

Case report

Solid Pseudopapillary Tumor of the Pancreas, an Unusual Neoplasm






DOI: 10.5377/alerta.v7i1.16357

Eduardo José Fuentes^{1*}, Mirian Liliana Pérez Beltrán², Karla Jeannette Contreras³, Beatriz María Guevara Vásquez⁴, Pedro Ábrego⁵

1-5. Rosales National Hospital, San Salvador, El Salvador.

*Correspondence

✉ ejfo75@gmail.com

1.  0000-0001-8424-3664
2.  0009-0007-8966-4258
3.  0009-0004-8247-5524
4.  0009-0006-4871-1778
5.  0009-0006-0351-5255

OPEN ACCESS

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Suggested citation:

Fuentes EJ, Pérez Beltrán ML, Contreras KJ, Guevara Vásquez BM, Ábrego P. TSolid Pseudopapillary Tumor of the Pancreas, an Unusual Neoplasm. Alerta. 2024;7(1):36-41. DOI: 10.5377/alerta.v7i1.16357

Received:

July 4, 2023.

Accepted:

January 11, 2024.

Published:

January 25, 2024.

Author contribution:

EJF¹, MLPB²: study design. EJF¹, MLPB², KJC³: manuscript design, literature search, data analysis. MLPB², KJC³, BMGV⁴, PA⁵: data collection. MLPB², KJC³: data or software management. EJF¹, MLPB², KJC³, BMGV⁴, PA⁵: writing, revising and editing.

Conflicts of interest:

The authors declare there are no conflicts of interest.

Abstract

Case presentation. A 26-year-old woman who presented with pain in the epigastrium and left hypochondrium, with increased abdominal perimeter and loss of 5 kg of body weight. Physical examination revealed a large mass in the epigastrium, with regular borders, slightly painful to the touch and non-mobile. Imaging studies revealed a mixed neoplasm in the pancreatic body and tail. **Treatment.** A corpo-caudal splenopancreatectomy was performed, with complete removal of the tumor. **Outcome.** The patient received specialized care and close postoperative surveillance in the intensive care unit, with no relevant complications. After hospital discharge, the patient reported a good general condition in the follow-up controls, which included a computed tomography scan performed after 12 months, where no tumor remnants or recurrences were evidenced.

Keywords

Pancreatic Neoplasms, Splenectomy, Pancreatectomy, Helical Computed Tomography.

Resumen

Presentación del caso. Se trata de una mujer de 26 años que presentó dolor en epigastrio e hipocondrio izquierdo, con aumento del perímetro abdominal y pérdida de 5 kg de peso corporal. En el examen físico se detectó una masa de gran tamaño en el epigastrio, con bordes regulares, ligeramente dolorosa al tacto y no móvil. Los estudios de imagen revelaron una neoplasia mixta en el cuerpo y cola pancreática. **Intervención terapéutica.** Se practicó una esplenopancreatectomía corpo-caudal, con extirpación completa del tumor. **Evolución clínica.** La paciente recibió cuidados especializados y vigilancia estrecha posquirúrgica en la unidad de cuidados intensivos, sin presentar complicaciones relevantes. Tras el alta hospitalaria, la paciente refirió un buen estado general en los controles de seguimiento, que incluyeron una tomografía realizada a los 12 meses, donde no se evidenciaron restos o recidivas tumorales.

Palabras clave

Neoplasia Pancreática, Esplenectomía, Pancreatectomía, Tomografía Computarizada Helicoidal.

Introduction

Solid pseudopapillary tumor of the pancreas (SPPT) is a rare exocrine pancreatic neoplasm, which represents approximately 2 % of all pancreatic tumors.ⁱ It was first described by Frantz in 1959ⁱⁱ and later characterized by Hamoudi in 1970, due to this, it received names such as "Frantz" or "Hamoudi" tumor, in addition to other terms related

to its histological appearance.ⁱⁱⁱ In 1996, the World Health Organization established its current designation as "solid pseudopapillary tumor" of the pancreas.^{iv}

SPPT has a tenfold higher incidence in women,^{iv} mainly in the second and third decade of life, with an average age of 22 years^{v,vi} and can develop in any part of the pancreas, however, it occurs more frequently in the distal part of the body and tail.

