



Solid Pseudopapilar Neoplasm of the Pancreas: Case Report and Literature Review

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ABSTRACT

Introduction: Solid pseudopapillary tumor of the pancreas is also called Frantz tumor. The World Health Organization classified it as a solid pseudopapillary neoplasm in 1996. It is more frequent in young or black Asian women, between 20 and 40 years of age, although there are isolated cases in children and men. **Case Report:** 24-year-old female. With a history of hemolytic anemia being treated by Hematology. The physical examination was unremarkable. Ultrasound revealed a tumor in the body and tail of the pancreas. In view of these findings, the radiological study was completed with a CT and MRI, having a suspected diagnosis of a pseudopapillary tumor, surgical treatment was performed with subsequent follow-up by medical oncology. **Discussion:** This type of tumor usually has nonspecific clinical manifestations, most of them caused by compression of the tumor to the

normal pancreatic parenchyma. About 15% of patients are asymptomatic. It has not been possible to clearly establish which cells are involved in their development; they seem to be derived from totipotential stem cells from the genital ridges that attach to the pancreas during embryogenesis. It has been related to mutations in chromosome 11q, which contains genetic material for the formation of proteins such as cyclin D1, FLI-1, CD56, and the progesterone receptor. Conclusions: Solid pseudopapillary neoplasm is a rare low-grade malignant entity that mainly affects young women. In most cases it is asymptomatic, so it is often diagnosed incidentally.

Keywords: Pancreas, pseudopapillary neoplasm, Diagnoses, Treatment.

INTRODUCTION

Solid pseudopapillary neoplasms are defined by the WHO Classification of Tumors of the Digestive System as low-grade malignant tumors composed of loosely cohesive epithelial cells that form solid, pseudopapillary structures and lack a specific line of pancreatic epithelial differentiation. It is an extremely rare epithelial tumor with low malignant potential. It accounts for less than 1-2% of all exocrine pancreatic tumors. It was first described in 1959, and since then it has been called by various names: papillary tumor of the pancreas or Frantz tumor, solid and cystic papillary epithelial neoplasm, or papillary cystic neoplasm. Since 1996 it has been called a solid pseudopapillary tumor of the pancreas. It is more frequent in young or black Asian women between 20 and 40 years of age (4), although there are isolated cases in less than 10% in children and men. They represent about 1%-3% of all resected cystic tumors. They are located mainly at the level of the body and tail of the pancreas, although it can also involve the head and neck. Between 1 and 1.8% have an extrapancreatic location, mainly in the retroperitoneum, omentum, mesentery, stomach, duodenum, colon, liver, adrenal gland, ovary, testis, and lung (5). As a general rule, the neoplasm presents as a solitary lesion, the multicentric manifestation is a curiosity (1-5).

The confirmatory diagnosis is made through immunohistochemical studies that mainly verify the mutation of the β -catenin protein. The immunohistochemistry panel proposed for diagnosis is positive for β -catenin and CD99 markers (dot pattern), and negative markers for chromogranin, trypsin, BCL10, and E-cadherin. Fine needle aspiration (FNA) guided by endoscopic ultrasound is useful for diagnosis, confirmed by immunohistochemistry on the cell block, which serves to exclude other pancreatic tumors (8). Endoscopic ultrasound-guided fine-needle biopsy has the advantage of providing an adequate tissue sample for greater diagnostic accuracy. The histopathological study and immunohistochemistry allow the evaluation of differential diagnoses (6).

CLINICAL CASE

A 24-year-old female with the following significant history: Hemolytic anemia diagnosed in November 2022 under treatment with prednisone 5 mg every 24 hours under hematology follow-up where she required the transfusion of packed red blood cells without presenting an adverse reaction. Surgical Instrumented uterine curettage in October 2022 with no reported complications. Other antecedents of no importance for current illness. The patient is referred from the hematology clinic during her follow-up by the service, it is reported as an incidental finding of a tumor of the body and tail of the pancreas by ultrasound performed in November 2022. Tomography is requested, and MRI: simple and contrasted CT of abdomen with a report

of a solid tumor of the pancreas in the body and tail, 84 x 81 mm, lymph nodes, hepatosplenomegaly, splenic collateral venous network. MRI (fig. 1).

