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Pancreatic Solid-Cystic Papillary Tumor: Clinical Features, Imaging Findings and Operative Management

Riccardo Casadei[1], Donatella Santini[2], Lucia Calculli[3], Raffaele Pezzilli[4], Nicola Zanini[1], Francesco Minni[1]

Departments of [1]Surgery, [2]Pathology, [3]Radiology, and [4]Internal Medicine,
S.Orsola-Malpighi Hospital. Bologna, Italy

Summary

Solid-cystic papillary tumors of the pancreas are very rare and, until today, 718 cases have been reported in a review of the literature. Four patients affected by solid-cystic papillary tumors, observed in our Institute between January 1985 and July 2005, are reported. The clinicopathological, operative and survival data of this tumor were reviewed comparing our experience with a review of the literature.

Solid-cystic papillary tumor have a preference for young women (age between 19-50 yrs) and show a large round, well-defined pancreatic mass(greater than 5 cm), clinically silent or with unspecific symptoms. The tumor is more frequently localized in the pancreas (80%) and is rarely a metastatic disease (20%). Surgical treatment with radical pancreatic resection of the tumor was performed in approximately 95% of the cases. In 467 patients, two-year survival was 97% (16 deaths) and 5-year survival was 95% (21 deaths).

In conclusion, preoperative diagnosis of solid-cystic tumors of the pancreas is difficult but knowledge of its characteristic findings can aid in reaching a proper diagnosis. Surgery is the treatment of choice; it should be conservative in localized tumors, and aggressive, in non-localized tumors. The prognosis is very good, with long-term survival also in patients with metastases or unresectable tumors.

Introduction

Solid-cystic papillary tumors (SCPTs) of the pancreas are very rare. Frantz first described this tumor in 1959 [1]. He reported four cases which had previously been misdiagnosed as non-functioning islet cell tumors and he established this tumor as a new entity which he called “papillary tumor of the pancreas” [1]. In a review of the literature, the tumor is reported with different terms: papillary epithelial neoplasm [2], papillary cystic neoplasm [3], solid and papillary epithelial neoplasm [4], solid and cystic acinar tumor [5], papillary and solid neoplasm [6], papillary cystic epithelial neoplasm [7], papillary cystic carcinoma [8], solid and cystic papillary tumor [9], solid pseudopapillary tumor or carcinoma [10]. Among these names, we chose SCPT because it best describes the pathologic features of the tumor: grossly the tumor showed solid tissue surrounding central hemorrhagic and cystic areas; histologically, a variegated pattern of solid, pseudopapillary and cystic growth is typical.

Table 1. Characteristics of the four patients observed in our Institute.

Case	Sex	Age	Site	Size	Pathology	Symptoms
1	Female	38 years	Tail	8 cm	Capsule infiltration No metastases	Pain, mass
2	Female	50 years	Head	4 cm	Solid-cystic capsule infiltration No metastases	Pain
3	Female	35 years	Body	3 cm	Solid-cystic capsule infiltration No metastases	Acute pancreatitis
4	Male	39 years	Body	5 cm	Solid capsule infiltration No metastases	None

Table 1. (continued)

Case	Preoperative diagnosis	Treatment	Follow-up
1	Tail tumor (US, CT)	Left pancreatectomy	Alive, disease-free (123 months)
2	Ductal carcinoma (US, CT)	Pancreaticoduodenectomy	Alive, disease-free (113 months)
3	Solid-cystic papillary tumor (US, CT, MR)	Central pancreatectomy	Alive, disease-free (54 months)
4	Neuroendocrine pancreatic tumor (US, CT)	Central pancreatectomy	Alive, disease-free (43 months)

US: ultrasound
CT: computed tomography
MR: magnetic resonance

In this study, we report our experience with this tumor comparing it to those described in the English literature in order to recognize some important features of the tumor useful in avoiding misdiagnosis and determining the optimal management and the role of conservative surgery.

