A 35 Year Old Female Presented with Solid Pseudopapillary Tumor of the Pancreas; Treated Successfully by Whipple's Procedure

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Abstract
Solid pseudopapillary tumor of the pancreas is one of the rarest forms of pancreatic neoplasm. It typically affects young females in their second or third decade of life. Although malignant, the malignant potential is low and aggressive surgical resection ensures a cure in a majority of patients. Here we present a case of a 35 year old female presented with abdominal pain and lump in the epigastric and right lumber region measuring about 7×5 cm having irregular surface, firm in consistency and does not move with respiration. CT scan of abdomen showed solid-cystic lesion measuring about 7×5 cm with heterogeneous intensity in the pancreatic head region. She underwent Whipple's procedure with en-bloc resection of tumor. Histopathology report showed solid pseudopapillary tumor. Her post-operative period was eventful and she was referred to oncology department for better management.

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Introduction

Solid pseudopapillary tumor (SPT) is a rare low-grade malignant cystic exocrine neoplasm of the pancreas of papillary architecture with a reported incidence of 0.13%–2.7% of all pancreatic tumors and is pathologically distinctive from other types of pancreatic tumors.[1]

The tumor has been referred to with multiple different names, including SPT of the pancreas, SPN, solid pseudopapillary epithelial neoplasm (SPEN), papillary cystic neoplasm of the pancreas, Hamoudi tumor, and Gruber-Frantz tumor (or just Frantz tumor).[2] It occurs predominantly in young women around second and third decades of age.[3] The predominant female preponderance has been hypothesized to be due to the close proximity of the primordial pancreatic cells to the ovarian ridge in the embryonic phase.[3] Although this tumor may occur anywhere within the pancreas, the pancreatic tail is the most common site of origin.[3] The entity was first described in 1959 by pathologist Virginia Kneeland Frantz and in 1996 reclassified by the World Health Organization.[4]

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In general, the diagnosis of SPT depends on the clinical and imaging features. Clinically, a slowly growing mass with or without vague abdominal pain may be the only patient's complaint and the blood tests are usually normal. These tumors may also be incidental findings during complementary studies, such as ultrasound and/or CT done for various reasons. The sonographic features of septations in the cystic portion are possibly due to prominent papillae projecting into the space of cystic degeneration.[5] CT shows a well-demarcated large encapsulated pancreatic mass.[6] The architecture of the mass varies from solid to mixed solid and cystic Calcifications and enhanced solid areas may be present at the periphery of the mass.[6]

Histologically, it shows fibrous septations and areas of degeneration forming white clefts; giving the tumor its pseudopapillary appearance.[7] Peripheral or central calcifications may be seen in solid areas.[7]

SPT of the pancreas is treated surgically and complete resection of the mass is usually curative. Whipple's procedure or distal pancreatectomy is performed most commonly, with en bloc resection of involved adjacent organs when indicated.[8] Complete margin-negative resection (R0) is associated with a long-term disease-free survival. About 10%–15% patients have metastases at the time of diagnosis or develop metastases at some point in the future.[9] Here we present a case of a 35 year old female presented with Solid Pseudopapillary Tumor in the head of pancreas in the Department of Surgery of Shaheed Suhrawardy Medical College Hospital which was treated successfully by Whipple's procedure.

Case Summary

A 35 year old female presented to the surgery department of Shaheed Suhrawardy Medical College hospital with the complaints of a slowly progressive lump occupying in the epigastric and right hypochondrium region for 3 months. She also mentioned about recurrent episodes of dull aching pain in the epigastric region which was gradual in onset, mild to moderate, intermittent, persisted for two to three hours with no specific aggravating and relieving factors. There were no history of jaundice, vomiting, hematemesis, melena, anorexia and weight loss. On examination her vitals were normal. Per abdominal examination revealed a non-tender mass measuring about 7×5 cm with irregular surface, firm in consistency and does not move with respiration. Her blood reports including complete blood count, renal and liver function tests, Chest x-ray and ECG all were within normal limits. Abdominal Ultrasound detected a mass in the pancreatic head region. (Fig-1) CT scan of abdomen showed solid cystic lesion measuring about 7×5 cm with heterogeneous intensity in the pancreatic head region. (Fig-2) Her CA 19-9 level was also normal. She underwent Whipple's procedure for complete resection of the tumor. (Fig-3) The specimen was send for histopathology and the report showed Solid Pseudopapillary Tumor. (Fig-4) Her post-operative period was uneventful and she was referred to oncology department for further management.

