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Case Report

Solid-pseudopapillary tumor of the pancreas (Frantz's tumor), a rare entity, regarding a case

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ABSTRACT

Frantz tumor is a rare solid pseudopapillary neoplasm of the pancreas. Represents 1-2% of all the exocrine neoplasm of the pancreas. Was described for the first time in 1959. Occurs insidiously in female, young patients. The diagnosis is made casually through images, it should be suspected in a tomography as the first differential diagnosis in the presence of a large tumor, located in the head and/or tail of the pancreas with defined contours with solid and cystic portions, and rare presence of metastasis. The treatment is surgical resection of the tumor with a very good prognosis, with a high survival rate and low recurrence. A case is reported of a 31-year-old female patient, with no significant history, presenting with abdominal pain of three months duration and subsequent early satiety, without other symptoms. Clinically with the presence of a palpable tumor in the right hypochondrium, without signs of peritoneal irritation, laboratory without alterations, imaging studies with presence of tumor in the head of the pancreas. A pancreaticoduodenectomy is done, with findings and definitive histopathological report of solid pseudopapillary neoplasia delimited to the pancreas, with good postoperative evolution. Frantz tumor is a rare entity in which the majority of solid papillary neoplasms behave in a benign manner, with complete surgical resection being the mainstay of curative treatment, which leads to better results that increase survival and reduces local and distant recurrence

Keywords: Pancreas, Pseudopapillary, Pancreaticoduodenectomy

INTRODUCTION

Frantz tumor is a rare solid pseudopapillary neoplasm of the pancreas. It represents 1-2% of pancreatic tumors, considered within cystic tumors and exocrine pancreas, it was described for the first time in 1959 by Virgina Kneeland Frantz in the Atlas of Tumor Pathology. In 1996, the World Health Organization call it a solid pseudopapillary tumor as a distinct tumor of the exocrine pancreas.[1–4] It is characterized by being large in size (>10 cm), with a low potential for malignancy, with a predilection for the female gender, with a male-female relationship. 1:9-10, in the second to fourth decade of life, with an average at age 30.[3,5] It has been observed predominantly in young black or East Asian women.[4]

CASE REPORT

31-year-old female, with no significant history. Condition begins with the presence of crampy abdominal pain, located in the epigastrium that radiates to the scapular region of three months of evolution, early satiety is added. In physical examination abdomen with the presence of a palpable tumor in the right hypochondrium

of approximately 5 cm. non-painful, with no signs of peritoneal irritation. Laboratory tests in normal ranges. Incidentally the presence of a tumor in the pancreas was observed on abdominal ultrasound. Therefore, a pancreatic-cholangiopancreatic magnetic resonance was decided to perform with the presence of a tumor of the head of the pancreas with a thickened capsule (Figure 1). A Trucut biopsy was taken, with a histopathological report: Solid pseudopapillary neoplasia.

The patient undergoes surgical treatment, performing a pancreaticoduodenectomy (Whipple Procedure), a complete piece is removed en bloc (Figure 2 and 3), reconstruction is performed with end-to-side hepaticojejunostomy with 3-0 monocryl suture and end-to-end pancreaticojejunostomy. telescoping, in two planes, with 3-0 PDS suture and 2-0 silk, after placement of a tutor with a 5 Fr feeding tube, and gastrojejunostomy and entero-entero Roux "Y" anastomosis with 3-0 PDS suture. During the immediate postoperative period, the patient is admitted to the intensive care unit management with NPO. Supported with total parenteral nutrition for 48 hours, with subsequent discharge to hospitalization for continue monitoring and mixed nutrition (enteral-parenteral) for 7 days, with favorable evolution, with adequate tolerance of the oral diet, with no evidence of bile, pancreatic or intestinal leakage through closed drainage (Blake type), so she was satisfactorily discharged home.

Definitive histopathological report: Head of the pancreas 10.5×6.0×5.3 cm with the presence of a nodular tumor, solid pseudopapillary neoplasia, well delimited, confined to the pancreas, with a partial capsule of connective tissue measuring 6.5×5.8×5.3 cm, light brown in color, does not invade pancreatic duct or bile duct, remaining surgical margins negative (Figure 4 and 5), according to TNM, classified pT3 pN0 pM0. Currently patient with good progress in follow-up outpatient visit with no data of recurrence.

