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FRANTZ'S TUMOR WITH ATYPICAL PRESENTATION: CASE REPORT

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Abstract: Objective: to report the case of a patient with Frantz's tumor, whose initial clinical presentation is very different from what is already reported in the literature, after all, he presented with clear signs of sepsis and necrohemorrhagic pancreatitis. Method: information was obtained by reviewing the medical records, interviewing the patient and companion, photographing the diagnostic methods to which the patient was submitted, and reviewing the literature. Conclusions: the reported case, editions and presentations made bring to light the discussion of the therapy of a rare and complex situation that is Frantz's tumor and demonstrate that, despite the rarity of the cases and the fact that it has a variable presentation, when conducts are well performed based on clinical and laboratory parameters, the surgical decision (exploratory laparotomy) is capable of obtaining satisfactory diagnostic and therapeutic results.
Keywords: Pain, pancreatitis, tumor, Frantz, laparotomy.

INTRODUCTION

Frantz's tumor, or also called solid pseudopapillary epithelial neoplasms (SPEN), is a rare pancreatic exocrine neoplasm with predisposition for age and gender. Theories say that these neoplasms develop from pluripotent pancreatic stem cells, or that it is from cells of female genital epithelial origin due to the increased prevalence in younger women.[13] Usually with nonspecific clinical manifestations with a favorable prognosis and, more frequently, it affects the pancreatic tail.[16]

SPEN are extremely rare pancreatic tumors, accounting for 1% to 2% of all exocrine pancreatic lesions.[10] It happens almost exclusively in the pancreas and in young women. Although their malignancy potential is low – 7 to 9% – these tumors tend to mimic other diseases and require meticulous investigation.[4] Most of these tumors present necrosis and hemorrhage, with cystic formation of lesions composed of solid portions of coarse appearance. In general, it is considered a slow-growing and non-aggressive tumor.[6] The standard treatment is total surgical resection, which has good results with a favorable long-term prognosis.[2,7]

With this case report we aim to show a rare case, with unusual clinical and laboratory manifestations. Male, 25-year-old patient with clinical presentation of necrohemorrhagic pancreatitis, whose diagnosis was Frantz's tumor located in the head of the pancreas, which was successfully treated and with complete outpatient follow-up until discharge from the hospital Guarujá-SP, Brazil.

CASE REPORT

R. T. S. N, male, brown, born and raised in the city of Guarujá-SP. Previous history of gastritis without medication or controls for approximately 3 years. No surgeries or previous pathologies.

Patient was seen at Hospital Guarujá for the first time in June 2020, whose reason for consultation was mild discomfort in the epigastric region. At that time he received analgesia without carrying out further studies.

In September of that same year, after repeated episodes of colic-like abdominal pain that subsided with simple oral analgesics (without medical prescription), the patient went to the emergency room at another service. At that moment, he presented cramping pain in the periumbilical region that had evolved a few hours ago, intensity 10/10. Ultrasonography and abdominal tomography without contrast were requested, and the patient was referred to the clinical gastroenterology service on an outpatient basis.

One month later, the patient was seen by a gastroenterologist who indicated upper digestive videoendoscopy and echoendoscopy.

Approximately 4 days after consultation with the specialist, the patient performs the studies requested by the specialist. Hours after the examination, the patient was admitted to Hospital Guarujá with excruciating abdominal pain in the periumbilical and epigastric region of 10/10 intensity. In addition to pain, there was also vomiting whose content was of gastrointestinal origin. At the emergency service, she received intravenous analgesic medication and hospitalization was chosen and laboratory and imaging studies were requested. The family member presented the result of the echoendoscopy previously performed to the medical service: mild enanthematous pangastritis, extrinsic compression in the antrum and second duodenal portion, solid peri-pancreatic lesion-biopsies.

The patient's initial clinical picture consisted of the following vital signs: Systemic Blood Pressure 80x50mmHg Heart rate 130bpm Temperature 38oC Respiratory rate 22irpm. On physical examination: Sweating, dehydration, mucocutaneous pallor added to laboratory parameters: leukocyte count 32,000/mm3 (RV 4,000-10,000/mm3); CRP 100.10 mg/L (RV<5.00 mg/L); Amylase: 1782 U/L (VR 25 to 125U/L); Lipase 1642U/L (VR 13 to 60 U/L). Which at that moment indicated a picture of acute pancreatitis. After hospitalization, an evaluation of the general surgery was requested, whose team followed the case for three days, and in view of the clinical and laboratory worsening with a very unstable clinical picture due to septic shock with the use of vasoactive drugs, an exploratory laparotomy was indicated.

