

Solid pseudopapillary tumor of the pancreas: Clinical features, diagnosis and treatment

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SUMMARY

Introduction: Solid pseudopapillary tumor of the pancreas (SPTP) is a rare neoplasm of low malignant potential with uncertain behavior, diagnosed mainly in young women.

Method: Our report comprises a series of cases of SPTP reviewed retrospectively, highlighting clinical, tomographic and immunohistochemical features, treatment performed and outcomes.

Results: Thirteen patients were found to have pancreatic [solid] masses on computed tomography scan measuring a mean diameter of 8.8 cm. All patients underwent complete surgical excision. Immunohistochemistry confirmed diagnosis in all cases.

Conclusion: SPTP occurs more frequently in young women. Diagnostic suspicion lies on the finding of a bulky, solid and cystic pancreatic mass. Imaging findings might provide diagnostic information before resection. Conservative approaches can be used in selected cases and survival rates are usually excellent following complete resection.

Keywords: pancreatic neoplasms, surgery, female, adolescent, immunohistochemistry.

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INTRODUCTION

Solid pseudopapillary tumor of the pancreas (SPTP) is a rare entity of low potential of malignancy and uncertain behavior. It comprises about 1 to 2% of exocrine tumors¹⁻³ and 13% of surgically excised pancreatic lesions.¹

Frantz was the first to report this entity, describing a papillary neoplasia with solid and cystic features, questioning its nature to be either benign or malignant, and diagnosing it as a non-functioning islet cell tumor.² Young women in the second and third decades of life are primarily affected,² with a ratio of 1:10 compared to men.³

Patients with SPTP more often present vague abdominal symptoms and a palpable mass. Computed tomography scan is useful to identify a cystic and calcified lesion, also determining its diameter and possibility of invading adjacent structures. Physicians have reported this neoplasm more frequently in the past few years, based not only on a better understanding of its clinical charac-

teristics, but also on immunohistochemical aspects. Its low malignant potential provides excellent long-term survival after surgical excision.^{4,5}

The present article describes clinical features and management of this uncommon pancreatic malignancy through a series of cases.

METHOD

From August 2001 to May 2015, we found records of 12 patients with SPTP managed in three different hospitals in the municipality of Aracaju, Sergipe, Brazil and one managed elsewhere. We conducted a retrospective descriptive analysis, including the following variables: sex, age, diameter and site of the lesion, clinical presentation, treatment performed, and follow-up. All tumors were resected, fixed in 10% formaldehyde, and then embedded in paraffin. Light microscopic analyses followed by immunohistochemistry confirmed the diagnosis.

RESULTS

Table 1 presents demographic, clinical and pathologic findings. All patients were female with mean age of 27 years, ranging from 16 to 39 years. Preoperative CT scans of the abdomen (Figure 1) showed lesions with various degrees of solid and liquid material and no invasion into surrounding tissues.

Eleven (11) patients underwent open surgical excisions and two had laparoscopic resections. Resected specimens revealed bulky, round, soft and brown tumors presenting solid areas and hollow spaces filled with debris on section analysis. Microscopic analysis revealed layers of uniform cells with both endocrine and exocrine features in arrangements resembling rosettes around a fibrous core. More frequently, immunohistochemical examinations showed positivity for progesterone receptors and α 1-antitrypsin, as shown in Table 2.

Patients had uneventful postoperative courses with no evidence of residual neoplasia, and, at the moment of this study, they are all well and free of disease.

DISCUSSION

All of the SPTP cases reported here were in females with mean age of 27 years, the youngest being 16 and the oldest 39 at admission. Most reports in the literature reported cases of women in the second and third decades of life. Few cases have been reported in men.^{6,7}

SPTP has been reported in the literature since 1933 and its incidence seems to be increasing mainly due to better diagnostic techniques, including reviews of past cases.^{3,8}

Patients often present bulky tumors, with mean diameter around 10 cm. Early signs and symptoms are vague and non-specific, leading to a delay in diagnosis.^{6,9-11} The same occurred with our patients, with nine presenting abdominal masses and tenderness at admission; one patient had associated vomiting due to gastric outlet obstruction; one had diabetes; and three other had no complaints and tumors were discovered in routine abdominal sonograms. The body and tail of the pancreas more often harbor these tumors and seldom do they invade adjacent tissues. Similarly, eight of our cases were in the body-tail region and five in the head of the pancreas.