It is asymptomatic, although in some cases it may present with a gradually growing abdominal mass and nonspecific abdominal pain or discomfort.^v

The mass is usually large, with a well-defined capsule and variable presence of necrosis, hemorrhage and cystic changes. Histologically, it is distinguished by tumor cells, characterized by areas of oriented cells surrounding delicate fibrovascular nuclei, resulting in the pseudopapillary structure.^{vi}

Different imaging modalities, such as ultrasound, computed tomography (CT), and magnetic resonance imaging (MRI), are essential for diagnosis and differentiation from other pancreatic lesions.^{vii} Complete resection of the tumor is curative in most cases.^v

This case report presents the clinical, radiological, and pathological findings of a patient with PSPT diagnosed at the Rosales National Hospital, El Salvador.

The main objective is to highlight the importance of recognizing and correctly diagnosing this clinical entity to ensure adequate treatment. Additionally, it makes it possible to determine the optimal therapeutic strategy and achieve successful results in this disease management.

Case presentation

A 26-year-old woman with a history of mild, cramping, intermittent, non-radiating pain located in the epigastrium and left hypochondrium of one year of evolution. She was relieved with oral analgesics and accompanied by a progressive increase in the abdominal perimeter in the left flank and epigastrium and a loss of approximately 5 kg of body weight. She denied the presence of vomiting, fever, acholia, and choluria. Furthermore, she had no personal

medical or surgical pathological history, and there was no family history of cancer.

The patient initially sought the assistance of a private physician, who subsequently requested abdominal ultrasonography, which revealed the presence of a mixed neoplasm in the retroperitoneum of the left hypochondrium; afterward, she was referred to the hospital.

During the physical examination, she presented without pallor or jaundice, with blood pressure of 120/70 mmHg, heart rate of 82 beats/minute, respiratory rate 18 breaths per minute, temperature of 37° and weight of 63 kilograms. The abdomen was globose, with moderate adipose panniculus, soft and depressible with a large mass, with regular edges, slightly painful to the touch and not mobile, located in the epigastrium, with no signs of peritoneal irritation.

Laboratory tests were performed (Table 1) and showed values within normal limits in the hemogram and liver tests. Special tests revealed normal levels of carcinoembryonic antigen, alpha-fetoprotein, and carbohydrate antigen 19-9.

A computed tomography (CT) scan showed a mass of 14.2 x 12.2 x 13.5 cm in its transverse, anteroposterior, and longitudinal axes occupying the pancreatic body and tail, with defined borders, polylobulated, and some capsular calcifications. The heterogeneous internal composition presented peripheral solid areas that performed with intravenous contrast material and the hypodense liquid component in its more central portions. No signs of infiltration to other organs or adenopathies were found (Figure 1). A solid pseudopapillary tumor of the pancreas was suggested as a preliminary possibility based on the imaging characteristics and the patient's age and gender.

Table 1. Laboratory values

Laboratory Tests	Result	Unit	Normal value	Laboratory Tests	Result	Unit	Normal value
Carcinoembryonic antigen	1.36	ng/mL	Smokers: 0-4.3 Non smokers: 0-3.4	Creatinine	0.60	mg/dL	0.4-1.5
Alphafetoprotein	6.37	ng/mL	0-7	Glucose	95	mg/dL	70-100
CA - 19-9	3.5	U/mL	0-39	Urea nitrogen	12	mg/dL	5-18
Hemoglobin	13.7	g/dL	12-16	Aspartate aminotransferase	26	UI/L	10-42
Hematocrit	41.1	%	36-48	Alanine aminotransferase	34	UI/L	10-40
Leukocytes	8.17	x 10 ³ /μL	5-10	Total Bilirubin	0.48	mg/dL	0.2-1.0
Neutrophils	66	%	55-65	Direct Bilirubin	0.11	mg/dL	0-0.4
Lymphocytes	20.9	%	20-40	Indirect Bilirubin	0.37	mg/dL	0.2-0.8
Platelets	312	x 10 ³ /μL	150-400	Alkaline phosphatase	55	UI/L	30-125

