

SOLID PSEUDOPAPILLARY TUMOR OF PANCREAS “FRANTZ TUMOR” CASE REPORT AND REVIEW

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TUMOR PSEUDOPAPILAR SÓLIDO DEL PÁNCREAS “TUMOR DE FRANTZ” INFORME DE CASO Y REVISIÓN

ABSTRACT

The solid pseudopapillary pancreatic tumor, also called “Frantz tumor”, was first described in 1959 by Dr. Frantz Gruber. Part of the classification of cystic neoplasms of the pancreas, it is a condition presented predominantly in young women, with a frequency of 90% of cases. A 24-year-old female patient complained of abdominal pain for 3 days, accompanied by nausea, vomiting, and abdominal distention. A double-contrast and a PET scan were performed that identified a lesion with a solid aspect in the pancreatic tail and in the liver segments VIII and V. A subtotal pancreatectomy with splenectomy and biopsy of liver lesions was performed, reporting positive for malignancy. It was decided to perform a right portal vein ligation. On the eighth postoperative day, right-sided two-stage hepatectomy was completed. On the first postoperative day of the second surgery, she presented clinical signs compatible with bleeding, confirmed with tomography as a hematoma in the surgical bed. Exploratory laparotomy was performed with hematoma drainage. She went to an Intensive Care Unit, where she evolved without complications. Postoperative bleeding in patients with hepatectomy varies from 4.2% to 10%, and there are several reasons for its occurrence. In our case, the diagnosis and control of bleeding on the surface of the resection were accurately performed. Postoperative bleeding is one of the most frequent operative complications of hepatectomy, so it is necessary to bring it under control.

Key words: Pancreatic neoplasm, Frantz tumor, metastasis, portal vein ligation, hepatectomy

RESUMEN

El tumor pancreático pseudopapilar sólido, también llamado “Tumor de Frantz”, fue descrito por primera vez en 1959 por el Dr. Frantz Gruber. Forma parte de la clasificación de las neoplasias quísticas del páncreas y se presenta predominantemente en mujeres jóvenes, hasta en el 90% de los casos. Una paciente de 24 años, con dolor abdominal de 3 días de evolución, acompañado de náuseas, vómitos y distensión abdominal. Se le realizó una tomografía computarizada que identificó una lesión con aspecto sólido del parénquima pancreático a nivel de la cola, y lesiones hepáticas en los segmentos VIII y V. Se le practicó una pancreatectomía subtotal con esplenectomía y una biopsia de las lesiones hepáticas, que resultó positiva para malignidad. Se decidió realizar una ligadura de la vena porta derecha. En el octavo día postoperatorio, se completó una hepatectomía derecha en dos etapas. En el primer día postoperatorio de la segunda cirugía, presentó signos clínicos compatibles con hemorragia, que se confirmó mediante tomografía computarizada como un hematoma en el lecho quirúrgico. Se realizó una laparotomía exploratoria con drenaje del hematoma. La paciente fue trasladada a la Unidad de Cuidados Intensivos, donde evolucionó sin complicaciones. El sangrado posoperatorio en pacientes sometidos a hepatectomía varía entre el 4,2 % y el 10 %, y las causas descritas son diversas. En nuestro caso, en relación con el sangrado en la superficie de la resección, el diagnóstico y el control se realizaron correctamente. El sangrado posoperatorio quirúrgico es una de las complicaciones operatorias más frecuentes de la hepatectomía, por lo que es necesario tenerlo en cuenta y no demorar su tratamiento para un control definitivo.

Palabras clave: Neoplasia pancreática, tumor de Frantz, metástasis, ligadura de la vena porta, hepatectomía

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Recepción: 11/10/2025
Aprobación: 29/05/2026
DOI: [10.48104/RVC.2026.79.1.17](https://doi.org/10.48104/RVC.2026.79.1.17)
www.revistavenezolanadecirugia.com

INTRODUCTION

The solid pseudopapillary tumor of the pancreas (SPT) also called "Frantz's tumor" was first described in 1959 by Dr. Frantz Gruber, however, it was in 1970 when Hamoudi described the histopathology and until 1980 that Kloppel described it as a particular clinical entity. In 1996 the World Health Organization (WHO) defined it as "Pseudopapillary solid neoplasm" ⁽¹⁾.