All of the tumors we report were completely excised. Authors have reported high resection rates and rarely any residual disease. Recurrence is low after complete resection, which is the only way to achieve a cure.¹²

In our series, correct diagnosis followed surgical resection. Definitive diagnosis usually occurs after surgical resection and pathologic examination of the specimen; however, sonography and tomography features of a bulky, solid and cystic tumor, especially in young women, lead to suspicion of a papillary cystic neoplasm.^{6,13} Some authors consider CT scan the most precise means of diagnosis.^{6,8,13} On the other hand, findings with magnetic resonance imaging appear to correspond more precisely to the pathological features.¹⁴ The cytological appearance of these tumors has been described and percutaneous needle aspiration can be used. However, this has been questioned because of the risk of needle-site implants.¹⁵ Whether the pancreatic masses are discovered after minor symptoms or

TABLE 1 Demographic, clinical and pathological features of 13 patients with solid pseudopapillary pancreatic tumor.

Cases	Sex	Age	Site	Diameter (cm)	Presentation	Type of resection	Follow-up
1	F	21	H	16	Mass	PD	165
2	F	16	B/T	15	Mass, pain	Distal P	156
3	F	22	B/T	10	Mass, emesis	Distal P + Spl	154
4	F	20	B/T	9	Mass	Distal P	151
5	F	39	B/T	14	Mass	Distal P + Spl	90
6	F	19	H	8	Pain	Enuc	89
7	F	17	B/T	6	Asymptomatic	VLP Res	85
8	F	32	B/T	8.5	Mass, pain	Distal P	81
9	F	34	B/T	7.7	Pain	Distal P	64
10	F	34	H	3.1	Pain	PD	29
11	F	37	H	7.5	Diabetes	PD	12
12	F	37	B	4.5	Asymptomatic	Partial R	9
13	F	37	H	5.2	Asymptomatic	PD	4
Mean	-	28	-	8.8	-	-	83.8

F: female; H: pancreatic head; B/T: body/tail of pancreas; B: pancreatic body; PD: pancreatoduodenectomy; Distal P: distal pancreatectomy; Spl: splenectomy; Enuc: enucleation; VLP Res: video-laparoscopic resection; Partial R: partial resection.

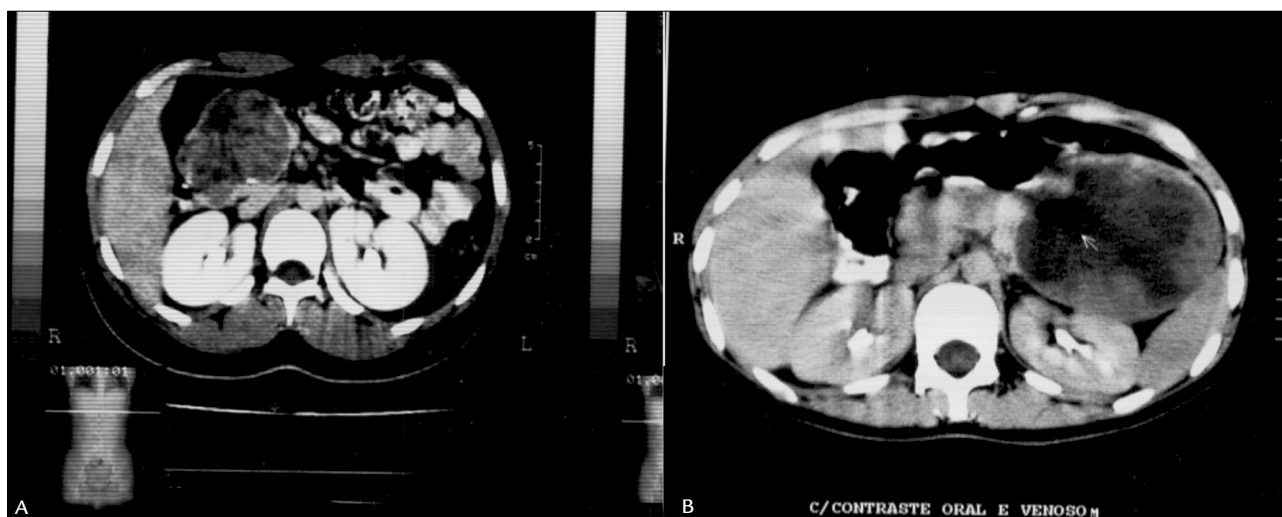


FIGURE 1 CT scan of pancreatic masses displaying (A) solid and (B) cystic features (cases 4 and 2).

TABLE 2 Immunohistochemical features of the tumors.

Marker	Cases												
	1	2	3	4	5	6	7	8	9	10	11	12	13
α 1-antitrypsin	+	+	+	-	+	+	+	+	+	NT	NT	NT	NT
Chromogranin A	-	-	-	-	-	-	+	+	+	-	+	-	-
Progesterone receptor	+	+	+	+	+	+	+	+	+	+	+	NT	-
AE1/AE3	NT	-	-	-	-	+	-	+	+	+	-	+	-
Synaptophysin	+	-	-	-	-	-	+	+	-	+	-	NT	-
Vimentin	NT	NT	NT	NT	+	NT	+	NT	NT	NT	NT	NT	+
CD 10	NT	NT	NT	NT	-	NT	+		NT	+	+	+	+
β -catenin	NT	NT	NT	NT	NT	NT	+	NT		+	+	+	+

NT: not tested; (+): positive; (-): negative.

incidentally, plain characterization of the tumors is paramount in order to identify them as benign, premalignant or malignant lesions. We recommend image characterization with contrast-enhanced helical CT scan. The decision to proceed with needle biopsy, as seen in case 13, should be made to rule out a benign neoplasm that could be managed without surgery. In addition, determination of CA 19.9 status might help defining management.

