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ScienceDirect

EJSO the Journal of Cancer Surgery

EJSO xx (2017) 1–5 www.ejso.com

Solid pseudopapillary neoplasm of the pancreas: Management and long-term outcome

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Accepted 1 February 2017

Available online ■ ■ ■

Abstract

Background: Solid pseudopapillary neoplasm (SPN) of pancreas is a rare pancreatic neoplasm with a low metastatic potential. Our aim was to study the clinical–pathological characteristics, and long-term outcome of this tumor.

Materials: Rretrospective single center study of patients operated for SPN of pancreas. Clinical and pathological data were collected.

Results: From 1995 to 2016, 1320 patients underwent pancreatic resection. SPN was confirmed in 32 cases (2.46%), including 29 (90.6%) female and three (9.4%) male, with a mean age of 28.4 ± 12.2 years. SPN was the most common pathology among female patients under age of 40 (72.4%). Abdominal pain was the most frequent presenting symptom (48%), whereas none of the patients presented with jaundice. Mean tumor diameter was 5.9 cm (range, 0.9–14 cm). All patients underwent margin-negative surgical resection. Two patients demonstrated gross malignant features, including liver metastases at presentation (n = 1), and adjacent organ and vascular invasion (n = 1). Microscopic malignant features were present in thirteen patients (40.6%). Recurrence occurred in the retroperitoneal lymph nodes (n = 1, 7 years post resection) and in the liver (n = 2, 1 and 5 years post resection). Mean follow-up was 49.2 months (range, 1–228 months). Five and 10-year disease-free survival was 96.5% and 89.6% respectively.

Conclusions: SPNs are low-grade tumors with a good prognosis. Margin-negative surgical resection is curative in most patients. However, almost 15% of patients demonstrate malignant features including invasion of adjacent organs or metastatic disease. Patients with malignant disease are still expected to have long survival, and aggressive surgical approach is advocated.

Keyword: Solid pseudopapillary neoplasm pancreas

Introduction

Solid pseudopapillary neoplasms (SPNs) of the pancreas were initially described by Frantz in 1959,[1] and included in the World Health Organization classification in 1996 by Klopper et al.[2] These tumors consist 0.3–2.7% of all pancreatic tumors.[3] Their recognition and diagnosis have recently been steadily increasing with more than 60% of total cases reported in the last 10 years.[4,5] Most SPNs are found in young female patients and are well circumscribed, large tumors frequently located in the distal part of the pancreas. Complete, margin negative surgical resection is considered curative in most cases.[6–12] However, 10–15% of SPN cases have malignant features, such as invasion of adjacent organs or distant metastases.[3,4,13–15] Locally advanced and even metastatic disease is often amenable to complete surgical resection, and complete resection is associated with long term survival.[4,6,7,13,15]

In this study we performed a retrospective study of patients operated for SPN of pancreas. The objectives of

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http://dx.doi.org/10.1016/j.ejso.2017.02.001

Please cite this article in press as: Lubezky N, et al., Solid pseudopapillary neoplasm of the pancreas: Management and long-term outcome, Eur J Surg Oncol (2017), http://dx.doi.org/10.1016/j.ejso.2017.02.001

this investigation were to describe the clinical–pathologic characteristics, surgical treatment, and long term outcome of SPN.

Materials and methods

The study was conducted in a high volume, tertiary hospital. All patients undergoing pancreatectomy from 1995 to 2016 were retrospectively reviewed and those with a pathological diagnosis of SPN of the pancreas were included in the analysis. Histological slides were reviewed by a single pathologist to confirm diagnosis of SPN. Records were obtained from the Hepatopancreatobiliary and Pathology database after obtaining approval by the institutional research ethics board. During this period, a total of 1320 patients underwent pancreatic resection. Diagnosis of SPN was confirmed in 32 cases (2.46%).

