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Solid pseudopapillary tumor associated with agenesis of the dorsal pancreas: a case report

Tumor sólido pseudopapilar associado à agenesia dorsal do pâncreas: relato de caso

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ABSTRACT

Solid pseudopapillary tumors (SPTs) of the pancreas are rare tumors with low potential for malignancy, uncertain lineage, and favorable prognosis in most cases. The SPT has an excellent prognosis, and the standard treatment is surgical resection. The agenesis of the dorsal pancreas (ADP) is an extremely rare type of congenital pancreatic malformation and is characterized by partial or total loss of the body and tail of the gland. Its association with SPT has been reported only in two studies. We report a case of SPT associated with total ADP. A 36-year-old woman was diagnosed with a complex mass on pancreatic head topography, measuring 7.8×5.5cm, associated with complete agenesis of the body and tail of the pancreas. She underwent gastropancreatoduodenectomy with a successful postoperative outcome. The anatomopathological examination suggested an SPT and it was confirmed by immunohistochemistry.

Keywords: Pancreatic neoplasms; Pancreatic cancer; Pancreatectomy; Pancreas; Agenesis.

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RESUMO

Os tumores sólidos pseudopapilares (TSPs) do pâncreas são tumores raros com baixo potencial de malignidade, linhagem incerta e prognóstico favorável na maioria dos casos. O SPT tem excelente prognóstico e o tratamento padrão é a ressecção cirúrgica. A agenesia dorsal do pâncreas (ADP) é um tipo extremamente raro de malformação congênita do pâncreas e é caracterizada pela perda parcial ou total do corpo e cauda da glândula. Sua associação com TSP foi relatada apenas em dois estudos. Relatamos um caso de TSP associado à ADP total. Uma mulher de 36 anos foi diagnosticada com uma massa complexa na topografia da cabeça do pâncreas, medindo 7,8×5,5cm, associada à agenesia completa do corpo e cauda do pâncreas. Ela foi submetida à gastropancreatoduodenectomia com sucesso pós-operatório. O exame anatomopatológico sugeriu TSP e foi confirmado por imunohistoquímica.

Descritores: Neoplasias pancreáticas; Câncer pancreático; Pancreatectomia; Pâncreas; Agenesia.

INTRODUCTION

The association of a solid pseudopapillary tumor (SPT) with agenesis of the dorsal pancreas (ADP), both considered as rare entities, has been reported only twice in the literature: in 2001 by Nakamura et al.[1] and in 2005 by Ulusan et al.[2] This article documents another rare case of this association.

The SPT is a rare type of neoplasm that accounts 1%-3% of all exocrine pancreatic tumors.[3] It was first described by a pathologist named Virginia Frantz, in 1959.[4] Since then, it has been given several different names, including "Frantz tumor," "solid and cystic tumor," "solid and papillary epithelial neoplasm," and "papillary-cystic tumor." In 1996, the World Health Organization established a new exocrine pancreatic tumor classification by histological type and defined this pathology as a solid pseudopapillary tumor.[5]

The SPT most commonly occurs in women, especially in the second and third decades of life.[6] It is considered a low-grade disease, with good prognosis in most patients. However, it can also develop into a malignant disease in a minority, with distant and lymph node metastases, vascular involvement, or recurrence. The most common symptom is abdominal pain, followed by total absence of symptoms and a noticeable abdominal mass.[7]

ADP is an extremely rare type of condition, and its prevalence remains unknown.[8] Pancreas development is a complex process and involves the fusion of two buds that grow from the foregut. The ventral bud gives rise to the most part of the head and uncinate process of the pancreas, while the dorsal bud gives rise to the cranial part of the head, body, and tail. ADP is caused by lack of regression of the dorsal bud during embryonic development and can be complete or incomplete.[8,9] In the complete form, the accessory papilla, duct of Santorini, the body, and tail are missing. In the incomplete form, only the tail of the pancreas is absent.[8] Some patients will be accompanied by other diseases, such as pancreatic tumors or pancreatitis.