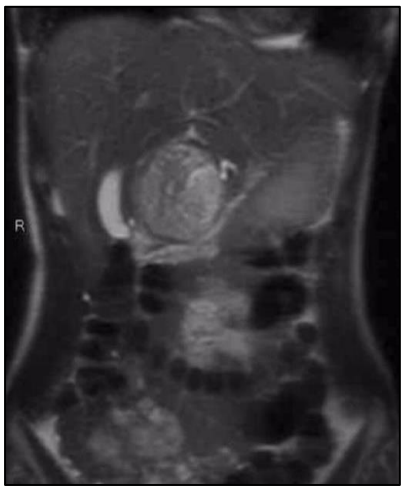

Figure 1: Magnetic resonance imaging with the presence of a tumor dependent on the head of the pancreas, which causes gastric compression and displacement.

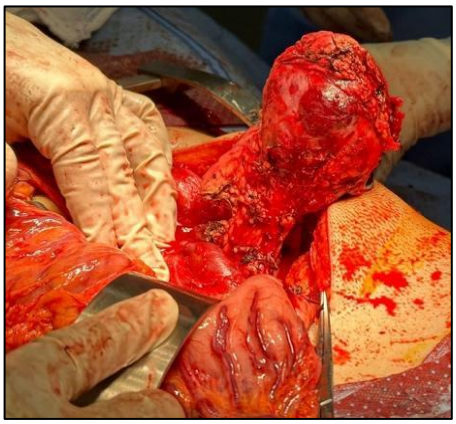

Figure 2: Tumor dependent on the head and body of the pancreas, approximately 5×5 cm, with regular edges, solid component, without involvement of the superior mesenteric artery or other structures.

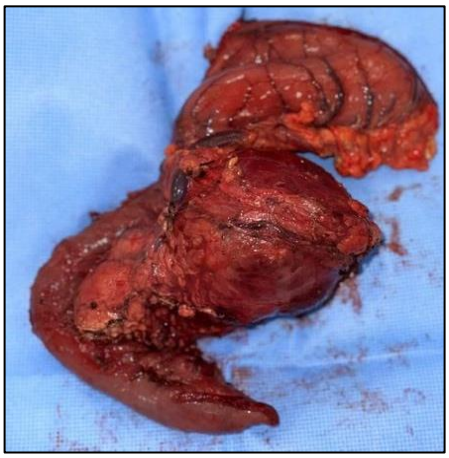

Figure 3: Product of pancreaticoduodenectomy (Frantz Tumor).

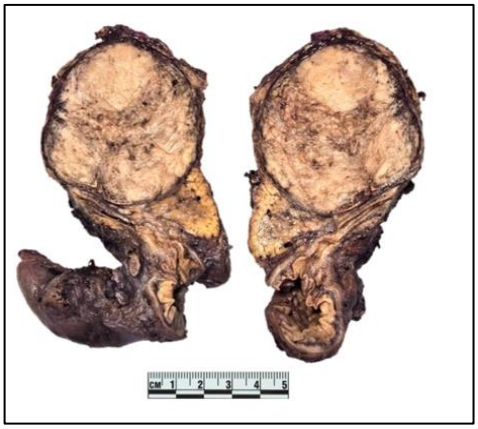

Figure 4: Macroscopic visualization of a pancreatic tumor, brown in appearance, well defined, with the presence of a capsule, confined to the head of the pancreas.

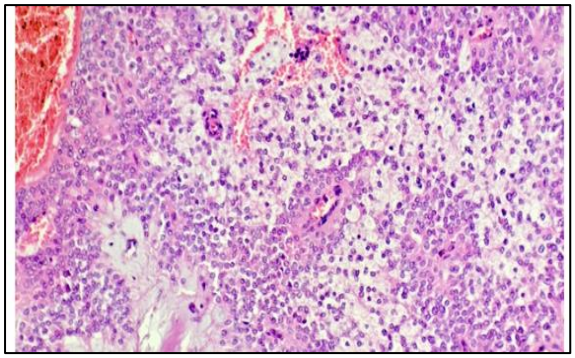

Figure 5: Histological section of pancreatic tumor located in the head, which shows: Pancreatic neoplasia with solid and pseudopapillary architecture formed by small, monomorphic cells, surrounding thin, hyalinized fibrovascular stems and rosetoid structures, with the presence of discohesiveness. The cells have eosinophilic cytoplasm, with intracytoplasmic hyaline globules, round nuclei with fine chromatin, without nucleoli or mitosis.

