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Case Report

Successful Whipple Operation for a Huge Pancreatic Tumor in a Young Woman

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Abstract **Solid pseudo-papillary tumor (SSPT) of the pancreas is a rare pancreatic tumor. It is distinct from other pancreatic tumors in its age and sex distribution. Following aggressive resection there is a favorable prognosis, with most patients going on to cure. This entity is widely reported in the surgical and pathological literature. We present a case of huge SSPT of the pancreas in a young woman who underwent successful Whipple's operation. She has been followed for two years without any evidence of recurrence. We also present a review of the literature on SSPT.**

Keywords: **Whipple operation, solid pseudo-papillary of the pancreas**

Introduction

Papillary and cystic tumor of the pancreas usually occurs in young women and behaves in a favorable manner in spite of histologic findings of apparent malignancy[1-4]. Frantz initially reported this tumor in 1959[5] and called it a "papillary tumor of the pancreas". Although the tumor sometimes attains enormous size, most can be resected. Surgical resection offers an excellent chance for long-term survival[6]. However surgical resection for a huge pancreatic tumor is still a challenge.

Case Report

A 19-year-old woman was found to have a huge right upper quadrant mass with severe abdominal pain six hours before admission. The mass has gradually enlarged in size for ten years without symptoms. On arrival, she had right upper quadrant mass with mild tenderness. All laboratory data were within the normal range except mild anemia. The abdominal CT scan showed a huge, well-defined arterial enhancing, mixed cystic-solid mass with internal calcification at the head of pancreas, measured about $10 \times 9 \times 14.2$ cm^3. Some foci within the solid portion at the superior aspect were hyperdense (50-60HU), probably representing internal hemorrhage. The duodenum was shifted laterally due to pressure effect of the mass. There was dilatation of the pancreatic duct, and focal areas of calcification at the most inferior aspect of the tumor (Figure 1).

At exploratory laparotomy, we found a $14 \times 9 \times 8$ cm^3 hard mass at the head of the pancreas, with adherence to the superior mesenteric vein. The mass caused widening of C-loop, with pressure effect to the

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duodenum. The pancreas at the body and tail was normal with mild dilatation of the pancreatic duct. There was no evidence of distant metastasis or invasion to the surrounding organs. Pylorus preserving pancreaticoduodenectomy (Whipple's operation) was performed without difficulty (Figure2). She was discharged seven days after surgery with uneventful recovery. The patient has been followed up for 12 months without evidence of recurrence.

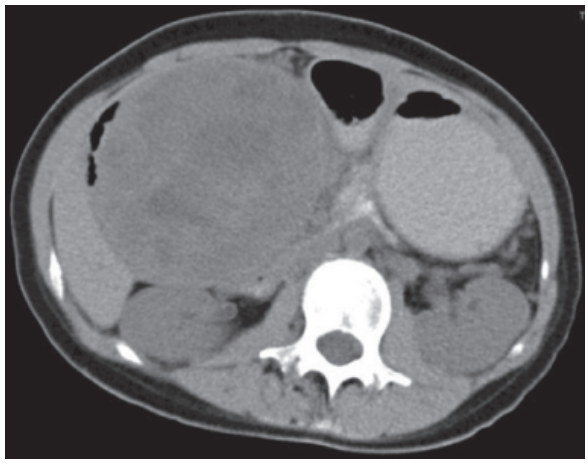

Figure 1 The abdominal CT scan showing a huge, well-defined arterial enhancing, mixed cystic-solid mass with internal calcification at the head of pancreas, measuring about $10 \times 9 \times 14.2$ cm^3.

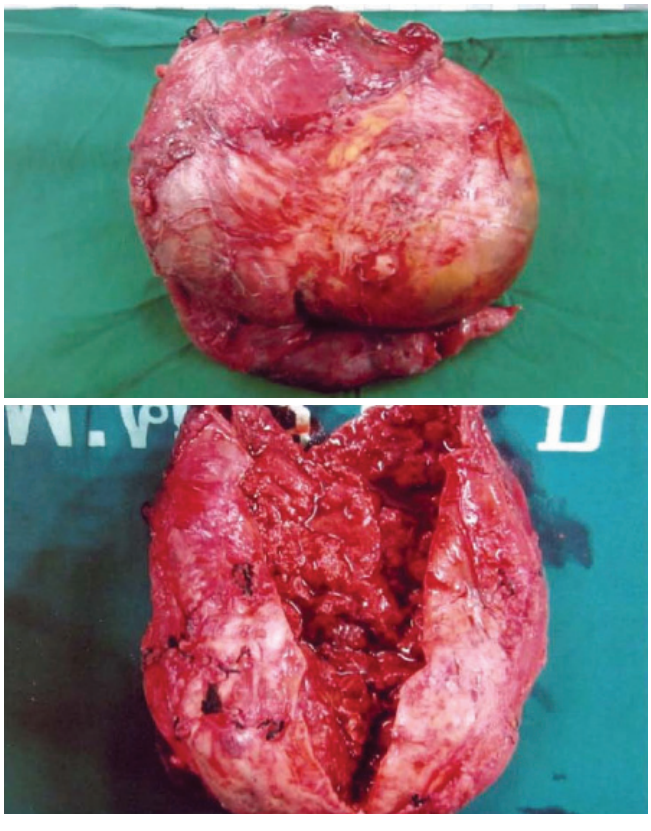

Figure 2 Showing the resected specimen - a well encapsulated soft tissue mass with internal papillary growth

