



Case Report

Frantz tumor in a 58 year old woman; case report and literature review

Monserrat del Carmen Valencia Romero^a, Adrián Morales Cárdenas^{b,*},
José Enrique Cabrales Vázquez^c

^a Fourth Year Resident of General Surgery, Hospital Civil Dr. Juan I. Menchaca, Mexico

^b Surgical Oncology, Regional Hospital IMSS 46, Mexico

^c General Director of the Institute Jalisciense of Cancerology, Guadalajara, Mexico

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ABSTRACT

Introduction: Frantz tumor or solid pseudopapillary neoplasm is a very rare tumor with low malignant potential, it constitutes 1–2 % of exocrine tumors of the pancreas.

Description of the case: We present the case of a 58-year-old female patient with a 4-month history of occasional abdominal pain in the epigastrium, the tomography detected a distal tumor of the pancreas. Is taken to distal pancreatectomy.

Discussion: It was first described by Franz in 1959 as a papillary tumor of the pancreas, in 2010 it was reclassified as a solid pseudopapillary tumor of low grade of malignancy. They appear in 8–16.6 % in patients before 13 years of age, in young women they present between the second and fourth decade of life. The treatment for excellence is surgical resection.

Conclusion: Pancreatic tumors represent a surgical challenge in any place of presentation.

1. Introduction

Frantz's tumor or solid pseudopapillary neoplasm is an extremely rare tumor with low malignant potential, it was first described by Dr. Virginia Kneeland Frantz in 1959 as a papillary cystic tumor of the pancreas in a 2-year-old child, in 1996 the WHO called them a solid pseudopapillary tumor, in 2010 it was reclassified as a solid pseudopapillary tumor of low grade of malignancy [1–4].

Frantz's tumor constitutes 1–2% of exocrine tumors of the pancreas, 5–10 % of cystic tumors of the pancreas, and 2–3% of all pancreatic tumors of all ages [1,5,6]. It occurs most frequently in young black or Asian women between the ages of 20 and 40, a small minority in childhood [2,7]. Most of the occasions they are detected incidentally, when there are symptoms these are usually nonspecific, such as abdominal pain or abdominal distention [7]. The diagnosis is established mainly with abdominal tomography or magnetic resonance imaging [8].

Only 15 % have a risk of being malignant neoplasms, the main site of metastasis is the liver. The treatment that provides the best results is surgery with excellent long-term control, the role of radiotherapy and chemotherapy are still under investigation, survival rate of 94–97 % [3,

6,7].

We describe the case of a 58-year-old female patient, at a rare age of presentation, based on the SCARE 2020 guidelines [9].

2. Presentation of case

We present the case of a 58-year-old female patient who comes to consultation, with a 4-month history of occasional abdominal pain in the epigastrium associated with decreased food intake as well as nausea. Father died due to peritoneal carcinomatosis, no genetic mutations detected in the family. Known with hypertension of 11 years of evolution, diabetes mellitus of 11 years of evolution, hypothyroidism of 38 years of evolution, previous surgeries; cholecystectomy at age 40, hysterectomy at age 42, allergy to dipyrone, no drug use. Physical examination without significant findings.

We performed tumor markers reporting ca 19–9 25 U/mL (0–37 U/mL), ACE 2 ng/mL (0–2.5 ng/mL). Abdominal ultrasound that reports a tumor in the retroperitoneum, abdominal tomography with intravenous contrast, which reports a tumor of the distal region of the pancreas of 8 cm (Fig. 1), in contact with splenic vessels (Fig. 2), renal hilum (Fig. 3).

After the diagnosis, she is offered to perform a distal pancreatectomy,

* Corresponding author.

E-mail addresses: dramontsevalencia@outlook.com (M.C.V. Romero), adrianmd05@gmail.com (A. Morales Cárdenas), cabrales007@gmail.com (J.E. Cabrales Vázquez).

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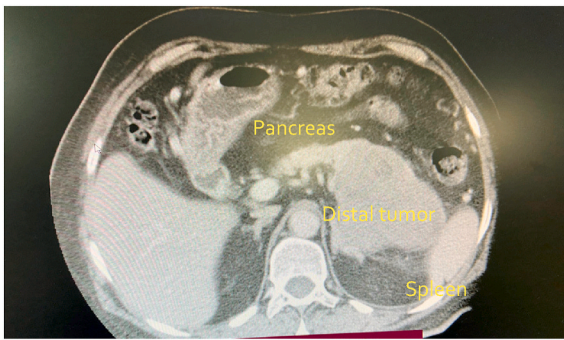


Fig. 1. Distal pancreas tumor.



