Solid pseudopapillary tumor of pancreas causing sinistral portal hypertension

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INTRODUCTION
Solid pseudopapillary tumor is an uncommon neoplasm that mainly occurs in women in the second to fourth decades of life. It is characterized by low potential for malignancy and a favorable prognosis. Since Franz [1] described this tumor in 1959 as a “papillary tumor of the pancreas, benign or malignant,” the number of reported cases has increased. Synonyms include solid and cystic tumor, solid and papillary epithelial neoplasm, papillary-cystic neoplasm, papillary cystic epithelial neoplasm, papillary-cystic tumor, and Franz tumor [1]. In 1996, the World Health Organization renamed this tumor solid pseudopapillary tumor for the international histologic classification of tumor of the exocrine pancreas [1]. Pathologic examination reveals that solid pseudopapillary tumor is usually a large, encapsulated mass composed of a mixture of cystic, solid, and hemorrhagic components. Both a capsule and intratumoral hemorrhage, are important clues to the diagnosis because these features are rarely found in other pancreatic neoplasms [1]. Deep pancreatic parenchymal invasion and angioinvasion are features of malignant SPT also called as solid pseudopapillary carcinoma [2]. Isolated splenic vein thrombosis due to adjacent pancreatic tail mass may result in sinistral portal hypertension, and is reported with pancreatic adenocarcinoma, cystadenoma and islet cell tumors [3]. Ours is the first reported case of SPT of pancreas with sinistral portal hypertension.

CASE HISTORY
A 23-year-old woman presented to the emergency department because of increasing upper abdominal pain and swelling for the last one month. She had a long history of vague upper abdominal discomfort and dyspepsia for the last 3 years. On examination there was a large epigastric mass on palpation. All laboratory parameters including serum amylase and liver function tests were normal. Carcinoembryogenic antigen (CEA), alpha-fetoprotein (AFP), and carbohydrate antigen (CA) 19-9 were all in the normal range. Ultrasound abdomen showed a large well-defined mixed solid-cystic mass inseparable from the pancreatic tail. Multidetector-row CT study of abdomen was performed next to characterise the mass. A large well-defined mass measuring 20cm x 15cm in size was seen closely abutting pancreatic body and tail. Non-contrast CT showed multiple areas of hyperdense (blood) attenuation within the mass, which showed mixed soft tissue and fluid attenuation. Dynamic contrast enhanced scan showed enhancing solid areas in the periphery of the mass and non-enhancing central cystic component. Venous phase of the study showed hypodense thrombus within the mid portion of the splenic vein with non-visualisation of the vein beyond this level. There was no plane of separation between the mass and the splenic vein compatible with splenic vein...
invasion. Spleen was mildly enlarged, there were no liver lesions or enlarged lymph nodes. Superior mesenteric vein and portal vein were normally outlined by the contrast. Multiple collateral vessels were seen extending from the splenic hilum and extending along gastric and colonic walls and communicating with patent SMV and PV. Prominent vessels were also seen in the wall of the stomach. Esophagogastroduodenoscopy showed varices in the fundus of stomach, there were no esophageal varices. Age and sex of the patient along with the CT imaging findings suggested a preoperative diagnosis of solid-cystic pancreatic mass possibly solid pseudopapillary tumor of pancreas with isolated splenic vein thrombosis resulting in sinistral portal hypertension. Percutaneous biopsy of the mass was performed under USG guidance for confirmation of the diagnosis. Microscopic examination revealed a solid, cystic and pseudopapillary growth pattern. Imaging and histopathologic findings were diagnostic of a pseudopapillary tumor of pancreas with splenic vein invasion causing sinistral portal hypertension.

**DISCUSSION**

Pancreatic tumors can be divided into three categories: exocrine tumors arising from acinar and ductal cells, endocrine tumors, and rare mesenchymal neoplasms. Pancreatic tumors are relatively less common than certain other malignancies, but they generally have an extremely poor prognosis. Solid pseudopapillary tumor (SPT) of the pancreas is a rare exocrine pancreatic tumor, which comprises only 1%-2% of all tumors of the pancreas, first described by Frantz in 1959[1]. In the last 10 years, there has been a steady increase in the number of these tumors. An apparent rise in the incidence is probably due to a better understanding of the pathology. The distribution of SPT of the pancreas is within the head in 32%, and body and/or tail of the pancreas in 68% of cases.

This uncommon, typically benign tumor is found mainly in young non-Caucasian women between the 2nd and 3rd decades of life. It seems to have a predilection for Asian and African-American women, although rare cases have been reported in children and men.

