Laparoscopic Surgical Treatment of a Solid Pseudopapillary Tumor of the Pancreas (Frantz's Tumor): Case Report

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Tratamento Cirúrgico Videolaparoscópico de Neoplasia Sólida Pseudopapilar do Pâncreas (Tumor de Frantz): Relato de Caso

Tratamiento Quirúrgico Videolaparoscópico de la Neoplasia Sólida Pseudopapilar del Páncreas (Tumor de Frantz): Informe de Caso

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ABSTRACT

Introduction: Solid pseudopapillary tumor of the pancreas (SPTP), also known as Frantz's tumor, is a rare neoplasm, accounting for approximately 0.17–2.7% of all pancreatic malignancies. The objective of this study is to report the case of an adolescent patient with SPTP who underwent laparoscopic surgery. **Case report:** A 13-year-old female, asymptomatic at presentation, was found to have a perisplenic lesion on routine abdominal ultrasonography. Follow-up imaging one year later demonstrated progressive enlargement of the lesion. Magnetic resonance imaging (MRI) revealed mass adjacent to the pancreatic tail. Laparoscopic resection was performed, for which histopathological evaluation combined with immunohistochemistry confirmed the diagnosis of Frantz's tumor. **Conclusion:** SPTP predominantly affects young females and remains a diagnostic challenge due to its rarity, nonspecific or absent symptoms, and broad differential diagnosis. When technically feasible, laparoscopic resection offers favorable oncological outcomes and provides the advantages of minimally invasive surgery, including reduced postoperative morbidity and faster recovery compared to open procedures.
Key words: Pancreatic Neoplasms/diagnosis; Pancreatic Neoplasms/surgery; Pancreas; Digestive System Surgical Procedures.

RESUMO

Introdução: A neoplasia sólida pseudopapilar do pâncreas, ou tumor de Frantz, é rara, correspondendo a 0,17-2,7% das neoplasias malignas do pâncreas. O objetivo deste trabalho é relatar o caso de uma adolescente com essa patologia tratada por cirurgia videolaparoscópica. **Relato do caso:** Detectou-se em uma mulher de 13 anos assintomática uma lesão periesplênica em ultrassonografia abdominal, com aumento após um ano visto em nova ultrassonografia. Ressonância magnética identificou massa adjacente à cauda pancreática. Optou-se pela exérese por videolaparoscopia, cujo histopatológico e imuno-histoquímica indicaram tumor de Frantz. **Conclusão:** A neoplasia sólida pseudopapilar do pâncreas afeta predominantemente pacientes jovens do sexo feminino, representando um desafio diagnóstico em razão da sua raridade, da ausência ou inespecificidade de sintomas na maioria dos pacientes e da numerosa lista de diagnósticos diferenciais. A realização do tratamento cirúrgico por videolaparoscopia, quando factível, é associada a um resultado oncológico satisfatório e, em comparação à cirurgia aberta, a uma melhor recuperação no pós-operatório.
Palavras-chave: Neoplasias Pancreáticas/diagnóstico; Neoplasias Pancreáticas/cirurgia; Pâncreas; Procedimentos Cirúrgicos do Sistema Digestório.

RESUMEN

Introducción: La neoplasia sólida pseudopapilar del páncreas, o tumor de Frantz, es rara, correspondiendo al 0,17-2,7% de las neoplasias malignas del páncreas. El objetivo de este estudio es reportar el caso de una adolescente con esta patología tratada por videolaparoscopía. **Informe del caso:** En una ecografía abdominal se detectó una lesión periesplénica en una mujer asintomática de 13 años, con un aumento al cabo de un año en una nueva ecografía. La resonancia magnética identificó una masa adyacente a la cola pancreática. Se optó por la exéresis por videolaparoscopía, cuya histopatología e inmunohistoquímica indicaban tumor de Frantz. **Conclusión:** La neoplasia sólida pseudopapilar del páncreas afecta predominantemente a pacientes jóvenes de sexo femenino, representando un desafío diagnóstico debido a su rareza, a la ausencia o inespecificidad de síntomas en la mayoría de los pacientes y a la numerosa lista de diagnósticos diferenciales. La realización del tratamiento quirúrgico por videolaparoscopía, cuando es factible, se asocia a un resultado oncológico satisfactorio y, en comparación con la cirugía abierta, a una mejor recuperación en el posoperatorio.
Palabras clave: Neoplasias pancreáticas/diagnóstico; Neoplasias pancreáticas/cirugía; Páncreas; Procedimientos Quirúrgicos del Sistema Digestivo.

