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Brief Report

Solid Pseudopapillary Tumor of Pancreas: A Clinicopathologic Report from a Single Institution in Southern Iran

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Abstract

Background: Solid pseudopapillary tumor of pancreas (SPTP) is a rare tumor of the pancreas which mostly occurs in young women. Since its first description in 1969, more than 500 cases have been reported. There have been just 2 case reports and 1 review (7 cases) from Iran. In this study, we reported our experience with 22 patients with SPT from the largest pancreaticobiliary center in Southern Iran.

Methods: During 6 years (2012–2017), 22 cases of SPTP were operated on in our center. All of these cases were recruited and after confirmation of the pathological diagnosis, clinical charts were evaluated and all the clinicopathologic findings as well as outcome of the surgery were evaluated.

Results: Among the 22 patients, 20 were female and 2 were males. The age range was 15–52 years and the tumor sizes were 3.5 to 17 cm. All of the tumors had preoperative diagnosis by imaging modalities and were operated on with no complication or recurrences. Just one case showed liver metastasis. All of the patients with SPTP in this study were alive and in good condition.

Conclusion: SPTP is not very rare in our center. In young patients presenting with pancreatic mass, especially in female patients, one of the most important diagnoses is SPTP. Conservative surgery and tumor excision is satisfactory and patients show excellent prognosis even after liver metastasis.

Keywords: Clinicopathologic findings, Solid pseudopapillary tumor of pancreas

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Introduction

Solid pseudopapillary tumor of pancreas (SPTP) is a rare tumor, accounting for 1%–2% of pancreatic tumors. It was first described by Frantz in 1959, and since then it has been called various names such as "pancreatic solid and cystic tumors", "solid cystic and papillary epithelial neoplasms" and "solid and papillary tumors". In 1996, its name was modified by the World Health Organization and was referred to uniformly as SPTP.[1]

Since its first description, there have been many case series and case reports of this tumor with experience from different centers' clinicopathologic, imaging, treatment and outcome reports of this rare tumor.[2] Our center is the largest pancreaticobiliary center in the South of the country and in this report we described our experience with this tumor for 6 years (2012–2017).To the best of our knowledge, this is the second and the largest reported series from Iran.[3-5]

Patients and Methods

During the last 6 years (2012–2017), in our center, as one of the largest referral centers of pancreaticobiliary surgery, 131 cases of pancreatic surgery has been performed, 22 (16.7%) of which have been finally diagnosed as SPTP by pathologic examination. Among these 22 cases, 20 (90.9%) were female and 2 were male patients. All of the clinical and paraclinical findings, imaging studies, treatment and surgical modalities as well as outcomes of the patients were extracted from clinical charts and direct contact of these patients. Also, pathological and immunohistochemical studies were collected from both the clinical charts and pathology reports.

Results

Twenty-two cases of SPTP were operated during 6 years (2012–2017). The age range was 15–52 (28.2 ± 13.4) years. Five patients were under the age of 18 (pediatric age group), all of which were females. There were 20 females and 2 male patients. The sizes of the tumor have been 3.5–17 (10.6 ± 4.3) cm. Majority of the tumors (16 cases) have been located in the head of pancreas (73%). Four cases have been located in the body and tail (18%) and 2 cases (9%) have been in the tail of pancreas.

The most common presenting symptom has been

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abdominal and flank pain which has been present in all of the patients. Only 2 patients (9%) had the chief complaint of weight loss and anorexia with abdominal pain. One patient (4.5%) presented with abdominal pain with nausea and vomiting. None of our cases have been incidentally detected.

Two patients have been known cases of diabetes mellitus (DM), otherwise the remainder of the patients didn't show any underlying disease.

Paraclinical findings of the patients showed normal tumor markers. In 13 patients tumor markers (Carcinoembryonic antigen and CA19-9) have been checked which were normal. Amylase and lipase have been evaluated in 15 patients. These 2 enzymes have been normal in all of the 15 cases. Range of amylase level has been 35–66 U/L (47.5 ± 11.5). This range for lipase has been 12–30 U/L (16 ± 7).

