Solid-Pseudopapillary Tumor of the Pancreas in Adults: A Case Report

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ABSTRACT

Solid-pseudopapillary tumor of the pancreas (SPTP) is an uncommon low grade exocrine pancreatic malignancy. We represented a 22 years old female with an abdominal mass of 4 years history and symptoms of weakness, loss of appetite, 7 kilograms weight loss and swelling for almost 2 months. She was suspected of pancreatic malignancy due her abdominal ultrasound and CT reports and therefore underwent explorative surgery that revealed a huge pancreatic tumor. The tumor was resected totally and hystopathological examination reported significant components of pancreatic solid pseudopapiller tumor features which was also the final diagnosis.

Keywords: Solid-Pseudopapillary Tumor; Pancreas

1. Introduction

Solid-pseudopapillary tumor of the pancreas (SPTP) is an uncommon low grade exocrine pancreatic malignancy, mostly seen in young women with an incidence of 1% of all neoplasms of the exocrine pancreas. There had been many hypotheses about the histogenesis of the tumor. The most commonly agreed upon theory propose the origin of tumor to be derived from a pluripotential epithelial stem cell from small pancreatic ducts with exocrine ductal differentiation [1]. Additionally, the elevated progesterone receptors in the absence of estrogen receptors in these tumors seem to explain the prevalence in reproductive age women. The body and the tail of the pancreas are more frequently affected. The clinical presentation of the tumor is usually nonspecific and its diagnosis is usually incidental during diagnostic tests or surgical explorations. As the tumor is usually asymptomatic, the diagnosis is difficult and incidental during diagnostic tests and surgical explorations.

The aim of the current report is to present a case of this rare disease that diagnosed and treated in our clinic.

2. Case Report

A 22-year-old woman admitted to our clinic with one and a half month history of abdominal pain in the right upper quadrant. She also had corresponding complaints of weakness, loss of appetite, 7 kilograms weight loss and swelling. She claimed that a cystic lesion of the liver was detected by an abdominal ultrasonography 4 years ago and was interpreted as a benign cyst with no further evaluation.

At initial admission to our center, she brought her abdominal ultrasound report performed by another clinic one month ago showing a 7 cm diameter low echoic or iso-echoic round liver mass, which was heterogeneous, composed of solid and cystic components. Intrahepatic bile ducts and related vascular structures were all found to be normal in this report. In addition, she had had a CT examination in the same clinic and the mass were interpreted as (the patient only had the written report not the CT images) a liver mass located in left lobe medial segment with exophytic protrusions through the pancreas.

She had tenderness on the right upper quadrant in her physical examination and the liver was found to be palpable 3 cm below the 12th costa. Other systemic examinations were all normal.

Her laboratory values including tumor markers: alpha-fetoprotein, carcinoembryonic antigen, CA-125, CA15-3, CA19-9 were found to be within normal limits except for high erythrocyte sedimentation rate of 66 mm/h (normal range: 0 - 20) and CRP: 17, 18 (normal range: 0 - 5).

Abdominal ultrasonography revealed a heterogeneous, iso-echoic tumor composed of solid and cystic components. She underwent an abdominal CT examination that
revealed a 9 × 8 × 7 cm heterogenous cystic-solid mass localized at the distal pancreas including the tail and corpus, protruding through liver left lobe medial segment, that has hypodense necrotic spaces which enhanced peripherally after intravenous contrast injection (Figure 1). The bile duct and pancreatic duct showed significant dilatation. The margins of the tumor and its relation with pancreatic tissue and other organs couldn’t be distinguished by CT images as the mass was huge comprising left liver lobe medial segment and the pancreas. The mass revealed a significant vascularization during the arterial phase of the CT. The CT findings were concordant with pancreatic carcinoma or SPT and interpreted accordingly.

The upper GIS endoscopy examination was normal except for an external gastric corpus and antrum compression from the direction of small curvature.

An angio-CT were also performed and revealed a huge avascular or hypovascular pancreatic mass.