On the basis of these data, we described the demographic, radiological, perioperative and pathological variables of the patients with pancreatic SPN who underwent surgery. All slides were reviewed by a single pathologist (G.G.), and histological features of aggressive biological behavior including cellular atypia, capsule invasion, lymph node metastasis, lymphovascular invasion, perineural invasion, and peripancreatic fat tissue invasion were documented.[16–18] Stage IV was defined as grossly detectable systemic metastasis or peritoneal seeding.[17] Immunohistochemical studies (IHC) were performed in 26 specimens (81.3%), and included staining for beta-catenin, CD10, neuroendocrine markers (neuron-specific enolase [NSE], chromogranin A, synaptophysin), progesterone, vimentin, Ki-67 and cytokeratin (AE1/AE3).

Table 1
Clinicopathological characteristics of resected pancreatic SPNs.

Clinical features	Frequency, mean ± SD, %
Sex	
Male	3 (9.4)
Female	29 (90.6)
Age, yr	28.4 ± 12.2
Symptoms	
No	9 (28.1)
Yes	23 (71.9)
Tumor location	
Head	10 (31.3)
Body and tail	22 (68.7)
Tumor size, cm	5.9 ± 3.7
Type of surgery	
Enucleation	3 (9.4)
Spleen preserving distal pancreatectomy	4 (12.5)
Distal pancreatectomy + splenectomy	17 (53.1)
Pancreaticoduodenectomy	8 (25)
Minimal invasive surgery	9 (28.1)
R status	
R0	30 (100)
R1	0 (0)
Microscopic malignant features	
Cellular atypia	8 (25)
Capsule invasion	11 (34.4)
Peripancreatic fat invasion	5 (15.6)
Perineural invasion	3 (9.4)
Lymphovascular invasion	3 (9.4)
LN metastases	0 (0)
Adjacent organ invasion	1 (3.1)
Stage IV	
Hepatic metastasis	1 (3.1)
Recurrence	
Hepatic	2 (6.25)
Peritoneal + lymphatic	1 (3.1)

Statistical analysis

Statistical analysis was conducted using SPSS, version 10 (SPSS Inc., Chicago, IL). Categorical data were compared using the χ^2-test with Yates' correction, and continuous variables were compared using the Man–Whitney *U* test. All analyses were carried out using GraphPad Prism 4 (v 4.02, San Diego, CA, USA) statistical software. A P-value of <0.05 was considered significant.

Results

General characteristics of resected pancreatic SPNs

Of 1320 pancreatic resections performed in our institution during the 22-year study period, the diagnosis of SPN was confirmed in 32 cases (2.46%). The general clinicopathological features of the patients with pancreatic SPN are listed in Table 1. There were 29 (90.6%) female and three (9.4%) male patients, with a mean age of 28.4 ± 12.2 years. SPN was the most common diagnosis (72.4%) among the female patients under the age of 40 years old undergoing pancreatic resection. In the first half of the study period (1995–2004), only 4 cases of pancreatic SPN were found, whereas in the second half, 28 new patients were diagnosed and treated surgically.

Approximately 28% of the patients were incidentally discovered without specific symptoms, whereas the rest of the patients presented with clinical symptoms including abdominal pain and discomfort (n = 15, 46.9%), weight loss (n = 5, 15.6%), back pain (n = 3, 9.4%), or vomiting (n = 2, 6.25%). None of the patients presenting with SPN of the pancreatic head presented with obstructive jaundice, diabetes or pancreatitis. In recent years, more SPNs were diagnosed in asymptomatic patients, and the tumor size tended to be smaller. Mean tumor diameter before and after 2010 was 7.7 and 4.43 cm respectively (P = 0.022).

The neoplasm was single in all patients and the most common location was the tail and body of the pancreas (68.8%). Mean tumor diameter was 5.88 cm (range, 0.9–14 cm). One patient had liver metastases at presentation.

All patients underwent a preoperative triple phase pancreatic CT scan. The classic appearance of SPN consisting of a heterogonous pancreatic mass with cystic and solid components and arterial phase enhancement was present in

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