5 males at mean age of 27.25years. Abdominal pain and lump were the most common presenting complaints. Median size was 10cms (range 6-17 cm). Distal Pancreatectomy (11pts) or Pancreaticoduo-denectomy (6pts) were the most common surgeries performed.

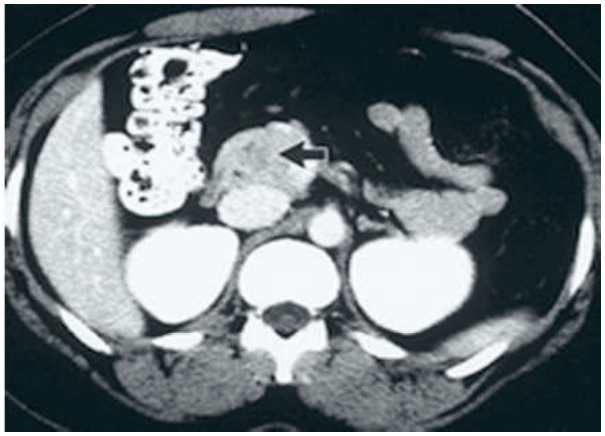

Fig. 1: Ct Scan View Of Solid Pseudopappilary Tumour Of Pancreas

In almostall cases, primarily, an abdominal sonography showed a cystic mass which was followed by a spiral abdominal and pelvic CT scan with oral and intravenous contrast that confirmed a cystic mass in the upper abdomen, posterior to the stomach with multiple solid components and mural enhancement.Laboratory data including complete blood count, blood chemistry, serum amylase level and coagulation profiles were normal in all cases.

Histopathology

Alm et al. gave the first cytological description of SPT about 21 years ago. Nadler et al. And Chavez et al. have published case reports on SPT diagnosed on radio logically guided fine needle aspiration cytology. In a series of 42 cases evaluated by fine needle aspiration, Nguyen reported that the most striking cytological features are the high cellularity and the presence of branching papillary fronds composed of central thin fibrovascular stalks covered by one to several layers of cuboidal or cylindrical tumour cells. These distinct cytological features are shared by all patients with SPT including older patients.

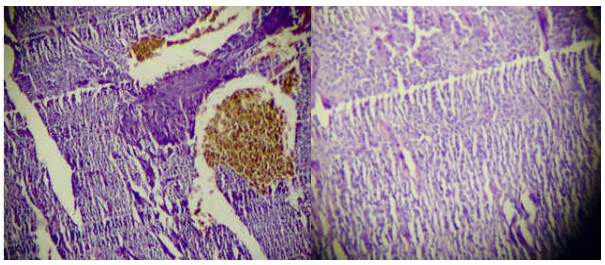

Fig. 2: Histopathological View of Solid Pseudopappilary Tumour

Results

From 2000 to 2010, twenty patients were operated in our center with final pathologic diagnosis of solid pseudopapillary tumor of pancreas. The group consisted of 15 females and 5males. The disease was presented as abdominal pain and lump in abdomen or fullness of the epigastric region.

None of them had positive drug history including hormonal medications. Physical examination revealed a well defined abdominal mass occupying the epigastrium and left hypochondriac region in three cases and the other two had no positive abdominal finding. In all cases, primarily, an abdominal sonography showed a cystic mass which was followed by a spiral abdominal and pelvic CT scan with oral and intravenous contrast that confirmed a cystic mass in left side of upper abdomen, posterior to the stomach with multiple solid components and mural enhancement. One of our patients, a small boy presented with pain in left loin and CT scan showed mass anterior kidneynear tail of pancreas. Solid pseudo papillary tumor of pancreas was mentioned as a possible differential diagnosis in the radiology reports. Laboratory data including complete blood count, blood chemistry, serum amylase level and coagulation profiles were normal in all cases. In macroscopic examination, there was a spherical or partially spherical lesion with maximum diameter of 12 cm (in two cases), tan grey in color covered by capsule and lobulated. In cut sections multiple cysts containing necrotic material or bloody fluid was seen. In serial sections for microscopic evaluation, a low grade invasive malignant neoplasm of pancreas characterized by solid cellular sheets with papillary fronds composed of rather uniform epithelial cells arranged around small and large vessels and cystic spaces with fibrinohyalinized borders were observed in all cases. Postoperatively patients were hospitalized for 8-14days and then

discharged without complications. Overall median survival after surgical resection is 90% in the long term. In our study Mean follow up was 44.4 months, 2 patients were inoperable and Gastrojejunostomy was done. In 1 patient only wide excision was done for tumour on ampulla.

