CASE REPORT

Metastatic Solid Pseudopapillary Tumor of the Pancreas: A Case Report

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ABSTRACT:

OBJECTIVE:

To report a case of Metastatic Solid Pseudopapillary Tumor of the pancreas (SPTP), in young age female patient after one year of distal Pancreatectomy, discovered during regular follow up, with review of literature.

CASE REPORT:

A 17 year old female diagnosed to have Solid Pseudopapillary Tumor of the pancreas (SPTP), by clinical and radiological methods, about 10x7cm affecting the body and tail of pancreas with involvement of splenic vein. The patient underwent extended left side Pancreatectomy and Splenectomy. Postoperative histology proves to be SPTP with complete resection R0 margin. After one year of regular follow-up, patient presented with infraumbelical palpable mass, 10 cm in diameter, shown to be related to the omentum by abdominal CT-scan. Patient underwent laparotomy, and multiple different size masses found in the greater omentum largest 10x12cm.complet omental resection was done. Normal ovaries and uterus. Histology shows Metastatic Solid Pseudopapillary Tumor of the pancreas (SPTP). Patient discharged at 3rd postoperative day.

CONCLUSION:

SPTP usually has benign behavior and treatment of choice consists of surgical resection, but it may spread outside the pancreas, particularly in peritoneal cavity. The outcome after surgical resection is excellent with 90% survival in the long term. Recurrence has been described in approximately 10% of the patients. Follow-up with CT is essential after surgical resection to detect recurrence or metastatic disease.

KEYWORDS: pancreas, metastatic solid pseudopapillary tumor, distal pancreatosplensctomy.

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INTRODUCTION:

Solid pseudopapillary tumor of the pancreas (SPTP) is a rare pancreatic neoplasm with uncertain etiology that usually occurs in young females. It afflicts young women with 10:1 women to men ratio, with average age 24 years [(1)].SPTP was first described in 1959 by Frantz [(2)]. Various terms have been used to describe this tumor: papillary solid-cystic pancreatic, tumor of Gruber-Frantz and solid pseudopapillary tumor of the pancreas. In 1996, the World Health Organization reached a consensus to name this clinical entity as solid pseudopapillary tumor of the pancreas. It is classified as a cystic tumor of the pancreas that is also among the exocrine tumors of the pancreas [(3)].These tumors are characterized by solid and cystic components, with cellular degenerative changes alternating with pseudopapillary formation [(4)]. SPTP is rare, accounting for 1% to 2% of all pancreatic tumors and 13% of surgically resected cystic lesions [(5)].The tumor can occur anywhere in the pancreas but more commonly tends to affect the body and tail. Most patients are cured after complete surgical resection, but 10% to 15% of patients present with synchronous metastasis or develop metastasis at some point [(6)] [(7)] [(8).] We present a case of young age female patient with omental metastatic SPTP.

CASE REPORT:

A 17 year old female patient presented with abdominal pain of few weeks duration associated with nausea, anorexia and weight loss. On examination there was vague abdominal mass, which was not tender. Abdominal Ultrasound (US) examination showed cystic lesion affecting body and tail of the pancreas, the size about 10X7 cm. Abdominal CT scan revealed non homogenous enhancing mass affecting the tail and body of the pancreas, the size about 10x7.5cm with possible invasion of the splenic vein. After two weeks of preoperative vaccination surgery was done through upper midline incision. After full exploration, finding was large cystic mass about 10x8 cm involving pancreatic tail, extended to the neck with involvement of the splenic vein at its junction with superior mesenteric vein. Extended left side

pancreatectomy done, with primary closure of pancreatic remnant (head stump) by 3-0 prolen with splenectomy, resection of the splenic vein at its junction with SMV, and suturing of the defect with 5-0 prolen. Postoperative period passed uneventful, and patient was discharged at 5th post-operative day. Histopathology report showed SPTP, with free resection margin (R0) resection. Behavior could not be assessed. One year later, during routine follow up, we discover the presence of pelvic mass. The mass was below the umbilicus, not tender, not mobile, and about 10 cm in diameter (**Figure 1)**. Abdominal CT scan and US showed multiple enhancing masses in the lower abdomen keeping with peritoneal and ovarian metastases, the largest one 10 x 9cm (Figure 2).After discussion in MDT, meeting the operation was decided. At exploration the finding was multiple masses involving the greater omentum; largest about 10X12 cm, another mass about 5X3cm and the third one was about 3 cm in diameter. These lesions extended by gravity to the pelvis with filmy adhesion to the peritoneum. Complete resection of the greater omentum, along with masses, and appendectomy was done due to adhesions of the mass to the appendix (Figure 3). Ovaries and uterus were normal. Postoperative period was uneventful and patient discharged at 3rd postoperative day. The result of histopathology prove to be metastatic SPTP, in Immunohistochemical staining result show positive for vimentin, s100,and ki67 in 20% of tumor cells (Figure 4).