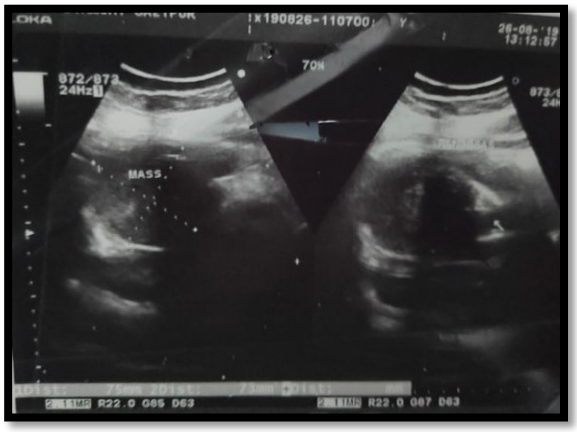

Figure 1. USG findings of abdomen showing the mass in the pancreatic head region

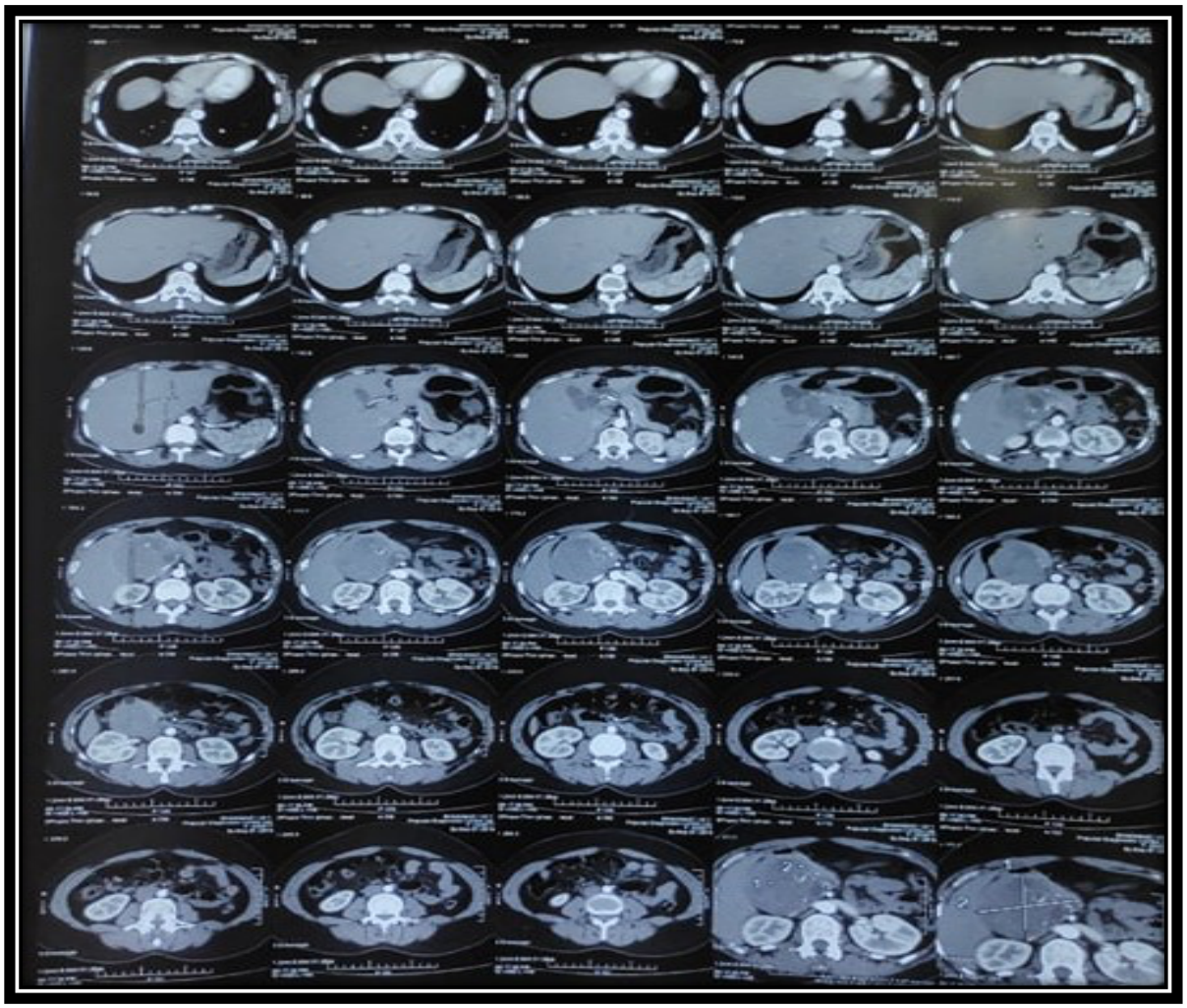

Figure 2. CT scan of abdomen showing a mass in the pancreatic head region

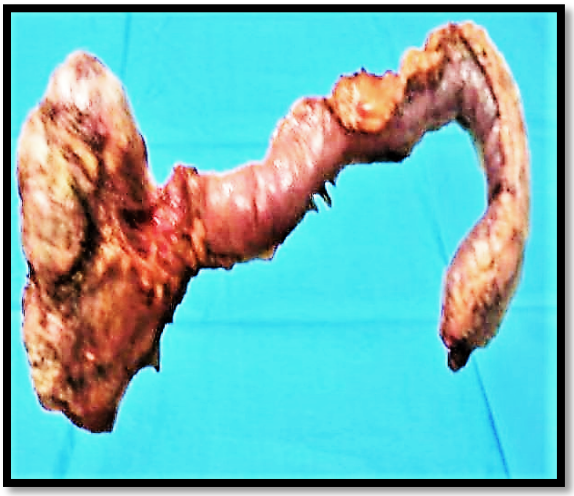

Figure 3. Resected specimen of the tumor

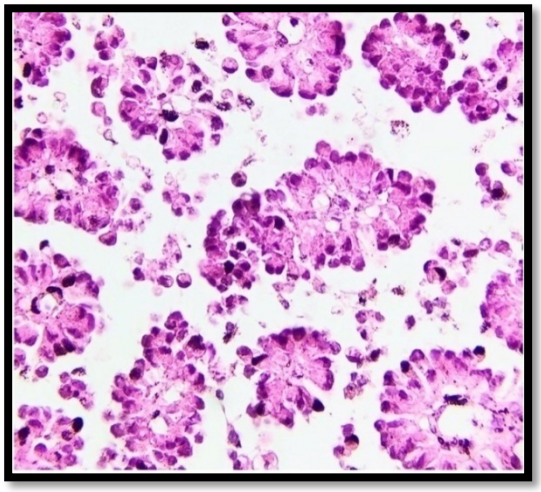

Figure 4. Microscopic appearance of SPT with characteristic pseudo-papillary growth pattern

Discussion

Solid papillary tumor (SPT) is an uncommon exocrine pancreatic neoplasm.[10] It is tumor characterized by its occurrence in young women and its limited malignant potential.[11] Our patient presented at 35 years of age which coincides age of presentation as reported by other studies.

Clinical presentation of SPTs is not specific. There was a considerable number of patients with no symptoms in whom SPT was found accidentally.[12] Patients mostly present with abdominal pain and occasionally mass in the abdomen which was also the presentation of our case.[12]

Usually, the serum tumor markers, such as alfa-fetoprotein, carcinoembryonic antigen, and CA 19.9 resides within normal ranges.[13] In our case, preoperative laboratory values and serum tumor markers were within normal range.

CT is the primary modality used to evaluate SPT. The tumor typically appears as a mixed-density lesion with solid component peripherally and cystic component more centrally.[14] Our patient had similar findings.

Papavramidis et al. reported the locations of SPT as the tail (35.9%), the head (34%), the body (14.8%) and the neck of the pancreas (1.01%), respectively.[15] YU et al.found that most of the tumors were distributed in the pancreatic head (39.8%), with the tail (24.1%), body and tail (19.5%), body (11.2%), and neck (3.6%).[16] In our study the lesion was present in pancreatic head region.

SPT is generally considered as a tumor with low malignant potential. Although resection of the tumor provides a 5-year survival rate more than 95%, local recurrence or distant metastases can occur.[17] Although enucleation is an adequate treatment in the majority of patients with SPT, we performed Whipple's procedure rather than enucleation. In different studies it has showed that Whipple's procedure has a better outcome in terms of recurrence than enucleation.[18]

Conclusion

SPT is a rare neoplasm which presents with vague symptoms which may have given rise diagnostic delay. So a high index of suspicion with accurate investigations can help to diagnose the disease timely. Whipple's procedure has a better outcome as it prevents further recurrence when the neoplasm has involved the head. So timely diagnosis and proper surgical intervention can provide a better prognosis to the patients suffering of SPT.

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