

FRANTZ'S TUMOR LOCATED IN THE HEAD OF THE PANCREAS: ENUCLEATION AND ITS COMPLICATIONS - CASE REPORT

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Abstract: Frantz's Tumor is a pre-malignant papillary solid-cystic neoplasm that predominantly affects young women, with a preferential location in the pancreatic body and tail, with a clinical course ranging from asymptomatic to progressing to cholestatic syndrome, depending on the location and effects. local compressions. The treatment is surgical, and the location and degree of extension with other structures are decisive factors for surgical planning. Pancreatic body-tail lesions can be addressed with corpocaudal (distal) pancreatectomy with or without splenectomy, and pancreatic head lesions can be addressed through pancreatoduodenectomy. A more conservative approach such as enucleation may also be an option, but the complications inherent to this strategy are also likely to occur. **Clinical Case:** Female patient, 23 years old, admitted to the Federal Hospital of Andaraí, Rio de Janeiro, diagnosed with Frantz Tumor in the head of the pancreas, underwent enucleation of the tumor, evolving with pancreatic fistula, intra-abdominal hemorrhage, being prolonged intensive support, multiple surgical approaches, and finally submitted to arteriography with embolization of a pseudoaneurysm of a gastroepiploic branch, with stabilization of the clinical picture. The objective is to show the existing outcomes also in the most conservative surgical approach, its management and resolution of these conditions.

Keywords: frantz tumor, pseudopapillary solid neoplasm, pancreatic fistula, gastroepiploic microbranch pseudoaneurysm.

INTRODUCTION

Papillary solid-cystic tumor (TSCP) of the pancreas is a rare neoplasm. It was first observed in 1927 in a 19-year-old woman. However, it was only described in 1959 by Frantz, who reported four cases that had been misdiagnosed as non-functioning islet cell tumors, and described this tumor as a new entity, naming it "papillary tumor of the pancreas". (1). It is a rare, idiopathic epithelial tumor with low malignant potential. (2,3). This tumor accounts for only 5% of cystic pancreatic tumors and 1 to 2% of all exocrine tumors of this organ. (6,7,8,9). In the recent past, there has been an increase in its frequency, partly due to greater knowledge of its existence and greater identification by immunohistochemical methods. (3). It usually affects young women (2-4). The symptomatology of this neoplasm has little specific characteristics. Often, the finding is incidental to physical examination or imaging studies. (2-5).

CASE REPORT

Patient A.M.S., female, 23 years old, sought surgery at the Hospital Federal do Andaraí in Rio de Janeiro due to a lesion in the head of the pancreas that was incidentally found on USG of the total abdomen in April 2022, after a car accident, and continued with the investigation clinic with T1, T2, T2 FS, diffusion and T1 FS Nuclear Magnetic Resonance, with venous contrast on 04/25/2022 (figures 1). The report describes an expansive, heterogeneous lesion, inseparable from the uncinate process and head of the pancreas, in the topography of the pancreaticoduodenal sulcus, with intervening cystic areas and extensive solid hypovascular component, measuring about 6.6x6.4cm, which did not determine invasion of vessels or surrounding structures, with the possibility of a solid pseudopapillary tumor being highly suspect; remainder of the pancreas with the same thickness and signal; Wirsung's duct

of normal caliber. She was asymptomatic, with no changes on physical examination, and no report of previous comorbidities. Hospitalization was chosen, and surgical procedure was performed on 07/27/2022 (enucleation of the pancreatic head tumor), with no intraoperative evidence of lymph node enlargement, ascites, peritoneal carcinomatosis or distant disease. The patient evolved in good general condition, asymptomatic, when on postoperative day 5, epigastralgia, nausea, hematemesis, elevation of pancreatic enzymes - amylase 271U/L, lipase 365U/L, CRP 28.46mg/dL (01/08/2022)), in addition to a change in the appearance and output of the Blake drain to a cloudy appearance of 550ml, with drain fluid (01/08/2022) showing amylase 36,355U/L, lipase 94,592U/L, configuring a pancreatic fistula. We proceeded with Upper Digestive Endoscopy (UGE) + passage of Nasoenteral Probe (SNE) on 01/08/2022, with conclusion of normal endoscopic examination. The patient maintained complaints of nausea and epigastric pain on D6, and a CT scan of the entire abdomen and chest with venous contrast was requested on 08/02/2022 (figure 2), showing bilateral pleural effusion, passive pulmonary atelectasis, greater on the left, mild ascites, gas foci and liquid occupying the surgical resection bed in the pancreatic pocket, NET entangled in the stomach; peritoneal drainage catheter with end between the stomach and spleen and adjacent subcutaneous fat thickening in the abdominal wall; normally distended gallbladder; absence of biliary dilatation; no observed mass with heterogeneous impregnation demonstrated on previous examination in head of pancreas; absence of lymph node enlargement. On D12, tachycardia and fever started, with a change in the appearance of the drain to hematic. there was evidence of diffuse parietal thickening of the gastric wall; slight increase in the