Our Experience

In the First Surgical Clinic of the University of Bologna between January 1985 and July 2005, 772 cases of pancreatic tumors were observed; of these, 623 (80.7%) were ductal adenocarcinoma, 76 (9.8%) islet cell tumors and 73 (9.5%) cystic pancreatic tumors. SCPT was observed in 4 cases, that is, in 0.5% of all pancreatic tumors and 5.5% of all cystic tumors. Clinicopathological, operative and survival data of each SCPT were reviewed and are summarized in Table 1.

Case 1

A 38-year-old woman was admitted to our Institute because of vague abdominal pain which had persisted for about 1 year. At physical examination, a palpable mass in the

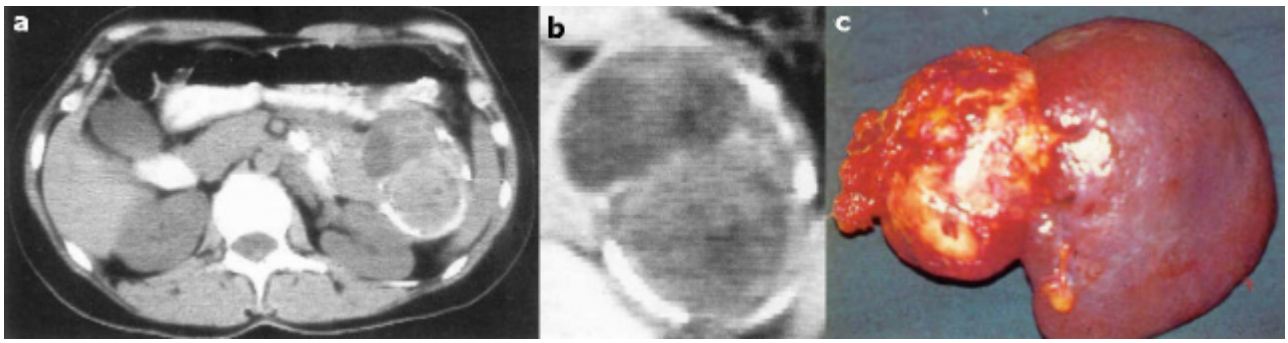

Figure 1. Case 1. A CT scan showing a round, encapsulated 8x5 cm mass of the pancreatic tail with non-homogeneous density (**a.**), a fibrotic capsule and multiple foci of central and marginal calcifications (**b.**). The tumor mass was completely removed with a distal pancreatectomy and splenectomy (**c.**).

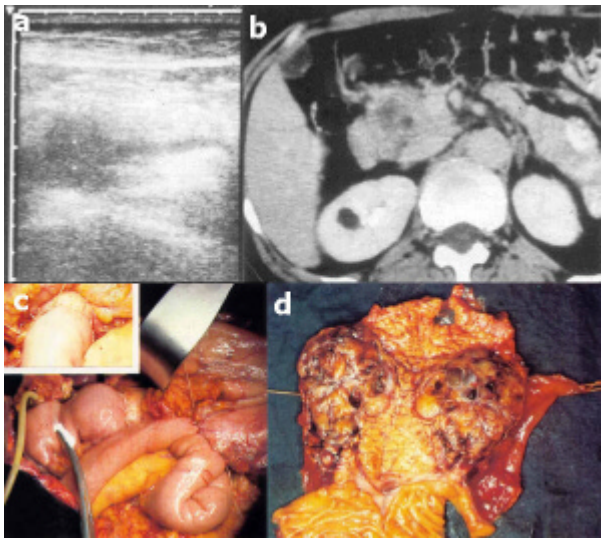

Figure 2. Case 2. US showing a solid mass of the pancreatic head, 4 cm in diameter, not well-defined and without an evident capsule, mimicking a ductal adenocarcinoma (**a.**). A contrast-enhanced CT scan confirmed the ultrasound findings and revealed, with a non-homogeneous mass of uneven soft-tissue density with central necrosis (**b.**). A pylorus-preserving pancreaticoduodenectomy was performed (**c.**) and the mass appeared as a solid and cystic lesion (**d.**). (Image **d.** is presented in another contribution by the same authors, published in these Proceedings [30], in order to describe aspects not related to those reported here)

