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Original Research Paper

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SOLID PSEUDOPAPILLARY NEOPLASM OF THE PANCREAS: PRESENTATION OF A CASE AND REVIEW OF THE LITERATURE

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ABSTRACT Solid pseudopapillary neoplasms (SPN) of the pancreas are rare tumors, occurring in 1-2% of all pancreatic neoplasms; of these 10 to 15% have an aggressive presentation. Most of the patients present disease localized to the pancreas, however, between 9-15% may present local invasion and metastasis. The clinical presentation in the vast majority of SPN is asymptomatic, even despite a large tumor size. We present a case of a 16-year-old female patient with a psedupapillary tumor of the pancreas (Frantz tumor) who was admitted due to abdominal pain and weight loss lasting 3 months.

KEYWORDS : pancreas, pancreatic neoplasia, neoplasia.

INTRODUCTION

Solid pseudopapillary neoplasia of the pancreas (SPN) is a rare tumor of the pancreas that can mostly be considered benign, however, a significant percentage can be aggressive and potentially malignant, usually located in the tail of the pancreas. This tumor can grow up to 8 to 10 cm in diameter, but given the rarity of the disease and the lack of research on it, its course remains unpredictable. SPN today have been diagnosed more frequently by the extensive use of studies such as computed tomography and magnetic resonance imaging, in which it can be found as an incidental finding [1,2]. We present a case of a solid pseudopapillary neoplasm of the pancreas, which was admitted due to abdominal pain and weight loss lasting 3 months.

Case Report

A 16-year-old female patient admitted to the emergency department due to a 3-month-long clinical picture consisting of epigastric pain of moderate intensity, a sensation of early fullness, weight loss and anorexia. The physical examination revealed a hemodynamically stable patient, without signs of respiratory distress, with abdominal distension and a palpable abdominal mass. A non-contrast tomography was performed, which revealed a round homogeneous lesion, which was restricted in the diffusion sequence represented by hyperintensity in the DW sequence and hypointense in the ADC map (**Figure 1**). Nuclear Magnetic Resonance Imaging (NMR) with contrast revealed a mass with enhancement in the early phase and its persistence in the late phases (**Figure 2**). At laparotomy, a well circumscribed and encapsulated tumor was found; in microscopic pathology, it is considered a polygon tumor with cells with moderate or abundant amphophilic cytoplasm. In immunohistochemical studies it is positive for alpha 1 antitrypsin and alpha 1 antichymotrypsin. The final diagnosis of a solid papillary tumor of the pancreas (Frantz tumor) is reached. Patient at 2 years of follow-up without relapse.

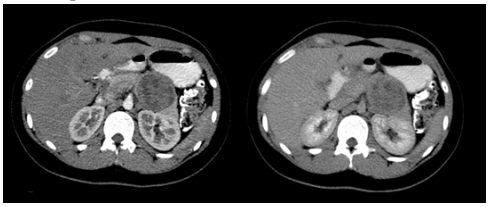

Figure 1. Abdominal axial tomography contrasted in arterial

phase, a space-occupying image is observed in the body of the pancreas, tumor-benign in appearance, located to the left of the mesenteric artery superior and without signs of arterial or venous vascular invasion, or involvement of the pancreatic duct, it has rounded morphology, with density and heterogeneous enhancement. Phase contrast tomography venous, heterogeneous enhancement is maintained.

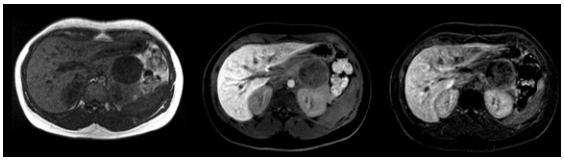

Figure 2. Abdominal, axial MRI, T1-weighted sequence, a homogeneous hypointense lesion is visualized. Abdominal, axial MRI, T1-weighted sequence after the administration of contrast medium, in arterial phase and subtraction technique, the lesion has homogeneous enhancement of the capsule and heterogeneous content, which is maintained in the late dynamic phases (not shown).