amount of fluid in the abdominal cavity; CRP 21.98mg/dL amylase 48U/L, lipase 149U/L, total leukocytes 25,680/mm³, rods 13/mm³, hemoglobin 7.3g/dL, hematocrit 23.4%; an empirical antibiotic regimen with piperacillin+tazobactam was started. The patient evolved on D13 with a change in the appearance of the drain to cloudy, maintaining a pancreatic fistula according to the biochemistry of the drain.

On D14, there was a lowering of the level of consciousness, signs of hemodynamic instability, a change in the appearance of the drain to hematic, and the patient was referred to the ICU, performed blood transfusion and intensive support. On D15, a surgical approach was chosen, exploratory laparotomy, showing unblocked pancreatic fistula, abundant amount of free liquid in the cavity (blood, clot, bile), bleeding of probable erosion of A.gastroduodenal; opening of about 1.5 cm in the second duodenal portion, presence of a candle flame lesion in the transverse colon; performing suture of the duodenum, section of the stomach at the level of the pylorus (Voughan) and performance of gastroenteroanastomosis; performed cholecystectomy with catheterization of the cystic duct with delaton for transcystic drainage of the bile duct, in order to reduce the debt of the duodenal fistula; and drainage of the cavity with two Blake drains (right positioned in a bloody area and left positioned subhepatically in an area of transcystic drainage); The patient evolved with stability of hemodynamic parameters until D19, maintaining leukocytosis at a plateau of 30,000 to 40,000/mm³, but with the onset of febrile episodes, with the decision to change the antibiotic regimen to Ertapenem, Tigecycline and Fluconazole (D0 08/15/2022). On D21, he maintained an active pancreatic fistula, drain fluid being amylase 32,180U/L, lipase 399,742U/L. On D22,

she developed hemorrhagic shock, needing emergency blood transfusion, and underwent exploratory laparotomy for damage control, showing the presence of a large amount of ascitic secretion, with clots in all quadrants, diffuse blood content, without active bleeding focus; perforation was identified in a previously ruffled duodenum, with output of hematicobilious secretion; normopositioned transcystic drain; duodenostomy was performed with Foley 20fr, and it was decided to keep the patient in peritoniosotomy for a second look. On D26, she was re-approached for revision of the peritoniosotomy, showing the presence of biliohematic secretion in the pancreatic pocket and seropurulent in a small volume, with few clots, without active bleeding; normopositioned transcystic drain; duodenostomy in a normal position, choosing to replace the blakes, with the right positioned in the pancreatic and subhepatic pocket and the left positioned in the pelvis, and proceeded with closure of the peritoniosotomy and follow-up in the ICU. On D29, it evolved again with hemorrhagic shock and change of appearance from blake to hematic drains, being re-approached - exploratory laparotomy, showing the presence of a moderate amount of biliohematic fluid with multiple clots throughout the cavity, with no focus of active bleeding; identified foley of the displaced duodenostomy, with opening in the duodenum and output of biliohematic secretion from the lumen; normopositioned transcystic drain; right blake drains positioned in the pancreatic pocket, and left blake in the pelvis, positioned normally; re-confection of the duodenostomy with 20Fr foley, tobacco purse suture and omentum patch over the duodenostomy; opted to keep in peritoniosotomy. Subsequently, surgical revision of the peritoniosotomy was performed on D33, D36, D40, D43, evolving on D50 with a drop in hematimetric indices, hemodynamic instability, being referred to