Patients with SPT of the pancreas are often clinically asymptomatic. They may 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 viscera [2,4]. Acute manifestations such as pancreatitis triggered by ischemia, distension or duct obstruction, or hemoperitoneum caused by the rupture of the tumor capsule are rare. There are usually no abnormalities in clinical laboratory tests (eg, serum amylase levels) or in pancreatic cancer markers (eg, CA19–9, carcinoembryonic antigen, -fetoprotein). The diagnosis is not uncommonly made incidentally at abdominal examination, ultrasonography (US), or CT performed for other reasons [2,4].

Although most SPTs exhibit benign behavior, malignant degeneration does occur in about 15% of cases evidencing as metastases or invasion of adjacent structures. The malignant pancreatic tumors were often older at presentation and had a male predilection. According to the WHO classification scheme, SPTs with clear criteria of malignancy (vascular and nerve sheath invasion or lymph node and liver metastases) are designated as solid-pseudopapillary carcinomas [2,4].

The splenic vein running along the posterosuperior aspect of pancreas is vulnerable to compression or infiltration to adjacent pancreatic lesions. This results in isolated splenic vein thrombosis (ISVT) and an unusual form of extrahepatic portal hypertension. Splenic vein thrombosis with normal superior mesenteric and portal veins results in splenoportal (gastroplenic) collaterals due to hypertension confined to the gastroplenic side of the portal venous circulation [3]. Esophageal varices are usually not seen in this form of EPHH, but may result when short gastric veins cannot cope with shunted splenic circulation or when the coronary vein drains into the splenic vein rather than the portal vein. Thus an extensive collateral circulation develops involving the segment of stomach drained by the short gastric vein This is called as left sided, sinistral and segmental portal hypertension which is characterized by gastric (fundal) varices, normal liver function and patent portal vein [5]. This carries a small risk of gastrointestinal bleeding (5%). Sinistral portal hypertension presenting as gastrointestinal bleeding has been reported with pancreatic adenocarcinoma, cystadenoma and islet cell tumor of pancreas [3,5]. Though our patient had documented sinistral portal hypertension, she did not report any incident of hematemesis or malena.
Figure 1

Fig. 1. Axial postcontrast CT image showing a large well-defined pancreatic body and tail mass having enhancing solid and nonenhancing cystic areas. Splenic vein posterior to the mass shows thrombus, portal vein is normal. Enlarged collateral vessels are seen in the splenic hilum and in the mesentry, compatible with sinistral portal hypertension.

Because of the low-grade malignant potential and good prognosis after complete resection of the tumor, it is important to make a correct diagnosis before operation. To the present, imaging studies or imaging-guided fine needle aspiration biopsy is the cardinal choice for preoperative diagnosis. Ultrasonographically, the tumor is well encapsulated, homogeneous or heterogeneous, and composed of solid echogenic and hypoechogenic cystic components. Adjacent major vein thrombosis, if present can be diagnosed with ultrasound doppler [6]. CT scans usually demonstrate a well-encapsulated and circumscribed retroperitoneal mass with various solid and cystic components owing to hemorrhagic degeneration.

Figure 2

Fig. 2. Axial non-contrast CT image showing a large well-defined mass in the region of pancreatic tail, with solid and cystic areas. More hyperdense areas (hemorrhage) are seen in the center of the solid-cystic mass.

Following administration of contrast material, enhanced solid areas are noted peripherally, whereas cystic spaces are centrally located [7]. MDCT has the advantage of fast scanning in different phases of contrast enhancement including dynamic arterial and delayed venous phases. This ensures visualisation of portal vein thrombus if present as in our case. Splenic vein thrombosis and or nonvisualization of splenic vein with splenoportal and gastrosplenic collaterals help in making the diagnosis of sinistral portal hypertension on CT in presence of patent portal vein and normal liver function.

Figure 3

Fig. 3. Axial post contrast CT image at the level of stomach shows enlarged vessels in the wall of the fundus of stomach compatible with gastric varices.

Esophagogastroduodenoscopy helps in confirming presence
of gastric varices due to left sided portal hypertension [3].

