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Case Report

Solid Pseudopapillary Tumor of the Pancreas: a Case Report and Review of Literature

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ABSTRACT

Solid pseudopapillary tumors of the pancreas (SPT) are rare tumors of the pancreas with low malignancy potential and a very good prognostic outcome after surgery. The outcome after radical resection is favourable. A case of solid-pseudopapillary tumor (SPT) of the pancreas in a 20-year-old woman is presented. The patient underwent resection of the mass in the pancreatic head and pancreaticoduodenectomy (Whipple procedure) with jejunostomy tube placement. We focus on the clinical features, imaging, and histopathological characteristics of solid-pseudopapillary tumors (SPT) of the pancreas.

Key words: **Pseudopapillary Tumor, Pancreas**

Introduction

Solid-pseudopapillary tumor of the pancreas was first described by Frantz in 1959(1). They are considered a rare pathologic entity with minimal malignant potential, affecting mainly juvenile females (2). It comprises 0.2% to 2.7% of all pancreatic tumors (3). In spite of the rise in identification, the pathogenesis and obvious therapeutic algorithm remain unclear. The growing numbers of this neoplasm is being diagnosed due to the extensive application of imaging examinations, such as computed tomography, magnetic resonance imaging, and ultrasonography. Most information proposes that patients with solid pseudopapillary tumor have a

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positive prognosis after surgical therapy (4).
Invasive tumor progression or distant metastasis has been seldom detected. Therefore, besides to conventional partial pancreatectomies, enucleation or local resection has been performed to treat these low-grade malignant tumors (5, 6). However, the best treatment for solid pseudopapillary tumor is a matter of debates (7).
Regarding to SPT is classified as a border line tumor in WHO fascicle, preopertative, intraoperative and postoperative appropriate diagnosis via imaging studies, cytology, histopathology and immunohistochemistry seems to be necessary for choosing a proper therapeutic policy (3). Thus, we decided to share our experiences concerning this rare tumor and describe its clincopathologic features.

Case report

A 20-year-old female presented with intermittent epigastric abdominal pain for 2 years to Afzalipour Hospital (Kerman, Iran). She also had a history of early satiety and postprandial epigastric pain that radiated to the back. The abdominal pain lasted for approximately 3 hours after eating, and there were no relieving factors. The patient had normal vital signs and was afebrile. Physical examination revealed tenderness at palpation over the epigastric region but was otherwise unremarkable The laboratory test including complete blood count , AST, ALT, alkaline phosphates, bilirubin, random blood sugar and Amylase were in normal range . Gastroscopy demonstrated no mural or mucosal abnormalities in the area of the duodenum, stomach, or esophagus. She underwent ultrasound of the abdomen, which revealed a cystic lesion about 4.5 cmx4 cm in size in the area of the pancreatic head. Abdominopelvic computed tomography (CT) with both orally and intravenously administered contrast material showed a well-encapsulated lesion with solid and cystic components (Fig. 1). The patient underwent ultrasound guided fin needle biopsy. Histologic examination and immunohistochemical analysis for CEA and Alfa feto protein was performed. At immunohistochemical analysis, the tumor cells were positive for CEA and Alfa feto protein. Based on histologic and immunohistochemical analysis two diagnoses, the Pancreatoblastoma or solid pseudo papillary tumor were made.
The patient underwent resection of the mass in the pancreatic head and pancreaticoduodenectomy (Whipple procedure) with jejunostomy tube placement. She had uneventful postoperative course except for wound infection, which was improved by drainage.
At gross examination, a soft, round, well-circumscribed 4.5 cm × 4 cm mass was identified in the pancreatic head. The cut surface was deep red purple, partly solid and partly cystic, focally with a spongy appearance (Fig. 2).
At histologic analysis, the tumor was composed of uniform polygonal cells with moderate to abundant amphophilic cytoplasm and arranged in solid nests with areas of degeneration characterized by separation of the cells into pseudopapillary aggregates with intervening accumulation of mucopolysaccharide rich ground substance. In the pseudopapillary regions, the nuclei were oriented away from vessels, resulting in a zone of cytoplasm that separated the capillaries from the nuclei. Aggregates of foamy macrophages were also evident between nests of polygonal cells (Fig. 3). Another focal findings in the microscopy included coagulative necrosis and peripheral blood lakes. There were also distinct intracytoplasmic hyaline globules measuring roughly 1 to 20 μm. The globules stained with periodic acid Schiff and were diastase resistant but alcian blue staining was negative in these globules (Fig. 4).
Immunohistochemistry revealed positive staining for vimentin, progesterone receptors , neurone-specific enolase along with a weakly positive cytokratin and negative results for synaptophysin, chromogranin and cytokeratin 7 (Fig. 5).
After two years fallow up, the patient is feeling well and no metastasis was detected.

