

CASE REPORT

Liver Metastasis Four Years after Whipple's Resection for Solid-Pseudopapillary Tumor of the Pancreas

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ABSTRACT

Context Solid-pseudopapillary tumor of the pancreas is a rare tumor which usually affects young females in their second and third decade of life. Metastasis is very rare after a resection of curative intent.

Case report We report a case of a 65-year-old white female who presented with metastasis to the liver four years after Whipple's resection for a solid-pseudopapillary tumor of the pancreas.

Conclusions Solid-pseudopapillary tumors of the pancreas can present with metastasis a long time after resection of the primary tumor. Long term close follow up of these patients should be done. The survival rate even after liver metastasis is good.

INTRODUCTION

Solid-pseudopapillary tumors of the pancreas are rare tumors of the pancreas of unknown cell origin and low malignant potential. It was first described by Frantz in 1959 [1] and was called by various names by different authors, but the present consensus name is solid-pseudopapillary tumor based on the characteristic histopathological appearance [2]. We are reporting a case of this condition which initially presented in the 6th decade of

life and metastasized to the liver and presented again with symptoms 8 years after Whipple's resection of the pancreas.

CASE REPORT

A 57-year-old white female patient presented to the emergency room in 1998 with a 3 day history of epigastric pain and vomiting. An ultrasound of the abdomen showed multiple gallbladder calculi with possible cholecystitis and a tumor lesion in the head of the pancreas. A computerized tomography scan (CT) of the abdomen showed a mass in the head of pancreas which measured 5x4 cm (Figure 1). The liver was normal. The mass displaced the right renal vein, inferior vena cava, superior mesenteric vein and artery, and portal vein without invading them. The

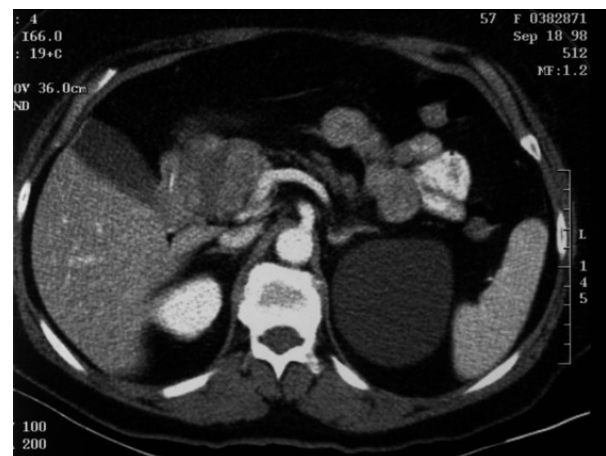


Figure 1. CT scan of abdomen from 1998 showing 5x4 cm solid and cystic tumor in the head of pancreas.

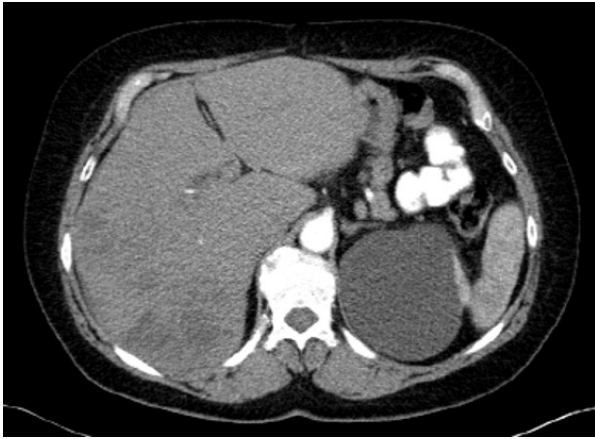


Figure 2. CT scan of abdomen from year 2002 showing small liver lesions.

patient underwent laparoscopic cholecystectomy with a biopsy of the pancreatic lesion. The biopsy of the tumor showed solid, cellular, hyper-vascular regions without gland formation and degenerative pseudopapillae consistent with the diagnosis of solid-pseudopapillary neoplasm of the pancreas. The patient underwent Whipple's procedure and the resected specimen confirmed solid-pseudopapillary tumor of the pancreas with invasion of the duodenum and perineural area. The resection margins were free of tumor and there was no lymphnode involvement. The patient was asymptomatic after the above procedure for 3 years and yearly CT scans of the abdomen for the first two post operative years were normal. The patient had a CT scan again in 2002 (Figure 2) that showed small lesions which might have been early metastasis to the liver. The patient refused liver biopsy at that time.