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INTRODUCTION

The solid pseudopapillary tumor (SPTP) is a low-grade malignant tumor primarily located at the pancreas, composed of poorly cohesive uniform epithelial cells forming solid and pseudopapillary structures[1]. This pathology was initially described in 1959 by Dr. Virginia Kneeland Frantz, reason for which it was named Frantz's tumor[2].

SPTP is a rare disease[3]. Most of the times, the symptomatology is unspecific and in nearly 30% of the patients can be asymptomatic with incidental diagnosis through imaging exams[4].

The objective of this study is to report a case of a young patient diagnosed with this pathology, whose surgical excision was performed by laparoscopy and discuss SPTP.

The Ethics Committee approved the study, report number 7,693,594 (CAAE (submission for ethical review): 82540224.5.0000.5047), in compliance with Directive 466/2012[5] of the National Health Council.

CASE REPORT

A 13-year old young asymptomatic woman was submitted to routine abdominal ultrasound which revealed a 4.2x3.3 cm round image with spleen-like eco-texture in close contact with this organ, potentially representing an accessory spleen. A new ultrasound one year later showed enlarged lesion (5.9 x 5.4 cm), being suggested nuclear magnetic resonance (NMR). Enhancement upper abdomen NMR identified expansive well limited heterogeneous formation, with predominance of high signal in T2 and post-contrast interposed enhancement located close to the pancreas tail, with insinuation to the splenic hilum, presenting areas of apparent enhancement, measuring 6.4 x 5.4 cm (figure 1) indicating hypothesis of Frantz's tumor.

At 15 years old, the patient was evaluated by surgical team and an excision was proposed due to potential malignancy. Carbohydrate antigen 19-9 (CA 19-9) and carcinoembryonic antigen (CEA) were requested pre-operatively, both were unchanged. Before the surgery, vaccination against encapsulated germs was performed due to the high likelihood of intraoperative splenectomy.

The procedure was conducted through videolaparoscopy, being found a lesion in the transition between the body and the tail of the pancreas (figure 2 A). The complete dissection of the upper and lower poles of the pancreas and isolation of the splenic vessels were made. Splenectomy was performed due to the contiguity of the vessels with the tumor at splenic hilum level involving the complete joint ligature of the artery and splenic vein associated with body-caudal pancreatectomy. The body of the pancreas was

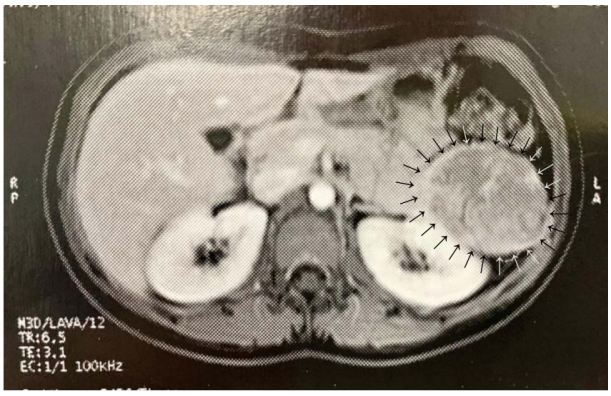

Figure 1. Nuclear magnetic resonance of abdomen (black/grey arrows: solid pseudopapillary tumor of the pancreas)

clamped with blue tape and reinforced with Caprofyl® 3-0. In addition, due to the macroscopic presence of enlarged lymph nodes at the peripancreatic region and unavailable frozen section biopsy intraoperatively, supra-pancreatic and infra-pancreatic lymphadenectomy, block resection of lymph nodes in splenic hilum and partial omentectomy close to the splenic angle were performed. Blake's drain was placed and the surgical pieces were removed through Pfannenstiel incision (figure 2 B).