Fig. 2. Splenic hilum affecting.

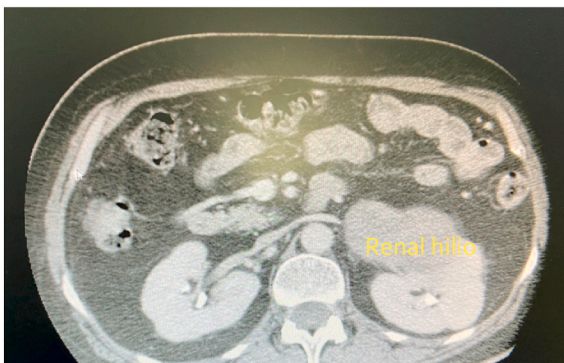


Fig. 3. Renal hilum affecting.

we comment on the risks and benefits of the procedure, the patient accepts and signs consent. The authors we carry out distal radical pancreatectomy plus splenectomy using an open approach (Fig. 4), surgical time 180 minutes, bleeding 700 mL, 2 days of hospital stay with no early complications, no pancreatic fistula formation 14 days after surgery, removal of the drain at 14 days after surgery.

Pathology report; Hematoxyllin-eosin stain, 8.5 cm × 7.9 × 7.3 cm pancreatic tumor. Malignant neoplasm of epithelial lineage composed of solid and pseudopapillary areas with polygonal cells with an irregularly ovoid nucleus, granular chromaticity with small nucleolus and some nuclear clefts (Fig. 5), scant eosinophilic cytoplasm poorly delimited, occasional mitosis (1 mitosis per 10 400X fields); Vascularized stroma with congestive areas, others hyalinized, focal necrosis, without lymphovascular permeation, the lesion is surrounded by a capsule of fibrous tissue, there is no invasion of adjacent organs, residual pancreatic tissue with reactive changes and vascular congestion, free surgical borders.

During the follow-up the patient is satisfied with the treatment, at the moment no recurrence data [9].

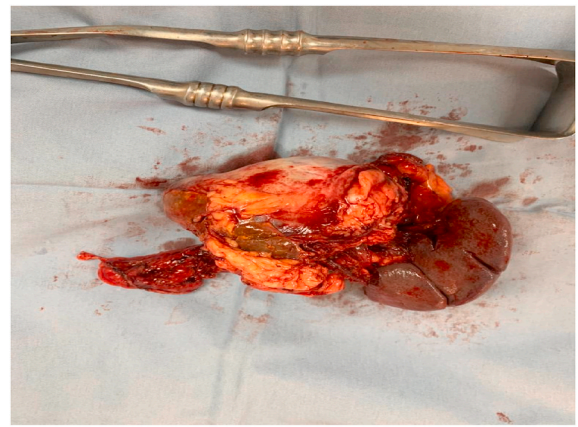


Fig. 4. Distal pancreatic resection.

3. Discussion

Solid pseudopapillary tumors of the pancreas are rare, accounting for 1%–2% of exocrine pancreatic tumors [1,2,7]. It was first described by Franz in 1959 as a papillary tumor of the pancreas, in 1966 the WHO reclassified it as a solid pseudopapillary neoplasm, in 2010 it was reclassified as a solid pseudopapillary tumor of low grade of malignancy [1,2,6].

They appear in 8–16.6% in patients before 13 years of age, in young women they present between the second and fourth decade of life [11]. It is 10 times more frequent in women than in men. The most frequent presentation is in the distal pancreas (85%) [1,7], with an average size of 9 cm, 10–15% present metastases at diagnosis [11,12]. In a study carried out at the National Institute of Neoplastic Diseases in Peru in 28 cases, the most common presentation site was in the head of the pancreas [13]. In our case, presentation was at 58 years of age, therefore it exceeded the most common presentation range [1,14].

Our case presented with early satiety and abdominal pain of 4 months of evolution as the main symptoms. According to the literature, the most common presentation is abdominal discomfort (36.8%), followed by abdominal pain, emesis and jaundice, in 12.3% of patients the tumor can be palpated. There are no specific tumor markers; in the case of the patient, there was no elevation of Ca 19-9 or carcinoembryonic antigen [1,8].

The diagnosis was established by means of an abdominal ultrasound as a suspicious study, followed by a tomography where we found a complex tumor with a well-defined capsule, adherence to the splenic vessels and renal hilum. In general, they are the most useful studies, however, magnetic resonance imaging is very useful to identify chemical and solid tumors in the pancreas, in T1 areas of hemorrhagic degeneration can be determined as well as a hypointense capsule [1,3]. Positron emission tomography in general is not very useful since it is not possible to determine whether it is a benign or malignant neoplasm [4,13,14].