the National Cancer Institute (INCA) to the radiointervention sector on D51. During the procedure, arteriography of the celiac trunk demonstrates the gastroduodenal artery with reversed flow; arteriography of the superior mesenteric artery demonstrates a pseudoaneurysm originating from the gastroepiploic branch, and embolization of this branch and the pseudoaneurysm was performed with histoacryl (figure 3). Control arteriography did not demonstrate opacification or pseudoaneurysm. Procedure without complications. The patient evolved with hemodynamic stability after the procedure, without recurrence of the bleeding events, opting in D58 for closure of the peritoniosotomy with separation of components and placement of marlex onlay mesh. On D65, she was referred to the ward, with optimization of nutritional support, respiratory and motor physiotherapy, where she remained until 10/31/2022, when she was discharged. The histopathology of the surgical specimen macroscopically shows a segment of the pancreas measuring 6.0x6.0x4.0cm and showing a yellowish and lobulated surface. The sections reveal an encapsulated, firm, white nodule, measuring 3.0x3.0x1.1cm and tangent to the radial surgical limit. The remainder of the parenchyma is yellowish and lobulated. Conclusion: solid pseudopapillary tumor of the pancreas. The lesion measures 30x30mm and is partially encapsulated. There are two foci where it microscopically invades the pancreatic parenchyma. Vascular and perineural invasion not detected. Surgical limit free of neoplasm, however there is a focus where the lesion is less than 1mm away from the limit. Pathological classification pT2. The patient is being followed up regularly, on an outpatient basis, by the General Surgery team.

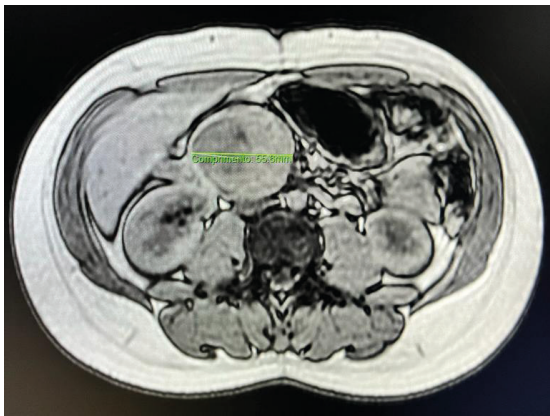


Figure 1: Abdominal MRI showing Frantz's tumor in the head of the pancreas, measuring 55.6mm in its longest axis.

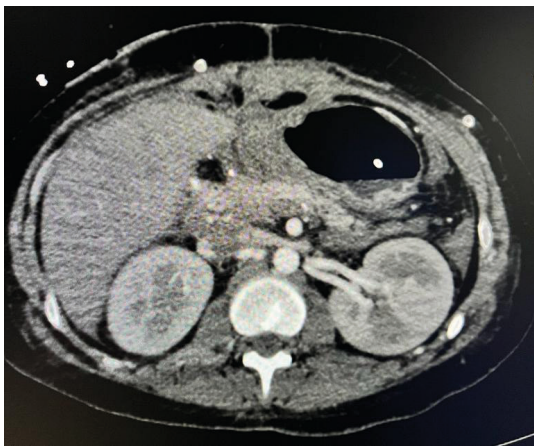


Figure 2: Full abdomen CT, arterial phase, post-enucleation of Frantz's tumor in the head of the pancreas.



Figure 3: Arteriography showing gastroepiploic microbranch pseudoaneurysm.

