CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 39 (2017) 225–230

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Solid pseudopapillary tumor of the pancreas: Experience at a tertiary care centre of Northern India

Namita Bhutani[a], Pradeep Kajal[b,*], Sham Singla[c], Vijender Sangwan[d]

[a] Deptt. of Pathology, PGIMS Rohtak, Haryana, India
[b] Deptt. of Pediatric Surgery, PGIMS Rohtak, Haryana, India
[c] Deptt. of Surgery, PGIMS Rohtak, Haryana, India
[d] Nirvana Diagnostic Centre, PGI Road, Medical Mor, Rohtak, Haryana, India

ARTICLE INFO

Article history:
Received 18 July 2017
Accepted 25 July 2017
Available online 24 August 2017

Keywords:
Case series
Solid pseudopapillary tumor (SPT)
Pancreas
Female
Young

ABSTRACT

INTRODUCTION: Solid pseudopapillary tumor (SPT) of the pancreas is rare, accounting for 0.13–2.7% of all pancreatic tumors. It is unique, has low malignant potential and predominantly affects young women. Radiological and pathological studies have revealed that the tumor is quite different from other pancreatic tumors. But the cell origin of SPT and tumorigenesis are still enigmatic. Abdominal mass is the most common presenting symptom. Due to the paucity of the number of cases, the natural history of the disease is not fully understood. This study was undertaken to examine the clinico-pathological characteristics of the disease and to evaluate the outcome of surgical intervention in a tertiary referral care centre.

MATERIALS AND METHODS: A retrospective analysis of all patients diagnosed and treated for SPN in our hospital over a period of 10 years (2005–2015) was carried out. A database of the characteristics of these patients was developed. In all, 11 patients were identified. A CT scan of the abdomen was performed in all the patients and the findings revealed a mass in the pancreas. The investigations performed included routine blood investigations, chest X-ray, CA-19-9 level and either an ultrasound or a CT Scan of the abdomen.

RESULTS: During the time period of 10 years, of 349 patients with pancreatic malignancy admitted to our department, only 11 were diagnosed as having SPN (3.15%). Ten patients were women (90%) and one patient was a man (10%). The patients had a median age of 27.6 years (range 17–41). The most common symptoms were abdominal pain and dullness. Eight patients (72.7%) presented with abdominal pain or abdominal dullness and three patient (27%) were asymptomatic. All the 11 patients were taken up for surgery. Three patients underwent distal pancreatectomy with splenectomy, three patients underwent the total mass excision and one patient underwent total pancreatic resection. Three required extended distal pancreatectomy with splenectomy. One underwent spleen-preserving distal pancreatectomy.

CONCLUSION: SPT is rare, but treatable pancreatic tumor. While clinical signs and symptoms are relatively nonspecific, characteristic findings on imaging and histology separate these tumors from the more malignant pancreatic tumors. The prognosis is favorable even in the presence of distant metastasis. Although surgical resection is generally curative, a close follow-up is advised in order to diagnose a local recurrence or distant metastasis.

1. Introduction

First described by Franz in 1959, Solid pseudopapillary tumor (SPT) of the pancreas is rare, accounting for 0.13–2.7% of all pancreatic tumors [1]. It is unique, has low malignant potential and predominantly affects young women [1,2]. Until it was defined by the World Health Organization (WHO) in 1996 as 'solid pseudopapillary tumor' of the pancreas, this tumor was described by using various names including 'solid cystic tumor', 'papillary cystic tumor', 'papillary epithelial neoplasia', 'solid and papillary epithelial neoplasia', 'papillary epithelial tumor' and 'Frantz's tumor', 'solid and papillary tumor', 'solid-cysticpapillary epithelial neoplasm', 'benign or malignant papillary tumor of the pancreas' [3].

Radiological and pathological studies have revealed that the tumor is quite different from other pancreatic tumors. But the cell origin of SPT and tumorigenesis are still enigmatic. The pathogenesis is thought to result from cells of the endocrine pancreas though some investigators have postulated origin from the exocrine pan-

* Corresponding author at: 23/8FM, Medical Campus, Rohtak, India.
E-mail addresses: namitabhutani89@gmail.com (N. Bhutani), pkajalsurg@gmail.com, drpradeepkajal@gmail.com (P. Kajal), drshamsingla@gmail.com (S. Singla), drvijendersangwan@gmail.com (V. Sangwan).

http://dx.doi.org/10.1016/j.ijscr.2017.07.064

creas. These tumours have a long asymptomatic period and are usually detected when they have grown to a large size [4–8].

Abdominal mass is the most common presenting symptom, with dyspepsia, early satiety, nausea, or vomiting being less common presenting symptoms. Up to 20% of patients are asymptomatic with tumors identified either incidentally on imaging or at operation for unrelated pathology [9,10]. Grossly, SPTs are identified as well demarcated, encapsulated tumors with extrapancreatic growth. Mixed solid and cystic components are evident with internal necrotic or hemorrhagic debris and lobulated, solid tissue at the periphery. Characteristic radiographic features include the presence of an encapsulated mass with solid and cystic components on either CT scan or MRI, with MRI notably better for identification of certain tumor characteristics such as the presence of a capsule, hemorrhage or cystic degeneration [10]. SPT should be added to the differential diagnosis in any patient with a solid and partly cystic mass of the pancreas especially in females under 35 years of age. Surgical resection is the treatment of choice for affected patients and is associated with an overall good prognosis [11].

