SOLID PSEUDOPAPILLARY TUMOR OF PANCREAS IN A 24-YEAR-OLD FEMALE: A DIAGNOSTIC CHALLENGE

Dr. Sonia Chhabra, Dr. Sunita Singh, Dr. Ashima Batra, Dr. Mansi Agarwal, *Dr. Nitesh Kumari, Dr. Rajeev Sen

Department of Pathology, PGIMS, Rohtak, Haryana India.

*Corresponding Author: Dr. Nitesh Kumari

Department of Pathology, Hostel PGIMS, Rohtak, Haryana India.

ABSTRACT

Solid pseudopapillary tumor (SPT) of the pancreas is a rare neoplasm usually missed clinically but diagnosed on either radiological or pathological examination. This tumor with malignant potential has been described in females in their third decade of life. The tumor is thought to have a ductal or acinar origin. Malignant potential and favourable prognosis, if diagnosed early, necessitate a timely intervention and a complete resection has been curative in most cases. Herein, we report a case of this rare tumor in a young morbidly obese female diagnosed solely on microscopic examination.

KEYWORDS: Solid pseudopapillary tumor (SPT), pancreas, female, young.

INTRODUCTION

Solid pseudopapillary tumor (SPT) of the pancreas is rare neoplasm, accounting for only 0.1-2.7% of all the pancreatic tumors.[1] Most reports of these tumors occur in female patients in the third decade of life with an age range of 12–79 years. Reports of the tumor in young children have also been made. The majority of patients present with vague abdominal symptoms, resulting in a delay in presentation and diagnosis. The pathogenesis is thought to result from cells of the endocrine pancreas though some investigators have postulated origin from the exocrine pancreas.[2] These tumors can be visualized in many imaging modalities, such as ultrasonog-raphy (US), computed tomography (CT) and magnetic resonance imaging (MRI), which can be used to differentiate it from other pancreatic lesions.[3] Complete resection of local disease is curative. Even patients with residual disease or metastases have been reported to have long-term survival following surgical treatment. Very few reports of the use of chemotherapy or radiotherapy for these tumors exist with only limited response.[2]

CASE REPORT

A 24 years female, a known case of type II diabetes mellitus presented with complaints of rapid weight gain since past 5-6 years and menorrhagia since past 1 year. The cycle lasted 7-30 days with 2-3 monthly cycle with presence of clots without dysmenorrhea. Her weight was 96 Kg. Per abdomen was soft with mild tenderness without any organomegaly. Her hemoglobin was 8 g%. Serum calcium was raised (9.5mg/dl). Rest of biochemical investigations were within normal limits. Ultrasound of abdomen and pelvis showed a heterogeneous, slightly echogenic and vascular ovoid mass, measuring 9x9x8 cms involving body and tail of pancreas. CECT abdomen revealed a 9.4×9.2×10.2 cms lesion arising from the body and tail of pancreas. (Figure 1A & B) Tumor markers (AFP, CEA, CA 1 9–9 and CA 125) were within normal limits.

Distal pancreatectomy with splenectomy with ADR was done. Per operatively, a 15x15 cm encapsulated highly vascular lump was seen originating from the body of pancreas. There was no free fluid and no lymphadenopathy or any other mass. Drain fluid amylase was markedly raised (925 U/L), although serum amylase was within normal limits (32.40 U/L).

We received a well encapsulated, well demarcated tumor in body and tail of pancreas. The cut surface revealed solid and cystic components with areas of necrosis and hemorrhage. (Figure 2A, B & C).

Microscopic examination revealed a well circumscribed tumor mass separated from pancreas by a fibrous capsule. The tumor cells were arranged in sheets, nests, pseudopapillary formations (tumor cells oriented around fine blood vessels). (Figure 3A & B) Areas of cystic change, necrosis and hemorrhage were also evident. Tumor cells had round to oval minimally pleomorphic nuclei, inconspicuous nucleoli and fine blood vessels. (Figure 3A & B) Areas of cystic change, necrosis and hemorrhage were also evident. The tumor cells were positive for PR (progesterone receptor) (Figure 3C), CD10 &
CD20 (Figure 3D). Immunostains CK, EMA, CEA, Vimentin, Synaptophysin, Chromogranin showed negative immunoreactivity. A diagnosis of solid pseudopapillary tumor of pancreas was rendered. The post-operative period was uneventful. The patient is still under follow-up since last one year.