Imaging studies including CT scan and ultrasonography (US) have been performed in all of our cases; however magnetic resonance imaging (MRI) has been done only in 10 patients.

Main findings in US have been presence of a solid and cystic well-defined and well encapsulated tumor. Rarely calcification has been reported however the tumor has no connection with the pancreatic ducts. CT scan showed more or less the same findings i.e. solid to cystic, as well as capsulated mass. MRI imaging was also the same except for better evaluation of the presence of hemorrhage and peri-tumoral invasion.

Overall all of the patients had preoperative diagnosis of SPTP with the help of imaging modalities, so none of our 22 cases had fine needle aspiration or biopsy before surgery.

The surgical procedure in 16 patients with the tumor located in the head of pancreas was pancreaticoduodenectomy. The remainder with the tumor in the body and tail had either tumor enucleation in 2 patients or distal pancreatectomy with or without splenectomy (in 4 patients).

Pathologic examination of the cases showed typical findings of SPTP. None of our cases showed atypia and capsular or vascular invasion.

Immunohistochemical staining showed 100% positivity for vimentin, nuclear β-catenin, progesterone receptor, and CD10. Focal and weak positivity for chromogranin was detected in one case. None of the 22 cases showed synaptophysin positivity. Cytokeratin was focally and weakly positive in 2 cases. Ki-67 positivity was very low or negative (<2%). None of our cases showed high proliferative activity.

In only one patient liver metastasis has been detected 2 years after the initial surgery and tumor excision. No lymphnode metastasis has been identified. None of our patients had any evidence of invasion to the surrounding tissues. Follow up of our patients for 12–72 months (40.3 to 16.5) showed no recurrence and all of our 22 patients have been alive at the end of 2017.

Table 1 shows the summary of the main findings in the 22 reported cases.

Discussion

SPT is a rare pancreatic tumor which was introduced about 60 years ago. Since then, more than 500 cases have been reported, however, so far, its exact origin has not been definitely clarified and still controversies exist. There are some reports about the origin from both pancreatic exocrine and neuroendocrine cells. Some other studies suggested that SPT may arise from centroacinar cells located between pancreatic ducts and acini.[6] The genetic profile of SPT is completely different from pancreatic ductal adenocarcinoma, and there are unique genetic mutations in this tumor such as β-catenin mutation that is not common in other pancreatic tumors.[7]

In our experience, the most common presenting symptom of SPT is abdominal pain mostly located in the left upper quadrant of abdomen accompanied by flank pain. In our experience, other signs and symptoms such as weight loss, nausea, vomiting and abdominal mass have been very rare. In other studies, the same result has been reported, however, there are reports of incidental discovery of the tumor during irrelevant imaging studies. There have been reports of seven fold increase in incidence of this tumor during the last 10 years, which is most probably attributed to more accurate and precise imaging modalities.[6]

This tumor is most commonly seen in females below the age of 25 years and it's very uncommon in men.[7] In our cases collected in 6 years and consisting of 20 female patients, only 2 males were identified to have this tumor. The most common location in previous studies has been

Table 1. Summary of the Main Clinicopathologic Findings of 22 Cases

Main Clinicopathologic Findings	Number
Age	15–52(28.2 ± 13.4) years
Female/Male	20/2
The most common presenting symptom	Abdominal pain (100%)
Location	
Head	16 (73%)
Body and tail	4 (18%)
Tail	2 (9%)
Tumor size	3.5–17 (10.6 ± 4.3 cm)
Surgical procedure	
Pancreaticoduodenectomy	16 (73%)
Enucleation	2 (9%)
Distal pancreatectomy	4 (18%)
Follow up in 2017	
Alive	100%
Recurrence	0 (0%)
Liver Metastasis	1 (4.5%)

variable but in our experience the most common location was the head of pancreas.[8-11]