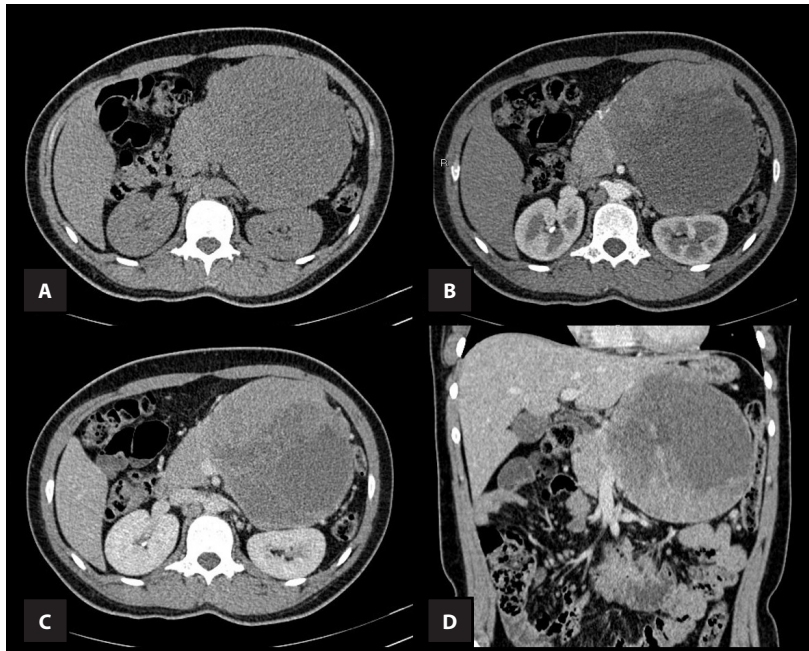


Figure 1. Multiphase helical CT of the abdomen with intravenous contrast. Axial (A-C) and coronal (D) slices in simple (A), arterial (B) and portal venous (C,D) phases

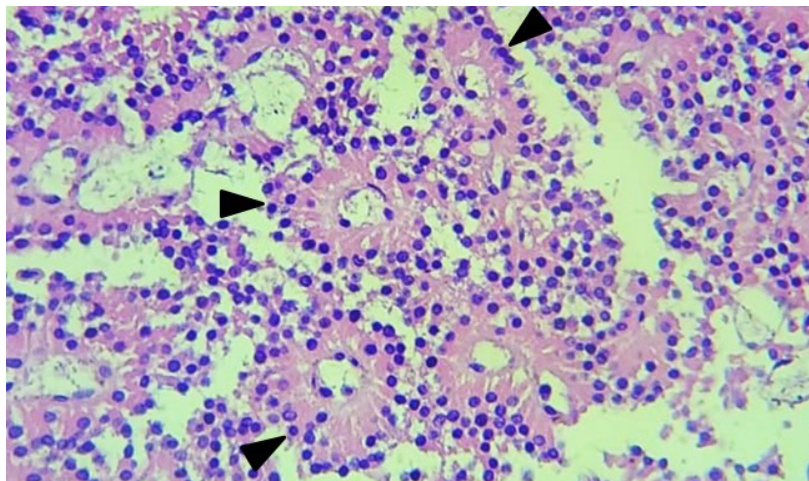


Figure 2. Microscopic examination of the surgical specimen. Histological section with hematoxylin and eosin stain (40x), showing monomorphic cells, poorly cohesive, with round, uniform, basophilic nuclei and eosinophilic cytoplasm, with intra- and extracytoplasmic hyaline globules that adhere to hyalinized fibrovascular cords in a pseudopapillary pattern with perivascular arrangement (black arrowheads). No vascular or perivascular invasion is evident in the capsule. Findings are consistent with a solid pseudopapillary tumor of the pancreas