CASE REPORT

The present paper reports a case of a 36-year-old woman who sought medical assistance due to a palpable mass in the epigastrium, which she noticed six months ago. The patient did not experience abdominal pain, weight loss, and jaundice, and had no history of pancreatitis. She had insulin-dependent diabetes mellitus for 10 years as the only comorbidity.

Imaging examinations were performed and tumor markers were measured. Abdominal computed tomography (CT) (Figure 1, 2) showed a large and complex mass with heterogeneous enhancement by intravenous contrast on pancreatic head topography, measuring 7.8×5.5cm, associated with complete agenesis of the body and tail of the pancreas. The mass tended to compress the duodenum, portal vein, and superior mesenteric vein, without signs of invasion. Abdominal magnetic resonance

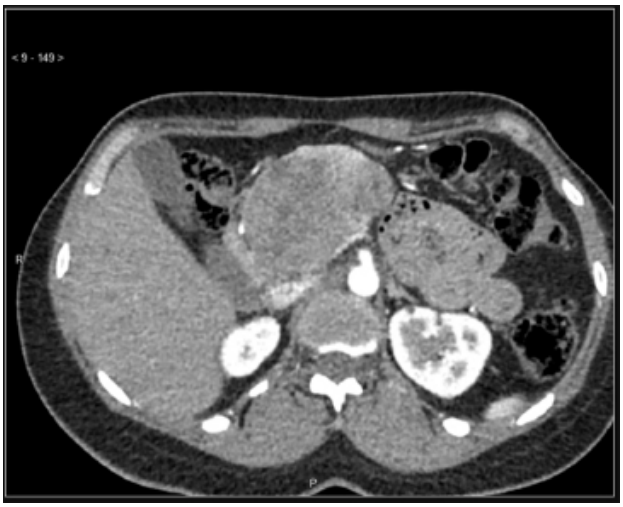

Figure 1. Abdominal CT showed a large and complex mass with heterogeneous enhancement by intravenous contrast on pancreatic head topography, measuring 7.8 × 5.5 cm, associated with complete agenesis of the body and tail of the pancreas.

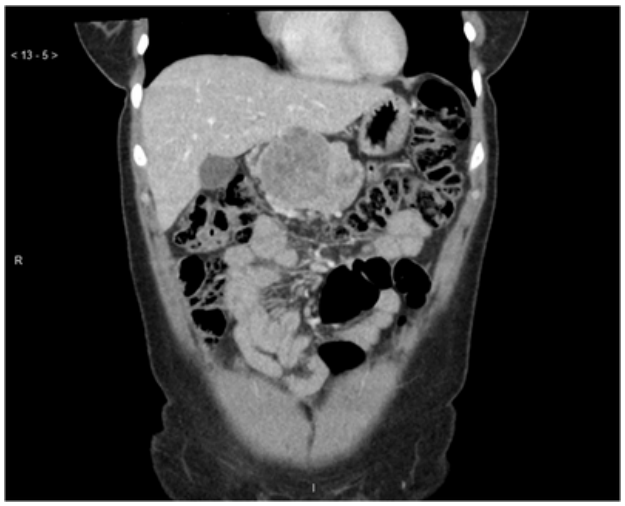

Figure 2. Abdominal CT showed a large and complex mass with heterogeneous enhancement by intravenous contrast on pancreatic head topography, measuring 7.8×5.5cm, associated with complete agenesis of the body and tail of the pancreas.