For the patient's surgical act under general anesthesia, the surgery started with a subcostal incision enlarged by planes, where free liquid was found in the abdominal cavity in a moderate amount. Presence of an extensive tumor mass measuring approximately 15 cm in the head of the pancreas, with an important compression mass effect on the duodenal arch. Necrotic liquid was found in the retrocavity of the epiplons, pus, intracavitary abscess with necrosis of the duodenum and parts of the pancreas due to compression of the splenic artery and some of its subsequent branches, denoting necrotizing pancreatitis. A gastroduodenopancreatectomy was performed due to the unfeasibility of the cephalic portion of the pancreas and the duodenal arch, and the piece was sent for analysis (Photo 1).

The result of the anatomopathological examination of the surgical specimen, reported by Dr. Ângelo Sementilli, from the Pathological Anatomy Service of Santos, indicated pancreatic tissue with necrosis, vascular thrombosis and hemorrhage (Photo 4), characterizing necrohemorrhagic pancreatitis. Tumor nodule showing solid areas with uniform cells, partially cystic central region and formation of some tubular structures (Photos 2 and 3). Immunohistochemistry with expression for progesterone, synaptophysin and vimentin receptors (Photos 6, 7 and 8).

After surgery, the patient remains in the Intensive Care Unit (ICU) improving for a few days. After this period, he developed hemodynamic destabilization and was monitored daily, with elevated CRP and rising leukocytosis, fever, tachycardia.

In the postoperative control tomography, an abscess was observed in the left hypochondrium, retrosplenic, requiring a new surgical approach (10/16/2020), laparotomy to drain the abscess. Under general anesthesia, a left subcostal incision was made with opening planes, findings of purulent secretion were found in the

retrogastric space and in the upper splenic pole without other evident alterations. Washing was carried out with warm saline solution and drainage in the cul-de-sac/ retrosplenic and retrogastric with a laminar tube and penrose.

The patient was hospitalized in a clinical ward for a few more days. His condition was quite stable and evolved favorably until he was discharged, with recommendations for outpatient follow-up.

Outpatient follow-up by the oncology department continued for 6 months, to assess whether there was metastasis or other complications; in addition to follow-up with a general surgery service. A liver mass was evidenced by tomography, which was biopsied guided by tomography. Biopsy result: absence of tumor representation in the histopathological sections examined.

In April 2021, the patient presented an incisional hernia - which was corrected with a new surgery, herniorrhaphy. No complications.

In May 2021, portal thrombosis was diagnosed due to clinical and laboratory parameters, based on which some imaging tests were indicated, such as contrast-enhanced abdominal tomography and magnetic resonance imaging, the result of which confirmed the diagnosis of thrombosis of segments V and VIII, partially recanalized and associated with Perfusional hepatic alterations - according to the Angio-MRI report with contrast. This pathology was clinically treated with Xarelto for a few months. After this period, a cure confirmed by control Angio-MR in the month of August 2021 was observed: complete regression of thrombosis affecting portal venous branches present in the previous examination.

One year after the surgery, after complete post-surgical follow-up with the oncology and surgery team, the patient was discharged and considered completely cured and without functional sequelae. A very rare case with unique presentations that completely deviate from the reports found in the literature, after all, a male patient, with an initial clinical presentation of acute necrohemorrhagic pancreatitis accompanied by septic shock on admission and a tumor located in the head of the pancreas.

DISCUSSION

This neoplasm was first described by Dr. Virginia Kneeland Frantz in 1959, which gave rise to its nomenclature. Since then, it has received several names, such as solid pseudopapillary tumor (SPT) of the pancreas, solid pseudopapillary neoplasm (SPN), cystic papillary neoplasm of the pancreas, Hamoudi tumor or Gruber-Frantz tumor.[15] Its etiology is still not fully known. It is a rare tumor, occurring in approximately 0.17%-2.7% of non-endocrine tumors of the pancreas.[21] There are currently about 452 reported cases of this tumor in the English literature.[9]

The clinical presentation is variable, depending on the time of evolution and the size of the tumor. In the asymptomatic form, it is usually an incidentaloma in search of other pathologies. Symptomatic cases may range from compressive symptoms due to the large volume, such as the abdominal pain, fullness with a palpable abdominal mass.[4] Laboratory tests are usually non-specific and these are usually normal - both biochemical and tumor markers.[1] In addition, normally not associated with other diseases, nor with neoplasms or paraneoplastic syndromes of hormone secretion.[4] In association with clinical and laboratory tests, tomography helps in the diagnosis.