Radiologic studies can be very helpful for the preoperative diagnosis of SPT. The most accurate method is CT scan, which shows thick-walled cystic and solid tumor with well-defined margin and encapsulation, sometimes with calcification and internal hemorrhage.[9] MRI is very similar to CT scan but can be helpful for the cases without a definite diagnosis and also for more accurate determination of invasion and delineation of the tumor before surgery. [12] Also endoscopic US and fine needle aspiration have been reported as an accurate method for preoperative diagnosis by performing immunohistochemistry in presence of adequate number of cells.[10] In our experience, this modality has not been performed in these 22 cases.

Paraclinical findings in SPT are not significant and no specific or sensitive biomarker has been introduced yet. Tumor markers (CEA, CA19-9, etc) are all negative and amylase and lipase are rarely increased. In our experience all the measured tumor markers and pancreatic enzymes were normal.[13]

According to the WHO definition, this tumor is a low grade malignant tumor and can show invasion and metastasis, therefore, the tumor should be resected. The procedure depends on the location of the tumor and tumor excision and enucleation, distal pancreatectomy and pancreaticoduodenectomy have been the procedures of choice.[14] There have been also reports of minimally invasive surgical procedures in small SPTs located in the distal part of the pancreas.[14-16]

Histopathologically, SPT classically consists of a large and encapsulated mass composed of cystic and solid areas. Intra-tumoral hemorrhage is very common and calcifications have been reported in one-third of cases. SPT is a cellular tumor with cystic degeneration and multifocal pseudo-papillary structures. The tumor tissue is composed of solid nests of medium-sized cells with low mitosis and no or minimal atypia.[17-21] Typical immunohistochemistry shows positive vimentin, nuclear β-catenin, progesterone receptor, and CD10. Epithelial markers such as cytokeratin (CK) are negative. Neuroendocrine markers such as chromogranin and synaptophysin are either negative or weak and focal. Important differential diagnoses of SPT in histopathology are pancreatic neuroendocrine tumors which are consistently positive for chromogranin and synaptophysin as well as CK, which are most commonly negative in SPT. There are reports of focal and weak staining for CG, however, the strong and diffuse positivity of neuroendocrine tumors have not been reported.[22]

Outcome of tumors with diagnosis of SPT is good although the tumor is considered a low-grade malignant tumor with the capacity for invasion and metastasis, however, even in the presence of lymph node and liver metastasis, the prognosis is good. Presence of lymphovascular invasion, large size, atypia and mitosis have been considered as signs of aggressive behaviour and potential for lymph node and liver metastasis. High Ki-67 positivity, indicating high proliferative activity, has also been considered as a poor prognostic factor.[18] All of the above-mentioned criteria can indicate worse outcome if the patient is male.[18] Our experience in 6 years with 22 patients who were followed for 1–6 years showed an excellent prognosis. None of our patients show any evidence of cellular atypia, mitosis or high proliferative rate. There was just one case with liver metastasis that is alive and completely well. Compared to previous reports from different parts of the world with reported aggressive behaviour, invasion and metastasis in up to 15%[18-20] it seems that SPT in our population has less potential for aggressive behaviour and metastasis (4.5%). Despite metastasis, the patients with SPT have excellent prognosis and more than 10 years of survival with metastasectomy and with no further chemotherapy.[21]

In conclusion, SPT should be considered as a most probable differential diagnosis of a pancreatic tumor in a young lady, and, in our center, it is not a rare tumor. Imaging studies are very accurate for preoperative diagnosis. Also, aggressive surgery is not necessary and conservative complete excision and enucleation with less invasive procedures is satisfactory even after metastasis.

Authors' Contribution

AK: Collecting the raw data. BG: Analyzing the data, writing the paper, submission. SN and SMH: Surgery.

Conflict of Interest Disclosures

The authors have no conflicts of interest.

Ethical Statement

Not applicable.

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