Figure 3. Helical CT of the abdomen, with oral and intravenous contrast. Axial section (A) and coronal section (B) in venous portal phase. Post-surgical changes consistent with a corpo-caudal splenopancreatectomy are evidenced. The pancreatic head is preserved. There is no evidence of tumor remnants or recurrences, nor intra-abdominal metastatic lesions or adenopathies

Tomographic sections of the upper abdomen showed a large encapsulated mass extending from the body to the tail of the pancreas, with lobulated borders that were well-defined and a heterogeneous internal structure with solid areas in the periphery showing enhancement with intravenous contrast material, together with a central liquid component of lower density. A mass effect was observed in the surrounding organs with a displacement of the same without finding signs of infiltration. (Figure 1)

Treatment

It was decided that surgical intervention was necessary, so five days after his admission, an open corpo-caudal splenopancreatectomy was performed. The tumor was removed from the body and tail of the pancreas, as well as the spleen, five days after admission.

During surgery, a tumor with mixed appearance and consistency was found in the body and tail of the pancreas, which exerted pressure on the portal vein and superior mesenteric vein but did not invade or compromise them, without invasion to neighboring organs. There were no complications during the procedure. The histologic report (Figure 2) showed an encapsulated pancreatic neoplasm consisting of a solid pseudopapillary tumor of the pancreas. Immunohistochemistry and hormone receptor test for progesterone was not performed due to lack of availability.

Outcome

Post-surgical recovery was performed in the Intensive Care Unit for three days, and the following ten days in the general surgery hospitalization area; finally, she was discharged without complications.

Three subsequent check-ups with specialists in general surgery and endocrinology were performed, in which she presented with good general health and no additional symptoms. After 12 months post-surgery, a body weight of 55 kg was recorded, and the follow-up tomographic study (Figure 3) showed no evidence of tumor residues, recurrences or intra-abdominal metastatic lesions.

Clinical diagnosis

Solid pseudopapillary tumor of pancreas.

Discussion

TPSP is a rare epithelial neoplasm of unknown origin that mainly affects women

in the second and third decades of life.^v Unlike other pancreatic neoplasms, it occurs in young people and even children, with a predilection for Asian and African-American women.ⁱⁱⁱ Its detection has increased thanks to the widespread use of imaging techniques such as CT and MRI, which allow the detection of smaller TPSP since many tumors are asymptomatic and found incidentally.^{viii,ix}

It generally develops in the body and tail of the pancreas (55 to 60 %), can also affect the head and neck (35 to 40 %)^{vix} and less frequently, extrapancreatic sites (1 to 1.8 %) such as the colon, mesentery, testis or retroperitoneum.^{ix}

Most have a benign clinical course, but 10 to 20 % have been reported to present malignant degeneration with metastasis and adjacent invasion.^{v-vii}

Five-year survival is excellent, ranging from 93.6 to 98.8 %. Tumors in the pancreatic head have a slightly less favorable prognosis, probably due to the surgical complexity of their anatomical location.^x

Most patients are asymptomatic at the time of diagnosis. When symptoms are present, abdominal pain is the most common^{v,xi}, in addition to other nonspecific symptoms such as nausea, vomiting, weight loss, and the presence of a mass in the right or left upper quadrant of the abdomen.^{vii,x-xii}

Laboratory tests are generally unaltered^{xiii}, and ACE, CA 19-9, and AFP tumor markers are rarely altered.^{xi,xii}

Imaging reveals an encapsulated tumor with solid and cystic components, and sometimes with capsular and intraparenchymal calcifications.^{iii,v,viii} Abdominal ultrasound is the initial method due to its easy access and non-invasive nature, showing solid lesions containing cystic areas or cystic lesions.^v In the presence of diagnostic suspicion, it is recommended to complement CT and MRI, which provide a better characterization of the lesions.