Light microscopy defines the histopathological findings, which comprises solid areas alternating with pseudopapillary formation, cellular degeneration, including cholesterol clefts and aggregates of foaming histiocytes, uniform cells showing endocrine and exocrine characteristics, and clusters of cells arranged around a fibrovascular core.^{4,16,17} More often, reported immunohistochemical aspects include positivity for progesterone receptors, vimentin, neuron-specific enolase (NSE) and α 1-antitrypsin.¹⁸ In our study, all cases tested were positive for progesterone

receptors. Some authors state that these receptors play an important role in tumor growth, but their etiology remains unclear.^{1,19} We also found positivity for vimentin, α 1-antitrypsin and synaptophysin. Tang et al. reported some histological findings suggesting unfavorable prognosis, such as necrosis, nuclear atypia, high mitosis rate, and sarcomatoid areas.¹⁹ None of these were found in our cases, probably anticipating good outcomes.

Due to the excellent prognosis after surgical excision, early reports proposed SPTP to be benign. Current opinion is that these tumors have low malignant potential.^{5,18,20} The possibility of metastasis is low and the outcome is usually favorable after complete excision, but there are reports of involvement of the liver, lymph nodes, and peritoneum. Adjuvant treatment is not indicated but there are a few reports on the administration of chemo and radiation therapy following incomplete resection.^{21,22} None of our cases had adjuvant chemo or radiation treatment.

Regarding the origin of these tumors, some researchers suggest that because of the early age of occurrence, there would be hormonal dependence, but only progesterone receptors have been found to be expressed. More recently, researchers have found nuclear and cytoplasmic accumulation of mutant genes for β -catenin.¹⁸

Since these are low-grade tumors and especially surrounded by a fibrous capsule, some surgeons advocate simple enucleation of the lesion. Nevertheless, pancreatoduodenectomy and distal pancreatic resection are the procedures most often employed. Lymphatic dissection is not recommended.^{3,6,23} In our series of cases, we performed head and distal resections, enucleation and two laparoscopic excisions. The decision to perform resection in all cases was based on the sole presence of a pancreatic mass and the addition of one core biopsy in one of the cases. Open surgery is more feasible in our institution, but eventually is surpassed by laparoscopic resection. However pancreatic laparoscopic procedures demand great expertise, that can be achieved by well-trained surgeons.

Preoperative core biopsy should be considered so that management of solid pseudopapillary tumors can be done surgically. Laparoscopic resection may be a better option but the choice of open pancreatectomy cannot be ruled out, especially in public health institutions. To date, all patients are alive and free of disease.

CONCLUSION

Both our case series and those reported in the literature revealed that SPTP occurs more frequently in young women. Diagnostic suspicion lies on the finding of a bulky, solid and cystic pancreatic mass. Imaging findings might provide diagnostic information before resection. Conservative approaches can be used in selected cases. Long-term survival usually follows complete resection.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

RESUMO

Tumor sólido pseudopapilífero do pâncreas: características clínicas, diagnóstico e tratamento

Introdução: O tumor sólido pseudopapilífero do pâncreas é uma neoplasia rara de baixo potencial de malignidade

e com comportamento incerto, diagnosticado principalmente em mulheres jovens.

Método: Uma série de casos dessa patologia é revisada retrospectivamente, focalizando os aspectos clínicos, tomográficos e imuno-histoquímicos, o tratamento realizado e o desfecho.

Resultados: Foram avaliadas 13 pacientes com massas pancreáticas, por meio de tomografia computadorizada, com diâmetro médio de 8,8 cm. Todas as pacientes submeteram-se a ressecção cirúrgica completa. A imuno-histoquímica confirmou o diagnóstico em todos os casos.

Conclusão: Esses tumores ocorrem frequentemente em mulheres jovens. Suspeita-se do diagnóstico ao encontrar uma volumosa tumoração sólido-cística no pâncreas. As imagens podem fornecer informações diagnósticas antes da ressecção. Podem-se empregar abordagens conservadoras em pacientes selecionados, e a sobrevida geralmente é excelente com a ressecção completa.

Palavras-chave: neoplasias pancreáticas, cirurgia, feminino, adolescente, imuno-histoquímica.

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