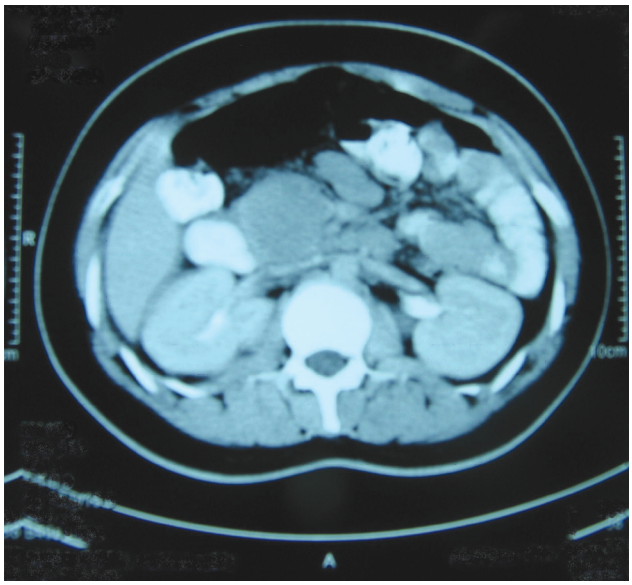

Fig. 1- Abdominal CT showing a well-encapsulated lesion with solid and cystic components

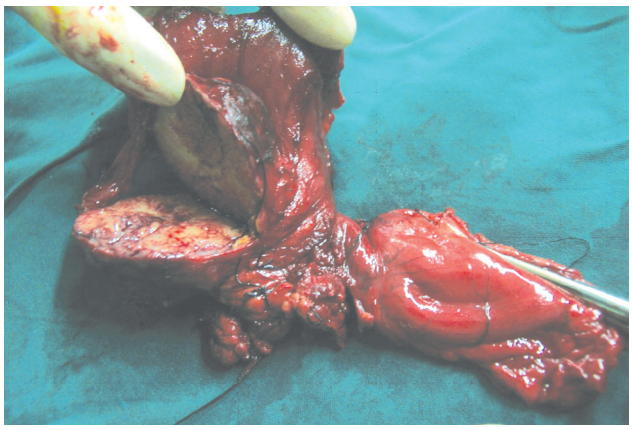

Fig. 2- The cut surface of tumor showing deep redpurple, partly solid and partly cystic appearance