DISCUSSION

Solid pseudopapillary neoplasms (SPN) of the pancreas are rare tumors, occurring in 1-2% of all pancreatic neoplasms; of these 10 to 15% have an aggressive presentation. This type of neoplasia occurs mainly in young women (90%), as in the case of our patient, its prevalence has increased significantly with the widespread use of images such as tomography and resonance to study other pathologies [1,2]. The mean age of onset is 35 years, most of them are asymptomatic and can have any location at the level of the pancreas and even extrapancreatic presentations [3].

The cellular origin of SPN has not yet been clarified, however, it has been associated that during the development of the pancreas, beta-catenin signaling is altered, which is necessary for the proper development and functioning of the pancreas in adult life. Some studies have suggested that beta-catenin mutations are significantly associated with SPN tumorigenesis [3]. Proteins that are associated with the cell cycle such as cyclin D1 and cyclin D2 have been found to be overexpressed in patients with SPN, associated with the dysregulation in the cell cycle that occurs in this pathology (2). These tumors have complex karyotypic changes, and are related to mutations on chromosomes 2, 4 or X, and strongly on chromosome 11 (1).

Most of the patients present disease localized to the pancreas, however, between 9-15% may present local invasion and metastasis. The 5-year survival for SPN is 97%, even in the presence of metastasis (4,5). however, unresectable or metastatic tumors have a negative impact on survival (1).

The clinical presentation in the vast majority of SPN is asymptomatic, even despite a large tumor size (4). It presents nonspecific symptoms such as abdominal pain, predominantly in the epigastrium, which is the most frequent symptom [6], nausea, vomiting, asthenia, back pain, early satiety, jaundice, a clinical picture similar to pancreatitis or initial diagnosis of pancreatitis and symptoms constitutional symptoms such as weight loss, symptoms presented by our patient (2).

The physical examination may find pain on palpation of the epigastrium, a palpable abdominal mass, jaundice or an enlargement of an abdominal mass that can be discovered by the patient, which may present compression of adjacent organs and symptoms associated with organ involvement corresponding according to the size of the lesion (5,6). Signs of an acute abdomen may present in about 8%, however the initial laboratories may be normal, therefore the diagnosis is mostly late (4,7).

The cystopapillary tumor of the pancreas is considered a low-grade tumor of malignancy, in all cases it is recommended to perform resection with negative surgical margins in order to achieve local control of the disease, prevent recurrence and metastasis, alleviate symptoms and ensure a good long-term prognosis. The location and local invasion generally determine the surgical technique. Surgical treatment depends on the location and extent of the tumor. Distal pancreatectomy, with preservation of the spleen, whenever possible, should be performed when it is located in the body and tail of the pancreas (8).

Differential diagnoses of SPN are cystic tumors such as cystic adenoma, microcystic adenoma, lymphandioma, cystadenocarcinoma, sarcoma, pancreatoblastoma, ductal and non-ductal acinar cell carcinoma, teratomas, hemangioendotheliomas and hydatic cysts, despite differentiating psepillary tumors of other pancreatic tumors, especially endocrine ones, is a challenge, although suspecting whether it is associated with leukemia, neuroblastoma or lymphoma, since in these cases PNS is the most common (2-4).

The prognosis of the disease with surgical treatment is good with a survival of 97% of cases, due to its low rate of malignancy and low probability of recurrence, around 10-15%. Mortality is approximately 1.5%, with perineural or vascular invasion, high degree of cellular pleomorphism, high mitotic rat and the presence of aggressive behavior indicators such as diffuse growth pattern, necrosis, as poor prognostic factors. extensive and significant nuclear atypia (9).

Ethical standards and patient consent

We declare that the patient described in this study gave informed consent prior to inclusion in this study.

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