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Case Report

Organ-preserving enucleation in a case of solid pseudopapillary neoplasm of pancreas

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ABSTRACT

Solid pseudopapillary neoplasm (SPN) is a rare pancreatic tumor affecting mostly females in the second or third decade of life. These tumors have a low malignant potential, and hence, surgical resection remains the main treatment. Large tumors may present with compressive symptoms such as abdominal pain or vomiting. Once resected completely, SPNs usually have an excellent long-term prognosis. We report a 16-year-old girl who presented with vomiting and abdominal pain localized to the right upper quadrant. Imaging showed an encapsulated tumor arising from the pancreatic head which prompted us to perform organ-preserving enucleation. Histopathology confirmed the tumor as an SPN.

Keywords: Solid pseudopapillary neoplasm of pancreas, Solid pseudopapillary tumor, Pancreatic tumor, Frantz tumor

INTRODUCTION

Solid-pseudopapillary neoplasm (SPN) of the pancreas was first described by pathologist Frantz in 1959.^[1] It is considered a rare pathologic entity with low malignant potential, affecting mainly young females. It accounts for 0.13–2.7% of exocrine tumors of the pancreas.^[2] In 1996, the World Health Organization (WHO) defined it as “solid pseudopapillary tumor” of the pancreas.^[3] Before that, this tumor was known by different names such as “solid cystic tumor,” “papillary cystic tumor,” “papillary epithelial neoplasia,” “solid and papillary epithelial neoplasia,” “papillary epithelial tumor” and “Frantz’s tumor,” “solid and papillary tumor,” “solid-cystic papillary epithelial neoplasm,” and “benign or malignant papillary tumor of the pancreas.” Although they have low malignant potential, they can invade locally. However, recurrence is unusual following complete surgical resection.^[2] We report a case of an 18-year-old girl who presented with abdominal pain and vomiting and was diagnosed with a pancreatic SPN which was successfully excised.

CASE REPORT

An 18-year-old girl presented with multiple episodes of non-bilious vomiting for the past 20 days. She also complained of vague, dull aching upper abdominal pain since 2 weeks. On clinical examination,

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there was a palpable lump in the right upper abdomen. On ultrasonography, there was a large well-defined solid cystic mass in the pancreatic head region, measuring 11 × 9.8 × 7.7 cm, with peripheral calcific foci. A contrast-enhanced computed tomography (CT) scan of the abdomen revealed a well-demarcated ovoid solid cystic lesion in the retroperitoneum, 14 × 10.5 × 14.9 cm in size, closely related to the pancreatic head. The tumor compressed the distal common bile duct with mild upstream prominence of the intrahepatic biliary radicals. A CT-guided biopsy from the mass lesion was suggestive of a solid pseudopapillary neoplasm of the pancreas. On exploration, we found a large solid, cystic neoplasm arising from the pancreatic head, adherent to the middle colic vessels in the transverse mesocolon, and the inferior mesenteric vessels [Figure 1]. The mass was enucleated in toto, carefully safeguarding all the vessels, and the mesocolic defect was closed [Figure 2]. Post-operative recovery was uneventful. The histopathological examination confirmed the presence of cells in a solid and pseudopapillary pattern which were positive for β -catenin, CD10, CD56, synaptophysin, chromogranin, and CD99. The tumor margins were clear with no extracapsular or lymphovascular spread and no involvement of surrounding lymph nodes. The patient is doing well after 2 years of follow-up.

DISCUSSION

Solid-pseudopapillary neoplasm, is an epithelial tumor of pancreas with low malignant potential that predominantly occurs in young women. The usual age of presentation is the second or third decade of life with a ten-fold higher incidence in females. Around one-fourth of the cases are seen in children.^[2]

The WHO classifies SPNs into two types – SPN with uncertain potential for malignancy and a solid pseudopapillary carcinoma (SPC). The criteria that distinguishes potentially malignant tumors, classified as SPC are perineural invasion, angioinvasion, deep invasion into the surrounding tissue, and distant metastases.^[3,4] The origin of this tumor is unclear. Kosmahl *et al.* suggested it to be extra pancreatic genital ridge angle-related cell origin rather than a ductal epithelial, and neuroendocrine origin or multipotent primordial origin as thought earlier.^[5]