This tumor belongs to the classification of pancreatic cystic neoplasms, being the least common, with an incidence of 1–3% among them. It predominantly affects young women in 90% of cases, with a mean age of 22 years (range: 20–30 years), although it has also been reported in men (mean age: 37 years) and children (under 12 years old) ^(1,2). The average size ranges from 6 to 8 cm, but some cases reach diameters of 15 to 22 cm ⁽⁷⁾. Furthermore, although it presents a low risk of malignancy, up to 15% of patients may develop metastases ⁽⁸⁾. SPT has been associated with mutations in exon 3 of CTNNB1 and alterations in Wnt/B-catenin and Hedgehog signaling pathways ⁽⁷⁾.

In this context, the ALPPS technique (Associating Liver Partition and Portal vein Ligation for Staged Hepatectomy) first described in 2012 ⁽⁹⁾, is designed to rapidly increase the volume of the future remnant liver (FRL), allows liver resections in two stages ⁽⁹⁾ It combines two established procedures: right portal vein ligation (PVL), which restricts blood flow to a portion of the liver, and in situ liver splitting, which allows for enhanced regeneration of the organ to facilitate resection of the remaining portion ⁽¹¹⁾.

ALPPS-induced liver regeneration is mainly attributed to portal hemodynamic alterations rather than to circulating proliferative factors. Hepatic transection and portal ligation generate an increase in portal flow in the FLR, which increases portal pressure and stimulates hepatocyte hypertrophy, reaching a growth of 65% to 110% in an interval of 6 to 15 days. This process involves the activation of signaling pathways such as JNK1-IHH, the production of nitric oxide by the sinusoidal endothelium and the induction of hypoxia in the FLR, which enhances cell proliferation. In experimental models, an increase of 60% in 24 hours and up to 134% in 72 hours has been reported. In addition, hepatic transection disrupts porto-portal collaterals, preventing venous outflow leakage and ensuring a more effective regenerative stimulus. These findings suggest that regeneration in ALPPS is predominantly driven by changes in hepatic hemodynamics and sinusoidal microcirculation, rather than by cytokines or growth factors ⁽¹⁰⁾.

ALPPS has been used mainly in patients with bilobar liver metastases or primary liver tumors initially considered unresectable, achieving success rates close to 99% in the second stage ⁽⁹⁾.

On the other hand, there are different types of ALPPS that are applied according to the clinical situation of the patient, including Classic ALPPS, which involves complete transection of the hepatic parenchyma and ligation of the portal vein, and Partial ALPPS, with partial transection of the parenchyma and ligation of the portal vein. Mini ALPPS is characterized by partial

parenchymal transection and portal vein embolization through the inferior mesenteric vein. In addition, Hybrid ALPPS combines complete transection with an anterior approach and two-stage portal embolization. Finally, Modified ALPPS preserves the portal pedicles during liver transection ⁽⁹⁾.

The accelerated hepatic regeneration promoted by the ALPPS technique allows complete resection in a shorter period of time, usually between 7 and 15 days between surgical stages; however, this technique also entails an increase in postoperative morbidity ⁽⁹⁾.

We are dealing with an infrequent entity with an even rarer presentation, so we consider it important to present the case and a review of the subject with the complications associated with surgical treatment.

CASE PRESENTATION

24-year-old female patient, with a history of peptic acid disease of 2 years of evolution under control with medical treatment and irritable bowel syndrome under control. She started her symptoms with abdominal pain of 3 days of evolution, of transictal type in the epigastrium of intensity 7/10, accompanied by nausea, vomiting and abdominal distension. On physical examination there were no specific findings, the initial laboratories were within normal parameters, so imaging studies were performed.

Investigations and Examinations

Double contrast tomography identified a solid-cystic lesion of the pancreatic parenchyma at the level of the tail with dimensions of 62 x 75 x 70 mm. In addition, in the liver there were lesions in segment VIII of 13 mm and segment V of 16 and 12 mm (Figure 1).

