Solid Pseudopapillary Tumor of the Pancreas in Child: A Case Report

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Solid pseudopapillary tumor of the pancreas is a very rare form of childhood pancreatic tumor. We report the case of an 11-year-old girl having a solid pseudopapillary tumor of the pancreas presenting with left upper abdominal pain. Imaging studies showed the lesion to be an ovoid solid mass arising from the body and the tail of the pancreas. The tumor was surgically resected and was histopathologically diagnosed as a solid pseudopapillary tumor.

Keywords: Pancreatic neoplasms; Pancreas computed tomography; Pancreas ultrasonography; Pancreas magnetic resonance

INTRODUCTION

Solid pseudopapillary tumor of the pancreas is a very rare benign or low-grade malignant neoplasm of childhood pancreatic tumor [1]. It is almost exclusively encountered in young women having a mean age of 26 years and has a male to female ratio of 1:9 [2,3]. Patients are often asymptomatic and the tumor is discovered incidentally on physical or radiological examination [2]. Patients may also occasionally present with an increasing abdominal mass associated with vague abdominal discomfort or may rarely present with an acute abdomen due to tumor rupture and hemothor- tion. We report a case of solid pseudopapillary tumor of the pancreas and describe the imaging findings of this rare tumor.

CASE REPORT

An 11-year-old girl presented the pediatric department with a chief complaint of left upper abdominal pain of two weeks duration. The patient had normal vital signs and was afebrile. Physical examination showed tenderness at palpation over the left upper abdomen. Laboratory examination revealed no abnormal findings. Ultrasonography (HD 5000, Philips Medical Systems, Bothell, WA, USA) showed an ovoid 2-cm size isoechoic mass at the body and the proximal tail of the pancreas (Fig. 1). Multidetector CT (Somatom Emotion 16, Siemens, Forchheim, Germany) revealed an ovoid 2.2 × 2-cm size solid mass with progressive fill-in (Fig. 2). MRI (1.5-T Sonata, Siemens) results demonstrated an ovoid 2-cm sized solid mass in the body and the proximal tail of the pancreas. The mass was low signal intensity on the axial fat suppressed T1-weighted image (Fig. 3A); slightly high signal intensity on the axial T2-weighted image (Fig. 3B). On the initial arterial phase contrast enhanced dynamic fat suppressed T1-weighted image, the mass was slightly hypointense (Fig. 3C), but on the portal venous and equilibrium phase images, it showed progressive filling and became almost imperceptible (Fig. 3D, E). Magnetic resonance cholangiopancreatography demonstrated mild dilatation of the pancreatic duct in the tail of the pancreas (Fig. 3F). The laparoscopic laparotomy was performed. There was a mass in the body and the proximal tail of the pancreas. Distal pancreatectomy was performed. At gross examination, the cut surface showed an ovoid, well-circumscribed 2 × 2-cm size solid mass at the body and the proximal tail of the pancreas (Fig. 4A). Microscopically, the tumor was not encapsulated and composed of pseudopapillary aggregates with several layers of epithelial cells and myxoid stroma with blood vessels (Fig. 4B). The vascular or perineural invasion, definite nuclear atypism, mitotic activity, and necrosis were not found. At immunohistochemical analysis, the tumor cells were positive for vimentin, α-1-antitrypsin, α-1-antichymotrypsin, neuron-spe-
cific enolase, chromogranin, and partly positive for progesterone receptor. The cells were negative for cytokeratin and carcinoembryonic antigen. These findings helped establish a diagnosis of solid pseudopapillary tumor of the pancreas.