Its origin is controversial. Several studies have reported evidence, including immunohistochemical evidence, of a possible ductal, acinar or neural origin

for this tumor. Due to the inconclusive findings, the hypothesis that it originated from a multipotent primitive pancreatic epithelial cell line, even though there is no conclusive information. Among some studies and research, the characteristic immunophenotype pattern for Frantz's tumor was identified: expression of vimentin, alpha-1 anti-trypsin, alpha-1 anti-chymotrypsin and neurospecific enolase.[8, 11] These findings confirm the opinion of most authors who agree with the theory that the tumor precursor cell comes from a primitive epithelial cell.[14, 21]

Usually, at the time of diagnosis, the tumors are already large. Despite this, invasion of vascular or biliary structures is rare, which makes resection possible in most patients. Note that the tumor preferentially affects the pancreatic body and tail.[11, 17, 18]

Surgical resection of the tumor results in virtually total survival (>95%) for those patients with tumors restricted to the pancreas at presentation.[19] Therefore, in most cases, the only sufficient treatment. The type of resection depends on the topography of the tumor and must aim to preserve adjacent structures. Duodenopancreatectomies with pylorus preservation and corpocaudal pancreatectomies with spleen preservation must be performed.[3]

In selected cases, enucleation is also a viable alternative, which was performed in one of the cases in the present series, without morbidity and with free surgical margins. Resectability rates are high because the tumor, when growing, displaces adjacent structures instead of invading them.[21] Most studies agree that wide resections or lymphadenectomies are not indicated.[18] Metastases must be resected, even when associated with tumor recurrence, which is more common in the elderly population[5, 12]

Advanced age can be a prognostic factor for the development of metastatic disease. Although the scarcity of reports of such cases precludes a definitive analysis of this concept, one series indicated that there was no age difference between metastatic and non-metastatic tumors and, in addition, review of all metastatic cases in the literature revealed a mean age of 29 years old.[14]

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ANNEXES

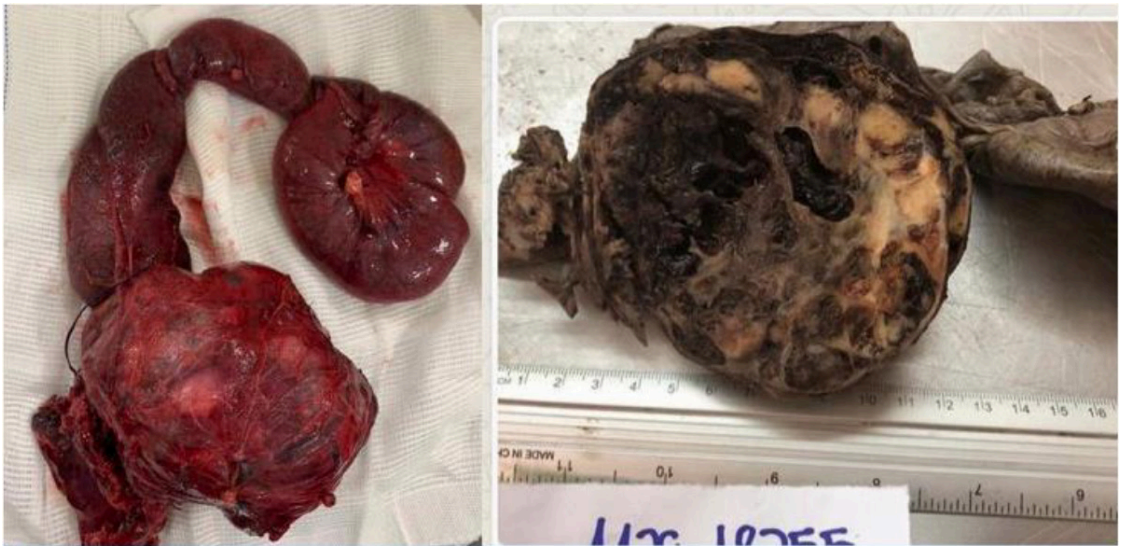

Photo 1: Surgical specimen with well-defined tumor mass between the enteric segment on the left. Pancreas fragment on the right.