DISCUSSION
The solid pseudopapillary tumor of the pancreas (SPT) is a rare, low-grade malignant tumor of unknown etiology accounting for 0.2-2.7% of all primary pancreatic tumors. This rare tumor seems to have a predilection for young Asian and African-American women. The male to female ratio is 1:10 and the mean age at presentation is 22 years.

The origin of solid pseudopapillary tumors is yet a matter of debate. Many investigators favor the theory that SPTs originate from multipotent primordial cells, whereas others suggest an extrapancreatic origin, from genital ridge angle-related cells.

The commonest presenting symptom is abdominal pain. Few authors have reported it to be an incidental finding during routine diagnostic imaging procedures. It may present as a completely asymptomatic palpable mass in young women. The classic CT features of solid pseudo-papillary tumor are a large well encapsulated mass with varying solid and cystic components caused by hemorrhagic degeneration. Calcifications and enhancing solid areas may be present at the periphery of the mass. MRI typically shows a well-defined lesion with a mix of high and low signal intensity on T1- and T2-weighted images. Literature supports USG and CT abdomen as the primary diagnostic modalities, however, in the present case, the diagnosis was made based on the pathological examination surgical excision of the mass.

Grossly, SPTs are identified as well-demarcated, encapsulated tumors with extrapancreatic growth. Mixed solid and cystic components are evident with internal necrotic or hemorrhagic debris and lobulated, solid tissue at the periphery.

The tumor is characterized by a fibrous capsule surrounding a varied portion of solid and cystic components showing hemorrhagic changes. Under light microscope, the pseudopapillary structures and degenerative changes, such as necrosis, hemorrhage, cholesterol clefts and foamy macrophages, are characteristic findings. Besides, the tumor cells are characteristically uniform with mild atypia and rare mitosis, indicating its benign behavior.

Solid-pseudopapillary tumors test positive for vimentin, neuron-specific enolase, α1-antitrypsin, and α1-antichymotrypsin and are negative for chromogranin, epi-thelial membrane antigen, and cytokeratin, insulin and glucagon. Depending on the tumor position (head, body or tail of the pancreas), the differential diagnosis include adrenal mass, pancreatic endocrine tumor, liver cyst or tumor, or a pseudocyst.

Figure 1. CECT abdomen revealing a 10.2x9.4x9.2 cm lesion arising from body and tail of pancreas. (1A & B).

Figure 2. Distal pancreatectomy specimen revealing a well circumscribed tumor in body and tail of pancreas (2A). The cut surface revealed solid and cystic components with areas of necrosis and hemorrhage. (2B & C).

Figure 3. Microphotograph revealing a well circumscribed tumor mass separated from pancreas by a fibrous capsule. (3A; H&E; 40X) The tumor cells were oriented around fine blood vessels, (pseudopapillary formations) (3B; H&E; 100X) The tumor cells were positive for PR (progesterone receptor) (3C; IHC; 100X) & CD10 (3D; IHC; 200X).
Surgical excision offers the best chance for cure and should always be attempted irrespective of the magnitude of resection involved. Few authors have reported an increased rate of resectability after chemotherapy. There are few case reports which have shown a survival benefit with radiotherapy. However, most authors agree that aggressive surgical resection is the best modality of treatment for achieving curative results and a better long-term survival.[5] In the present case, the patient has been doing well after surgery alone with no chemotherapy or radiotherapy planned as yet.

Despite the locally aggressive features, the tumor has a low-grade malignant potential and tends to have a favorable prognosis, even in the presence of metastatic disease. Overall 5-year survival is as high as 97% in patients undergoing surgical resection.[5] Metastasis occurs in a small number of cases, the most common site being the liver.[6]

To conclude, owing to the vague symptoms and, sometimes, even the concealed occurrence, the extreme rarity and the potential to develop into a malignancy, a high index of suspicion is required on the parts of clinician and radiologist. The diagnosis made histologically can be confirmed on immunohistochemical analysis. However, the entity requires still more recognition to become a frequent differential for the abdominal masses in young females.

REFERENCES