Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

# Frantz's tumor as incidental finding during Heller myotomy for achalasia: case report and review of literature

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**Introduction:** Solid pseudopapillary tumor is a low-grade malignancy of the pancreas and predominantly affects young women. This neoplasm is a rare pancreatic entity with vague clinical presentation. Diagnosis is often incidental through imaging or even during surgical approach for another condition.

**Case presentation:** A 22-year-old Brazilian female with gastrointestinal symptoms was diagnosed with achalasia and underwent Heller myotomy. Intraoperatory findings included an enlarging mass in the distal pancreas. During follow-up for the surgical approach of achalasia, a hypothesis of Frantz's tumor was stated, and spleen-preserving distal pancreatectomy was performed.

**Discussion:** The pathological pathways of Frantz's tumor is still unclear, and its connection with chromosomal abnormalities is under investigation. Although the tumor has been reclassified over the years to solid pseudopapillary tumor, surgical resection remains the standard treatment.

**Conclusion:** Despite a surgical challenge, surgery presents a great prognosis in these patients and long-term survival. High suspicion and proper investigation are fundamental to diagnosis and early treatment.

**Keywords:** achalasia, Frantz's tumor, pancreas, pancreatectomy, solid pesudopapillary tumor, solid pseudopapillary neoplasm

## Introduction

Solid pseudopapillary tumor (SPT) of the pancreas was first described in 1959 by Virginia Frantz[1]. Nowadays, Frantz's tumor is classified as a solid pseudopapillary neoplasm of low-grade malignancy, and it represents about 1–3% of all exocrine pancreatic tumors[2–4]. It affects more women than men at a young age, with a preference for Black and Asian females[2,5,6]. Despite its low malignant potential, metastasis may still occur in 15% of the patients[3,7,8].

The tumor presents as a solid-cystic enlarging abdominal mass with origin in any part of the pancreas, slow-growing and low metastatic rate[4,6,9]. Clinical presentation may vary from asymptomatic patients to vague gastrointestinal symptoms or intestinal obstruction[6,10]. Diagnosis is usually incidental through imaging due to nonspecific symptomatology[6].

We report a case of a 22-year-old Brazilian woman with an incidental diagnosis of a solid-cystic pancreatic tumor during Heller myotomy in a reference center in the Amazon.

This case report is being reported in line with the SCARE 2023 criteria[11].

### HIGHLIGHTS

- Solid pseudopapillary neoplasm is a rare condition of the pancreas, and it usually affects women between the second and third decades of life.
- Despite low malignant potential, metastasis may still occur in 15% of the cases.
- Surgical resection is the standard treatment for Frantz's tumor presenting a great prognosis and long-term survival.

## Case presentation

A 22-year-old female was first admitted in our digestive surgery service for surgical treatment of achalasia. She complained of a ten-month history of progressive dysphagia and post-prandial vomiting, associated with sialorrhea mostly at night. In the moment of first admission, she presented with dysphagia even for water, retrosternal pain and involuntary weight loss of 15 pounds in 6 months. She had no significant past surgical history, absence of comorbidities or drug allergy.

At first examination, the patient was hemodynamically stable, afebrile, acyanotic, anicteric and hydrated. There were no respiratory or cardiovascular abnormal findings. The abdomen

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Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

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Annals of Medicine & Surgery (2024) 86:4861–4864

Received 2 April 2024; Accepted 3 June 2024

Published online 17 June 2024

http://dx.doi.org/10.1097/MS9.0000000000002273

was flat, soft, non-tender and non-distended with no palpable masses or signs of peritonitis.

Esophageal manometry revealed the absence of effective contraction, unrelaxed internal esophageal sphincter and integrated relaxation pressure in 100% of deglutition, caractherizing type 2 achalasia. She was also submitted to an esophagus, stomach and duodenum seriography showing dilated thoracic esophagus of 3.5 cm and distal esophageal narrowing. Two upper gastrointestinal endoscopies with histopathological analysis were performed, the first one identifying mild gastritis and the presence of *Helicobacter pylori*. The second one after lansoprazole, clarithromycin and amoxicillin scheme with no evidence of the bacteria.