Preoperative diagnosis was decided to be a pancreatic carcinoma and explorative surgery was planned. During the surgical exploration; a heterogeneous, irregular approximately 10 cm diameter pancreatic mass originating from uncinate process of the pancreas was observed. The tumor was ascending on the duodenum through the choleduct, hemi circulating on the anterior wall of duodenum with no invasion. The tumor seemed to repress the pancreatic head and tail below the duodenum and was lateralizing the stomach. In addition, invasion of the proximal choleduct above the duodenum was present. The invasion through the choleduct was recognized while the mass tried to be dissected below the duodenum with its capsule. As the tumor was originated from uncinate process a total excision including uncinate process of pancreas was performed. As the proximal choleduct was invaded, a choledochotomy followed by a choledocoduodenostomy was also performed with a feeding tube replaced within. The mass was completely resected without any complications during the surgery.

The postoperative course was uneventful and no adjuvant therapy was given.

The patient was discharged from the hospital on postoperative 15th day.

The pathological investigation showed a 90 × 85 × 95 cm pancreatic tumor partially demarcated by a fibrous capsule. Heterogeneous solid pseudopapiller regions had areas of extensive necrosis and hemorrhagic material.

Microscopic histopathological examination demonstrated that the cells that form solid and papillary regions were uniform without significant atypia and very few mitotic activities were observed. The histopathological findings concluded the diagnosis as pancreatic solid pseudopapiller tumor (solid-cystic papillary neoplasm; Gruber Frantz Tumor) (Figure 2).

3. Discussion

Solid-pseudopapillary tumor of the pancreas (SPTP) is an uncommon low grade exocrine pancreatic malignancy that rarely metastases and named after Frantz VK who first reported the disease as “papillary tumor of the pancreas, benign or malignant” [2]. The body and the tail of the pancreas are more frequently affected (64%) than the head [3]. But these huge tumors may repress other parts of the pancreas concluding with atrophy. Unlike the most previous reports our patient had SPTP originating from the head of the pancreas and specifically uncinate process of the head which is very rare.

Review of the literature by different authors reported...
ever hepatic tumors should also be considered in different age, gender and clinical course might help. How-tumors, cystadenocarcinomas, pancreoblastoma, etc. Butions like inflammatory pseudocysts, mucinous cystic were observed. It is challenging to make an accurate dif-

These tumors distinguish from other exocrine pancre-

Our patient total tumor size was approximately 10 cm as the reported size of these tumors vary widely between 0.5 cm through 25 cm with a mean of 9.3 cm [4]. In general they are big, round, well circumscribed with a pseudo-
capsule, full of mucinous secretion, and multilocular at times [7]. Microscopically, cysts are made up of an epithe-
lial column (cystadenomas) or a combination of epithe-
lial column and atypical epithelial cells (cystadenocarcinomas) [1]. The solid regions composed of nests and sheets of uniform epithelial cells alternating with cystic spaces and pseudopapillae. The cells are rather discohe-
sive with cytoplasmic delicacy and nuclear membrane irregularities. Pseudopapillae formed by tumor cells around central vessels are commonly seen. Pleomor-

Immunohistochemistry evaluations of the pancreatic tumor in the diagnostic process of STPD’s are somewhat informative for differentiating diagnosis from other solid and pancreatic lesions like inflammatory pseudocysts, mucinous cystic tumors, cystadenocarcinomas, pancreoblastoma, etc. But young age, gender and clinical course might help. How-
ever hepatic tumors should also be considered in differential diagnosis. Our patient was thought to have a be-
nign cystic liver CT shown lesion 4 years before admis-
sion and the tumors features observed during preopera-
tive diagnosis supported a liver mass, as well.

CT scans are also useful of showing a well circum-
scribed retroperitoneal mass with various solid areas and calcifications around hyper dense cystic components owing bleeding areas and hemorrhagic degeneration [16]. We performed CT and angio-CT imaging techniques which showed those typical findings of pancreatic tumor or SPTD appearances with good localization of the origin and was very helpful in our diagnosis.