The patient was admitted to complete the study protocol, PET CT was requested and reported a lesion in the tail of the pancreas, of heterogeneous density, solid, with lobulated and well-defined border, measuring 67 x 83 mm in its major axes, associated with metabolism with a maximum SUV of 14.5. The liver with multiple hypodense lesions in segments V, VI, VII and VIII, the largest lesion in segment 6, measuring 17 mm, and associated with increased metabolism with maximum SUV of 7.1, with splenomegaly (Figure 3). Tumor markers were requested, which were reported in normal ranges; Carcino Embryonic Antigen of 5, Alpha-feto protein of 1.1, Ca 19.9 of 2. Subsequently, an endoscopic ultrasound biopsy was performed, which reported a solid pseudopapillary tumor of the pancreas (TSP), so she was scheduled for surgery.

Treatment

She was scheduled for subtotal pancreatectomy with splenectomy (Figure 4), biopsy of hepatic lesions in segments IV, VI, VII reported positive for malignancy so it was also decided to perform ligation of the right portal vein. Without complications.

The pathology reported: Pancreas; solid pseudopapillary carcinoma without neoplasia in surgical edge. Liver biopsy; solid



Figure 1. Double contrast CT-Scan identified a solid-cystic lesion on the pancreatic parenchyma at tail's level, measuring 62 x 75 x 70 mm, with heterogeneous reinforcement and mass effect towards the greater curvature of the stomach and involvement of splenic arterial and venous branches with engorgement and collateralization of its branches. In addition, the liver showed solid hypodense lesions with diffuse heterogeneous reinforcement during the contrasted phases suggestive of secondary deposits; in segment VIII of 13 mm and segment V of 16 mm and 12 mm

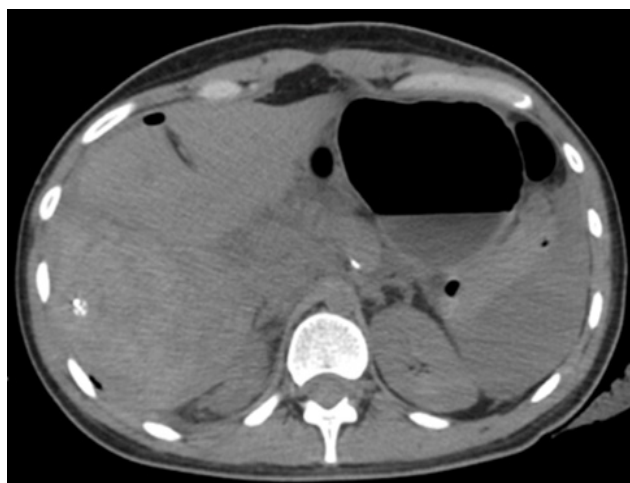


Figure 2. Extensive hematoma is observed in surgical bed, it extends from the subphrenic region to the flank, with approximate dimensions of 20 x 9 x 7.8 cm, with a 730 cc estimated volume. The hematoma had heterogeneous density ranging from 20 to 66 HU, due to different stages of bleeding

pseudopapillary carcinoma metastatic in two hepatic wedges.

RESULTS

On the 8th postoperative day a right hepatectomy was performed and the transoperative findings were reported; Hypertrophy of the left hepatic lobe with metastasis of pseudopapillary carcinoma of the pancreas limited to the right liver, without evidence of invasion by transoperative ultrasound.

On the first postoperative day of the second procedure, she presented with hemorrhage confirmed by tomography (Figure 2) as a hematoma in the surgical site. She underwent exploratory laparotomy in which a 1300 ml hematoma drainage and hemostasis of the surgical site was performed. She was transferred to the Intensive Care Unit due to past reports of hemorrhage and for close monitoring, she evolved without complications and was discharged on the ninth postoperative day.

DISCUSSION

Solid pseudopapillary tumor of the pancreas is a rare tumor and requires surgery for its treatment. Among its frequent postoperative complications are biochemical leakage and pancreatic fistula, peripancreatic collections and abscesses, hemorrhage and hematoma, pancreatic pseudocyst, intestinal occlusion, cholangitis and surgical wound infection ⁽³⁾.