Figure 3. CT scan of abdomen from year 2006 showing liver metastasis.

The above patient, now 65-years-old, presented to the hospital in October 2006 with abdominal pain, anorexia and weight loss. On examination, she had an abdominal mass in the right upper quadrant suggestive of an enlarged liver and her laboratory tests revealed elevated liver enzymes. A CT scan of the abdomen showed a markedly enlarged liver containing multiple varying sizes of lesions predominantly in the right lobe of the liver (Figures 3 and 4). Some of the lesions demonstrated central low attenuation areas suggestive of central necrosis. Multiple large heterogeneous mesenteric lymphadenopathy was also noted within the mid abdomen (Figure 5). The largest lymph-node measured 3x3 cm. A liver biopsy was performed which revealed infiltration of the liver by metastatic solid-pseudopapillary tumor of the pancreas. The tumor consisted of solid, cystic and pseudopapillary areas. The cells adjacent to the blood vessels were attached to the stroma and to one another but those which were further away were detached from one another

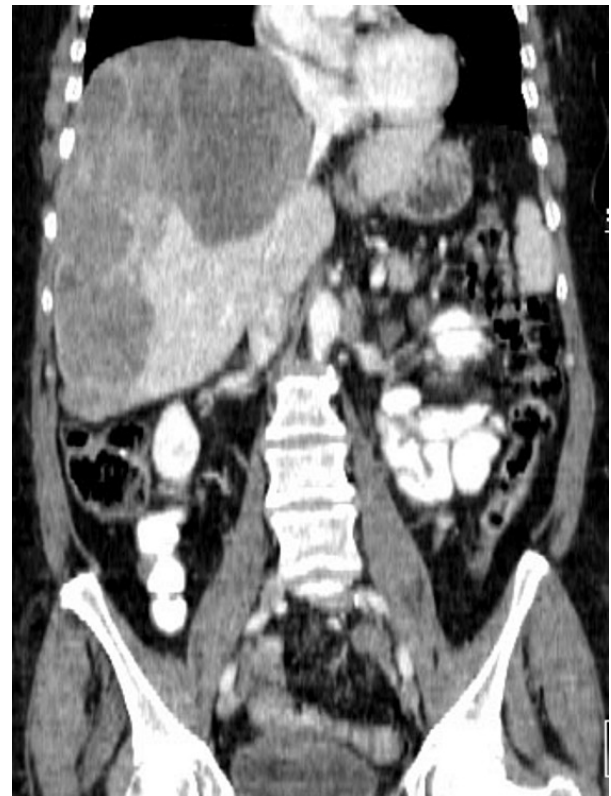


Figure 4. Coronal reconstruction CT scan image from year 2006 showing extensive involvement of right lobe of the liver.



Figure 5. CT scan of the abdomen from year 2006 showing large lymphadenopathy in the mesentery of the mid-abdomen.

forming pseudopapillae. Tiny slits separated the pseudopapillae (Figures 6, 7, and 8). The treatment options for this patient were limited because of extensive involvements of the liver and the mesenteric lymphnodes. The patient's daughter was willing to donate her liver for a living donor transplantation of the liver if that had any hope. Although we referred the patient to the transplantation center for consideration for liver transplantation, the patient was only offered symptomatic treatment at the transplantation center.

DISCUSSION

Solid-pseudopapillary tumors are a very rare neoplasm of the pancreas with low malignant potential usually occurring in young females in their second or third decade of life. Characteristic histology is needed for diagnosis which may need open biopsy.

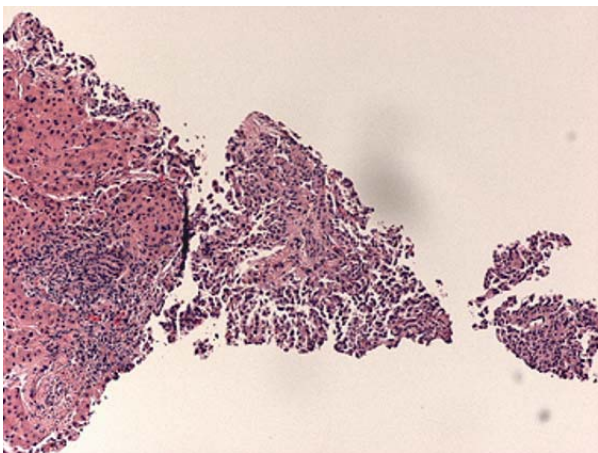


Figure 6. Low power micrograph showing the normal liver tissue and the infiltrating tumor tissue.