MRI provides similar findings but reported to be more informative for differentiating certain tissue characteristics like hemorrhage, cystic degeneration or the presence of capsule [17].

Preoperative endoscopic ultrasonography-guided fine needle aspiration biopsy for preoperative detection of tumor are supported by some authors as imaging tech-
niques are sometimes confusing due to similarity of find-
ings among cystic lesions of pancreas [18]. In contro-
versy this might be unacceptable by some because of uncertainty in diagnosis and the possible spread of tumor cells [18,19].

Tumor resectability consideration preoperatively is often provided by improved imaging techniques that offers a more accurate extension diagnosis. Vascular infil-
tration is the main factor for non-resectability. The sensitivity of helical CT for a successful assessment of such vascular infiltration is reported to be only 60% - 89% causing undetected non-resectability. However volumetric angio-CT is superior by demonstrating the anatomic relation to the vascular tree, thus allowing a more appropriate planning for surgery [1].

SPT has a low grade malignant potential and has a very slow growing pattern. The doubling time of this malignancy were reported to be 677 days, 240 days and 765 days in different studies [20]. The first probable tumor of our patient as we informed in words, was observed incidentally 4 years before any symptomatic disease, unfortunately, we were not able to find the genuine USG report of that time. She also noted that, the incidentally observed tumor was misdiagnosed as a liver benign cyst with no further evaluation recommended.

Surgery is the only valid option with proven effectiveness for SPTP [21,22]. The accurate diagnosis of this low potential malignant tumor is important as the completeness for SPTP [21,22]. The accurate diagnosis of this low potential malignant tumor is important as the complete resection of the tumor reported to provide a 97% survival rate [23].

The surgical method depends on the size and location of the tumor and its relationships with the surrounding organs. A careful exploration during surgery should be done as to assess various arterial and venous vascular structures. STD s usually has a benign clinical course. Some challenging cases may present with local recurrence or distant metastases. In addition a locally advanced or metastatic disease at initial presentation is also possible in minority of the patients [20]. The common sites of metastasis include liver, peritoneum and lymph nodes. In addition it might invade adjacent tissues and vascular structures. In a review of 497 patients, 26 of them had (5%) portal vein invasion and 44 of them had (9%) other organ invasions (spleen, colon, duodenum, etc.) at the initial surgery [5].

Resection should be considered in case the upper mesenteric artery, hepatic artery and portal vein is observed to be tumor free. Well-encapsulated tumors, as reported here, usually enable simple enucleating of the neoplasm.

For advanced local invasive disease, to preserve the third and fourth duodenal portions, treitz ligament and first jejunal loop with careful dissection is extremely important [24]. For tumors of the pancreatic head, duodenopancreatectomy is suggested with recommendation of pylorus preservation for fewer digestive disturbances [19,25]. In our case, fortunately the tumor seem to be well-capsulated and originated from the uncinate process of the pancreatic head and simple resection of the uncinate process was enough with dissection of the tumor anterior to duodenum. As no local invasion to the duodenum were present, no resection of duodenal tissue was necessary for full resection. Only invasive focus was recognized on the choleduct and full excision of the invasive portion of the choleduct were performed with following choledochoduodenostomy. Body-tail pancreatectomy including splenectomy is indicated for tumors located in the body or tail of the gland. Depending on the low recurrence rate and prolonged survival after complete local resection, aggressive attempts at complete resection seem to be warranted, even if accompanied by metastases [21,26-28]. But neither extensive lymphatic dissection nor adjacent structure resection is reported to be reasonable [24].

Recurrence after apparently radical resection of a SPT can occur in 10% - 15% of cases and liver is the common site [5,23,29-31]. Resection of these metastasis long term effects are still unpredictable but seem to improve survival rates [20]. The role of chemotherapy, radiotherapy and other medical treatment options are still under study with no proven efficiency of benefit or vice versa [32, 33].

REFERENCES