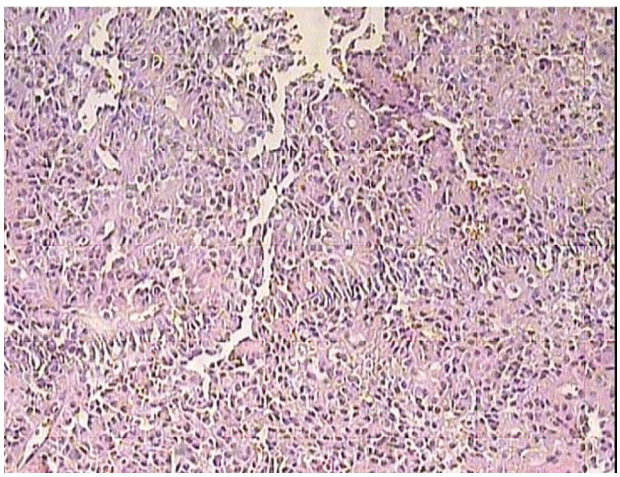

Fig. 3- Pancreatic tumor showing uniform polygonal cells with pseudopapillary aggregates (Hematoxylin and eosin staining ×40)

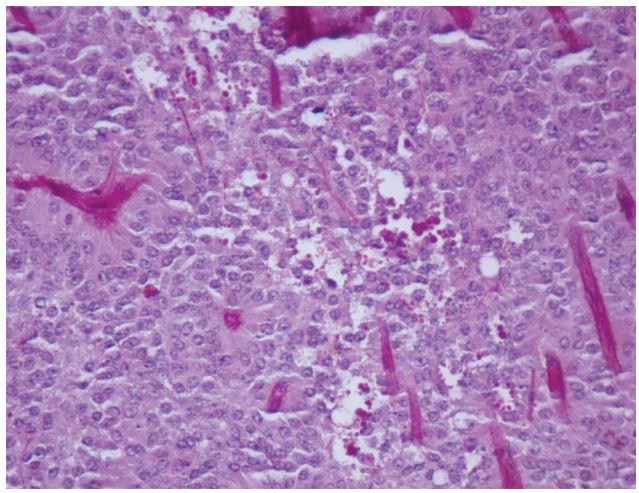

Fig. 4- Pancreatic tumor showing intracytoplasmic hyaline globules (Periodic acid Schiff staining)

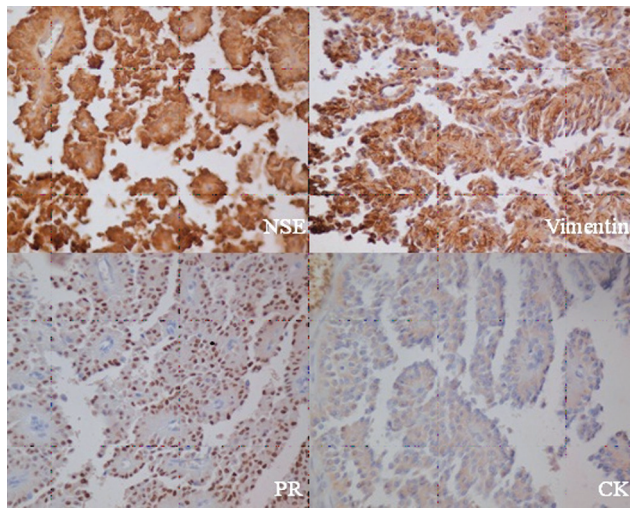

Fig. 5- Pancreatic tumor showing positive staining for Neuron-specific enolase (NSE), Vimentin, Progestrone receptor (PR), and a weakly positive reaction for Cytokeratin (CK)

Discussion

A SPT is a very rare entity that was first described by Frantz in 1959. SPT has been categorized as a borderline tumor of the pancreas by WHO, 1996. An SPT often presents as imprecise nonspecific symptoms. The most common symptoms are mild abdominal pain or discomfort. Patients can also present with fullness associated with nausea and early satiety, which is secondary to a mass effect. Approximately 15% of patients are asymptomatic (3). The differential diagnosis of SPT of the pancreas includes solid or cystic