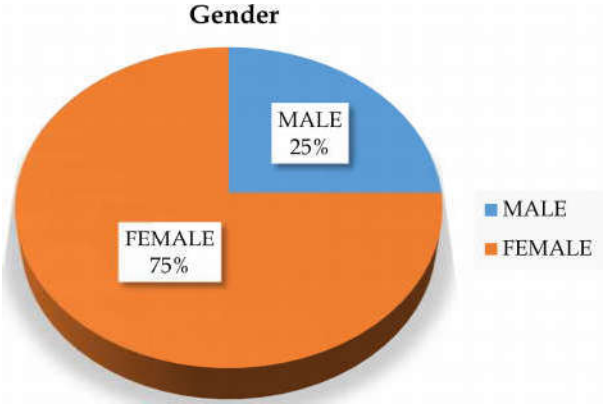

Table 1: Male-Female Ratio (5:15)

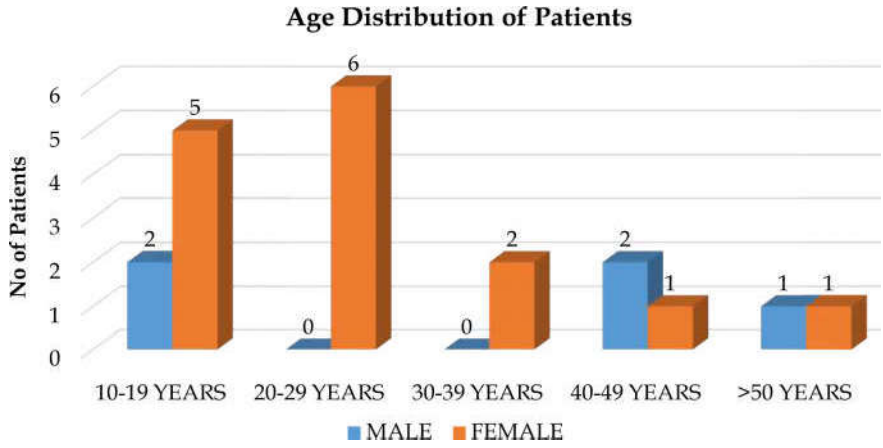

Table 2: Age at presentation

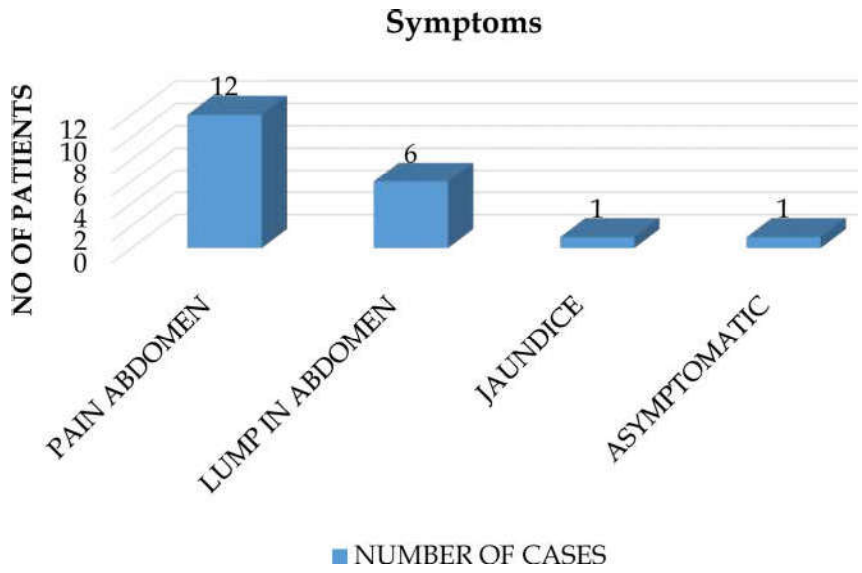

Table 3: Symptoms

Table 1: Type of surgery

Type of Surgery	No of Patients
Distal Pancreatectomy	11
Pancreaticoduodenectomy	6
Local Wide Excision	1
Inoperable	2

