

Solid Pseudopapillary Tumor of the Pancreas

Abstract

Solid pseudopapillary neoplasia of the pancreas is a rare tumor. It accounts for <1%–2% of exocrine pancreatic tumors. This relatively indolent tumor has the propensity to affect young women. We herein report a case of solid pseudopapillary tumor of the pancreas in a 24-year-old woman; for which curative distal pancreatectomy with splenectomy was performed. Postoperative period was uneventful, and during a follow-up of 2 years, she remains asymptomatic.

Keywords: Frantz tumor; pancreatic tumor; solid pseudo-papillary neoplasm

Introduction

Solid pseudopapillary tumor (SPT) of the pancreas is a relatively rare tumor. It accounts for <1%–2% of the exocrine pancreatic tumors.^[1] The SPT predominantly affects young women.^[2] This tumor typically originates in the tail of the pancreas. In spite of their low malignant potential, they can invade the nearby structure or metastasize to distant organs. Local recurrences are unusual after complete resection and their long-term outcomes are favorable. Herein, we report a case of a 24-years woman with SPT involving the pancreatic body and tail managed satisfactorily after complete curative resection.

Case Report

A 24-year-old female presented with a lump in the left upper abdomen for the past 4 months. There were no other complaints. Her family and history were also not significant. Abdominal examination revealed a firm, nontender, well-defined lump of around 12 cm × 12 cm, occupying the left hypochondrium, left lumbar, and umbilical regions. The hematological and biochemical parameters were unremarkable. Abdominal ultrasound revealed a solid heterogenous mass lesion of 12 cm × 10 cm × 11 cm in the left upper abdomen, probably arising from the pancreas. Contrast-enhanced computed tomography (CT) of the abdomen showed a well-defined heterogenous encapsulated

mass of size 12.4 cm × 10.5 cm × 11 cm originating from the body and tail of the pancreas [Figure 1]. The small bowel was displaced medially, and the left renal vein was stretched by the tumor. Surrounding fat planes were well maintained. There was no evidence of distant metastasis.

Thus for curative as well as for diagnostic intent exploratory laparotomy with distal pancreatectomy along with splenectomy was performed. Spleen could not be preserved as the splenic artery and vein were embedded on the superior surface of the tumor. Intra-operatively there was a well-encapsulated tumor involving the body and tail of the pancreas reaching up to the anterior abdominal wall between the stomach and transverse colon [Figure 2]. There was no evidence of peritoneal and other visceral metastases. The resected pancreatic stump was over-sewn and a drain placed in the splenic fossa. Postoperative period was uneventful, and the patient discharged on the 6th day. During a follow-up of 2 years, the patient remains asymptomatic.

Histopathological examination revealed a tumor encapsulated by a thick capsule, the tumor was comprised cells arranged in pseudopapillary, pseudomicrocystic, and solid patterns [Figure 3]. However, the most striking pattern was the pseudopapillary pattern formed due to the separation of vascular cores along with adherent cells from surrounding structures as a result of degeneration. This pattern was especially prominent toward the periphery

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Figure 1: Contrast-enhanced computed tomography abdomen showing the pancreatic tumour with adjacent pancreatic tissue