After the procedure, the patient was discharged six days after surgery. In the 18th day post-operation presented fever and leukocytosis. Computed tomography (CT) showed collection of 300 ml at the left subphrenic region, the patient was admitted for antibiotic therapy and tentative CT-guided punction. Because collection reduced and symptoms remission the punction was not required. She was discharged at the 9th day without new complications.

The histopathological and immunohistochemistry analysis of the lesion was consistent with the diagnosis of SPTP (Chart 1).

The patient was referred to a specialized oncology service, she is in follow-up with no complaints or signs of relapse.

DISCUSSION

SPTP accounts for 0.17-2.7% of the pancreatic malignant tumors[3], located in 66-74% in the body or tail and in 26-34% of the cases at the head of the pancreas[6]. Rarely it can occur in the stomach, duodenum, omentum, mesentery, retroperitoneum or ovary[1].

The pathology affects women mainly, at a rate of 1.7:1 to 10:1 in relation to men, being diagnosed between 23 and 35 years[3]. One of the explanations for the predominance in women is the presence of progesterone receptors in the tumors. Kosmahl et al.[7] in a study with patients with SPTP, revealed that 100% of the neoplasms evaluated had progesterone receptors.

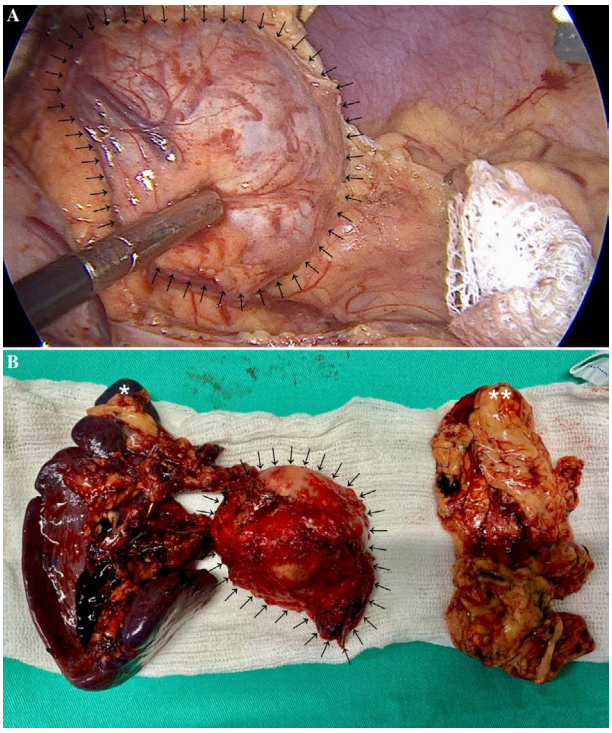

Figure 2. A: Videolaparoscopy imaging (black arrows: solid pseudopapillary tumor of the pancreas); B: Image of the excised surgical pieces (black arrows: solid pseudopapillary tumor of the pancreas; *: Spleen; **: greater omentum)

The pathogeny of Frantz's tumor is still uncertain. It is known that there are genetic mutations related to SPTP that favor the accumulation of beta-catenin in cellular cytoplasm and, later, inside the nucleus, to which it binds to key-elements of DNA regulation as the genes regulating the cyclin growth D1 and C-MYC[8]. Furthermore, it is suggested that the carcinogenesis of SPTP is associated with the incorporation of primitive ovary cells inside the pancreatic parenchyma during the embryonic development. However, it does not explain the occurrence of this neoplasm in men[7].

The main clinical findings in symptomatic patients are pain, abdominal discomfort and palpable abdominal mass. Some patients evolve with abdominal distention, nausea and/or vomits, early satiety, hyporexia, fatigue, fever and/or anemia. Weight loss, jaundice and acute pancreatitis are rare manifestations. In addition, the literature reports hemoperitoneum due to rupture of tumor capsule[1-4, 6-13].

Lymph node metastases occur in nearly 1.6% of the cases and distal metastasis in 7.7% of the cases, including liver, lungs, peritoneum and mesentery of the small and large intestine. SPTP can evolve with direct infiltration of adjacent structure, for instance, spleen, duodenum and portal vein[1,9].

