

Case Report

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Solid Pseudopapillary Neoplasm of the Pancreas in Males: A Rare Case and Literature Review

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Abstract

Introduction: Solid Pseudopapillary Neoplasm of the pancreas is a malignant tumor and very rare in males.

Case presentation: In this case report we present a 23-year-old male patient, with nonspecific abdominal complaint due to a lesion with calcification in the body of the pancreas. The staging ruled out the possibility of distant illness. Surgical resection of the lesion was performed via laparoscopic distal pancreatectomy.

Conclusion: Unusual neoplasm, quite rare in males. Surgical resection enables cure and good prognosis, promoting long and disease-free survival.

Keywords: Solid pseudopapillary tumor; Frantz tumor; Pancreas neoplasms; Malignancy; Survival; Surgery treatment

Introduction

Even with the increase in recent years in the diagnosis of Solid Pseudopapillary Neoplasm of the Pancreas (SPN) [1,2] it is still considered a rare pathology and represents around 2% of pancreatic tumors [3,4]. Its prevalence is higher in females between the third and fourth decades of life, in a male ratio of 10:1 [5], which makes it even rarer among men [6]. It is described by the World Health Organization (WHO) as malignant. Its origin remains uncertain [6] and despite its indolent evolution, it has aggressive potential and can cause symptoms, invade other organs, vascular structures and can even cause distant metastases [1,7,8]. The curative treatment for SPN is surgical and the prognosis with survival > 5 years and disease-free survival depends on complete tumor resection [9]. The authors present here a case report of surgical resection of SPN in a young male patient conducted in a single center with extensive experience in pancreatic surgery.

Case report

23-year-old male patient, hypertensive, followed up at the upper digestive tract surgery service of Hospital da Cidade

in Salvador - Bahia - Brazil, for nonspecific abdominal pain. Abdominal ultrasound image of the pancreas body of irregular contour, heterogeneous, measuring 4.9 x 4.0 cm and without vascular flow. Tomographic follow-up was performed, showing a solid lesion in the pancreatic body, measuring 5 cm in diameter and the presence of peripheral calcifications (Figure 1A). Distal atrophy of the pancreatic parenchyma and chronic thrombosis of the splenic artery were also observed (Figure 1B).

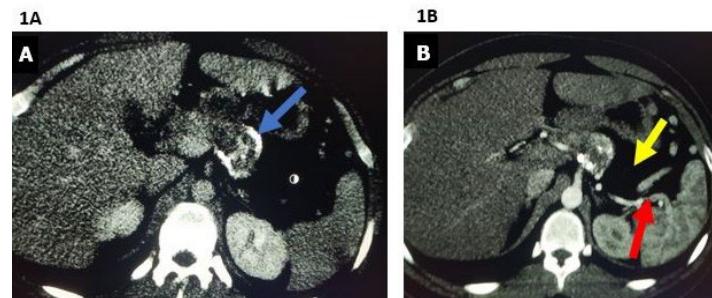


Figure 1: A) Solid lesion with peripheral calcifications, 5 cm in diameter, located in the body of the pancreas (blue arrow). B) Atrophy of the pancreatic parenchyma (yellow arrow). Chronic thrombosis of the splenic artery (red arrow).

In a multidisciplinary discussion with specialized radiology of the digestive tract, the main diagnostic suspicion was SPN, the staging was complemented with a chest tomography, which was normal. During the perioperative period, the patient was vaccinated for encapsulated germs, entered into the ERAS (Enhanced Recovery After Surgery) protocol, with emphasis on a supplemental immunomodulatory diet 7 days before surgery, use of a supplement with maltodextrin to shorten fasting 2 hours before surgery. Submitted to laparoscopic distal pancreatectomy (LDP) plus splenectomy without retroperitoneal lymphadenectomy, surgical time of 150 min, without the use of blood products, abdominal drain placed next to surgical remnant, pancreas head. Surgical piece removed by Pffanenstiel incision. Immediate postoperative period in the ward, with opioid-free analgesia. Six hours after surgery, the use of chewing gum, diet with clear liquids and walking were started. Abdominal drain amylase was collected on the 1st, 3rd and 5th postoperative days (POD) with respective results of 19,000u/l, 8,000u/l and 1,900u/l. Hospital discharge on the 5th POD without an abdominal drain, with a mild oral diet, no abdominal pain and usual bowel rhythm. There was no readmission. The pathological anatomy showed free surgical margins (Figure 2A&2B), absence of angiolympathic invasion, 4 lymph nodes free of neoplasia. Immunohistochemistry confirmed the diagnosis of Solid pseudopapillary neoplasm of the pancreas. In outpatient follow-up for 18 months, without disease recurrence and with good quality of life.