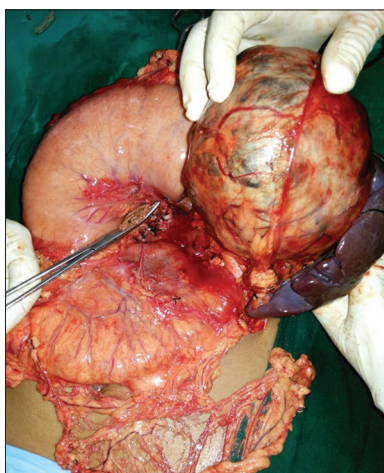


Figure 2: Intra-operative picture of the tumour with spleen

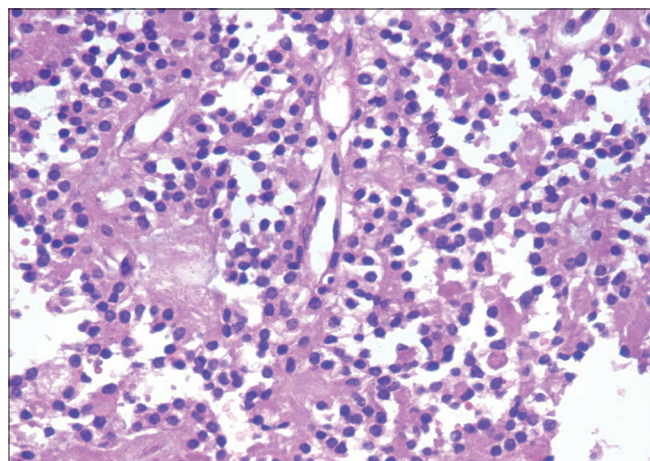


Figure 3: Pseudo-papillary pattern with presence of fibrovascular cores (H and E, $\times 400$)

of the tumor; there were no areas of necrosis, mitosis, pleomorphism or vascular, and capsular involvement. The final diagnosis of solid pseudopapillary neoplasm of the pancreas was rendered.

Discussion

One of the earliest descriptions of this rare entity was reported in 1959 when Virginia Frantz described a “papillary cystic tumor of the pancreas”^[3] and hence, it is also known as FRANTZ tumor. Until its inclusion as “SPT” of the pancreas in the World Health Organization (WHO) classification of pancreatic tumors in 1996,^[4] this tumor had been described using different nomenclature in the literature; such as “papillary epithelial neoplasm of the pancreas,” “solid and cystic tumor of the pancreas,” “adenocarcinoma of the pancreas of childhood,” “papillary-cystic tumor”, and “solid and papillary epithelial neoplasm;”. In the current WHO classification, SPT is defined as a low-grade malignant neoplasm of the exocrine pancreas.^[5] In the light of established nomenclature of this relatively rare tumor, two-thirds of the 700 reported cases have been described in the past 10 years.^[2]

The SPT has a tendency to affect young women.^[2] Men are rarely affected by this tumor.^[6] The clinical presentation of the tumor is usually nonspecific. Abdominal discomfort or vague pain is the most common initial symptom.^[1] It can present as gradually enlarging abdominal lump and associated compressive symptoms induced by the large tumor. In the series by Yu *et al.*, where 553 patients with SPT were reviewed retrospectively, nearly one-third were asymptomatic (31.7%), 37.6% presented with abdominal pain, 35.9% with lump and 32.8% with abdominal discomfort.^[7] Therefore, it is not uncommon that the tumor is detected only when it has reached a remarkable size.

The etiology and differentiation status of SPT remains enigmatic owing to its uncertain histogenesis.^[8] Although the liver and peritoneum have been found to be affected by metastases, these are only rarely seen.^[8] Malignant transformation of this tumor is usually characterized by angioinvasion, perineural invasion, and deep invasion of the surrounding pancreatic parenchyma.^[9]

Preoperative diagnosis of these lesions is not always possible; however, image-guided fine-needle aspiration cytology may be diagnostic in some cases. Radiological examination, particularly CT scan is helpful in determining its site, size, relation to surrounding structures, and metastasis. Complete surgical excision is almost curative for tumors limited to the pancreas. For tumors involving the body and tail of the pancreas, distal pancreatectomy with or without splenectomy is advocated.^[9] As these tumors are usually encapsulated and have a relatively indolent behavior more radical surgery or lymphadenectomy is not indicated.^[10] Even in the presence of metastatic disease, debulking of the tumors seems to have a beneficial effect. Chemotherapy and radiotherapy can be used in advanced cases.

Conclusion

To conclude, SPT is a relatively rare tumor with low malignant potential and unclear histogenesis, which mainly

affects young women without significant symptoms and has a good prognosis after complete surgical resection.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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