pancreatic disease, entities such as inflammatory psuedocyst, mucinous cystic tumor, microcystic adenoma, islet cell tumor, cystadenocarcinoma, and pancreatoblastoma (8). The radiologic features on ultrasound, CT, or magnetic resonance imaging (MRI) are that of a usually well-defined big mass that can be mostly a thick-walled cystic construction or a mainly solid mass with some cystic elements. The solid portions enhance mildly on contrast-enhanced CT, but overall it enhances less than the normal pancreas (9). These tumors usually have vague clinical presentation and may form very giant masses before recognition (10). Despite of unspecific laboratory tests, CT and fine needle aspiration means are useful to diagnose the disease (11). Ohtomo describe the diagnostic yields of MRI in this tumor. In presented case abdominal CT was use for imaging and it seems to be sufficient for preoperative imaging (12). Nakagohri *et al.* reported that accurate identification is made preoperatively in 71% of patients (13). When a solid pseudopapillary tumor did not contain a cystic element, the diagnosis seems to be not easy.

We did not have facility of FDG-PET, in malignant form of SPT the stronger accumulation of FDG than surrounding pancreatic parenchyma on FDG-PET is observed (13).

Sometimes the exact diagnosis of SPT is not simple. Bektas *et al.* reported a SPT case that the final histological specimen was initially assessed differently by two departments of pathology: one classified the tumor initially as endocrine, the other as a solid pseudopapillary lesion. In our case the ultrasound guided biopsy diagnosis could not shows the exact diagnosis of the tumor (6).

Abdominopelvic computed tomography (CT) of our patient showed a well-encapsulated lesion with solid and cystic components. In lesion in pancreas, pseudocyst represents 80% of cystic lesions. Primary cystic neoplasm for instance serous cystadenomas, mucinous cystic neoplasms, intraductal papillary mucinous tumor, and SPTs comprises 20% (14). Infrequently ductal adenocarcinoma and islet cell tumor demonstrate secondary cystic transformation (2). The several entities in this unit arise more commonly in female but vary in age of appearance. Mucinous cystic neoplasm appears in a wide age range. Cystadenoma is seen in much older patient. Islet cell tumor of pancreas is detected mainly in adults and no sex difference is demonstrated. On the contrary, SPTs present principally in adolescent girls and young women (2, 15). In presented case sonography guided FNA was performed, which showed two diagnoses as the Pancreatoblastoma and SPT. A preoperative precise cytological diagnosis could be useful because it permits the preservation of segment of the uninvolved pancreas and keeps away from development of succeeding diabetes mellitus (2). Since these tumors are very vascular, preoperative precise diagnosis can prevent complications such as intraoperative bleeding (16). The highly cellular smears demonstrate abundant papillary tissue sections with slender branching fibrovascular stalks, which are attributed to this tumor. Pseudorosette configurations are also described (2).

The pathologic diagnosis of SPT is made primarily based on the distinct solid and cystic arrangement and typical pseudopapillary characteristics under the microscope (17). On the cut surface, a variegated manifestation is seen with variable arrangement of solid hemorrhagic and cystic-necrotic parts. The microscopic features of SPT are solid areas which alternate with a pseudopapillary pattern composed of a fibrovascular stalk surrounded by several layers of epithelial cells (18). In our cases, tumor cells showed similar histopathology (18).

Immunohistochemically, SPT cells were typically positive for vimentin, a1-antitrypsin, a1-antichymotrypsin, epithelial markers (CK and EMA), CEA, alfa feto protein, neuron-specific enolase and progestron receptor. Chromogranin is never detected but focal expression of synapto-

physin in a few tumors can be observed. Immunohistochemical staining of Ki-67 is positive in some patient (18, 19). In presented case the immunohistochemical staining was positive for vimentin, progesterone receptors, neurone-specific enolase , CEA and alphafeto protein.

The histopathologic differential diagnosis of this tumor includes psedocyst of pancreas, well-differentiated pancreatic endocrine neoplasm and acinar cell carcinomas (20).