The patient was submitted to laparoscopic Heller myotomy by digestive surgery service. Intraoperative findings included a pancreatic mass involving the tale of the pancreas and firm adherence with the spleen. There were no additional abnormal findings. During the postoperative recovery, she experienced improvement in gastrointestinal symptoms. Progressive meal planning was conducted with a multidisciplinary team involving speech therapy, nutritionist and physical therapist. She was discharged four days after surgery, oriented to maintain an oral diet with pasty food initially, and continue outpatient investigation of pancreatic mass during follow-up.

Before being discharged, she was submitted to an abdominal and pelvic computed tomography (CT) that revealed an expansive heterogeneous mass of solid and liquid components in the tail of the pancreas, measuring $7.8 \times 8.0 \times 8.0$ cm displacing adjacent structures. She evolved with satisfactory recovery after surgical treatment of achalasia, and despite orientations on the need for surgery for the pancreatic lesion, the patient did not continue with medical follow-up and after 1 year of lost contact, she returned to our service. An abdominal CT was performed revealing enlargement of the pancreatic mass to $11.5 \times 8.8 \times 8.6$ cm, now involving the body and tail of the pancreas (Figs. 1 and 2), dislodging the stomach and being in close contact with the fourth duodenal portion, with clear cleavage planes between the tumor and the left kidney and its vein. Other vessels were not affected by the mass. CA 19-9 was negative.

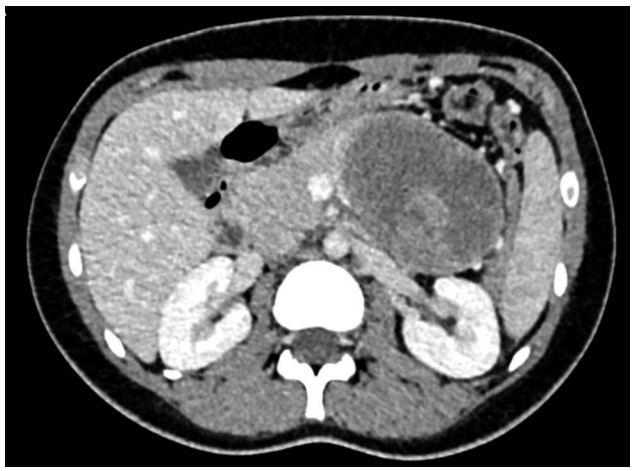

**Figure 2.** Axial abdominal contrast-enhance computed tomography revealing distal pancreatic mass containing solid and cystic components.

She was submitted to an open spleen-preserving distal pancreatectomy (Fig. 3) with clean margins performed by a digestive surgeon and general surgeons, then sent to ICU for close postoperative monitoring as it is routinely done in our service. In first postoperative day, the patient evolved with tachycardia, pallor and hematocrit drop. The surgical drain was assessed, noticing a high bloody output of 700 millimeters. Reoperation was performed and hemostasis of retroperitoneal dissection was carried out. The patient remained in ICU for one day and, with no further events, hemodynamically stable, was sent to the ward, evolving with mild pain on the drain incision, responsive to analgesics, progressive reduction of drain output and good oral dietary intake. No fever or vomiting were reported. Until the present moment (6 months later), the patient remained asymptomatic, fully recovered, with great satisfaction on her treatment and no signs of recurrence on imaging evaluation.