left upper abdomen was found. All laboratory parameters, including tumor markers, were normal. Ultrasound (US) revealed a large round, well-demarcated mass (8 cm in diameter) in the upper left abdomen with non-homogeneous echogenicity, fibrotic capsule and calcifications. A computed tomography (CT) scan showed a round, encapsulated 8x5 cm mass of the pancreatic tail with non-homogeneous density, a fibrotic capsule and multiple foci of central and marginal calcifications (Figure 1a,b). Preoperatively, a diagnosis of pancreatic tail neoplasm or pancreatic hematoma was suspected. Intraoperatively, a large, well-defined mass of the pancreatic tail was found. There was no metastatic spread to the liver, peritoneum or lymph nodes nor was involvement of the adjacent organs or tissues detected. Radical resection (R0) of the tumor by means of distal pancreatectomy and splenectomy was performed (Figure 1c) Histologically the characteristic findings of a borderline SCPT, according the WHO classification, were identified.
The postoperative course was uneventful and the patient is alive and disease-free at a follow-up of 123 months.

Case 2

A 50-year-old woman was referred to our Institute because of vague abdominal discomfort and dyspepsia which had persisted for 3 years. All laboratory parameters were normal. US showed a solid mass of the pancreatic head, 4 cm in diameter,, not well-defined and without an evident capsule, mimicking a ductal adenocarcinoma (Figure 2a); a contrast-enhanced CT scan confirmed the ultrasound findings and revealed a non-homogeneous mass of uneven soft-tissue density with central necrosis (Figure 2b). Preoperatively, a diagnosis of ductal adenocarcinoma or serous cystic tumor of the pancreatic head was suggested. At laparotomy, a solid mass of the pancreatic head was found. There was no metastatic spread to the liver, peritoneum or lymph nodes nor was involvement of the adjacent organs or tissues detected. A pylorus-preserving pancreaticoduodenectomy was performed (Figure 2cd). The definitive diagnosis was of borderline SCPT with capsule infiltration and involvement of the surrounding pancreatic parenchyma. The postoperative course was uneventful and the patient is alive and disease-free at a follow-up of 113 months.

Case 3

A 35-year-old woman was admitted to our Institute after several recurrences of acute pancreatitis with typical pancreatic abdominal pain and hyperamylasemia. US showed a round, well-defined, non-homogeneous mass of the pancreatic body, 3x3 cm in diameter, (Figure 3a). Endoscopic ultrasound, CT scan and magnetic resonance (MR) confirmed the ultrasound findings (Figure 3bc). Preoperatively, a diagnosis of SCPT or serous

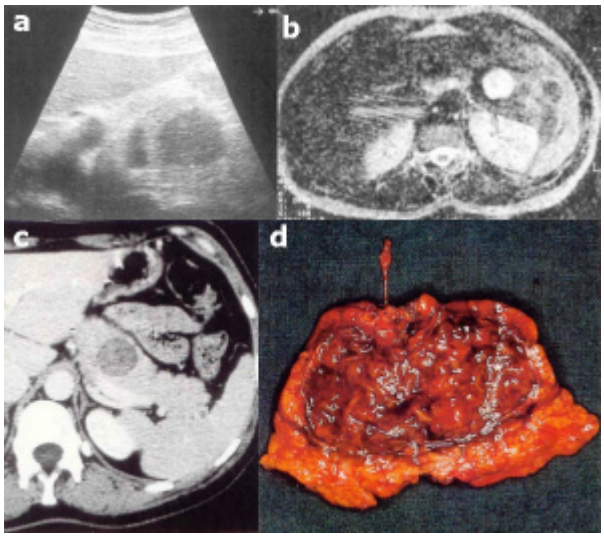

Figure 3. Case 3. US showing a round, well-defined, non-homogeneous mass of the pancreatic body, 3x3 cm in diameter (**a.**). Magnetic resonance (**b.**) and a computed tomography scan (**c.**) confirmed the ultrasound findings. A conservative, radical pancreatic resection was performed with a central pancreatectomy including the neoplasm (**d.**). (Image **d.** is presented in another contribution by the same authors, published in these Proceedings [30], in order to describe aspects not related to those reported here)