Clinical and microscopic finding are very helpful for discriminating SPT from pseudocyst of pancreas. Pseudocysts are more prevalent in male sex. Elevated serum amylase and recurrent pancreatitis attacks are another clues which are in favor of pseudocyst. In the contrary, SPT have a more incidence in women and the serum amylase level is usually normal. Microscopically pseudocysts are deprived of epithelial lining , but SPT have eosinophilic or clear neoplasitc cells (21). In this case, normal serum amylase presence of epithelial cells and female sex were suggestive for SPT.

Well-differentiated pancreatic endocrine neoplasm is the next differential diagnosis. The tumor has similar cellular features with SPT such as homogenous round and oval cells with uniform nuclei. However speckled chromatin favors well differentiated pancreatic neoplasm, whereas presence of solid areas admixed with pseudopapillae foamy macrophages, cholesterol crystals and eosinophilic hyaline globules are suggestive for SPT. Immumohistochemistric studies show strong staining for chromogranin and synatpophysin and often a pancreatic hormone such as insulin glucagon and somatostatin for well differentiated pancreatic endocrine neoplasm. Markers such as neuron-specific enolase and CD56 for both of these tumors are in common. Nevertheless most SPT only focally and weakly express synaptophysin never chromogranin instead SPT strongly is positive for CD10 , alpha-1 antitrypsin and vimentine (20). In this case, positive staining for vimentin and negative ones for synaptophysin and chromogranin was in favor of SPT.

Acinar cell carcinoma also falls into the differential diagnosis of SPT but distinction between these two entities is usually obvious. The cell arrangement in acinar cell carcinoma is usually more cohesive forming solid areas. Pleomorphism and mitotic activities are frequent. Focal lumen formation can be seen. Single prominent nucleolus and granular cytoplasm are also evident in this tumor .In contrast, SPT, as in this case, are usually cystic ,the cells are very uniform, lumen formation is never encountered at light microscopic level, they lack mitosis, nuclei are grooved, the nucleoli are not prominent ,and neoplastic cells aggregate around delicate vessels rather than lumens (20, 21).

In presented case, the tumor was in pancreatic head but SPT can detected in any part of pancreas. Hu and colleagues reported a 19 year olds female patient with huge mass in distal of pancreas pushed the stomach. They undertook the patient distal pancreatectomy and splencetomy (1). The distal pancreas tumor may not be detected before mass enlargement. In our patient, the Whipple procedure was necessary because of involvement of pancreatic head.

In presented case pancreatico- duodectomy was needed because of the size of tumor and involvement of pancrearic head. Bektas reported a case of SPT in a young woman presented with unspecific complaints in the upper abdomen (6). They detected a mass in the area of the pancreatic head. They could resect only the tumor without a major procedure (6). In pancreatic head tumor if tumor is not large and the adjacent organs are not involved this procedure is possible.

Our patient had elective operation , Potrc reported a young man patient with SPT presented with sings of intra abdominal hemorrhage after trauma who was treated with Whipple's procedure (22).

In our case, Whipple's procedure was performed; however, standard pancreatoduodenectomy or distal pancreatectomy gives rise to a significant

deficit of normal pancreatic parenchyma and may produce destruction of exocrine and endocrine role. In addition, even though pancreatoduodenectomy can be conducted with a little mortality rate, morbidity is still common. Consequently, a number of authors have suggested enucleation or local resection for solid pseudopapillary tumors (5, 6). Chemotherapy may extend survival in Solid-pseudopapillary carcinoma with unresectable metastases (13).

In most patients, surgical therapy is curative and neither chemotherapy nor radiotherapy should be combined. In the few cases where surgery is not feasible, radiotherapy can be applied since these tumors seem to be radiosensitive (14).

Our limitation in this case report is the short time of postoperative fallow up, which was two years, it seems if it would be longer the consequence of treatment could be more reliable.

The overall prognosis of SPT of the pancreas is good because of their favorable biologic manifestations. Proper preoperative diagnosis is required since these patients may be definitively cured with sufficient surgical resection.

Acknowledgments

The authors declare that there is no conflict of interests.

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