Case Report

Solid pseudopapillary neoplasm of pancreas

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ABSTRACT

Solid pseudopapillary neoplasm (SPN) of the pancreas is a rare neoplasm of low malignant potential that mostly affect young women. These tumors are of unclear pathogenesis, are slow growing, and can become considerably large before causing symptoms. Complete resection is curative in most cases. Metastases are frequently amenable to resection. Favorable prognosis with long-term survival has been shown even in patients with metastatic disease. We report a case of SPN of pancreas in a 15-year-old female.

Key words: Pancreas, pseudopapillary, solid

INTRODUCTION

Solid pseudopapillary neoplasms (SPN) of the pancreas are rare tumors and predominantly occur in young women in the second and third decades of life, it is usually a benign tumor with low-grade malignant potential.[1-3]

CASE REPORT

A 15-year-old female had preoperative complain of a single episode of severe pain abdomen. Ultrasonography of abdomen revealed large hypoechoic ovoid space occupying lesion (SOL) in pancreatic body and tail region (10.2 cm × 7.2 cm) - Pancreatic mass associated with moderate ascites. Contrast-enhanced computed tomography scan of whole abdomen identified a fairly well-defined mildly heterogeneously enhancing lesion in the left hypochondriac region (measures 9 cm × 7.8 cm) superiorly reaching up to anterior abdominal wall, inferiorly compressing pancreas, bowel loops displaced medially and laterally. No calcification and vascular extension is seen. Fat planes with adjacent structures are maintained - retroperitoneal SOL is likely possibility. Moderate ascites was present [Figure 1].

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A diagnosis of solid pseudopapillary neoplasm (SPN) of the pancreas was suspected. Exploratory laparotomy was done which revealed a retroperitoneal mass arising from body and tail of pancreas without any features suggestive of rupture or hemorrhage. Excision of mass is done. Histopathology of tissue from pancreatic SOL demonstrated it to be an SPN [Figure 2]. Immunohistochemistry further showed it reactive for vimentin, CD10, CD56, and synaptophysin confirming it to be an SPN of the pancreas.[1] Ascitic fluid was sent for cytological examination that came out to be negative for malignant cells.

DISCUSSION

SPN of the pancreas is an uncommon and enigmatic pancreatic neoplasm first described by Frantz in 1959.[2,3] This lesion usually has a low malignant potential and tends to occur primarily in young women. Metastases, with an incidence of 15%, most of which are hepatic, and local recurrence have rarely been reported in the long-term follow-up of patients with SPN.[2-4] Over time, different names were used to describe this tumor, including Frantz or Hamoudi tumor; solid and papillary tumor; papillary cystic tumor; solid-cystic tumor; solid, cystic, and papillary epithelial neoplasm; and adenocarcinoma of the pancreas in childhood.[3-6] The origin of solid pseudopapillary tumors has not yet been clarified. Many investigators favor the

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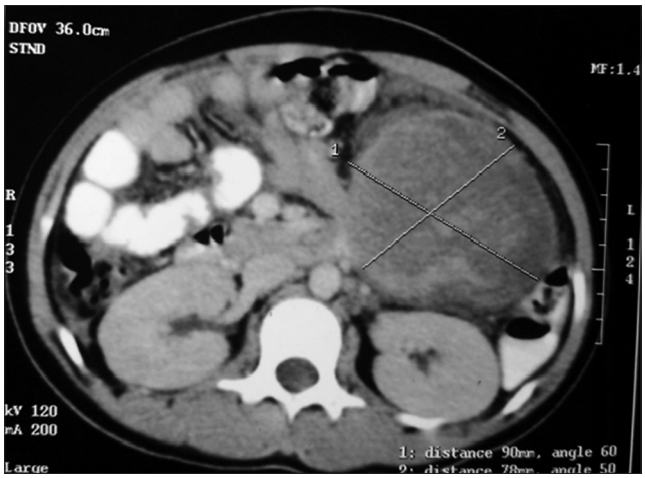

Figure 1: Contrast-enhanced computed tomography whole abdomen showing well-defined mildly heterogeneously enhancing lesion in left hypochondriac region

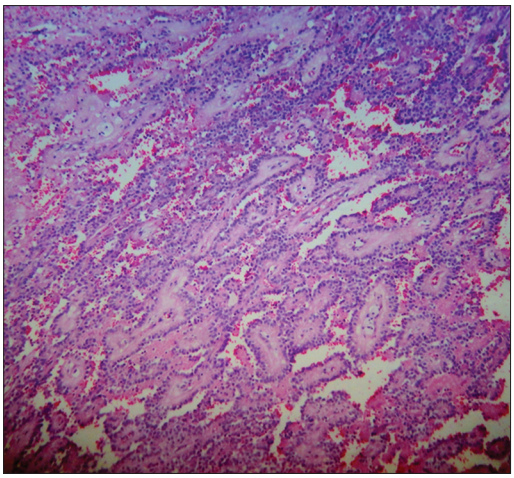

Figure 2: Higher power view of the mass showing pseudopapillae lined by several layers of epithelial cells having ovoid nuclei with inconspicuous nucleoli along with a population of clear to foamy cells

theory that SPNs originate from multipotent primordial cells, whereas others suggest an extrapancreatic origin from genital ridge angle-related cells.[4-7] Seven-hundred eighteen patients with SPNs have been reviewed in the English literature; female dominance has been found with a ratio of 10:1, and the mean age at presentation is 22 years.[3,8] The majorities of these tumors are located in the pancreatic body and tail.[9] Most patients present with unclear clinical features. Examples of symptoms include increased abdominal girth, abdominal discomfort, abdominal pain, poor appetite, and nausea, which can be caused by tumor compression on the stomach and other adjacent organs.[5,7] Complete resection of local disease is curative. Rarely vascular invasion of the superior mesenteric artery or portal vein are encountered and may limit resectability. The surgical bypass may be the only feasible option in patients with large tumors where the risks of attempting resection or debulking may be associated with overwhelming morbidity. Even patients with residual disease or metastases have been reported to have long-term survival following surgical treatment. Overall 5-year survival is as high as 97% in patients undergoing surgical resection.[5] Very few reports of the use of chemotherapy or radiotherapy for these tumors exist with only limited response.[10,11]

CONCLUSION

A high index of suspicion is necessary to diagnose SPN. Because of the indolent nature of these tumors and the low malignant potential, aggressive attempts at complete surgical resection are warranted. Large tumors are usually resectable and size does not predict the outcome. Patients have an excellent prognosis.

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Conflicts of interest

There are no conflicts of interest.

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