

Solid pseudopapillary tumor of the pancreas. A case report

Tumor sólido pseudopapilar del páncreas. A propósito de un caso

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Summary

The solid pseudopapillary tumor of the pancreas is an extremely rare neoplasm of epithelial origin that represents less than 1 - 2 % of exocrine pancreatic tumors, being more prevalent in the female population, between the second and third decade of life. We present the case of a 20-year-old patient who consulted a local hospital in the city of Bogotá, Colombia, due to recurrent abdominal pain and sensation of a mass in the mesogastrium and left flank, with imaging and histological findings of a solid pseudopapillary tumor of the pancreas.

Resumen

El tumor sólido pseudopapilar del páncreas es una neoplasia de origen epitelial, extremadamente rara, que representa menos del 1-2 % de los tumores pancreáticos exocrinos. Es prevalente en la población femenina, entre la segunda y la tercera décadas de la vida. Se presenta el caso de una paciente de 20 años de edad que consulta por dolor abdominal recurrente y sensación de masa en el mesogastrio y el flanco izquierdo, con hallazgos imagenológicos e histológicos de un tumor sólido pseudopapilar del páncreas.

Introduction

Pseudopapillary solid tumor of the pancreas is a benign neoplasm of epithelial origin, whose etiology is unknown and represents less than 2% of exocrine pancreatic tumors, with prevalence in young women between 25 and 35 years of age.

Since this is a low incidence neoplasm, the key aspects of this pathology will be discussed, as well as the usefulness of the different imaging modalities that play an important role in its diagnosis and treatment; for this purpose, the clinical case of a patient who was admitted to a local hospital in Bogotá, Colombia, with clinical and imaging findings that suggested the diagnosis, which was later confirmed by histology, is presented.

Case report

A 20-year-old female patient consulted at a local hospital in Bogota, Colombia, for two months of recurrent abdominal pain, involuntary weight loss and postprandial emesis, associated with a sensation of mass in the mesogastrium and left flank; the patient was admitted to the emergency department with extrainstitutional ultrasound showing a solid-cystic mass in the left flank, of undetermined origin. Physical examination revealed pain on palpation in the left upper quadrant of the abdomen and a solid mass in that location. Paraclinical tests showed elevated transaminases and direct hyperbilirubinemia.

The plain abdominal radiograph (Figure 1) shows a mass in the left hypochondrium and flank, with dimensions of 12 × 15 cm, with apparent displacement of the stomach to the right, which is confirmed in the computed axial tomography (CT) of the abdomen (Figure 2) showing a predominantly solid lesion, rounded morphology and defined margins, with heterogeneous enhancement, apparently dependent on the body and tail of the pancreas, which configures the "claw sign", with no clear cleavage plane with the left adrenal gland. In magnetic resonance imaging (MRI) (Figure 3) the lesion had a low signal in T1, with areas of microcystic appearance with high signal in T2 and progressive heterogeneous enhancement with contrast medium.

Distal pancreatectomy plus splenectomy was performed, and the pathologic report evidenced solid pseudopapillary neoplasm, involving the body and tail of the pancreas, without involvement of the left adrenal gland. The neoplastic cells were positive for A1 antitrypsin, cyclin D1, CD56, vimentin, progestin receptor and CD10; the cell proliferation index evaluated with Ki67 is 1 %. Regional lymphadenectomy was not required.

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Discussion

Solid pseudopapillary neoplasm of the pancreas is a rare entity, accounting for less than 2% of pancreatic tumors. Any part of the pancreas can be the site of origin; although it is prevalent in the tail and body. It occurs in both sexes, with female predominance and mean age of 25-35 years. Its etiology is unknown and the possibility of a multifactorial origin with ductal, epithelial, neuroendocrine, pluripotential cell components has been described, and an extrapancreatic origin has even been considered (1, 2).