Because of its superior contrast resolution, MRI is better than CT for distinguishing certain tissue characteristics of SPT, such as hemorrhage, cystic degeneration, or the presence of a capsule. Typical MRI demonstrates a well-defined lesion with heterogeneous high or low signal intensity on T1-weighted images, and heterogeneous high signal intensity on T2-weighted images. On dynamic examination, the most common enhancement patterns were high signal intensity on T1-weighted imaging and slightly progressive heterogeneous peripheral contrast enhancement after administration of gadolinium. Compared with the lesion, the capsule is commonly seen as a hypo-dense rim on both T1- and T2-weighted images. After enhancement, it is characterized by early and more intense or identical enhancement [8]. Differential diagnosis should be made between microcystic adenoma, mucinous cystic neoplasm, nonfunctioning islet cell tumor, pancreatic carcinoma, pancreaticoblastoma, and calcified hemorrhagic pseudocyst [8]. Microcystic adenoma usually found in the elderly may be cystic or solid with irregular enhancement after contrast study. It tends to have a central scar with a patchy-like or sunburst-type pattern of calcification, which is characteristic but uncommon. Mucinous cystic neoplasm often seen in elderly women is round or oval, non-lobulated, smoothly demarcated, and protruded from the pancreas or surrounded by pancreatic tissue, pushing the nearby organs. Its attenuation is similar as water or as soft tissue if there are new hemorrhagic components. The cystic wall and wall nodules may be slightly enhanced on contrast images. Nonfunctioning islet cell tumor occurs in an elder age group than SPT and does not predominate in females. This is a solid tumor with more homogeneous appearance and obvious enhancement. Pancreaticoblastoma is a disease of childhood. Owing to its malignancy, hepatic metastasis is often seen. Pancreatic adenocarcinoma is also seen in older patients. It does not grow as large as SPT. Calcification is quite unusual and cystic or hemorrhagic degeneration is uncommon. The tumor may lead to dilation of the bile duct and pancreatic duct in early stage, and invade the vessels nearby. Enhanced adenocarcinoma is not common. The patients with calcified hemorrhagic pseudocyst usually have a history of pancreatitis and usually older than those with SPT. The calcification of pancreatic parenchyma is irregular or diffused or at the periphery of the pancreas. The cystic wall may be enhanced, but there should not be enhanced papillae or flocules [4,8].

In conclusion, in the appropriate clinical setting, CT and MR imaging features can be highly suggestive for the diagnosis of solid pseudopapillary tumor. This tumor should be considered when a well-marginated, large, encapsulated, solid and cystic mass with areas of hemorrhagic degeneration and progressive peripheral or slightly heterogeneous contrast enhancement, seen after gadolinium administration on dynamic examination, is detected in the pancreas of a young woman [8].

The pathologic diagnosis of SPT is mainly based on the well-defined solid and cystic structure and characteristic pseudopapillary features under the microscope [4,9]. On the cut surface, a variegated appearance is evident with variable combinations of solid hemorrhagic and cystic-necrotic areas. The microscopic features of SPT are solid areas which alternate with a pseudopapillary pattern composed of a fibrovascular stalk surrounded by several layers of epithelial cells. Immunohistochemical studies are frequently performed to confirm the diagnosis. SPT is typically positive for vimentin, antitrypsin and are negative for trypsin and chymotrypsin. They may also show focal immunoreactivity for neuron-specific enolase (NSE) and cytokeratin [4].

Because the tumor is usually surrounded by a pseudo-capsule and exhibits benign or low-grade malignancy, conservative resection with preservation of as much pancreatic tissue as possible is the treatment of choice [9,10]. When the tumor is well encapsulated and it is technically possible, simple enucleation of the neoplasm can be performed. Distal pancreatectomy with or without splenic preservation is feasible for tumors in the body or tail of the pancreas. For tumors of the pancreatic head, duodenopancreatectomy can be done, preferably with pyloric-preserving measures rather than with the Whipple technique [4,9] Localization of the tumor at the neck and body of the pancreas allows surgeons to resect the midportion of the pancreas, including the mass, and to preserve the rim of the head, the uncinery process, and the tail portion. Laparoscopic or laparoscopy-assisted surgery seems to be attractive in patients treated with enucleation or distal pancreatectomy. Extensive lymphatic dissection or more radical local approaches are not indicated [4,9,10].

Malignant SPT designated as a solid-pseudopapillary carcinoma occurs in 15% of adults and 13% of children with a significant increase in the elderly and male patients. According to the WHO classification system, the criteria for distinguishing malignant tumors form of the tumor “SPT carcinoma” comprise angioinvasion, perineural invasion,
and deep invasion of the surrounding pancreatic parenchyma and metastasis. As for metastases, the most common site is the liver; lymph node and peritoneal spread are rarely reported [4,9,10]. As for the treatment, splenectomy with distal pancreatectomy may be preferred in presence of splenic vein thrombosis with sinistral portal hypertension to eliminate the risk of bleeding from the gastric varices [3]. The presence of metastasis at the first diagnosis does not exclude primary surgery because clinical benefits are associated with tumor resection. Complete resection of both primary tumor and metastases is important. Specifically, when liver metastases are present, operative excision (lobectomy or enucleation) is used in most patients. Adjuvant therapy is used only in a small number of patients because of the high resectability of SPT [4,9,10].

In conclusion, imaging findings of solid-pseudopapillary tumor of the pancreas are characteristic, and the diagnosis can be made preoperatively with the combination of clinical features. CT also helps in the diagnosis of sinistral portal hypertension, which may be a rare occurrence with SPT. In such a case distal pancreatectomy with splenectomy should be performed to achieve complete tumor excision and to reduce the risk of gastrointestinal bleeding.

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