DISCUSSION

The solid pseudopapillary tumor of the pancreas was first described in 1959 by the pathologist Virginia Kneeland Frantz ⁽⁴⁾ and reclassified in 1996 by the World Health Organization ⁽¹⁰⁾. The etiopathogenesis of the disease is still unknown. Many authors agree on the theory that the tumor originates from epithelial cells. It is postulated that the tumor originates from primitive acinar pancreatic cells by the presence of a well-developed Golgi complex and the presence of alpha-1-antitrypsin, or from female genital tubercle cells. ^(11,12). The occurrence of this tumor in young women with the presence of progesterone receptors may indicate relationships between tumor development and female hormones. ⁽¹³⁾. These tumors typically occur in women (10:1 ratio) between the second and fourth decade of life and about 20 to 25% of cases are seen in the pediatric age group. ^(14,11). Frantz's tumor is located more typically in the body and tail of the pancreas, and may also reach the head ⁽¹⁵⁾. The clinical presentation of the tumor is nonspecific. Generally, women present with abdominal discomfort, moderate abdominal pain, and signs of tumor-induced compression. In some patients, the tumor is detected incidentally on physical examination or imaging studies ^(14,11). Tumor growth is indolent and although it extends beyond the pancreatic limits, it rarely invades adjacent organs or vascular structures. Thus, surgical resection is feasible and the first treatment option. Adjuvant therapies such as chemotherapy and radiotherapy are not recommended for this type of tumor. ⁽¹⁶⁾. Survival after surgical resection in 5 years is around 97% and death directly associated with the disease is low. ⁽¹¹⁾. Despite recent advances in pancreatic surgery, fistulas are common and remain a major post-pancreatectomy complication. ⁽¹⁷⁾, also being feasible in other surgical approaches to the pancreas. In the

present case, due to the peripheral location in the pancreatic head and the characteristics of the lesion compatible with Frantz's tumor, we opted for a smaller procedure, the enucleation of the lesion. However, the patient had grade C pancreatic fistula, followed by duodenal fistula, and erosion of vascular structures, with severe hemorrhage and need for radiointervention approach for angioembolization of pseudoaneurysm. According to the IGSPS (International Study Group of Pancreatic Fistula), the grading of pancreatic fistulas occurs in three levels: Grade A, defined as biochemical extravasation, with amylase three times higher than the serum level, on the third PO day. These fistulas do not alter the patient's prognosis and allow expectant management. Grade B, in which there are systemic alterations related to the fistula and/or the need to reposition the abdominal drain using non-invasive methods. Or grade C, with fistula-related organ failure and/or surgical re-approach⁽¹⁸⁻¹⁹⁾. Up to 30% of patients undergoing pancreatectomy may develop fistulas, increasing morbidity, length of stay and complexity of hospital medical care (20). Another prevalent postoperative complication is hemorrhage, also presented by the patient in this case. According to the IGSPS definitions, these post-surgical hemorrhages must be categorized in order to better monitor the evolution of patients. With regard to time, bleeding can be early (before 24 hours after surgery) or late (after 24 hours after surgery). Early bleeding is associated with failure in hemostasis during the surgical procedure. Late ones are related to endothelial injury caused by fistulas or abscesses, as in the case in question. As for location, these bleeds can be intraluminal, within the digestive tract, or extraluminal, into the abdominal cavity. Severity is defined as mild, if the amount of volume lost is low and there are no clinical repercussions, need for intervention or drop

in hemoglobin (Hb) greater than 3 g/dL. On the other hand, severe bleeding will manifest itself clinically after a large volume loss, Hb drop greater than 3 g/dL and require intensive treatment. Based on these characteristics, the IGSPS grades hemorrhages into A, B and C. "A" hemorrhages are early and mild, without clinical repercussions or therapeutic approaches. "B" hemorrhages, early and severe or late and mild, require diagnostic investigation with CT, angiography or endoscopy, in addition to surgical, endoscopic or embolization treatment. Finally, late and severe "C" grades require diagnostic and therapeutic evaluation like grade "B" grades and pose a threat to life.⁽²¹⁾.

CONCLUSION

Frantz's tumor presents indolent growth, benign evolution and surgical treatment in the vast majority of cases. This case addresses complications that are difficult to manage clinically, such as pancreatic fistula and hemorrhage episodes, which are conditions that can occur even in more conservative and smaller approaches, such as enucleation of the pancreatic tumor. It is worth emphasizing the importance of radiointervention, being in this case the main measure for hemodynamic stabilization, through angioembolization of the described pseudoaneurysm, which contributed to the positive outcome of the case. Although several surgical strategies have been incorporated in recent years to manage pancreatic tumors, the present case calls into question the more conservative approach, since the outcome regarding complications such as pancreatic fistula and hemorrhage are also present in this approach. The subject remains controversial, requiring constant updating, especially in relation to serious complications such as those described.

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