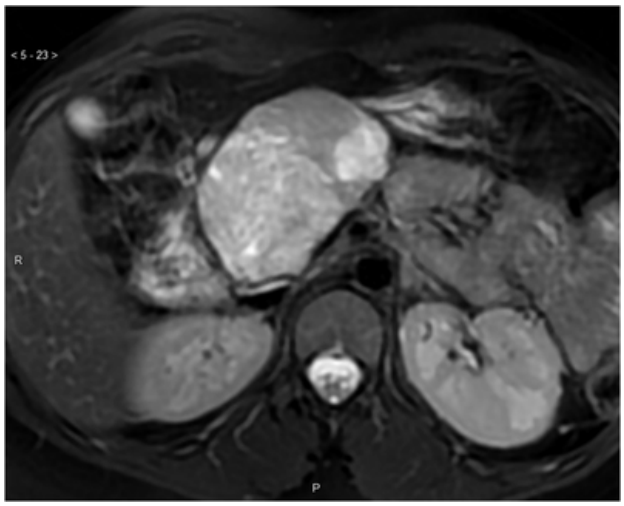

Figure 3. Abdominal magnetic resonance imaging showed similar characteristics of a large and complex mass pancreatic head topography.

(Figure 3, 4) imaging showed similar characteristics. The radiologists suggested a Frantz tumor or neuroendocrine tumor as a possible diagnosis. The tumor markers measured were carcinoembryonic antigen, carbohydrate antigen 19-9, cancer antigen 125, chromogranin A, and 5-hydroxyindoleacetic acid, which showed normal values.

The patient was referred for surgical treatment. During laparotomy, the body or tail of the pancreas was missing, and a large mass was noted at the head of the pancreas with invasion of the second part of the duodenum and adherence to the superior mesenteric and portal veins. Hence, gastroduodenopancreatectomy with complete resection of the pancreas was performed (Figure 5). The resection was particularly difficult because of the adherence to superior mesenteric vein. It was necessary minutious dissection, which started with vascular isolation and preservation. A Roux-en-Y reconstruction was carried out. Latero-lateral gastrojejunal anastomosis was performed in the alimentary loop, while latero-lateral hepaticojejunal anastomosis was performed in the biliary loop. The patient was transferred to the intensive care unit (ICU) immediately after surgery and was eventually moved to a regular room on postoperative day 2. She showed satisfactory improvements on the first few days, with good pain control, early ambulation, and good tolerance to oral feeding. The only complication that the patient developed was a low-output biliary fistula, which was treated with prophylactic abdominal drainage. The patient was discharged from the hospital on postoperative day 6 in excellent condition. The patient returned to the hospital after one week, and the drain was removed; then, she was instructed to visit the outpatient clinic within a few months for follow-up checkup.

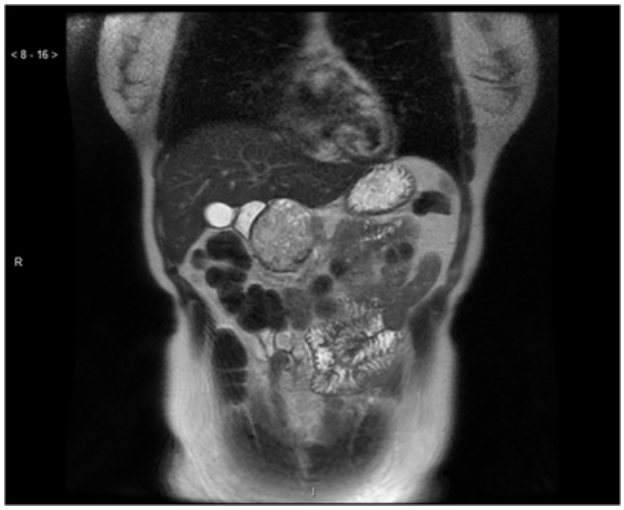

Figure 4. Abdominal magnetic resonance imaging showed similar characteristics of a large and complex mass pancreatic head topography.

The anatomopathological examination suggested an SPT with 18 lymphatic nodes without atypical findings. This finding was confirmed by immunohistochemistry, which revealed diffuse positivity for beta-catenin, cyclin D1, CD-10, and progesterone receptor, and focal positivity for synaptophysin.