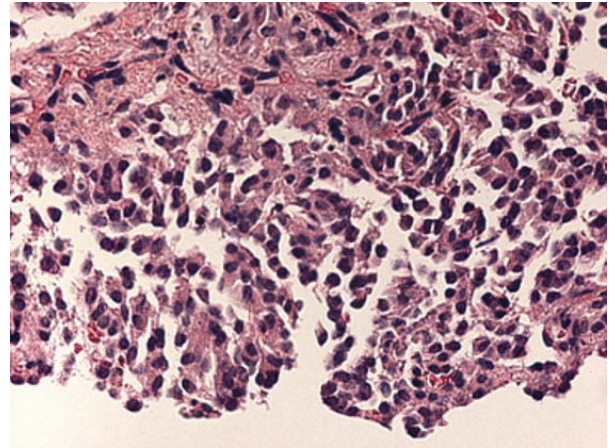


Figure 7. High power micrograph showing characteristic pseudopapillae.

Immunohistochemistry can help in some cases. Although resection of the tumor when possible is curative in most cases, local recurrences or distant metastasis can occur. The prognosis for solid-pseudopapillary tumors is excellent after curative resection (more than 90% survival at 5 years). Metastasis develops in less than 15% of cases and the liver is the most common site of metastasis [3, 4]. Resection of liver metastasis is possible if the involvement of the liver is limited. Survival even after metastasis is good even without definitive treatment because of the indolent nature of the disease [5]. This patient probably had liver metastasis at least 4 years without any symptoms before she presented to the hospital again in 2006. The histological features which predict metastasis include invasion of blood vessels, peri-neural clefts and adjacent organs, a high degree of

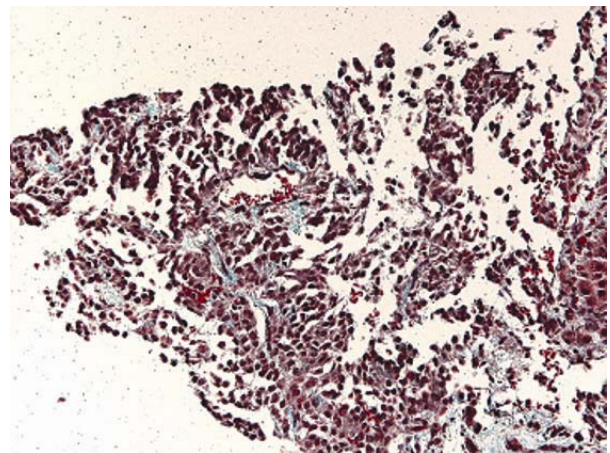


Figure 8. Trichrome staining clearly showing the vascular stalk of the pseudopapillae.

cellular pleomorphism and an elevated mitotic rate [6]. One study which compared the histological features between malignant and nonmalignant tumors supported the assumption that venous invasion, nuclear grade, and prominent necrobiotic nests are useful as histologic parameters to detect the malignant potential of solid-pseudopapillary tumors [7]. Another study concluded that the older the age of presentation, the more likely the tumor is to harbor malignant cells. The authors suggested that the origin of solid-pseudopapillary tumors is totipotential primordial cells found in the embryonic pancreas [8]. Local invasion on initial surgery may also be a feature which might predict metastasis at a later date [9]. This case had perineural invasion and duodenal involvement in the resected specimen which are known markers for aggressive behavior. Benefits of chemotherapy and radiotherapy for this condition are unknown although there are some anecdotal reports of benefit [10, 11, 12, 13]. Although liver transplantation can improve survival in metastatic neuroendocrine tumors which share many similar growth and other characteristics of solid-pseudopapillary tumors, the benefits of liver transplantation for metastatic solid-pseudopapillary tumors is unknown [14].

CONCLUSIONS

This case proves that solid-pseudopapillary tumor of the pancreas can present later in life and metastasis can occur long after the resection of the primary tumor. Therefore, long term close follow up for recurrence should be performed. The survival rate even with liver metastasis is good, but the possibility of liver transplantation as a potential cure needs to be explored.

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