**DISCUSSION**

Solid pseudopapillary tumors of the pancreas are very rare in children and typically diagnosed in young women, especially black or East Asian women and have a female predominance [4,5]. Synonyms include solid and cystic tumor, solid and papillary epithelial neoplasm, papillary-cystic neoplasm, papillary cystic epithelial neoplasm, papillary-cystic tumor, and Franz tumor. In 1996, the World Health Organization renamed this tumor as solid pseudopapillary tumor for the international histologic classification of tumor of the exocrine pancreas [6]. It is most commonly detected incidentally, but may occasionally present with a gradually enlarging abdominal mass or complain of vague abdominal pain or discomfort. The abdomen is usually nontender on palpation, but obstructive symptoms may occur if the tumor grows large enough to compress adjacent organs. There are usually no abnormalities in clinical laboratory tests or in pancreatic cancer markers. The mass may occur in anywhere in the pancreas but is most frequently found in the head or tail. At gross examination, the mass is usually large and well encapsulated and contains varying amounts of necrosis, hemorrhage, and cystic change [4]. At microscopic analysis, there are two distinct types of cellular arrangements: solid and papillary. The hallmark histologic pattern occurs when the tumor cells from papillary configurations composed of a fibrovascular stalk surrounded by several layers of epithelial cells. Solid areas containing necrosis, foamy macrophages, cholesterol granulomas, and calcifications may also be seen [5]. The pathogenesis of these tumors is still controversial [7]. However solid pseudopapillary tumor shows immunohistochemical and ultrastructural evidence of both a neuroendocrine and an acinar-ductal differentiation, suggesting that this tumor arises from a pluripotent stem cell [7,8]. Solid pseudopapillary tumors are typically positive for vimentin, neuron-specific enolase, α-1-antitrypsin, and α-1-antichymotrypsin and negative for chromogranin, epithelial membrane antigen, and cytoker-

![Fig. 1. Ultrasonographic transverse scan shows an ovoid isoechoic mass at the body and the proximal tail of the pancreas.](image1)

![Fig. 2. (A) Contrast-enhanced arterial phase computed tomography (CT) scan demonstrates an ovoid peripherally enhanced low attenuation mass (arrows) in the body and the proximal tail of the pancreas. (B) Contrast-enhanced portal phase CT scan shows progressive fill-in of the mass (arrows).](image2)
atin. In our case, the tumor cells were immunoreactive for vimentin, α-1-antitrypsin, α-1-antichymotrypsin, neuron-specific enolase, chromogranin. The above-mentioned immunohistochemical features are strongly suggestive of solid pseudopapillary tumor of the pancreas. Sex hormones may play a role in the pathogenesis of growth of solid pseudopapillary tumors. Nearly all studies demonstrate no evidence of estrogen receptor. However progesterone receptors are present in many cases [5]. In our case, the cells were positive for progesterone receptor. At sonography, the appearance of solid pseudopapillary tumor is variable and lacks correlation with gross pathology [9]. In our case, ultrasonography showed isoechoic mass. Contrast-enhanced computed tomography (CT)
plays a major role in the diagnostic evaluation of neoplasms of the pancreas. However, when compared with magnetic resonance (MR) imaging, CT has inherent limitations in showing certain tissue characteristics, such as hemorrhage, cystic degeneration, or presence of capsule. On contrast-enhanced CT, solid pseudopapillary tumor typically presents as a large heterogeneous mass. Our study showed relatively homogenous mass on CT. Solid pseudopapillary tumor of the pancreas is heterogenous high or low signal intensity on T1-weighted, heterogenous high signal intensity on T2-weighted images, which reflects the complex nature of the mass. Our case is somewhat atypical due to the small size of the tumor, which may the lack of significant cystic change or hemorrhage at MR imaging. On gadolinium-enhanced dynamic MR imaging, the most common enhancement pattern of solid pseudopapillary tumor consists of early, peripheral, and heterogeneous enhancement during the arterial phase with progressive but heterogeneous fill-in of the lesion during the portal venous and equilibrium phases [10]. Our case showed also peripheral enhancement with progressive fill-in on gadolinium-enhanced dynamic MR imaging, which suggests a solid pancreatic neoplasm and helps distinguish solid pseudopapillary tumor from other pancreatic neoplasms, such as neuroendocrine tumors, that typically enhance more than the pancreas. Solid pseudopapillary tumors posses a malignant potential risk of 5 to 10% and must therefore be resected completely and aggressively [2]. Unlike pancreatic ductal adenocarcinoma, complete resection of a solid pseudopapillary tumor is usually curative and patients can survive a long period after the operation. Death due to tumor growth, liver metastases, or peritoneal seeding is rare. In children, the differential diagnosis of this lesion included neuroendocrine tumor, pancreatoblastoma. Although neuroendocrine tumors occur in patients who are older and do not have the female predominance observed with solid pseudopapillary tumor, neuroendocrine tumors may appear cystic, contain calcifications, and show areas of internal hemorrhage. Pancreatoblastoma is more aggressive than solid pseudopapillary tumor and often presents with liver metastases at the time of diagnosis.

In conclusion, in the pediatric age group, solid pseudopapillary tumor of the pancreas is a very rare tumor. Awareness of the imaging findings will allow accurate diagnosis and appropriate management to be undertaken.

REFERENCES

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