Profile of all the cases in given in the below box in detail

S. No.	Age	Gender	Presentation	Imaging studies	Location	Surgery	Outcome
1	15Y	Female	Lump in abdomen	CT scan→ 125/89/120 mm heterogeneous mass	Head and uncinate process	Whipples surgery	Died after 17 months of surgery cause unknown
2	20Y	Female	Lump in abdomen	CT scan→ 17/10/16 mm heterogeneous mass with central necrotic area	Body and tail	Distal Pancreatectomy	Asymptomatic after 1 Year of surgery
3	20Y	Female	Epigastric pain	CT scan→ 55/57/50mm lobulated heterogeneous moderately enhancing mass	Head and neck	Whipples Surgery	Asymptomatic after 8 years of surgery
4	58Y	Male	Epigastric pain, recurrent attacks of jaundice	CT scan→ calcification in the head and 55/30/80mm heterogeneously enhancing mass MPD and CBD dilated	Ampulla	Wide excision	Asymptomatic after 7 years of surgery
5	52Y	Female	Epigastric pain	CT scan→ well defined cystic lesion with internal calcifications and septations 28/26mm	Tail	Distal pancreatectomy and splenectomy beyond the sm vessels	Asymptomatic after 7 years of surgery
6	35Y	Female	Epigastric lump	CT scan→ 33/44 mm cystic lesion heterogeneously enhancing lesion	Body and tail	Distal Pancreatectomy	Alive after 6 years of surgery
7	30Y	Female	Pain in abdomen	CT scan→ well defined heterogeneously enhancing lesion, 40/60/70 mm	Body and tail	Distal Pancreatectomy	Asymptomatic after 2 years
8	40Y	Male	Pain in abdomen	USG : Well defined lesion with heterogeneous echotexture,60/40/30mm	Body and tail	Distal Pancreatectomy	Asymptomatic after 3 yrs
9	12Y	Male	Pain in abdomen	USG: 86x59 mm mass lesion ant to LT. kidney near tail of pancreas. CT Scan: 110x50 mm hypodense lesion near tail of pancreas. USG guided Bx s/o Germ cell Tumour	tail	Distal pancreatectomy	Asymptomatic after 8 years
10	16Y	Female	Pain+Lump in abdomen	CT scan→ Well defined heterogeneously enhancing 80/65 mm lesion UsgBx→SPEN	Head	Inoperable mass, Bx done	Lost to follow up
11	24Y	Female	Lump in abdomen	CT scan→ Well defined heterogeneously enhancing 100/80 mm lesion	Tail	Distal pancreatectomy + splenectomy	Asymptomatic after 8 years
12	45Y	Male	Pain in abdomen	CT Scan→ cystic lesion heterogeneously enhancing lesion 40/50/40 mm	Tail	Distal Pancreatectomy	Asymptomatic after 8 years
13	29Y	Female	Asymptomatic	CT Scan→ cystic lesion heterogeneously enhancing lesion,70/80/60mm	Tail	Distal Pancreatectomy	Asymptomatic after 8 years
14	20Y	Female	Pain in abdomen	CT scan→ Well defined heterogeneously enhancing 100/60/80mm lesion	Head & body	Inoperable mass, G-J &Bx done	Lost to follow up
15	16Y	Female	Pain in abdomen	CT scan→ heterogeneously enhancing 76/70mm lesion USG Bx→SPEN	Tail	Distal pancreatectomy + splenectomy	Asymptomatic after 4 months than lost to follow up
16	26Y	Female	Pain in abdomen	CT scan→ Well defined heterogeneously enhancing, predominantly cystic mass lesion 30/40/30 mm USG→60/75mm exophytic mass in pancreatic head	Head	Whipples surgery	Revision G-J for stomal obstruction after 1 month, later follow up is asymptomatic

17	17Y	Female	Pain+Lump in abdomen	CT scan → Well defined encapsulated heterogeneously enhancing 120/100 mm lesion USG→100/130mm hypo echoic with internal cystic component CT guided Bx→ SPEN	Head	Whipples surgery	Asymptomatic after 3 year than lost to follow up
18	11Y	male	Pain in abdomen	CT scan -> solid papillary neoplasm of head of pancreas. 70x60x50mm. USG -> mass 2nd part of duodenum with areas of necrosis. CT guided Bx and FNAC ->pancreatoblastoma	Head	Pylorus preserving Pancreaticoduodenectomy	Asymptomatic after 3 years
19	35	female	Pain in abdomen	CT scan-> heterogeneously enhancing lesion in C loop. 100x80x60 mm. USG-> NAD CT guided Bx -> papillary cystic neoplasm	Head	Pylorus preserving Pancreaticoduodenectomy + right hemicolectomy	Asymptomatic after 3 years
20	24Y	female	Pain in abdomen	CT scan -> 66x65 mm solid mass in body and tail of pancreas with necrotic areas in it. No calcification USG-> 66x60 mm well defined mix echogenic mass lesion in tail of pancreas with necrotic areas within it.	Body +tail	Distal pancreatectomy+ splenectomy	Asymptomatic after 2 tears