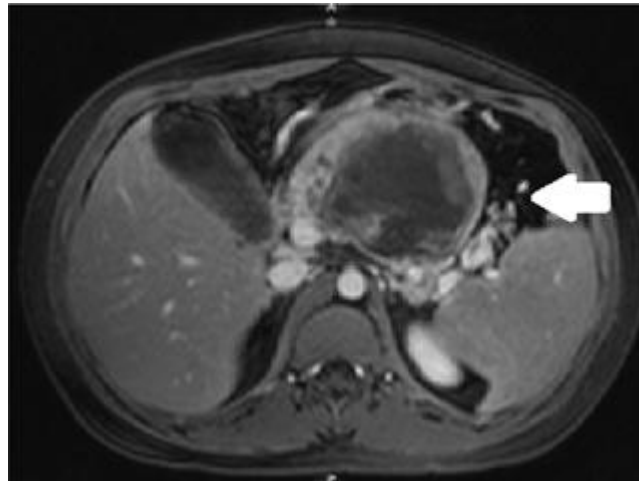


Figure 1: Solid pseudopapillary tumor of the body and tail of the pancreas, splenomegaly, and tortuous dilatation of the splenic vein.

With a report of probable pseudopapillary solid tumor of the body and tail of the pancreas, splenomegaly, tortuous dilatation of the splenic vein (Fig. 2).

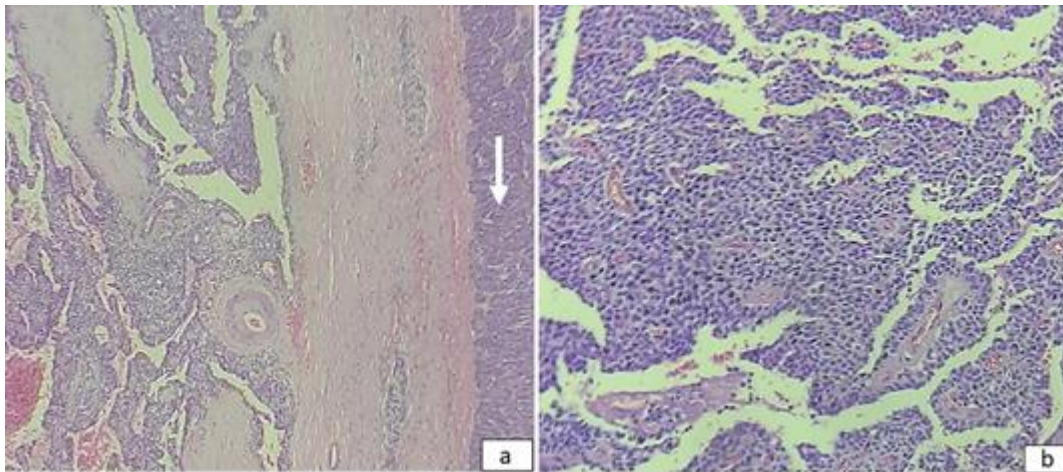
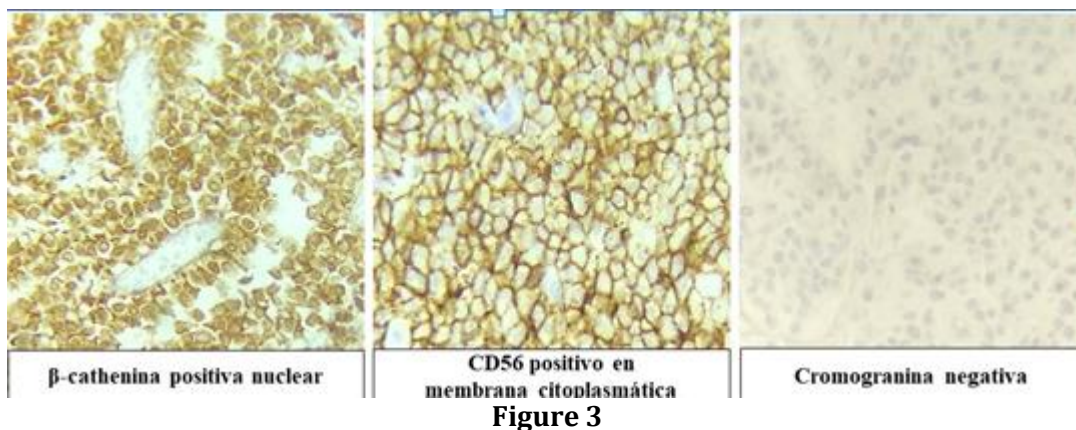


Figure 2: Solid pseudopapillary neoplasm delimited by dense fibro connective tissue, adjacent pancreatic parenchyma (arrow); b). Neoplasm with a solid and pseudopapillary growth pattern made up of monotonous cells with eosinophilic cytoplasm with a round nucleus with well-defined edges, homogeneous basophilic chromatin.

Endoscopy is requested with a study report of normal appearance, with biopsy taking without evidence of alteration data, control laboratory studies and tumor markers are requested: blood chemistry, liver function tests, serum electrolytes, blood count: within normal ranges. AFP 1.91; CA 19-9 2.5; CEA 0.97; which do not show elevation.