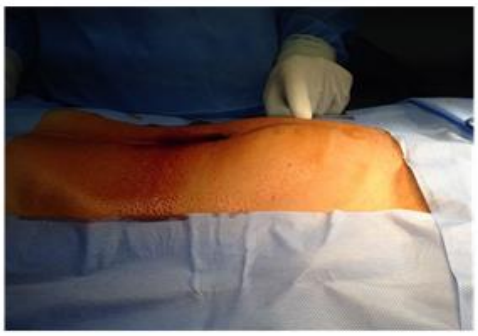

Figure (1) Visible pelvic mass

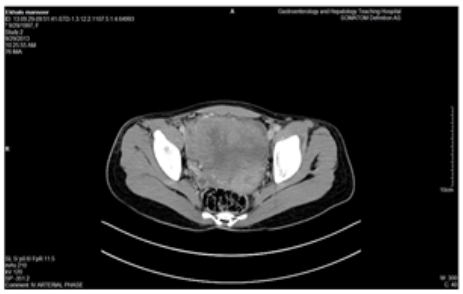

Figure (2) CT-scan of the abdomen shows pelvic mass

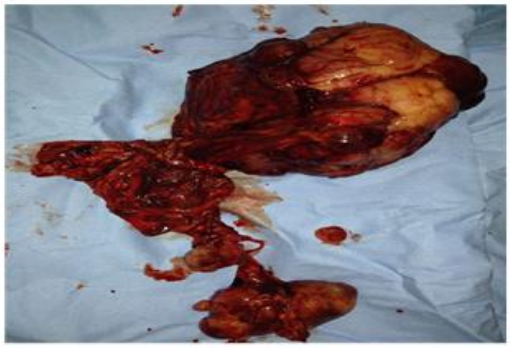

Figure (3) Gross specimen

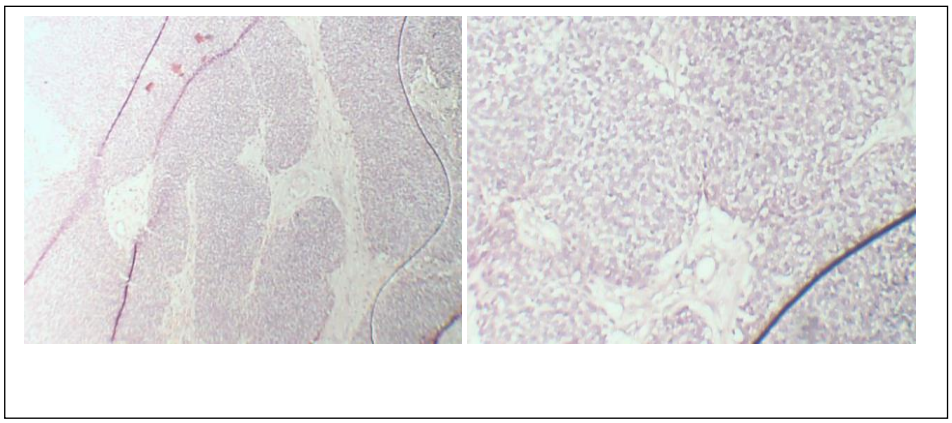

Figure 4: Shows the histology result of the mass.

DISCUSSION:

SPTP is a rare tumor and it represents approximately 1% of all pancreatic neoplasm [4] [5]. Recently, Papavramidis et al reviewed 210 English language papers published from 1933 to 2003, and found 718 well-documented cases[6]. In the last 10 years, there has been a steady increase in the number of these tumors. An apparent rise in the incidence is probably due to a better understanding of the pathology. In the 718 patients with SPTP, more than 90% were female aged 22 years on average.

Clinically, the patients with SPTP usually present with vague abdominal pain or a mass. About 15.5% of the patients are reported to have been asymptomatic [7]. Acute manifestations such as pancreatitis triggered by ischemia, distension or duct obstruction, or hemoperitoneum caused by the rupture of the tumor capsule are rare, but should be kept in mind. Frequently, no abnormalities are found by clinical laboratory tests, such as the levels of serum or urine amylase levels, pancreatic cancer markers and blood sugar [7].

As part of the general investigation, US show a well-circumscribed nonhomogeneous mass in the epigastrium. Abdominal CT usually shows heterogeneous enhancement with progressive central filling and late enhancement of the capsule [3]. MRI reveals an encapsulated mass with solid and cystic components as well as hemorrhage without obvious internal septum, SPTP should be highly suspected [8]. Although some image characteristics are suggestive of SPTP, fine needle aspiration biopsy (FNAB) can be used to obtain a possible preoperative histological diagnosis; however, some researchers have suggested that FNAB should be avoided because of the potential risk of tumor spillage [9].