CT shows an encapsulated mass with cystic and solid components due to necro-hemorrhagic degeneration and calcifications in its periphery.^{xii} On the other hand, MRI provides further characterization of the lesion. The body appears weaker on T1-weighted pictures but more intense due to internal bleeding on T2-weighted images. Typically, the solid component enhances poorly, and minimal thickening with capsular enhancement is seen.^{iii,v,viii} Larger lesions enhance heterogeneously, while smaller lesions enhance homogeneously.^{vii,viii} Internal hemorrhage is a characteristic finding, reported in a range of 29 to 88.9 % of cases. In most tumors larger than 3 cm in size, a peripheral capsule is observed.^{viii}

Lanke *et al.* propose a management algorithm after detection by transabdominal ultrasound; CT and MRI are recommended; and in cases of low suspicion of TPSP, a fine needle puncture by endoscopic ultrasound with immunohistochemistry is suggested, which can be useful for preoperative diagnosis, even in patients with high suspicion.^{xiv}

From the macroscopic point of view, they are large tumors with a diameter between 2 and 16 cm and an average size of 5 cm.ⁱⁱⁱ They usually have a heterogeneous macroscopic appearance and are characterized by being round, well-defined, encapsulated, and having a mixed composition, both cystic and solid, in variable proportions.^{viii,xi}

Histologically, it is characterized by a neoplasm with cells arranged in several layers around fibrovascular stalks, resulting in the formation of a pseudopapillary structure.^{x-xv} The pseudopapillary architecture composed of hyaline globules, cholesterol clefts, foamy macrophages and nuclear grooves with absence of neuroendocrine chromatin (salt and pepper) are characteristic.ⁱⁱⁱ

There are molecular alterations, such as karyotypic changes on chromosomes 2, 4, or X, with loss of heterozygosity in the HRAS gene and differential expression of genes, including those associated with tumors. In addition, up-regulation of p27 and p21 is observed, but no mutations in p53 or K-ras are observed, and additionally, ErbB and GnRH signaling pathways are affected.^{ix,x,xv} They also have β -catenin mutations and overexpress cyclin D1 without becoming malignant, and there is evidence of FLL-1, CD56, and progesterone receptor expression, whose genes are on chromosome 11q.^{xvi}

The differential diagnosis of TPSPs includes pancreatic adenocarcinomas, which are the most common cancers in the pancreas. These cancers exhibit a high level of aggressiveness and possess the potential for local invasion and metastasis. Furthermore, cystadenomas and cystoadenocarcinomas must be in the differential diagnosis.^{viii} Although the papillary features of these cystic lesions may resemble those of solid pseudopapillary tumors, their biological behavior and prognosis differ.^{v,vii,viii}

Complete surgical resection is the treatment of choice, and the approach depends on the location of the tumor.^{iii,v,vi,x,xiii,xvii} In tumors located in the head of the pancreas, pancreatoduodenectomy is an option associated with an overall good prognosis, although recurrence can occur^{xii} and related complications have been reported, including pancreatic fistula, postoperative bleeding, delayed gastric emptying and infection.^{viii} A laparoscopic approach can be used to treat

tumors located within the body and/or tail, allowing splenic preservation. Laparoscopic surgery has been shown to cause less blood loss, better postoperative recovery, shorter hospital stay, and lower risk of complications than laparoscopic surgery.^{xii,xiii,xvii}

Two percent of patients experience recurrence after resection.^{xviii} The risk may be increased by factors such as tumor size >5 cm, lymphovascular invasion, lymph node metastases, synchronous metastases, and positive margins.^{xix}

This case includes the clinical, imaging and histologic features of a TPSP, with complete surgical resection, which generated a recovery without recurrence or malignancy.

In conclusion, the presence of a large pancreatic mass with large size mixed solid and cystic nature, encapsulation, and presence of hemorrhage in a young woman must generate suspicion of a TPSP.

Ethical aspects

This case reflects information obtained from clinical records respecting patient confidentiality. Informed consent was obtained from the patient together with a family witness. Data in the publication have been used for academic purposes.

Acknowledgements

To the staff in the Surgery, Radiology and Imaging and Pathology Service of the Rosales National Hospital for their contribution to the care and diagnosis of the case.

Funding

The authors declare no financial support was received for this work.

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