cystic tumor was suspected. Intraoperatively, a well-defined solid, 3x3 cm mass was detected without metastasis or adjacent organ involvement. An intraoperative biopsy resulted in a diagnosis of SCPT of the pancreatic body.
A conservative, radical (R0) pancreatic resection was performed with a central pancreatectomy including the neoplasm (Figure 3d). The definitive diagnosis was borderline SCPT with capsule infiltration but without pancreatic parenchyma involvement. The postoperative course was uneventful and the patient is alive and disease-free at a follow-up of 54 months.

Case 4

A 39-year-old man was admitted to our Institute because an US, performed for other reasons in the absence of abdominal symptoms, incidentally revealed a round, well-defined, non-homogeneous, mass of the pancreatic body (5x4 cm in diameter) having a thin capsule and multiple central foci of calcifications (Figure 4a). A CT scan confirmed ultrasound findings (Figure 4b). Preoperatively, diagnosis of non-functioning islet cell tumor was suggested. A solid mass in the pancreatic body was detected at laparotomy. No liver or peritoneum or lymph node metastases were detected and the tumor did not involve the surrounding tissues or organs. Thinking it was a non-functioning islet cell tumor, a central pancreatectomy was performed (Figure 4c). The diagnosis was borderline SCPT with capsule infiltration but without pancreatic parenchyma involvement. The postoperative course was uneventful and the patient is alive and disease-free at a follow-up of 43 months.

Discussion

A solid-cystic tumor of the pancreas is a rare tumor which represents about 2% of all

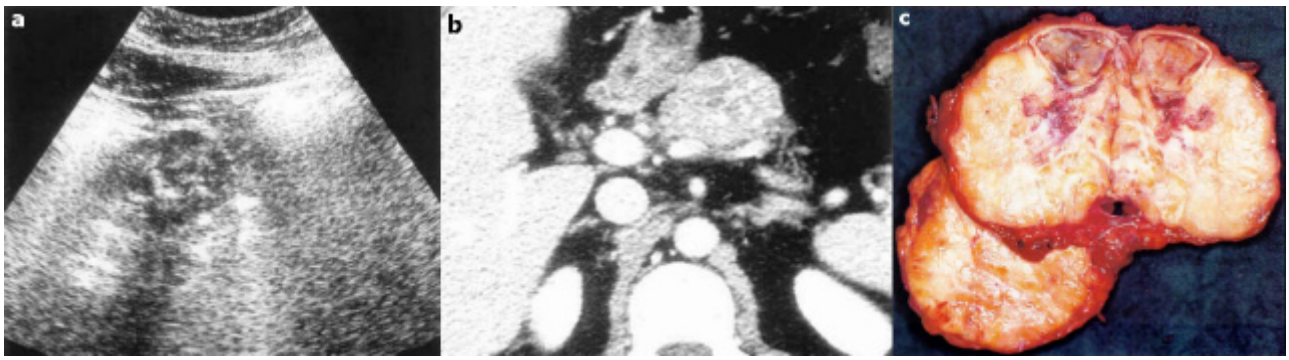

Figure 4. Case 4. US revealing a round, well-defined, non-homogeneous mass of the pancreatic body, 5x4 cm in diameter, with a thin capsule and multiple central foci of calcifications (**a.**). A computed tomography scan confirmed the ultrasound findings (**b.**). A central pancreatectomy was performed and the mass appeared as a solid and cystic lesion (**c.**). (Image **c.** is presented in another contribution by the same authors, published in these Proceedings [30], in order to describe aspects not related to those reported here)