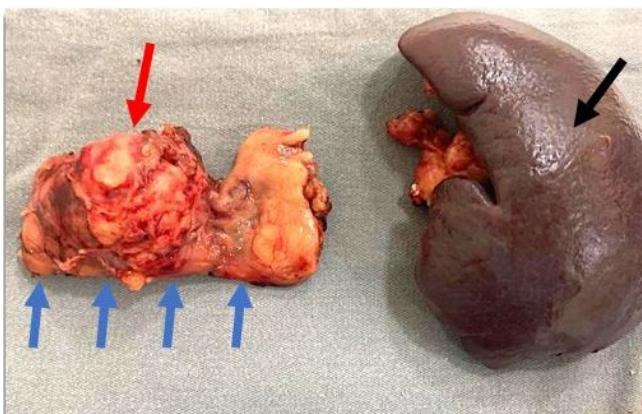


Figure 2A: Surgical parts. SPN (red arrow), body and tail of the pancreas (blue arrows) with free margins. Spleen (black arrow).



Figure 2B: Surgical piece. Posterior view showing ligature of splenic vessels with hemoclips (yellow arrow).

Discussion

Incidence and prevalence of SPN

The Solid Pseudopapillary Neoplasm of the pancreas has been described in the literature as having a rare incidence and with more than 700 well-reported cases [10]. It is believed that due to advances in radiological methods, well-established tumor nomenclature and better histopathological evaluation, more than two thirds of the cases were seen in recent years [10]. Silano F et al. [9] in a specialized center, showed an increase in SPN in the last decade (Table 1). With a sample composed exclusively of females, these authors believe that greater accessibility to specialists, such as gynecologists, with screening tests, has collaborated with incidentalomas, since 85.7% of the patients were asymptomatic [9]. The prevalence of this neoplasm is much higher in females compared to males, with a proportion in the literature review between 9 and 10:1 [5,11-21] and there are few reports in males, around 12% of all SPN [22]. β -estrogen and progesterone receptors found in SPN may be the cause of the predominance of this pathology among young women, including tumor growth in a favorable hormonal environment, such as pregnancy [23,24]. The controversy is the occurrence in men. It appears that the pancreas is an organ capable of converting androgen into estrogen through aromatase - 36 promoting an increased estrogenic microenvironment in the pancreatic tumor tissue [25]. In our pancreatic surgery service, analyzing 17 patients, only 1 (5.8%) was male, corroborating the literature and the rarity of this neoplasm among men.

Year of surgery	N	Follow-up (yr)	Disease free	Conduct
1998	1	13	Yes	MD
1998	1	1	Yes	Death ¹
2012	4	5	Yes	MD
2013	1	5	Yes	MD
2014	2	5	Yes	MD
2015	2	5	Yes	MD
2017	1	3	Yes	SG
2018	2	2	Yes	SG

Table 1: Follow-up of patients with pseudopapillary neoplasms of the pâncreas. Silano F et al. Yield Surgery in SPN. Mean Follow-up: 4.7 years; Medical discharge: 10 patients; 3 patients still under follow-up, and one died a year after surgery. ¹Patient remained free of disease and died 1 year later from pneumonia, unrelated to the pathology. CD: Conduct; SG: Follow-up; MD: Medical discharge.

Symptoms

Symptoms may be related to tumor size and location. Mass effect with compression of organ and/or neighboring structures can cause jaundice in those with pancreatic head injury and gastric symptoms in the body and tail SPN [9]. In the literature review, the presence of abdominal pain ranged between 7.1% and 87.5% and incidental findings of SPN ranged between 0% and 85.7% (Table 2). Some authors showed patients with tumors > 10 cm in the head of the pancreas and without jaundice [26]. It appears that SPN does not have a specific pattern for the development of symptoms. In this case report, the patient reported a nonspecific abdominal discomfort that did not seem to be related to SPN discovered by abdominal ultrasonography, but rather an incidentaloma.

Ref.	Year	n	Study	Incidental (%)	Ap (%)	Other findings (%)
Papavramidis et al. [11]	2006	718	R	15.5	46.5	Mass 34.8
Lubezky et al. [12]	2017	32	R	28	48	Unspecific 24
Song et al. [13]	2017	53	R	39.6	37.7	Mass 30.2
Wright et al. [8]	2019	78	R	30.7	42.3	Nauseas 14.1
Lin et al. [14]	2019	60	R	65	31.7	Strain 2.2
Torres et al. [15]	2019	16	R	-	87.5	Mass 12.5
Liu et al. [16]	2019	243	R	63.4	19	Unspecific
Farhat et al. [17]	2020	10	R	20	40	Mass 40
Silano F et al. [9]	2021	14	R	85.7	7.1	Jaundice 7.1

Table 2: Incidental and symptoms in patients with solid pseudopapillary neoplasms, literature data; R: Retrospective study; Ap: Abdominal pain.