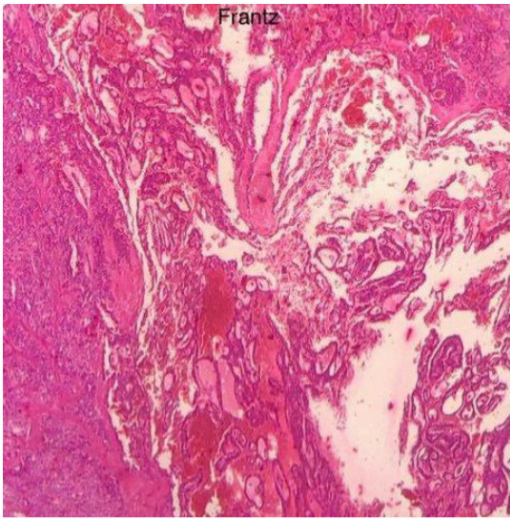

Photo 2: Histology of the tumor nodule showing a solid area at left and partially cystic central region.

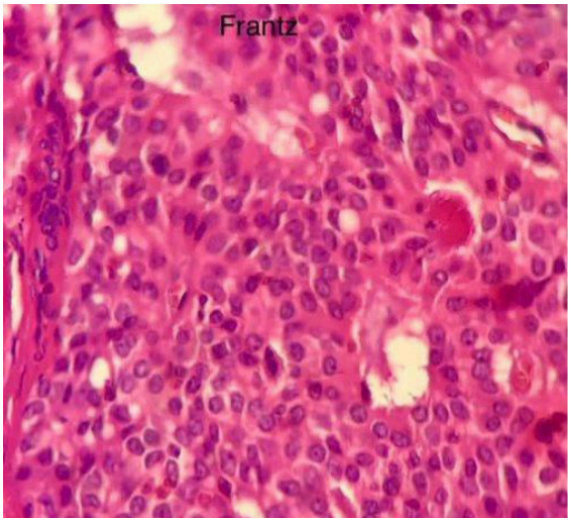

Photo 3: Solid tumor area with uniform cells and forming some tubular structures.

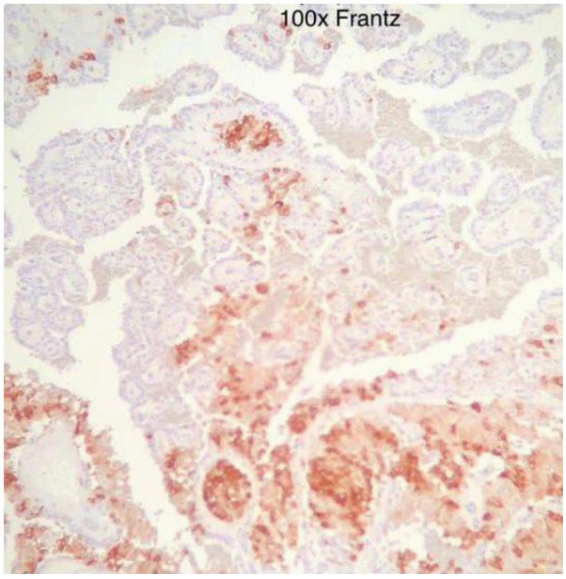

Photo 7: Expression for Synaptophysin in areas of neuroendocrine pattern.

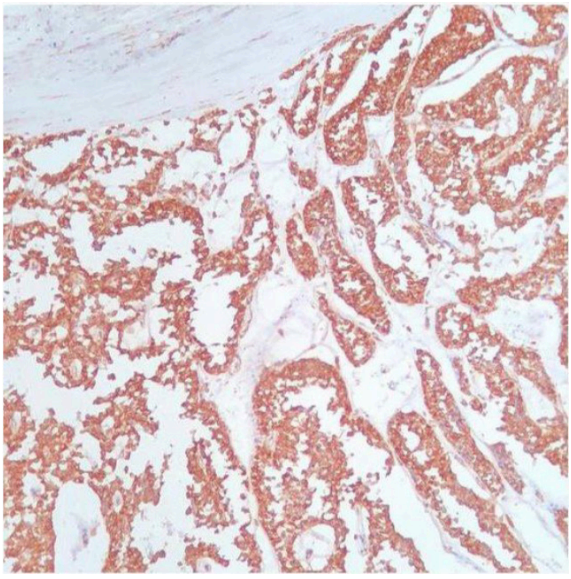

Photo 8: High expression for vimentin in tumor cells.