pancreatic tumors [9, 11, 12, 13, 14, 15, 16] and 9.3% of cystic pancreatic tumors [11, 13, 14, 17, 18, 19, 20]. In our experience, SCPTs represent 0.5% of all pancreatic tumors (4/772) and 5.5% of cystic pancreatic tumors (4/73). There has been an increasing incidence of this entity in recent years; In 1979 Cubilla and Fitzgerald [16] reported an incidence of SCPTs of 0.17%, in 1987, Morohoshi *et al*. [9] of 2.7% and in 2004, Koshmal *et al*. [12] of 6.1% of all pancreatic tumors. Martin *et al*. [11] noted that, in the cases reported, more than two-thirds of the total had been described in the last 10 years. Finally, a review of the literature has revealed an increase in the number of cases observed: in 1995, Mao *et al*. [21] reviewed 292 cases, in 1999, Lam *et al*. [13], 452 and in 2005, Papavramidis and Papavramidis [22] reported 718 cases of SCPTs. It should be pointed out that only a few authors have reported more than 10 cases while many authors have reported a few cases or, often only a single case report.

The main preoperative problem is the misdiagnosis of an SCPT of the pancreas. Until recently, SCPTs had been identified as one of several other types of tumors: adenocarcinoma, islet cell tumors, cystadenomas, papillary cystadenocarcinomas or cystadenocarcinomas. Le Borgne *et al*. [17] reported a multicenter study of the association of French surgeons about SCPTs and noted that only 6/22 (27.3%) underwent surgery with a suspected diagnosis of SCPT; Panieri *et al*. [23] describing 12 cases, suggested a diagnosis of SCPT in 4 cases (33.3%); Cheng *et al*. [24] observed that in 22 cases of SCPTs, the misdiagnosis rate was 45.5%. In our experience we suspected an SCPT in only 1 of the 4 cases (25.0%). An evaluation of both a review of the literature and our experience may be of assistance in recognizing some important features which could help in the preoperative diagnosis. The majority of SCPTs of the pancreas have been diagnosed in young women, more than 70% of them between 19-50 years of age [22]. In our experience, 3 of 4 cases (75%) were female and all were between 19-50 years. Symptoms, when they do occur, are often vague and non-specific. These symptoms are abdominal pain (46.5%) and palpable mass (34.8%); asymptomatic cases were reported in 15.5% of cases [22]. In our experience, 3 of 4 patients (75.0%) presented with symptoms: two cases had vague abdominal pain, associated in one with a palpable mass; in the third case, recurrent acute pancreatitis with characteristic abdominal pain was observed. One case (25.0%) was asymptomatic and discovered incidentally during screening examination. As a result of these subtle symptoms, tumor presentation can be quite large (mean diameter of 6.1 cm) with more than 75% greater than 5 cm in diameter [22]. SCPTs are more frequently localized in the pancreatic tail or head (70%); they rarely have extrapancreatic localization (1%). In our experience, 2 cases were greater than 5 cm in diameter, while the remaining were less than 5 cm.

Imaging studies of SCPTs of the pancreas include abdominal ultrasound, endoscopic ultrasound, spiral CT, MR and, in some cases, fine needle aspiration biopsy (FNAB) or cytology. Characteristic imaging findings include large, round, well-encapsulated, combined cystic and solid masses; calcifications or internal septations are detected in some cases. The mass was sometimes seen as a pure solid-looking mass. In our cases, we always performed abdominal ultrasound and spiral CT, and, in one case, MR and endoscopic ultrasound. In three cases, we noted a large, round and well-encapsulated mass; in only one was a suspected diagnosis of SCPT made. In one case, the mass was not well-encapsulated, with irregular margins mimicking a ductal adenocarcinoma. Fine needle aspiration biopsy or cytology is reserved for selected cases: in a review of the literature, only 52 cases of FNAB were reported as being positive for SCPT, [22] while, in our experience, we never performed a FNAB.