DISCUSSION

SPT is a rare type of exocrine tumor, which typically grows slower and is associated with fewer or any symptoms. It is usually detected when it has grown into a large tumor. The rate of diagnosis has been increasing in the last 20 years, probably due to better accessibility to imaging examinations.[6] Among 2,744 patients with SPT who were identified by Law et al.

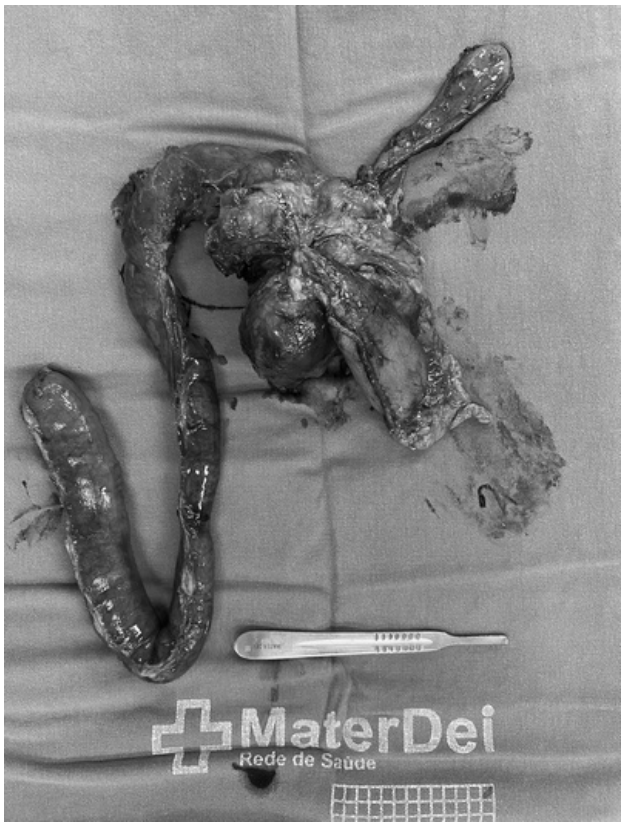

Figure 5. Specimen after resection.

(2014)[6] in a systematic review, 2,410 of them have been reported between 2000 and 2012, and only 334 were reported between 1959 and 1999.[7]

The typical clinical picture is a young woman complaining of abdominal pain, which occurs in 63.6% of the cases. According to Law et al. (2014),[7] 38.1% of the patients with SPT are asymptomatic, 32.5% develop an abdominal mass, and 19.7% experience nausea and vomiting. Less frequent symptoms include weight loss, jaundice, and pancreatitis. Women account for 87% of the total patients with SPT, and the average age was 28.5 years. The average tumor size was 8.6cm; in 59% of the cases, tumors commonly developed in the body and tail of the pancreas. However, tumor localization is not very useful for identifying SPT, as the tumor can occur throughout the pancreas.[7]

Given the excellent prognosis of SPT, establishing the correct preoperative diagnosis is essential for proper therapeutic planning. Abdominal CT findings usually show a hypodense retroperitoneal mass that is well defined, encapsulated, and mostly composed of solid and liquid components, with peripheral calcifications and central areas of hemorrhagic degeneration that undergo heterogeneous enhancement after administration of venous contrast, better identified in the arterial phase. The composition could also be solid or all cystic in a minority of cases. Magnetic resonance imaging (MRI) can be considered better than CT in some aspects, such as identifying areas of cystic degeneration, hemorrhage, and the tumor capsule.[10,11] Despite the availability of these imaging tools, the pre-operative diagnosis is complicated by the similarities of the findings between the SPT and other malignant pancreatic cystic neoplasms. Due to the fact that the final diagnosis of SPT is based on histopathological examination, some authors suggest the performance of endoscopic ultrasound-guided fine-needle aspiration biopsy.[12] However, this method remains controversial because of the risk of spreading tumor cells from a possible malignant lesion.