The clinical presentation of SPNs is with non-specific symptoms such as abdominal discomfort, mild abdominal pain, or asymptomatic palpable lump or incidentally diagnosed abdominal mass on imaging. Some patients may present with obstructive symptoms such as nausea, vomiting as in our case or jaundice.^[2,6] CT scan, ultrasonography, endoscopic ultrasound, and magnetic resonance imaging are the various imaging modalities used to diagnose SPN. The typical appearance on CT is that of a large encapsulated pancreatic mass with well-defined borders that contain areas of calcifications as well as intratumoral hemorrhage.^[7] The most common location of SPN is the tail of the pancreas,

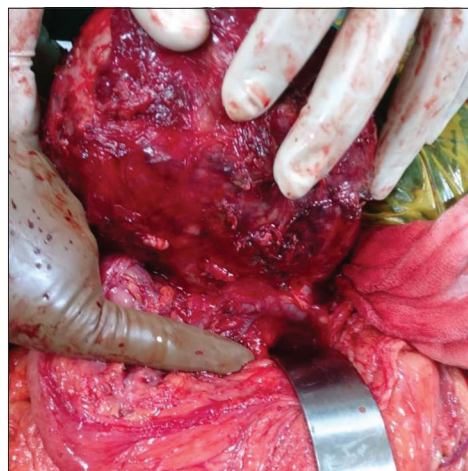


Figure 1: Pancreatic mass closely adhered to middle colic and inferior mesenteric veins.

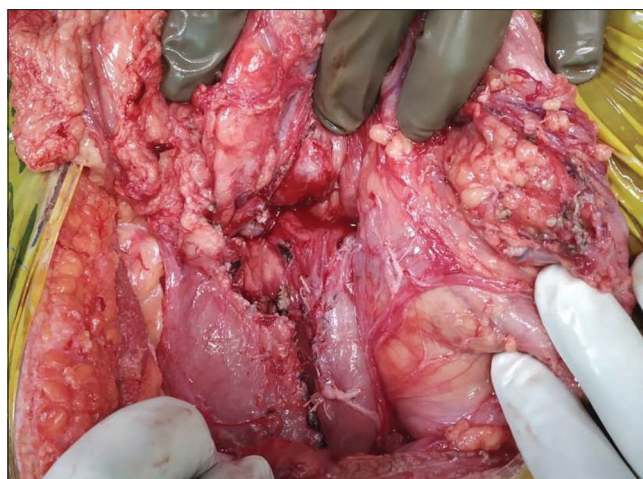


Figure 2: Pancreatic bed after enucleation of the mass.

followed by the head and the body of pancreas.^[2,7] About 85% of SPNs are localized to the pancreas while around 15% have already metastasized at presentation.^[7,8] Unusual presentations include multicentric tumors in the pancreas and extrapancreatic sites, such as the mesocolon, retroperitoneum, omentum, liver, and duodenum, possibly representing synchronous tumor spread. Hematological investigations and tumor markers are usually unremarkable.^[9,10]

Complete surgical resection is the treatment of choice. In view of its low malignant potential, organ preserving surgery is preferable. Even though SPNs may present as large sized masses due to delayed diagnosis, tumor size is not a predictor of resectability. The various surgical options include Whipple's pancreatoduodenectomy, distal pancreatectomy (with or without splenectomy), middle pancreatectomy, or enucleation depending on the size, location, and invasion of adjacent vessels and organs.^[2,9,10] Follow-up becomes essential

for early detection of any local recurrence or metastasis, that may occur in the liver, regional lymph nodes, mesentery, omentum, or peritoneum.

Patients with SPN have a good prognosis even in the setting of local recurrence and/or metastasis. The overall 5–8 year survival rate of SPN patients is up to 95%.^[2,10] Positive tumor margins do not affect the final outcome of patients. Hence, enucleation, rather than radical resection, is advisable whenever feasible.^[6,9,10] Although the role of chemotherapy and radiotherapy is unclear, it has been used in some cases with aggressive disease.^[11,12]

CONCLUSION

SPN is a rare pancreatic neoplasm that typically affects young females with vague symptoms. Appearance on cross-sectional imaging is characteristic and a pancreas-conserving surgical approach is advisable as these patients have an excellent long-term survival, even with positive resection margins, local recurrence, and/or metastasis.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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