Histopathological Findings

The specimen measured $14 \times 9 \times 8$ cm^3 and had a well-circumscribed solid-cystic pancreatic mass (Figure 3). Light microscopy showed solid, cystic and papillary regions. The solid portions were composed of fairly uniform, small to medium-sized cells with moderate amounts of eosinophillic cytoplasm. The nuclei were round to ovoid, with fine chromatin and prominent nuclear folds. Nucleoli were small and

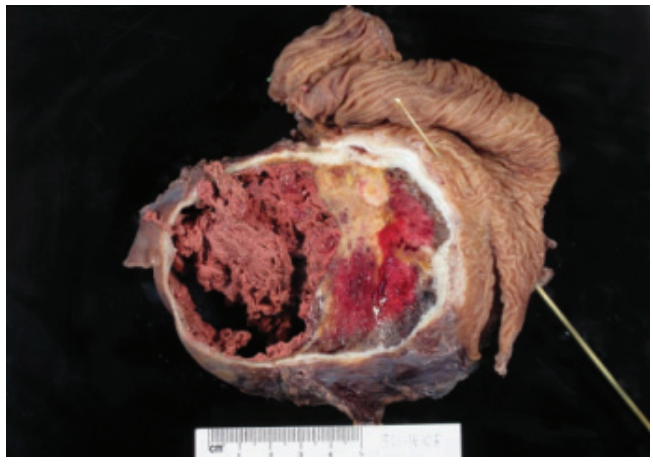

Figure 3 The specimen measured 14 x 9 x 8 cm^3 and had a well-circumscribed solid-cystic pancreatic mass. The cut section revealed solid, cystic and hemorrhagic areas.

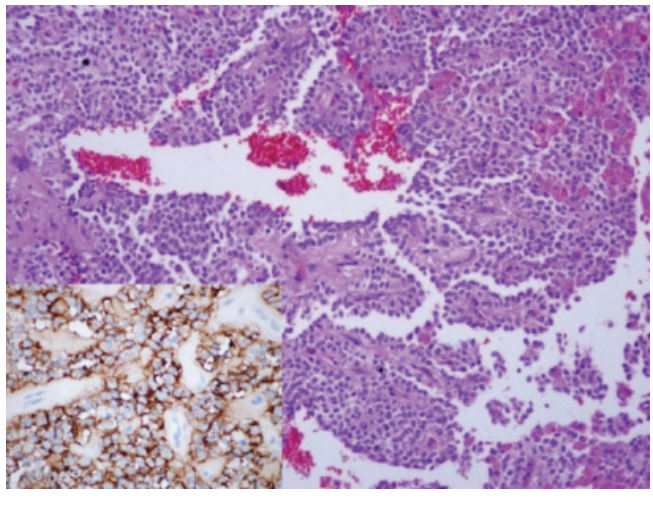

Figure 4 Immunostaining demonstrated that the neoplastic cells were positive for vimentin, CD56, progesterone receptor (PR), and weakly positive for synaptophysin, but negative for AE1/AE3 and chromogranin A.

inconspicuous. No mitosis was evident. The papillae were composed of one or more layers of identical cells arranged around delicate fibrovascular stalks that appeared as pseudo cores when cut in cross-section. The cystic areas were filled with necrotic debris and many histiocytes. The results of immunostaining support the diagnosis of solid pseudopapillary neoplasm (Figure 4).

Discussion

Frantz first described solid pseudo-papillary cystic tumor (SPPT) of the pancreas in 1959[8,9]. The precise incidence of pancreatic SPPT is not known as it is rare. A review of the English language literature showed that altogether 452 cases have been reported[8-11]. Pancreatic SPPTs are tumors of adolescents and young women. Only thirty cases were found in men[12]. In addition, 66 of 452 tumors (15%) were malignant, showing evidence of metastases or invasion of adjacent structures. Pancreatic SPPTs were evenly distributed throughout the pancreas. The tumors were usually large, encapsulated, and with cystic areas.

Plain films of the abdomen may show a calcified mass in the upper abdomen[1]. Abdominal ultrasonography and CT scan examinations appear to be the most effective methods for diagnosis. On abdominal ultrasonography the pancreatic mass is heterogeneous, with cystic components. Abdominal CT scan usually shows a well-demarcated heterogenous mass with calcification. Angiography will show few or no vessel supplying the tumor. The features shown by the MRI in the present case were unusual and seldom described in the literature[7].

The main differential diagnosis on microscopic examination was pancreatic endocrine tumor. All pancreatic endocrine tumors were positive for one of six markers, listed in Figure 4[8,13]. Thus negativity to these markers and neurosecretory granules together with the papillary and solid histological pattern and predominant occurrence in young females are features that help differentiate pancreatic SPPTs from pancreatic endocrine tumors[13].

SPPT is usually diagnosed after surgical resection[12,13], but fine-needle aspiration seems to be an alternative method for preoperative diagnosis[14-18]. Most SPPTs are benign or behave like very low grade malignancy. Distant metastases to the liver, lung, and skin occur predominantly in older women and carry a higher death rate[2]. Complete resection is the treatment of choice. However, chemotherapy[17], radiotherapy[13], and hormone therapy have been reported to give good results.

Because the patient is usually young, the operation should include recent modifications of pancreatic surgery which reduce postoperative morbidity, in particular, pylorus preservation in pancreaticoduodenectomy[18], to avoid dumping syndromes and diarrhea associated with gastrectomy, enabling the patient to gain weight to levels before illness[12,19].

Conclusion

Solid pseudo-papillary tumor (SPPT) of the pancreas is rare. They usually occur in young females, but they may also present in older women. Preoperative diagnosis and surgical resection are very important, because these neoplasms are potentially curable.

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