In general, SPT is classified as a neoplasm of epithelial origin, but the findings so far are unable to determine the specific cell line. The pathophysiology of SPT, therefore, remains unknown, and among the possible precursor lines identified by immunohistochemistry are exocrine epithelial cells (acinar and ductal), neuroendocrine cells, and stem cells.[13]

The SPT has an excellent prognosis, and the standard treatment is surgical resection. Normally, the absence of invasion to other organs allows resection with tumor-free margins. Due to the presence of capsule and the low grade of malignancy, conservative surgery must be performed. If it is located in the body or tail of the pancreas, distal pancreatectomy may be performed with spleen preservation if there are no signs of invasion. Enucleation should be considered for small tumors.[6] Approximately 95% of the patients were reported to be disease free after surgical removal of the tumors. Law et al. (2014)[7] identified only 29 deaths caused by SPT, and the mortality rate was less than 2%. The recurrence rate was 4.4%, and the incidence of lymph node metastases was 1.6%. For this reason, large lymphadenectomies are not recommended.[7] The use of chemotherapy and radiotherapy in the treatment of SPT is limited, and this may be due to the success of the surgical treatment. The small number of studies regarding this subject makes it difficult to draw any conclusions.[6,7]

A systematic review published in 2009 by Schnedl et al.[14] identified 53 cases of ADP published between 1911 and 2008. They found that 53% of the patients had diabetes and 53% had abdominal pain. Pancreatitis was detected in 30% of the patients.[14] In another systematic review, Cienfuegos et al. (2016)[9] identified another 53 cases published between 2008 and 2015, with a total of 106 cases. Approximately 43% of the patients had diabetes, 28% had abdominal pain, and 11% had pancreatitis. Other less frequent symptoms were exocrine pancreatic insufficiency, jaundice, weight loss, and back pain. Similar to SPT, the increase in the rate of diagnosis since 2008 is attributed to better accessibility to imaging examinations.[9]

Due to the functional reserve of the pancreas, several SPT patients can remain asymptomatic. The tail of the pancreas contains most of the islets and consequently the β-cells, which justifies the appearance of diabetes in the ADP. Abdominal pain might be caused by exocrine pancreatic insufficiency or chronic pancreatitis.[14,15] The

association between acute pancreatitis and ADP has been reported in several studies; among the mechanisms that could explain this association are dysfunction in the Oddi sphincter, pancreatic head compensatory hypertrophy, increased pancreatic juice secretion, and pancreatic duct hypertension.[15] Another important consideration is the possibility of pseudoagenesis of the dorsal pancreas, which refers to the fatty replacement of pancreatic tissue, secondary to inflammation caused by severe or recurrent pancreatitis. This hypothesis should be excluded in order to accurately diagnose ADP. The patient in this case had no history of acute or chronic pancreatitis, which suggests a real ADP.

Some authors have suggested the association between ADP and the occurrence of pancreatic tumors, but this has not been proven yet. The possible mechanisms remain unclear, and one of the hypotheses is that ADP increases the risk of chronic pancreatitis and consequently tumor appearance.[14,15] Erotokritou et al. (2018)[16] published a review and reported a case that demonstrated the association between ADP and a neuroendocrine tumor. They found 15 similar cases with different tumors: 8 cases with pancreatic adenocarcinomas, 1 with intraductal papillary mucinous neoplasm, 3 with periampullary adenocarcinoma, 1 with cystic without cellular atypia, and 2 with SPT.[16]

The two cases showing the association between ADP and SPT were already mentioned in this article. Nakamura et al. (2001)[1] published the first case, but they suggested an incomplete type of ADP since the Santorini duct was detected on endoscopic retrograde cholangiopancreatography. The second case was published by Ulusan et al. (2005),[2] and they suggested a complete ADP, which was examined by conducting a CT scan. In our case, structures from the dorsal pancreas were not seen on abdominal MRI or CT, which indicate that it was a true ADP case associated with an SPT.