Discussion

There is a variety of cystic neoplasm in the pancreas. These include benign serous cystic neoplasm, benign and malignant mucinous cystic neoplasms, and benign and malignant forms of intraductal papillary-mucinous neoplasms. It is important not to assume that all fluid-filled pancreatic abnormalities represent pseudocysts, or that a dilated pancreatic duct represents only chronic pancreatitis. The presence of a solid component in a cystic lesion, septations within the cyst, and the absence of a clinical history of pancreatitis are factors that should alert the surgeon to the possible presence of a neoplasm

However, SPPT is the preferred term for a distinctive type of pancreatic tumor, also known as papillary and solid epithelial neoplasm, papillary-cystic neoplasm and cystic-solid papillary carcinoma. Solid-pseudopapillary tumor of the pancreas has a tendency to predominantly affect young women aged between 15 and 35 years [6,7,8]. Age is reported to range from 8to70 years [2]. In our series age of presentation ranged from 11 to 58 years. This tumor rarely affects men and is characterized by a long asymptomatic course and nonspecific symptoms. Therefore, it is not uncommon that solid-pseudopapillary tumor is detected only when it has grown to a remarkable size (8-10cm) [8]. A tumor size of 20 cm in diameter has been reported once in the literature [9]. One feature of this tumor is its low malignant potential. On cross-section it often contains areas of hemorrhage and necrosis. Most cases are surrounded by a well developed capsule, but in some instances, the edges are those of a solid infiltrative neoplasm. Although the liver is found to be the mostly affected site by metastases, these are only rarely seen [5,9]. Furthermore, there are only few reports about invasive growth [2,7]. Survival time has been reported to reach up to 21 years [1].

Mao in a cumulative review of the literature found that 90% of the patients were female with a mean age of 23.9 years and male to female ratio of 1:9 [5]. In our series 88% of patients were female, withmale to female ratio was 1:9, and the median age was 25 years.

Rebhandle et al. reported 4 girls 12-16 years of age presenting with abdominal pain and mass (diameter 7-15 cm), located in tail (n=2), body and tail (n=1) and head (n=1) of pancreas [10]. In our series tumor was located in body and tail (n=9), head (n=6), ampullary region (n=1). Generally the tumor tends to be fairly benign in young females but appears more aggressive in older males whose mean age is about 10 years older than women [11].

Pseudopapillary tumor may spread outside the pancreas, particularly in the peritoneal cavity. Metastatic spread may be promoted by trauma, including tumor biopsies which should never be performed [12].

In our series two cases have shown peritoneal metastasis with encasement of superior mesenteric artery.

Given the good prognosis of the disease, it is important to make the diagnosis preoperatively, if possible so that adequate resection will be undertaken. Therefore, imaging studies should be carefully assessed; fine needle aspiration (FNAC) is sometimes considered [12].

In our cases, Abdominal ultrasound and CT-scan show a well encapsulated complex mass with both solid and cystic components and displacement of nearby structures. There may be calcifications at the periphery of the mass and intravenous contrast enhancement inside the mass suggestive of hemorrhage.

Surgery is the mainstay of treatment that is usually curative for localized disease. There are evidences for prolonged survival after adequate surgical resection even with metastases. Intra-operative frozen section may be helpful to ascertain the adequacy of the resection margins. There have been only a few reports regarding the use of radiotherapy or chemotherapy [13], so it's difficult to judge the value of such measures.

Conclusion

SPPT of the pancreas is a rare indolent neoplasm with an unclear origin that typically occurs in young females. The diagnosis depends on histological confirmation, but its appearance on imaging is fairly characteristic. We believe that SPT of the pancreas should be treated aggressively, with attempts made for complete resection, even if this requires metastatectomy. Long-term survival can be achieved with an aggressive approach to both the primary lesion and to the synchronous or metachronous metastatic lesion, predominantly found in the liver.

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