Diagnosis

The use of Endoscopic Ultrasound – Guided Fine Needle Aspiration (EUS-FNA) biopsy is part of the arsenal cited in the literature for preoperative histological diagnosis [27]. Tumor markers such as Carbohydrate antigen 19-9 (CA19-9) and carcinoembryonic antigen (CEA) do not seem to be useful because they are usually within the normal range for SPN [28]. Referring to imaging diagnosis (tomography and/or resonance of the abdomen), this neoplasm may have the following characteristics: Solid heterogeneous cystic areas, well-defined capsule and when they are small in diameter, they can be described as a solid nodule. They usually do not cause

pancreatic duct dilation and in some cases they may present calcifications [29]. In our surgical service, we do not perform preoperative biopsy using EUS-FNA. This exam is costly and has low accessibility, in addition to being able to rupture the tumor capsule with dissemination of neoplastic cells and, in our understanding, preoperative histology would not change the surgical approach. Regarding tumor markers, in addition to being usually normal for SPN, there is no specific marker, which we do not use in our strategies for diagnosing this neoplasia. Silano F et al. [9] reported in a retrospective study with 14 patients that 92.8% had a strong radiological suspicion of SPN preoperatively after analysis of specialized radiology in the digestive tract and only 1 patient, transferred from another service, underwent EUS-FNA for diagnosis. In all 13 operated patients, pathological analysis and immunohistochemistry confirmed SPN (Figure 3). In the case report presented, the abdominal tomography (Figure 1) was discussed with specialized radiology. The lesion was 5 cm in males, a differential diagnosis of pancreatic neuroendocrine tumor was listed. However, for the case presented, both diagnoses would be surgical without the need for biopsy.

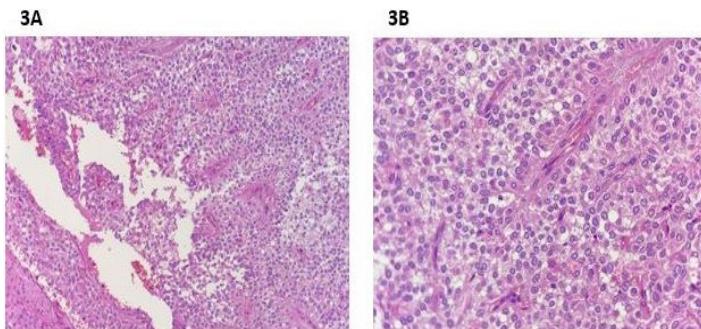


Figure 3: A) Solid monomorphic pattern, with blood vessels in between. B) Neoplastic cells with eosinophilic or clear vacuolated cytoplasms with round nuclei.

Staging

SPN metastases are not common. This neoplasm has a more indolent behavior when compared to other pancreatic tumors and tumor aggressiveness occurs in 10% - 15% of cases, being detected already in the diagnosis or in the evolution of the disease [12,21] where metastasis to the liver could occur, invasion of neighboring organs and vascular invasion [21]. Therefore, we follow the usual pattern of imaging staging for digestive tract neoplasms, chest and abdomen tomography. In our patient, the chest tomography was normal and in the abdomen there was no metastatic disease.

Treatment

Surgery is the gold standard in the treatment of SPN and in more than 95% of cases R0 resections are possible. Surgery can be curative even in scenarios with distant metastasis [16]. It is important that at the time of diagnosis, especially for incidentalomas that are

more likely to be small tumors and therefore without symptoms, surgical treatment be scheduled, avoiding neoplastic growth and the need for large resections of neighboring organs and even large vessels. In the patient reported, shortly after imaging diagnosis, he underwent Laparoscopic Distal Pancreatectomy without wider resections as the tumor remained small, 5 cm.

Follow-up

The patient reported has been under outpatient follow-up for 18 months. Karnofsky Performance Status (KPS) 100% and good quality of life. We did not use tumor markers in the follow-up. We performed imaging exams, abdominal tomography in the 1st year, and subsequently abdominal ultrasonography can be performed, depending on the clinical evolution. Patient continues without disease recurrence.

Conclusion

Solid Pseudopapillary Neoplasm of the pancreas is extremely rare in males and surgical treatment is its main pillar of cure.

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