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REFERENCES

1. Nakamura Y, Egami K, Maeda S, Hosone M, Onda M. Solid and papillary tumor of the pancreas complicating agenesis of the dorsal pancreas. J Hepatobiliary Pancreat Surg. 2001;8(5):485-9.
2. Ulusan S, Bal N, Kizilkilic O, Bolat F, Yildirim S, Yildirim T, et al. Solid-pseudopapillary tumour of the pancreas associated with dorsal agenesis. Br J Radiol. 2005 Jun;78(929):441-3.
3. Cienfuegos J, Lozano M, Rotellar F, Martí P, Pedano N, Arredondo J, et al. Solid pseudopapillary tumor of the pancreas (SPPT). Still an unsolved enigma. Rev Esp Enferm Dig. 2010 Dec;102(12):722-8.
4. Frantz VK. Papillary tumors of the páncreas: benign or malignant? Tumors of the pancreas. Atlas of tumor Pathology, 1st ed. Washington DC, Armed Forces Institute of Pathology 1959: 32-3
5. Klöppel G, Solcia E, Longnecker DS, Capella C, Sobin LH. Histological typing of tumors of the exocrine pancreas. 2nd ed. Berlin: Springer; 1996.
6. Costa-Neto G, Amico E, Costa G. Tumor sólido-cístico pseudopapilar do pân- creas (tumor de Frantz). Estudo de quatro casos. Arq Gastroenterol. 2004 Dez;41(4):259-62.
7. Law JK, Ahmed A, Singh VK, Akshintala VS, Olson MT, Raman SP, et al. A systematic review of solid-pseudopapillary neoplasms. Are these rare lesions? Pancreas. 2014 Apr;43(3):331-7.
8. Guimarães AB, Guimarães CA, Manso JEF. Agenesia ou pseudoagenesia do pâncreas dorsal. Rev Col Bras Cir. 2015;42(5):352-5.
9. Cienfuegos JA, Rotellar F, Salguero J, Benito A, Solórzano JL, Sangro B. Agenesis of the dorsal pancreas: systematic review of a clinical challenge. Rev Esp Enferm Dig. 2016 Aug;108(8):479-84.
10. Antoniou E A, Damaskos C, Garmpis N, Salakos C, Margonis GA, Kontzglou K, et al. Solid pseudopapillary tumor of the pancreas: a single-center experience and a review of the literature. In Vivo. 2017 Jul/Aug;31(4):301-510.
11. Cerron-Vela CR, Navarro PM, Araujo-Banchon WJ. Rev Facul Cienc Méd Córdoba. 2020;77(1):1-6.
12. Salla C, Chatzipantelis P, Konstantinou P, Karoumpalis I, Pantazopoulou A, Dappola V. Endoscopic ultrasound-guided fine-needle aspiration cytology diagnosis of solid pseudopapillary tumor of the pancreas: a case report and literature review. World J Gastroenterol. 2007 Oct;13(38):5158-63.
13. Santini D, Poli F, Lega S. Solid-papillary tumors of the pancreas: histopathology. JOP. 2006 Jan;7(1):131-6.
14. Schnedl WJ, Piswanger-Soelkner C, Wallner SJ, Reittner P, Krause R, Lipp RW, et al. Agenesis of the dorsal pancreas and associated diseases. Dig Dis Sci. 2009 Mar;54(3):481-7.
15. Mei W, Cao F, Li F. Two cases of agenesis of the dorsal pancreas and a review of the literature. BMC Gastroenterol. 2020 Apr;20:94.
16. Erotokritou A, Gerharz CD, Sagir A. Agenesis of dorsal pancreas associated with pancreatic neuroendocrine tumor: a case report and review of the literature. J Med Case Rep. 2018 Jun;12(1):185.