The management of SCPTs of the pancreas is related to the extension of the disease.

Surgical removal of the tumor is usually the treatment of choice; in fact, Papavramidis and Papavramidis [22] reviewed 553 cases of which 522 (94.4%) underwent pancreatic resection.

With localized disease (400 out of 497cases, 80.5%), complete surgical removal of the tumor is possible in 100% of cases. Moreover, the non-aggressive behavior of the tumor and the presence of a dense capsule, allows us to perform conservative, radical resection. In Japan, 35% of SCPTs originating in the pancreatic head have been treated with enucleation [25]; a tumor in the neck or in the body of the pancreas can be removed with midportion resection of the pancreas, including the mass, preserving the rim of the head, the uncinery process and the pancreatic tail; a tumor in the tail can be resected with a spleen-preserving distal pancreatectomy. Extensive lymphatic dissection or more radical approaches are not indicated when the disease is localized. A laparoscopic approach, instead, should be considered in SCPTs of the pancreatic body-tail.

The surgical removal of non-localized disease was evaluated in 62 (63.9%) out of the 97 cases, and it was possible in 51 of these cases (82.3%). Local invasion and metastases are not contraindications for resection. Portal vein resection is advocated when there is evidence of tumor invasion. For the metastases, surgical debulking should be performed, in contrast to other pancreatic malignancies. Liver metastases can be removed with enucleations or lobectomies. From a review of the literature of 23 cases of SCPTs with liver metastases, 18 (78.3%) were resected and 5 (21.7%) were not treated surgically. In our experience, all cases were localized to the pancreas and were treated surgically; in two cases, a radical but conservative pancreatic resection (central pancreatectomy) was carried out and, in the remaining two, a major pancreatic resection (left pancreatectomy with splenectomy and pylorus-preserving pancreaticoduodenectomy) was performed.

Experience with therapy other than surgery has been used in only a small number of patients because the resectability rate for SCPTs of the pancreas is so high. Adjuvant chemotherapy and radiotherapy are reported in some unresectable cases with good results [26, 27]. Neoadjuvant chemotherapy is also reported with success in a few cases [28, 29].

The prognosis of SCPTs of the pancreas is good. Papavramidis and Papavramidis [22] reported an overall 2-year survival rate of 97% (patients with and without metastases), and a 5-year survival rate of about 95%. SCPTs limited to the pancreas are cured by complete surgical excision. In patients with non-localized tumors, long-term survival is also possible, and it seems that excisional therapy offers the best chance. Survival time for SCPTs with treated liver metastases is good ranging from 6 months to 17 years. However, in the few unresectable cases, in which chemotherapy or radiotherapy were used, the results were encouraging. In our experience , all patients, with localized disease are alive and well at a mean follow-up of 83 months (range 43-123).

In conclusion, a diagnosis of SCPTs should be considered in young women presenting with a large, round, well-defined pancreatic mass. SCPTs of the pancreas should be treated surgically: conservative radical, pancreatic resections in localized tumors and aggressive treatment, with complete resection of both the primary tumor and metastatic lesions, in non-localized disease. The prognosis is also good, with long-term survival in non-localized treated tumors or in the few unresectable cases.

Keywords Neoplasms; Pancreas; Pancreatectomy; Pancreatic Neoplasms

Abbreviations SCPT: solid-cystic papillary tumor

Correspondence
Riccardo Casadei
Dipartimento di Scienze Chirurgiche e Anestesiologiche
Chirurgia Generale - Prof. Minni

Policlinico S.Orsola-Malpighi
Via Massarenti n. 9
40134 Bologna
Italy
Phone: +39-051.341.541
Fax: +39-051.341.483
E-mail: casadei@aosp.bo.it

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