After completing the surgical protocol for surgical treatment with surgery performed based on distal pancreatectomy plus prophylactic splenectomy. With finding of liver without evidence of tumor activity, pancreas with a tumor in the body and tail of 12 cm in diameter, firmly adhered

to deep planes, and superior mesenteric and splenic artery, characteristics of an indurated tumor, with smooth edges, well defined, with macroscopic characteristics of mucinous appearance, lymph nodes around the celiac trunk, 1.5 cm, Spleen without data of tumor activity, 18 x 9 x 9 cm, requiring transfusion of 6PFC and 6CE together with platelet apheresis requiring management in the Intensive Care Unit of one day post-surgery, without vasopressor support or ventilatory support. After staying on the floor, he showed clinical improvement with his subsequent discharge to continue management externally, where he attended with a histopathological study report (fig. 2) which mentioned: Product of subtotal pancreatectomy, splenectomy plus omentectomy. Solid pseudopapillary neoplasm in the body of the pancreas confined to the pancreas with a tumor size of 11x10x8. 5cm, 80% necrosis; lymphatic and vascular invasion are not evident. Resection edges without neoplastic and 22 lymph nodes with follicular hyperplasia, compatible with type 4 pancreatic heterotopia in adipose tissue of the splenic hilum with a benign behavior. Positive immunohistochemical report of B catherin, positive CD56 in the cytoplasmic membrane and negative Chromogranin (Fig. 3). After evaluating the pathology and post-surgical results, the patient is sent for evaluation to receive treatment by medical oncology.



DISCUSSION

This type of tumor usually has nonspecific clinical manifestations, most of them caused by compression of the tumor to the normal pancreatic parenchyma. About 15% of patients are asymptomatic. It has not been possible to clearly establish which cells are involved in its development; they seem to derive from pluripotent stem cells from the genital ridges that join the pancreas during embryogenesis has been linked to mutations on chromosome 11q, which contains genetic material for the formation of proteins such as cyclin D1, FLI-1, CD56 and the progesterone receptor (6). Despite the fact that the solid pseudopapillary neoplasm expresses the progesterone receptor, which makes it sensitive to hormones, which could explain its prevalence in women, its role in tumorigenesis has not been proven. Additionally, there are mutations in exon 3 of the β -catenin gene, through which the Wnt signaling pathway is activated, which is enhanced by the BCL9 gene, which is present in this type of tumor, and this increases the transcriptional activity and oncogenesis (7).

Solid pseudopapillary neoplasm is usually composed of different elements, such as hemorrhagic areas and calcifications, and presents a fibrous capsule. Additionally, there is a

monomorphic, uniform cell population, of small to medium size, on a clean or hemorrhagic background, and papillary structures (8).

One of the common elements that can be seen in diagnostic images is an encapsulated lesion, with solid and cystic components, intramural hemorrhage, fibrous capsule, and eventually calcifications. A large tumor is evident at the level of the body or tail of the pancreas, with defined contours with solid and cystic portions without internal partitions. One study found that on multislice computed tomography images; generally, have smaller lesions with a greater solid component and calcifications than women (9). Grossly, sharp demarcation of pancreatic tissue is typical, facilitating surgical removal. Sometimes it may be surrounded by a delicate or thick capsule. On palpation it feels rubbery, and the cut surface has a characteristically spongy appearance. When the tumor is voluminous, extensive necrotic and hemorrhagic areas can be seen (10).

Where possible, one should always try to preserve the spleen, even when the splenic vascular bundle is compromised and must be ligated. Splenic preservation can be achieved by splenic vessel-sparing distal pancreatectomy or by the Warshaw surgical technique. This technique consists of ligating the splenic artery and vein preserving the circulation of the spleen by preserving the short gastric vessels and the left gastroepiploic vessels (11). A meta-analysis that compared the preservation of splenic vessels with the Warshaw technique showed that this technique has a significantly higher incidence of splenic infarction and gastric/epigastric varices, in addition to more patients with splenic infarction (12).

In general, it is accepted that most of the tumor follows a benign course. In large-scale studies, 2-23% has been shown to be malignant based on local recurrences or metastases. The 5-year survival is excellent, reaching 94-99%, and even the 10-year disease-specific survival rate is 96% (13). The local recurrence rate is low (less than 10% after 5 years of resection). Women and men have the same prognosis and the multicentric forms have similar clinical and pathological characteristics to the solitary forms. However, when the tumors are located in the head, the disease-free and overall survivals are significantly shorter than those of other locations. When metastases do develop, usually occur late, 8 to 15 years after complete resection of the primary tumor (14).

CONCLUSION

Solid pseudopapillary neoplasm is a rare low-grade malignant entity that mainly affects young women. In most cases it is asymptomatic, so it is often diagnosed incidentally. Clinical findings, radiological images and cytology constitute the basis of its diagnosis. Surgical resection of the tumor continues to be the mainstay of its treatment. Preferring conservative surgery with subsequent follow-up.

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