Solid pseudopapillary neoplasm of the pancreas: a single institution experience of 14 cases

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Abstract

Solid pseudopapillary neoplasm of pancreas (SPN) is a rare entity; therefore proper diagnosis, evaluation and formulation of treatment protocols is difficult. A total of 14 cases were diagnosed and treated at our institute over a 10-year period (1992–2002). A retrospective study of all these cases was performed with respect to age group most frequently affected, sex ratio, common presenting symptoms and signs, investigative protocols, pathological features, treatment offered, outcome and prognosis. All the 14 patients were females, with a median age of 20 years (range 13–45 years). The commonest presenting symptom was abdominal pain. All these patients underwent surgical exploration; 13 patients underwent resection of the tumour. The average length of postoperative hospital stay was 10.3 days. Significant postoperative morbidity was seen in only one case. There was no postoperative mortality. All these patients who underwent resection were disease-free on follow-up ranging from 6 to 96 months (median 36 months).

Key Words: Solid pseudopapillary neoplasm of pancreas, papillary epithelial neoplasm of pancreas

Introduction

Solid pseudopapillary neoplasm of pancreas (SPN) is a rare entity. It is almost exclusively seen in females and occurs in the second or third decades of life [1–3]. Due to the paucity of the number of cases seen, the natural history of the disease is not fully understood.

There has been a steady increase in the number of diagnosed cases of SPN recently, with more than two-thirds of the total cases described in the last 10 years [4]. 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.

Patients and methods

A retrospective analysis of all patients diagnosed and treated for SPN in our hospital over the past 10 years (1992–2002) was carried out. The clinico-pathological, radiological, operative and survival data were obtained and analysed. In all, 14 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 4/14 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.

Results

All the patients were females in the age group of 13–45 years (median 20 years). Ten patients presented with a dull aching pain in the abdomen. Two presented with a painless abdominal mass (14%) in the epigastric region. Two patients had non-specific abdominal symptoms for which they had been investigated (14%). In five patients the tumour was located in the pancreatic head (36%), in two it was located in the body (14%), and in the remaining seven patients it occurred in the tail (50%). The tumour size ranged from 3 to 18 cm (average 6.8 cm). Four patients had undergone a preoperative FNAC: in three patients the FNAC correctly diagnosed SPN while one was reported as non-Hodgkin’s lymphoma (however, the final HPR revealed the tumour to be SPN).

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resection was performed. Six patients underwent distal pancreatectomy with splenectomy, two underwent a pylorus-preserving pancreatectoduodenectomy, and two patients required right hemicolectomy along with a Whipple resection. Two required subtotal distal pancreatectomy with splenectomy. One underwent spleen-preserving distal pancreatectomy. None of these patients had distant metastasis.

One patient suffered from a major morbidity. This patient had undergone a subtotal-distal pancreatectomy with splenectomy, and had suffered an inadvertent injury to the intra-pancreatic portion of the common bile duct (CBD) during the excision. This patient required a T-tube placement in the duct. Postoperatively, a stent was placed endoscopically to facilitate the removal of the T-tube. The stent was removed 3 months later with no further problems. This was the only complication in the 13 patients who underwent resection.

The average postoperative hospital stay was 10.3 days. There was no postoperative mortality. All 13 patients were free of disease in a median follow-up period of 36 months (range 6–96 months).

Discussion

SPN is predominantly encountered in young, female patients (first three decades of life), but has also been reported in males and in children [1,3]. In our series, all the patients were females with a median age of 20 years.

These tumours may be discovered by chance during diagnostic imaging procedures or may be suspected in the presence of an asymptomatic palpable mass in young women. In our series, 10 patients presented with dull aching abdominal pain, while 2 patients presented with an abdominal mass.

Depending on the tumour position (head, body or tail of the pancreas), the differential diagnosis includes adrenal mass, pancreatic endocrine tumour, liver cyst or tumour, or a pseudocyst [5].

CT scan (Figure 1), 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 [6,7]. Magnetic resonance imaging (MRI) can be diagnostic. Typically, a large, well-defined, encapsulated lesion with heterogeneous high or low signal intensity on T1-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 [8]. In our series, we relied on CT imaging for the preoperative work-up.

FNAC has been used for the preoperative cytological diagnosis of SPN [6,9]. The cytology specimen is usually highly cellular and is characterized by the presence of epitheloid cells that present singly or in aggregates containing fibrovascular cores. No evidence of pleomorphism or mitotic activity is seen in the cells. The most conclusive criterion for identification of SPN is the pseudopapillary arrangement with bland appearing tumour cells. EUS-guided FNAC has been reported, and this can help in correctly diagnosing SPN pre-operatively [10]. In the current series, 4/14 patients had a pre-operative percutaneous US-guided FNAC, and the diagnosis of SPN was made correctly in 3 patients. It is not necessary to have a tissue diagnosis pre-operatively, and surgery can be advised on the basis of radiological imaging.

The histogenesis of these tumours is unknown but they possibly originate from the primordial cells and lack definite endocrine and exocrine differentiation.

On gross examination, SPN is a well encapsulated tumor. On cut section it shows solid and cystic areas with necrotic and haemorrhagic patches. Some tumours also demonstrate firm, fibrotic regions within the tumour. On microscopy, there are solid areas composed of polygonal epithelioid cells with intervening stroma. There is evidence of cellular degeneration. Aggregates of foamy histiocytes, cholesterol clefts and cytoplasmic vacuolization can be seen. Despite being an encapsulated tumour, the microscopic interface between tumour and adjacent normal pancreas does show an infiltrative growth pattern [1,2,4,6,11–13] (Figure 2).

Immunohistochemical studies have shown that SPN is reactive for vimentin, antitrypsin, cytokeratin, S-100 protein and neuron-specific enolase. Flow cytometry shows aneuploidy. C-Ha-as oncogene presumably is linked to the development of the tumour. No pathologic factor is of proven prognostic importance [11–13]. Laboratory values are not contributory, although a few cases do show raised levels of CA19-9.

SPN is considered to be a tumour of low-grade malignant potential [14]. The logical conclusion is that complete surgical excision is the best option for patients who have SPN. Thus surgery should always be attempted in a suspected case of SPN even if it implies that major resections (like pancreaticoduode-