Postoperative bleeding in hepatectomy patients ranges from 4.2-10%, and the described reasons for its occurrence are

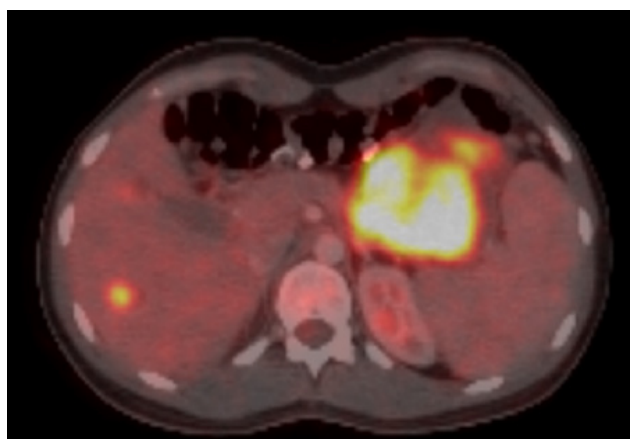


Figure 3. The liver presented multiple hypodensity lesions in segments V, VI, VII and VIII, the largest lesion on segment VI with 17 mm, associated with an increased metabolism with a maximum SUV of 7.1 and splenomegaly. No mesenteric, retroperitoneal adenopathy, iliac chains or inguinal were identified

threefold: 1) bleeding from the resection surface, 2) incomplete hemostasis in the transoperative period, and 3) loss of tension or release of vascular staples or sutures ^(4, 5).

In this case, the hemorrhage was related to the first cause described, was diagnosed and optimally controlled. It is increasingly feasible to diagnose and treat these tumors at a wider range of stages, however, complications arising from pancreatic and hepatic resections represent a significant challenge for the surgeon and the medical team. The morbidity and mortality of these patients can increase rapidly, so these procedures should be



Figure 4. Product of the distal pancreatectomy with splenectomy

performed in specialized centers by a multidisciplinary team with experience in the management of these complications.

Hepatopancreatectomies are highly complex procedures that require meticulous surgical planning. In this case, in order to optimize the hypertrophy of the future remnant liver (FLR) and to allow a safe two-stage resection, we chose to perform a Mini-ALPPS, a modified variant of the classic ALPPS. Unlike the latter, where hepatic transection is complete in the first stage, Mini-ALPPS employs partial parenchymal transection and portal vein embolization to stimulate hepatic hypertrophy in a more controlled manner, avoiding extensive manipulation of the hepatic hilum and reducing the physiological impact of the procedure ⁽¹²⁾.

In previous studies, it has been shown that liver regeneration in Mini-ALPPS occurs mainly due to hemodynamic changes that increase portal flow to the FLR, favoring a more rapid hypertrophy than that observed in conventional techniques such as portal embolization ⁽¹²⁾. However, although Mini-ALPPS allows accelerated liver regeneration, it has also been reported that it may be associated with higher postoperative morbidity and mortality compared to less invasive techniques, particularly in patients with advanced liver fibrosis ⁽¹²⁾.

In comparison with other reports of Mini-ALPPS, in this case we were able to complete the resection in two times with adequate FLR growth and without major postoperative complications. These findings reinforce the usefulness of this technique in patients with liver metastases from pancreatic tumors, in whom the remaining liver volume may be a limiting factor for complete oncologic resection ⁽¹²⁾.

CONCLUSION

Frantz tumor is a rare entity, with greater frequency in young women, surgery resection with free margins is the therapeutic option of choice. Routine lymphadenectomy has not proven to be useful. Metastases of this tumor are present in 20% of the cases at the time of diagnosis and the cases in which a resection of these metastases can be performed in the same surgical time are exceptional ⁽⁶⁾.

The frequency of complications faced peri-operatively due to resection of the primary tumor in the pancreas or metastases in the liver have a direct influence on the patient's prognosis.

AUTHORS' CONTRIBUTIONS

All authors contributed substantially to the development of this manuscript. Case conception, data collection, literature review, and drafting of the report were shared responsibilities. All authors reviewed and approved the final version of the manuscript.

CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest related to this publication.

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