Histopathological analysis revealed heterogeneous lesions with solid and pseudopapillary areas, connective stroma and calcification. SPT was confirmed, measuring $11.0 \times 8.0 \times 7.0$ cm with clean margins and no signs of lymphovascular invasion. The

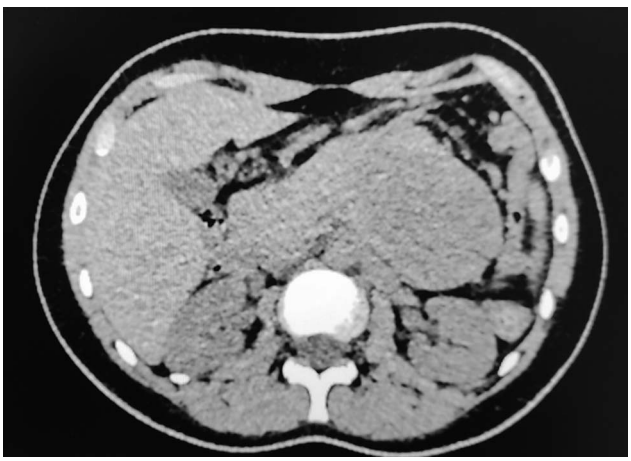

**Figure 1.** Axial abdominal computed tomography showing distal pancreatic heterogeneous mass.

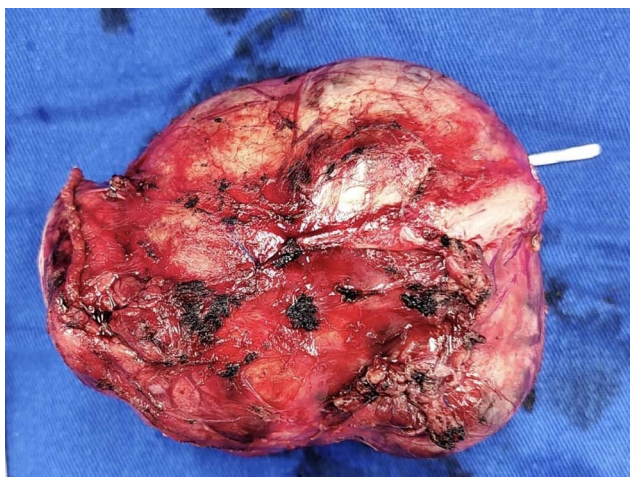

**Figure 3.** Product of distal pancreatectomy.

largest cystic cavity of 9 cm with bloody clots inside were described, associated with a solid area of 5.0 × 1.4 cm.

## Discussion

SPT of the pancreas is a rare entity, and its definition and classification has been discussed since 1959[5,8,12]. Nowadays, it is classified as borderline features according to the WHO, and it can be found in the literature as solid-cystic tumor, solid papillary tumor, papillary-cystic tumor and solid-cystic acinar tumor of the pancreas[13,14]. It is more common in women, especially in younger individuals between the second and third decade of life, accounting for 1–3% of all exocrine neoplasms of the pancreas[3,4]. Our patient was female and her age falls within the range at diagnosis.

Clinical presentation is generally vague and most of the cases are asymptomatic or cause nonspecific symptoms according to the mass size. Abdominal distention, pain, nausea, vomiting and early satiety are some common symptoms[5,9,14]. Although some patients may present nonspecific gastrointestinal symptoms, a slowly enlarging abdominal mass might be palpable in the upper abdomen. However, diagnosis is usually incidental[9,14]. In our case, the patient was previously diagnosed with achalasia and during laparoscopic Heller myotomy, a pancreatic mass involving body and tail of the organ was identified. In post-recovery and follow-up, hypothesis of SPT of the pancreas was stated.

Pathogenesis of SPT of the pancreas is still unclear. Currently, chromosome abnormalities are the theme of study to determine their role in the pathological process[8,14]. Complex changes in karyotype such as breakpoints, bands or monosomy have been reported as molecular alterations in chromosomes 2, 4 and X[4,12,16].

