Solid Pseudo Papillary Epithelial Neoplasm of Pancreas

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Abstract

Solid pseudo papillary epithelial neoplasm (SPEN) usually located in the body and tail of the pancreas are slow growing tumors of the pancreas. They are considered to be a rare pancreatic tumor and are typically present in women in their third decade of life. These tumors have a low malignant potential and rarely metastasize. However, very few cases respond to chemotherapy, but are efficiently resectable by surgery. Here we report a case with the above features.

Key words: solid pseudo papillary epithelial neoplasm(SPEN)

Introduction

Solid pseudo papillary epithelial neoplasm of pancreas(SPEN) are rare tumors and account for 1-2% of all primary tumors of pancreas. A total of 450 reports of solid papillary tumor of the pancreas have been reviewed in the literature since it was first described by Frantz in 1959[1]. In the above case,first diagnosis was papillary carcinoma of the pancreas which should have responded to chemotherapy. But that didn't happen and in fact the size of the tumor increased during the course of treatment. Later considering the age factor and other CT scan features, it was suspected to be a Solid pseudo papillary epithelial neoplasm of the pancreas (SPEN). Intraoperatively even though the tumor looked larger, with meticulous dissection it was completely removed without affecting the adjacent structures to which it was adhered. These are the unique features of the above case.

Case Report

A female patient Asian origin aged 22 years, reported to us with a history of pain in the abdomen, general weakness and weight loss over a period of 4 months. The patient had consulted a physician with the above complaints and was asked to undergo CT scan of abdomen and pelvis. The CT scan showed mass arising from the pancreas. A CT guided biopsy report confirmed it to be papillary carcinoma of the pancreas. The patient received 3 cycles of chemotherapy (Etoposide and Cisplatin) and repeat scan after completion of chemotherapy confirmed the chemoresistant nature of the tumor. Consequently she was referred to us for surgical management.

On examination of the patient there was a mobile mass occupying the right hypochondriac, epigastric, left hypochondriac, & left lumbar quadrants of the abdomen. CT scan was again repeated. Considering the age, sex, clinico-pathological features and the refractory nature of the tumor to the conventional chemotherapy, we suspected it to be SPEN and planned a surgery. To affirm the location and any impending complications, we also consulted radiologists to rule out the involvement of the portal and superior mesenteric vein before initiating the laparotomy.

The surgical procedure with roof top incision was begun. Thorough exploration led us to the considerably huge mass of growth arising from the body and tail of the pancreas, extending to the subhepatic region, displacing the stomach superiorly and invading the transverse mesocolon and splenic hilum[Fig1].Exploration of the liver also confirmed that there were no metastatic lesions on the surface. With meticulous dissection whole mass along with the spleen was removed and evaluation of the physical features revealed that the mass measured 13 x 10 cms[Figure 2]. The head of the pancreas was preserved and pancreatic duct was suture ligated.

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The specimen was sent for histopathological examination and the report confirmed that the tumor was indeed a SPEN and had characteristic features like small to medium polygonal cells elongated with ovoid nuclei arranged in solid pseudopapillary, microcystic, trabecular pattern and without lymphadenopathy.

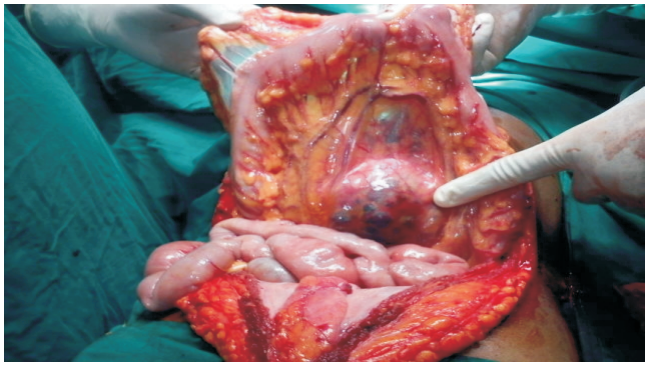

Figure 1. Mass seen adherent to transverse mesocolon

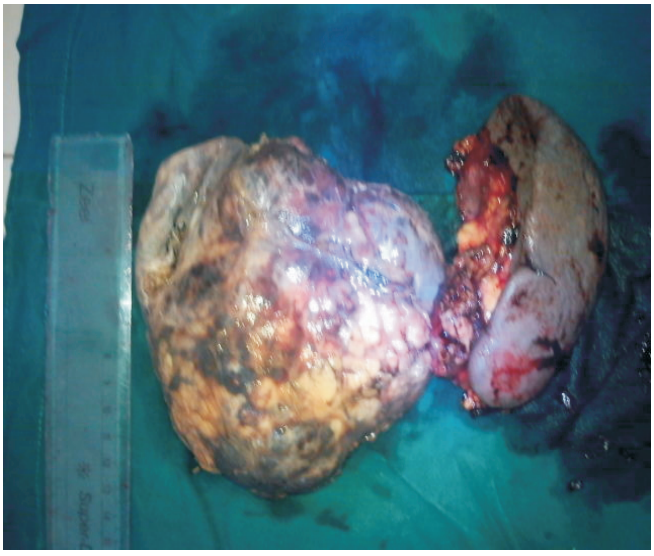

Figure 2. Resected specimen consisting of pancreas and the spleen

Discussion

Solid pseudo papillary epithelial neoplasms of pancreas are an internationally accepted entity and have been identified by a number of synonyms including Frantz's tumour, solid and cystic acinar tumor, papillary epithelial neoplasm, and solid and papillary epithelial tumor or neoplasm. The tumor is thought to arise from the ductal or acinar origin and has observed predominantly in young women in their third decade of life. However reports also exist for these tumors afflicting young children [2,3,4].

These are slow -growing tumors with indolent course, are commonly located in the body and tail of pancreas and the process of genesis is unknown. Some authors have reported that it is probably a neoplasm of uncommitted cells with most cells similar to intercalated duct cells or centroacinar cells[5]. Most of the SPTs are large with solid and cystic areas. These tumors begin as solid masses in which there are many poorly supported tiny vessels and then the cells farthest from the small vessels undergo swelling and degenerative change, whereas the cells next to the vessels remain intact. This results in a pseudopapillary pattern and cystic spaces[6].

The tumors exhibit low-grade malignant potential and rarely metastasize i.e.., 15% metastatic rate involving liver and peritoneum and lymph nodes. Even in the presence of disseminated disease, the clinical course is usually protracted and overall 5-year survival rate is 97% [7]. In the absence of possible metastasis these tumors, even of large size are localized and easily resectable.

Majority of patients present with vague abdominal symptoms, resulting in a delay in presentation and diagnosis. Most of the symptoms are due the size of the tumor compressing on adjacent structures. Symptoms or signs due to metastasis to other organs are rare. The differential diagnosis of SPEN is an endocrine neoplasm, cystic tumor of the pancreas and pancreatic pseudocyst and pancreatoblastoma in children [8]. Complete resection of local disease is curative. Even patients with residual disease or metastases have been reported to have long -term survival follwoing surgical treatment. Very few reports of the use of chemotherapy or radiotherapy for these tumors exist with only limited response.

Acknowledgments

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