It is a benign neoplasm with a very good prognosis, even in the context of malignant transformation, which occurs in 18.3% of adult patients and 43.1% of pediatric patients (3), with survival rates of up to 97% at 5 years (4). Factors associated with a worse prognosis, predictors of recurrence or metastasis, include male gender, age less than 13.5 years, locoregional, perineural and/or lymphovascular invasion, lesion size greater than 5 cm, positive resection margins and capsule invasion. Although metastases are rare, they are more frequent in liver and peritoneum (2, 3). The diagnosis is usually incidental; however, when patients present with symptoms, these are non-specific, such as recurrent abdominal pain, sensation of fullness, involuntary weight loss and postprandial emesis; likewise, laboratory tests in most cases are normal, including CA19-9 and carcinoembryonic antigen, so they have no prognostic value (1, 3).

The imaging presentation is quite characteristic: in ultrasound it is observed as a solid, heterogeneous mass with defined margins, with peripheral vascular flow in the color Doppler scan and cystic areas in its interior of variable size due to necrosis or hemorrhage.

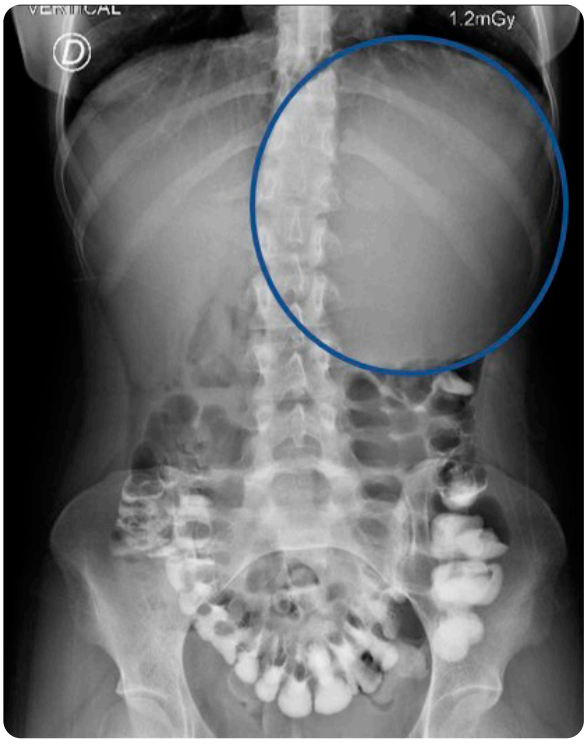


Figure 1. Abdominal X-ray. Increased radiopacity in the left hypochondrium and flank, displacing adjacent structures, with dimensions of 12 × 15 cm. Remains of contrast material in colic frame.



Figure 2. CT of the abdomen with contrast medium, axial, sagittal and coronal sections. Mass in the left hypochondrium and flank, with defined margins and heterogeneous enhancement, measuring approximately 139 × 135 × 97 mm (L × AP × T). The lesion displaces adjacent structures and configures the "claw sign" (blue line) with the body of the pancreas, suggesting pancreatic origin.