Chart 1. Histopathological and immunohistochemistry analyses of the pieces excised

Microscopy and diagnosis		
1) Pancreas: neoplasm composed of poorly cohesive uniform epithelial cells displayed in delicate interspersed sheets, sometimes papilla-like with xanthomatous macrophages. The neoplasm has 5.5 cm in its longest axis. The nucleus is rounded, regular borders, with homogeneous chromatin and little evident nucleolus. The cytoplasm is poorly limited, sometimes eosinophilic or clear. Cystic-hemorrhage areas were observed and interspersed fibro-hyalinized stroma. Absence of mitosis figures.		
2) Surgical margins: neoplasm-free		
3) Peripancreatic lymph nodes: Absence of neoplasm. 11 neoplasm-free lymph nodes		
4) Spleen: neoplasm-free		
5) Splenic hilum: neoplasm-free		
6) Omentum: neoplasm-free. Two neoplasm-free lymph nodes. Neoplasm-free omentum		
Panel of antibodies		
Antibody	**Clone**	**Interpretation**
CK AE1AE3	AE1AE3	Positive in neoplasm
Chromogranin A	Polyclonal	Negative in neoplasm
Beta catenin	Polyclonal	Positive in neoplasm (nuclear pattern)
Vimentin	V9	Positive in foci of neoplasm

The final pre-operation diagnosis of SPTP is challenging due to its rarity, unspecified symptoms and broad differential diagnoses that encompass benign cystic lesions and other pancreatic neoplasms[8-10]. Abdominal ultrasound, CT with intravenous contrast and NMR are useful for pre-operation diagnosis[9,10]. Endoscopic ultrasound-guided fine-needle aspiration biopsy (FNAB) can also be utilized because it allows a histopathological and immunohistochemical analysis of the neoplasm[11]. Regardless of low incidence of adverse events, Yamagochi et al.[14] described a case of metastatic implant of SPTP in the stomach of a patient post endoscopic ultrasound-guided FNAB with distal gastrectomy.

The prognostic and diagnostic utility of tumor markers as CA 19-9 and CEA is limited because the values are unchanged for most of the patients[6].

Surgical resection is the treatment of choice with preservation of pancreatic tissue as much as possible without losing the oncologic efficacy to avoid pancreatic failure. In tumors of the head of the pancreas, duodenopancreactectomy with pyloric preservation is better than conventional Whipple surgery. In lesions of the neck of the pancreas, the middle portion of the organ can be resected when there is local extra-pancreatic extension involving the removal of the portal and inferior vena cava or even of the inferior mesenteric vessels if necessary[8]. Additionally, enucleation avoids extensive resections[13].

Further to the rarity of SPTP, the utilization of videolaparoscopy is still uncommon, which prompted the authors to describe this case. A systematic review by Law et al. analyzed pancreatic resections performed in 2,285 patients with SPTP. In 1,903 cases, the surgery was open, in 39, laparoscopy was utilized and in 343, the method was unknown[12]. Laparoscopy is a treatment option associated with least bleeding, short hospitalization and better esthetic result compared with open surgery. Regardless of these advantages, laparoscopy approach should be avoided in large tumors, especially when there is proximity to vascular structures[13].

Pancreatic fistula is the most common complication in surgery in addition to potential delay of gastric emptying, pancreatitis, hemorrhage and/or infection[6,8].

So far, radiotherapy and chemotherapy are therapeutic options of uncertain efficacy due to the scarce experience with SPTP[8].

Overall, SPTP has good prognosis, more than 95% of the patients submitted to full exeresis of the lesion achieve disease-free survival[12]. Recurrence and local invasion are not contraindications for resection and in selected cases, metastases can also be managed surgically. In cases of tumor irresectability, cytoreductive surgery can be considered, depending on the case[10].

CONCLUSION

Predominantly, SPTP affects young female patients, a challenging diagnosis due to its rarity, absence of or non-specific symptoms in most of the patients and broad differential diagnoses. When feasible, surgical treatment through videolaparoscopy is associated with satisfactory oncologic outcome with better post-operative recovery than open surgery.

CONTRIBUTIONS

All the authors contributed substantially to the conception and design of the study, analysis and interpretation of the data, writing and critical review and approved the final version to be published.

DECLARATION OF CONFLICT OF INTERESTS

There is no conflict of interests to declare.

DATA AVAILABILITY STATEMENT

All the content underlying the text is contained in the manuscript.

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None.

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