Initial investigation starts after proper physical examination through ultrasonography (US) and CT scan. Heterogeneous solid mass that may contain hypoechoic cystic areas is present in US, while CT reveals large well-delimited mass with solid and cystic components with heterogeneous enhancement in contrast exam[5,15,17]. Cystic elements are found to be more central and solid components more peripheral[17]. Laboratory tests and tumor markers are usually within normal values[4,15]. Some authors defend fine-needle (FNB) biopsy; however, other authors are discordant due to possible tumor cell spread and the fact that FNB does not differentiate between Frantz tumor and pancreatoblastoma[15]. Our patient was submitted to an abdominal CT scan with contrast study to analyze the pancreatic mass and establish surgical planning.

The most common sites affected by this condition are the distal body and tail, followed by the head of the pancreas[4,14,17]. Nevertheless, some studies pointed out a different predominance for the head (30–40%), body (32%) and tail (28%), respectively[6]. Median size at diagnosis is 3.6 cm (range of 0.9–15 cm); however, tumor larger than 30 cm have been reported[4,9,14]. In our case, Frantz's tumor was first confined in the body of the pancreas; however, our patient failed to continue follow-up for almost a year and tumor was posteriorly involving pancreatic body and tail.

Frantz's tumor management is based on surgical resection and attempt to be as conservative as possible[14]. Most tumors are diagnosed in localized stage and with its low malignant potential, surgery remains the best chance of long-term survival. Surgical planning depends on the tumor location, varying from distal pancreatectomy to Whipple's surgery. Currently, there are less aggressive techniques in advance such as enucleation, central pancreatectomy, Kimura's and Warshaw's techniques[4,6,18]. Splenectomy is frequently performed in cases of tumor mass close to the splenic hilum; however, the laparoscopic approach is more related with the possibility of spleen preservation[14]. Our patient was submitted to an open spleen-preserving distal pancreatectomy.

Histopathological analysis reveals two types of tissue in macroscopy: solid areas mixed with pseudopapillary components and hypervascularization[12,15]. When the tumor cells disconnect from the arterial blood and form fibrovascular tangles, associated with necrosis and hemorrhagic cavities in microscopy[4,14]. Immunohistochemistry usually reveals positivity for vimentin in all cases and beta-catenin, cytokeratins, CD10, CD56, α1-antitrypsin and α1-chymotrypsin in most cases[4,6,9,14].

Despite low malignant potential of Frantz's tumor, metastases occur in 10–15% of the cases[6,8]. The most common metastatic location is the liver and survival rate remains between 94 and 97%, even in 5-year survival in metastases[2,5]. Differential diagnosis of SPT must include pseudocysts, hydatid cysts, cystic tumors such as cystadenoma, cystadenocarcinoma and lymphangioma[2,6,9].

Additionally, Frantz's tumor presents borderline characteristics once it is a malignant neoplasm with benign progression in most cases. Further molecular, genetic and histological analysis is necessary to understand its pathogenesis. Follow-up is conducted with CT scan or MRI every 3 months in the first year, every 6 months in the second year and once a year for 5 years[2].

## Conclusion

Frantz's tumor is an uncommon but relevant condition of the pancreas, which may be incidentally found in image studies or even during surgery. High suspicion and proper investigation are essential to diagnose this entity in a timely manner. Pancreatic tumors are a surgical challenge and planning with image evaluation is necessary.

## Ethical approval

This study was exempt from ethnical approval.

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Source of funding

Not applicable.

## Author contribution

T.R.M.P.: study conception, data collection, writing the paper, design. G.L.d.C.: study conception, data collection, writing the paper and design. D.C.R.: study conception and data collection.

R.A.d.S.N.: study concept, data analysis, supervision and final revision. R.A.d.S.J.: study concept, data analysis, supervision and final revision.

## Conflicts of interest disclosure

The authors declare that the article content was composed in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

## Research registration unique identifying number (UIN)

Not applicable.

## Guarantor

Thaís Regina Moreira Printes.

## Data availability statement

Not applicable.

## Provenance and peer review

Not commissioned, externally peer-reviewed.

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