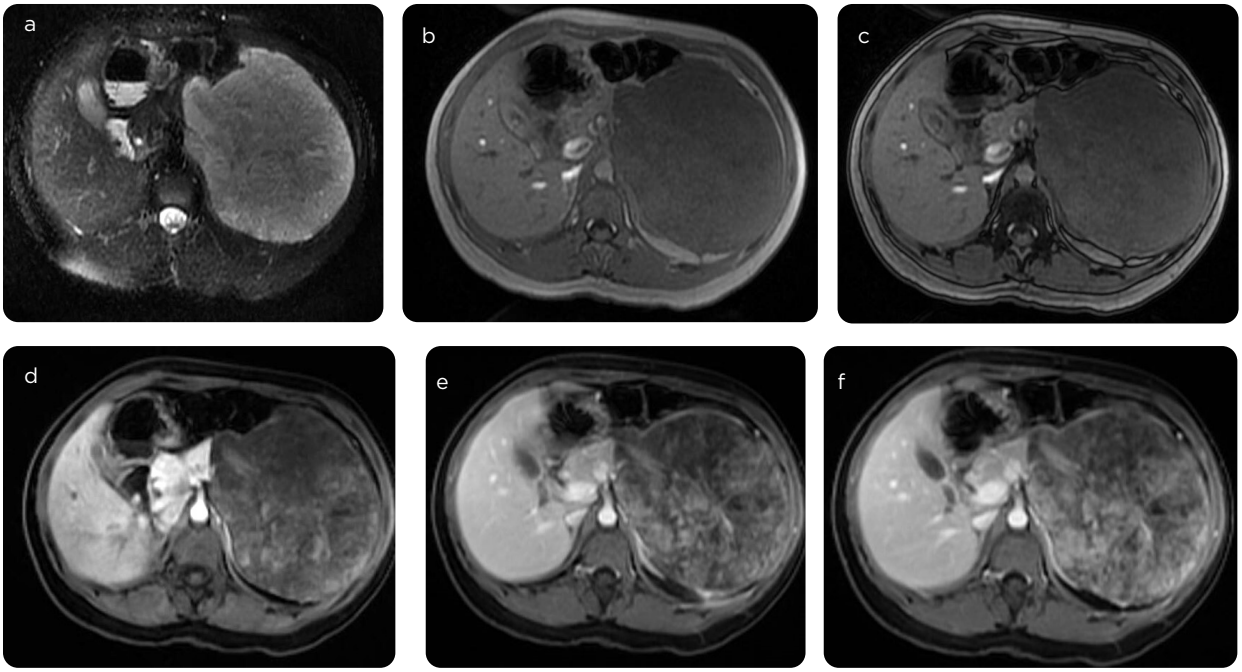


Figure 3. MRI of the abdomen with contrast medium. a) axial T2 FS, b) axial T1 in phase, c) axial T1 in opposite phase, d) axial T1 with contrast, arterial phase, e) axial T1 with contrast, portal phase, f) axial T1 with contrast, late phase. Heterogeneous mass, predominantly solid, dependent on the tail and body of the pancreas, with low signal in T1, microcystic areas in T2, without signal drop in opposite phase suggesting fat content, with progressive heterogeneous enhancement with contrast medium.

In CT they are well encapsulated lesions, of round or oval morphology, with a solid-cystic component (with areas of hemorrhage more frequent in tumors larger than 5 cm), peripheral calcifications (approximately 20%) and heterogeneous enhancement. In MRI they have a heterogeneous behavior in T1 sequences (high signal in areas of hemorrhagic degeneration), high signal/heterogeneous in T2, with early and slowly progressive enhancement in T1 sequences with contrast medium (5).

Among the differential diagnoses are hypervascular neoplasms or with cystic component, such as serous cystadenoma, neuroendocrine tumors and metastases (6); however, the pseudopapillary solid tumor must be considered in an adequate clinical and epidemiological context.

The definitive diagnosis is made by histology and immunohistochemistry. It is characteristically positive for α1antitrypsin, CD56, CD10 and vimentin. The treatment of choice is surgical resection, even in cases of distant metastasis or local recurrence (7).

In the clinical case presented, radiography, CT and MRI were available, which allowed correlating the findings with those described in the literature. In addition, knowledge of the clinical history and a multidisciplinary management team ensured an adequate approach and satisfactory results. The patient is currently under follow-up, without complications or signs of tumor recurrence. The importance of considering this diagnostic possibility in young patients with an appropriate clinical and imaging picture is emphasized.

Conclusion

Pseudopapillary solid tumor of the pancreas is a rare neoplasm that characteristically presents in young, asymptomatic women, with an excellent long-term prognosis; the role of imaging is fundamental, since its correct interpretation guides the diagnosis, which then requires histological confirmation.

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