In the case of our patient, based on the findings of the tomography, we decided to take the patient to surgery. It is difficult to make the decision to take a biopsy before surgery, especially in people of this age range due to the not so low risk of malignancy, as well as the possibility of dissemination, the best option for taking a biopsy by endoscopic ultrasound, without however, it remains controversial [4,14].

The differential diagnosis after 40 years is with cystadenomas, cystadenocarcinomas, intraductal papillary mucinous neoplasms, teratomas and neuroendocrine tumors [2,8]. Depending on the location, a pancreaticoduodenectomy, a left pancreatectomy, or a central pancreatectomy may be chosen [9]. Due to the tumor size greater than 8 cm, we decided to carry out open surgery, we ruled out performing the procedure by minimally invasive, tumors smaller than 5 cm, it was possible to perform surgery by minimally invasive [1,9].

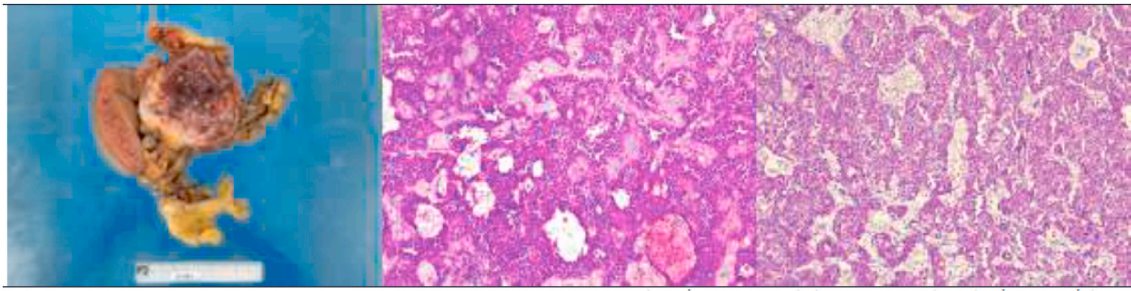


Fig. 5. Histopathological report: malignant epithelial neoplasia consisting of solid areas and pseudopapillary with polygonal cells of irregularly ovoid core cells.

The final histopathological study reported a malignant neoplasm of epithelial lineage composed of solid and pseudopapillary areas without vascular or perineural invasion of low malignant potential (1 mitosis per 10 fields). This type of tumors are of unknown etiology, an epithelial and neuroendocrine ductal origin, a primordial pluripotential cell and even extrapancreatic origin have been proposed [2].

Most are tumors with low malignant potential, the treatment of choice is surgical resection trying to be as conservative as possible, however enucleation or incomplete resection is associated with local recurrence, poor prognosis and death, the recommended margin is of 3–5 mm [7,11,13]. We perform a radical resection to achieve adequate control [7]. In patients with tumors near the splenic hilum and splenic vessels, en bloc splenectomy is most often chosen. Lymphadenectomy is generally not recommended [10,14].

The final pathology report shows free margins. Global 5-year survival is 95–97 % as long as metastatic disease is not present. 10–15 % present with metastatic disease, the liver being the most frequently associated [1,8,11]. Associated poor prognostic factors are a tumor larger than 8 cm, cellular atypia, invasion of the capsule, lymphovascular invasion, and perineural invasion [10,14].

The follow-up of the patients is carried out every 3 months with abdominal tomography or magnetic resonance imaging every 3 months in the first year, every 6 months in the second year, then every year for 5 years. In general, adjuvant treatment is not recommended unless there is evidence of metastatic disease, the most commonly used agents are gemcitabine plus cisplatin. In our case we did not consider the use of adjuvant [1,7,14].

4. Conclusions

Pancreatic tumors represent a surgical challenge in any place of presentation, timely diagnosis is vital to offer an effective treatment, in the case of Frantz tumors, surgical resection represents the best chance of cure.

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Ethical approval

No require ethical 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.

Author contribution

Adrian Morales Cardenas. Concept and design, data collection, drafting, revision and approval of final manuscript. Montserrat del Carmen Valencia Romero. Concept and design, data collection, drafting, revision and approval of final manuscript. Jose Enrique Cabrales Vazquez. Data collection, drafting, revision and approval of final manuscript.

Registration of research studies

N/A.

Guarantor

Montserrat del Carmen Valencia Romero.
Adrian Morales Cardenas.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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