Surgical resection is now considered the most efficient treatment option for patients with SPTP, because it offers a good chance of long-term survival [10].The most common sites of metastasis are the liver, regional lymph nodes, mesentery, omentum, and peritoneum [11]. Among 17 published cases of metastatic SPTP (there were 6 cases with peritoneal carcinomatosis), three of them with history of abdominal trauma with peritoneal carcinomatosis. [12]. Metastatic disease is rare, with local recurrence being more likely. The incidence of recurrence, local invasion or metastatic disease has been calculated to range between 10 and 15 %[6] [7]. Of the 452 cases referred to by Lam et al. and Rebhandl et al., only 66 showed recurrence, local invasion or distant metastases. Metastatic spread has been reported in the liver, lung, skin, peritoneum, omentum and lymph nodes. Only two reports of children with metastatic disease were found. Eleven other children were reported to have local invasion, and a further three cases were reported to have developed recurrence or metastatic disease in later life. Peritoneal carcinomatosis has been reported in only three adults with SPTP. A single case of a child with SPTP, lymphadenopathy and an omental metastasis has been reported. [13].Mozaffar M reported 4 cases (3 girls and a boy) 13-16 years of age presented with peritoneal masses following blunt abdominal trauma [14]. In review of 553 chines patients with SPT Peng-Fei Yu et al found that only 15% have metastasis, of them only 7 patients (1.4%) with omental metastasis [15].

The location and local invasion determine the surgical technique. For tumors that affect the body and tail of the pancreas, pancreatectomy is indicated, performing a distal procedure with spleen-sparing, except for cases of vascular involvement or splenomegaly [16]. Subjects with tumors involving the pancreatic neck, central pancreatectomy are performed with pancreaticojejunostomy or pancreaticogastrostomy. Central pancreatic resection is particularly indicated in young patients to avoid secondary sequelae of diabetes [17]. Tumors in the pancreatic head require cephalic pancreatoduodenectomy performed with pyloric preservation. Small tumors can be removed by enucleation. In general, SPTP can be removed laparoscopically because they are generally benign and have thick fibrous capsules. However, the decision to perform laparoscopic surgery should be made carefully to avoid the risk of rupture [18]. Lymphadenectomy is not performed because extension to lymph nodes is extremely rare; there is no scientific evidence for the value of this greater procedure [19]. The surgical approach to liver and peritoneal metastases is surgical resection. In cases of unresectable liver metastases, treatment is controversial and the results are not encouraging [20]. Chemotherapy and radiotherapy have been used infrequently because most SPTP can be successfully resected, although some reports indicate the significant success of chemotherapy and radiotherapy for treating the advanced lesions that were not completely resected . Trans-arterial chemoembolisation (TACE) has been reported as a useful treatment modality for liver metastasis; however, additional studies for determining the usefulness of TACE for treating SPTP liver metastasis are needed[21].

The pathologic diagnosis of SPTP is mainly based on the well-defined solid and cystic structure and characteristic pseudopapillary features under the microscope. Solid areas alternating with pseudopapillary formations, evidence of cellular degeneration, nuclear grooves and aggregates of hyaline cytoplasmic globules are found, at least focally, in every case[22]. Immunohistochemical studies are frequently performed to confirm the diagnosis. SPTP is typically positive for vimentin and antitrypsin and are negative for trypsin and chymotrypsin. They may also show focal immunoreactivity for neuron-specific enolase (NSE) and cytokeratin [23].

The differential diagnosis of SPTP of the pancreas includes any solid or cystic pancreatic disease entity, such as mucinous cystic tumor. Micro cystic adenoma, islet cell tumor, cystadenocarcinoma, acinar cell carcinoma, inflammatory pseudo cyst, mucus secreting tumor, pancreatoblastoma, and a vascular tumor-like hemangioma. The first four are usually seen in older patients and have no particular gender preponderance [24].

The prognosis of SPTP patients even with local recurrence and metastasis or invasion is good. It has been reported that the overall 5-year survival rate of SPTP patients is about 95 %. Due to the favorable prognosis and long survival rate of SPTP patients with local recurrence or metastasis, it is difficult to identify the predictive factors for their survival time [24].At present; there are no established clinical or histological criteria to predict the biological behavior of SPTP. While invasion of blood vessels (as in our patient with splenic vein invasion), perineural infiltration, invasion of adjacent structures, high degree of cellular polimorfism, and elevated mitotic rate and proliferative index, assessed by Ki-67immunoreactiv are suggested to be associated with metastases and recurrence. However, absence of these features does not preclude malignant behavior [25] [26]. Recurrence, local invasion, and limited metastasis are not the contraindications for resection, and some patients with unresectable SPTP may also have a long survival time [27].

We report a patient with pancreatic tail SPTP resection, confirmed histologically complete resection. Complicated one year later by omental recurrence, operated on with complete omentectomy and proved to be a pseudopapillary tumor. The malignant behavior of this case can be predicted by vascular (splenic vein) invasion and high Ki67 immunoreactivity. For the metastases, there is also general consensus that surgical debulking should be performed. Recurrent disease and metastases in patients with SPTP demonstrate that it does not always behave in a 'benign' manner. Follow-up with CT is essential after surgical resection to detect recurrence or metastatic disease.

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