Due to the paucity of the number of cases seen, the natural history of the disease is not fully understood. This study was undertaken to examine the clinico-pathological characteristics of the disease and to evaluate the outcome of surgical intervention in a tertiary referral cancer centre.

2. Material and methods

A retrospective analysis of all patients diagnosed and treated for SPN in our hospital over the past 10 years was carried out (2005–2015). A database of the characteristics of these patients was developed, including age, gender, tumor location (data were derived from radiological investigations or surgical records) and size (data were derived from radiological investigations or surgical records and finally confirmed by pathology), treatment (data were derived from the medical records, including the types of surgery), and histopathological and immunohistochemical features. In all, 11 patients were identified. A CT scan of the abdomen was performed in all the patients and the findings revealed a mass in the pancreas. Pre-operative fine needle aspiration cytology (FNAC) was performed in 2/11 patients. All the patients who underwent resection were followed up every 6 months. The investigations performed included routine blood investigations, chest X-ray, CA-19-9 level and either an ultrasound or a CT Scan of the abdomen.

3. Results

During the time period of 10 years, of 349 patients with pancreatic malignancy (which does not include ampulla vateri, distal choledocal and duodenal tumor) admitted to our department, only 11 were diagnosed as having SPN (3.15%). Ten patients were women (90%) and one patient was a man (10%). The patients had a median age of 27.6 years (range 17–41). The most common symptoms were abdominal pain and dullness. Eight patients (72.7%) presented with abdominal pain or abdominal dullness and three patient (27%) were asymptomatic with the diagnosis made by an incidental finding on routine examination. Abdominal CT and/or magnetic resonance imaging (MRI) showed the typical features of solid pseudopapillary neoplasm in seven (63.6%) of the patients (Fig. 1). Usually, the tumors appeared as, well circumscribed lesions with a mixed cystic and solid component but were almost entirely solid or else cystic with thick walls. In two patients the tumor was located in the pancreatic head (18%), in three patients in the body (27%) and in the remaining six patients in the tail (54%).

All the 11 patients were taken up for surgery. Three patients underwent distal pancreatectomy with splenectomy, three patients underwent the total mass excision and one patient underwent total pancreatic resection. Three required extended distal pancreatectomy with splenectomy. One underwent spleen-preserving distal pancreatectomy. The mean diameter of the tumor was 6.9 cm (range 3–12 cm). Patient characteristics are summarized in Table 1. In seven cases lymph node dissection was done in a number between 5 and 14, whereas no dissection was needed for four patients. No lymph node metastasis was present in any patient.

At histopathological examination, tumor mass separated from pancreas with a fibrous capsule was seen. Pseudopapillary, cystic and solid growth patterns were seen in the tumor mass. Tumor cells had an ovally shaped, small and centrally localized nucleus and large eosinophilic cytoplasm. Tumors consisted of pseudopapillary structures made of cells aligned around fine vessels, solid areas, hemorrhagic areas and cystic areas of different size (Fig. 2). No mitosis was seen in eight cases, whereas minimal mitosis was present in one case (2/10 per high powered field) and multiple mitosis were present in two cases (20/10 per high powered field) (Table 2). The immunohistochemistry profiles are summarized in Table 3 showh in Fig. 3 Capsular invasion was present in three cases (case numbers 2, 6 and 10), spleen invasion wasalso present in case number 2. These three cases were considered as malignant SPN and treated with six courses of gemcitabine + cis-platinum chemotherapy. Multiple liver and omentum metastases developed in case number 2 at the seventh postoperative month; this patient died at the ninth month. Multiple liver and omentum metastases developed in case number 6 at the 20th postoperative month and she died at the 24th month. The other eight cases have been followed up closely and no recurrence or metastasis has been seen. The average postoperative hospital stay was 10.3 days.

4. Discussion

Solid pseudopapillary tumor of the pancreas (SPT) is an uncommon and enigmatic pancreatic neoplasm first described by Frantz in 1959 [1]. This lesion usually has a low malignant potential [2]. The tumor has been identified by a number of synonyms including solid and cystic tumor, solid and papillary epithelial neoplasm, papillary cystic neoplasm, papillary cystic epithelial neoplasm, papillary cystic tumor, and Franz tumor. In 1996, the World Health Organization (WHO) renamed this tumor as SPT for the international histologic classification of tumor of the exocrine pancreas [3]. This uncommon, typically benign tumor is found mainly in young non-Caucasian women between the 2nd and 3rd decades of life. It seems to have a predilection for Asian and African- American women, although rare cases have been reported in children and men [12]. Female predominance has been attributed to the proximity of primordial pancreatic cells to the ovarian ridge during development [13].

The differential diagnosis of suspicious neoplasms should include microcystic adenoma, mucinous cystic neoplasm, nonfunctioning islet cell tumor, pancreatic adenocarcinoma, pancreaticoblastoma, cystic degeneration of solid neoplasm and calcified hemorrhagic pseudocyst [14]. Abdominal discomfort or vague pain is the most common symptom, followed by a gradually enlarging mass and compression signs induced by the tumor. Some patients are completely asymptomatic, with the tumor detected incidentally by imaging studies or routine physical examination [1]. CT scan, ultrasonography (US) and endosonography (EUS) have been used with variable success in diagnosing SPN. CT scan and EUS are more sensitive and specific and have shown more accuracy in diagnosing SPN [15]. Magnetic resonance imaging (MRI) can be diagnostic. Typically, a large, well-defined, encapsulated lesion with heterogeneous high or low signal intensity on T1-weighted,