DISCUSSION

Frantz tumor is a rare pancreatic neoplasm, usually measuring between 6.08 and 10.5 cm. It is characterized by being a large cystic tumor, circumscribed and surrounded by a capsule with marked degenerative changes due to hemorrhage and necrosis.[4] It is located in any part of the pancreas: head and neck 34%, body 14.8-25%, tail 35.9- 40% and uncinate process 0.43%, some extrapancreatic sites described are: mesocolon, mesentery, liver, greater omentum, peritoneum, duodenum, jejunum, ovary and retroperitoneum.[2,5,6]

It has affinity for female in 95%, young people between 17-35 years old. It presents as a slow-growing abdominal tumor, with very non-specific symptoms, abdominal pain, nausea, vomiting, weight loss, dyspepsia, may occur. stomach heaviness or sensation of mass, 30-50% are asymptomatic.[4–7] Its symptoms are secondary to tumor compression of adjacent organs, jaundice is rare even in patients with lesions that compromise the head of the pancreas.[4,8] The diagnosis is made incidentally through images, it is a benign-looking tumor, it can have malignant behavior with metastasis to the liver, lung and skin (around 15%).[3,5,8,9]

The pathogenesis has not been clearly described. Its predominance in young and female patients indicates the possibility of a hormonal influence on its development, only progesterone receptors have been demonstrated in these tumors and some reports suggest the presence of beta form of estrogen receptor.[10] According to its histological pattern, there are two theories about its origin: exocrine theories due to its papillae, ducts and acini and another endocrine due to neurosecretory granules. They are slow-growing tumors, doubling their size in 2 years. Initially organized with a radial distribution of cells from fibrovascular stems, subsequently presenting papillary degenerative changes of hemorrhage and necrosis that form pseudocysts. Perineural invasion raises suspicion of possible malignancy. In immunohistochemistry, the solid papillary tumor is positive for vimentin, æ-1 antitrypsin, œ-1 antitrypsin and neuronal specific enolase.[4,6] From a genetic point of view, due to the activation of β-catenin and its white cells, but unlike pancreatic ductal adenocarcinoma, Frantz tumor is not associated with alterations in the K-ras, p53 or DPC4 genes.[10]

Its imaging characteristics are different; on ultrasonography, it is identified as a large, circumscribed tumor, well delimited, with a fibrous capsule of high echogenicity. In their internal architecture they usually have cystic and solid components.[4,7,9] Tomography findings are heterogeneous, hyperdense tumor with a defined capsule, in 10% of cases it contains fluid and in 30% capsular or peripheral calcifications are observed, with an attenuation coefficient of 20 to 50 hounsfield units due to blood products and cellular debris seen in approximately 20% in these tumors. Tomography represents the best diagnostic tool in the evaluation of cystic lesions of the pancreas, however, MRI helps in differentiation from other pancreatic tumors, provides better visualization of the tumor capsule and the extent of hemorrhagic necrosis due to its superior resolution and contrast capacity.[4] T2-weighted images show fibrous capsule, displaying a discontinuous border of low signal intensity; If gadolinium is applied, heterogeneous uptake is seen in the periphery, enhancement and progressive filling in the solid areas.[2,4]

Frantz tumor should be considered as the first differential diagnosis, when observing a large lesion in the head or tail of the pancreas with defined contours with solid and cystic portions, without internal septa with rare presence of metastases in young women.[2,4,6] These tumors, since they are not functional, present negativity to endocrine markers and neoplastic markers such as CEA and CA 19-9.[4,6] Needle aspiration cytology has been shown to be useful in preoperative evaluation but does not provide a specific diagnosis.[5]

The treatment consist in total surgical resection of the tumor, with a good prognosis with a high survival rate of up to 94-97% at five years and low recurrence, even in the case of metastasis and/or local recurrence.[10] Adjuvant therapy is unnecessary, unless there is evidence of metastatic disease.[3,7] Surgical procedures depend on its location and the size, which are enucleation, distal pancreatectomy and pancreaticoduodenectomy.[1,5–8] In case of location of the head and neck, pancreatoduodenectomy must be performed, as was the case of our patient. In tumors that involve the body or tail and have some type of vascular compromise, a distal pancreatectomy should be performed, which is preferred to be performed with splenic preservation, because it is associated with less morbidity, fewer infectious

complications, and a lower incidence of fistula. pancreatic disease and shorter hospital stay.[11] No difference in prognosis is found between the different techniques, as long as the resection is complete, which is recommended in all patients. Lymph node dissection is unnecessary due to its low rate of lymphatic dissemination.[3–6,9,10] In the case of unresectable tumors, drainage of the lesion and cystoenterostomy with adjuvant radiotherapy can be performed, since the tumor is radiosensitive.[1] Local invasion and metastases are not considered contraindications to surgical resection, and even patients with unresectable tumors can survive more than ten years after surgical cytoreduction.[7] Patients are followed every 3 months with abdominal CT or MRI during the first year, every 6 months in the second year, then annually for 5 years.[9] It has been reported that the recurrence rate after resection surgery is 3 to 9%.[7]

CONCLUSION

Frantz tumor is a rare entity in which the majority of solid papillary neoplasms behave in a benign manner, so obtaining the differential diagnosis and surgical planning, to optimize complete surgical resection, is the cornerstone of curative treatment. which leads to better results that increase survival and reduce local and distant recurrence.

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REFERENCES

1. Miguel TCL, Mercedes GLF, Azucena RSL, Margarita RGI, Andrés RC, Javier MBL. Tumor de Franz. A propósito de un caso. Rev AVFT 2020; 39(6):701-8.
2. Baltazar-Ramos JI, Martínez-Reyes G, Pérez-Corro MÁ, Denis-Rodríguez E, Melo-Santiesteban G. Solid pseudopapillary neoplasm. Report of three cases and review of the literature. Rev Med Inst Mex Seguro Soc 2023;61(2):212–9.
3. Miranda-Castillo F, Auria-Aspiazu D, Plaza-Vélez J, Villacís-Muñoz M, Benites-Ordinola C. Tumor de Frantz-Gruber, un tumor sólido quístico pseudopapilar del páncreas infrecuente. Cirujano General. 2020;42(3):223–7.
4. Tafur Anzola A, Suarez Jiménez D. Tumor de Frantz: el tumor de las mujeres jovenes. Correlacion Radiologico-patologica de dos casos en tomografia revision de la literatura. Rev Med. 2017;25(1):70–7.
5. Zumarán-Cuéllar DO, Romero-Hernández DT, Blanco-Benavides DR, Ramírez-Aceves DR. Tumor de Frantz-Gruber: reporte de un caso. Rev Gastroenterol Mex 2004;6:9.
6. Jáquez-Quintana JO, Maldonado-Garza HJ, Zubía-Nevárez CI. Gruber-Frantz tumor: a rare pancreatic neoplasm. Rev Esp Enferm Dig 2022;114(3):172-3.
7. AlQattan AS, Alshaqaq HM, Al Abdrabalnabi AA, Alnamlah M, Alanazi AA, Alqahtani MS. Huge solid pseudopapillary tumor of the pancreas "Frantz tumor": a case report. J Gastrointest Oncol 2020;11(5):1098–104.
8. Erguibi D, El Berni Y, Moufakkir A, Boufettal R, Saad RJ, Chehab F. Solid pseudopapillary neoplasm of the head of the pancreas: A case report. Annals of Medicine and Surgery 2021;69:102708.
9. Romero MDCV, Morales Cárdenas A, Cabrales Vázquez JE. Frantz tumor in a 58 year old woman; case report and literature review. Ann Med Surg (Lond). 2021;68:102625.
10. Perea Cosío RA, Norberto Rodríguez A, Menchaca Alanís LM, Sarmiento Huizar V, Mendoza García MÁ, Chapa P, et al. Tumor sólido pseudopapilar de páncreas o "'Tumor de Frantz'". Presentación de dos casos clínicos. Cirujano General. 2021;43(4):265–70.
11. Padrón Pardo OL, Salamanca Chaparro WH, González Salebe VM, Gutierrez Arias PC, Ramírez Moreno JS, Lúquez Mindiola AJ. Neoplasia sólida pseudopapilar de páncreas: una serie de cinco casos y revisión de la literatura. Rev colomb Gastroenterol 2022;37(4):466–77.

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