CASE REPORT

Solid-Pseudopapillary Tumors of the Pancreas

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BACKGROUND:
Solid-pseudopapillary or solid-cystic papillary tumors are rare tumors of the pancreas which has special characters that they rarely metastasize and the patient usually enjoys long term survival. This is a case report of a fifty years female who had cystic tumor involved the pancreatic tail and she underwent distal pancreatectomy with splenic preservation, the histology study revealed solid pseudopapillary tumor of pancreas.

KEYWORDS: solid pseudopapillary tumor, pancreatic cystic tumor, distal pancreatectomy.

INTRODUCTION:
Solid-pseudopapillary tumors (SPTs) of the pancreas were first described by Frantz in 1959[(1)]. Since being reclassified by World Health Organization WHO in 1996, SPTs of the pancreas are an internationally accepted entity [(2)]. However, these tumors are rare and account for 1-2% of all primary tumors of the pancreas [(3)]. They are also called papillary cystic neoplasm, solid-cystic papillary tumor or Gruber-Frantz tumor, and they confined to the pancreas in 85% of patients [(4)]. Most of these tumors are found in young women in the second or third decade. Rarely these tumors were seen in children [(5)].
SPTs are histologically, clinically and prognostically quite distinct from the more common pancreatic ductal adenocarcinoma. These neoplasms are of low malignant potential and their prognosis is extremely good unlike other tumors of the pancreas, and even the 10-15% of patients with liver or peritoneal metastasis from SPTs commonly enjoy long term survival [(6)]. This paper was designed to report a case of SPT of the pancreas to increase our knowledge about this rare tumor.

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CASE REPORT:
A fifty years old female, admitted to the surgical department of the Gastroenterology and Hepatology Teaching Hospital Baghdad-Iraq, presented with vague upper abdominal pain of one year duration, otherwise she had good appetite, no fever or weight loss with normal bowel habit. The physical examination revealed tender vague left hypochonderial mass. Biochemical and hematological investigations were unremarkable.
Abdominal US revealed a well defined complex mass mainly cystic component with septations 70 × 75 mm. in diameter at pancreatic tail, raising the suspicion of mucinous tumor of the pancreas. Abdominal CT scan showed a multiloculated cystic mass involve the body and tail of the pancreas which has peripheral enhancement after IV contrast (Figure1).
On exploration there was a well defined cystic mass involving the body and tail of the pancreas intimately contact to splenic hilum (Figure 2). After careful separation of the mass from the splenic capsule and vessels with ligation and division of some short gastric vessel, distal pancreatectomy with splenic preservation was then possible.
Macroscopically the mass has well defined capsule containng brownish hemorrhagic fluid. On cut section there was a grayish white papillary projection. Microscopic examination revealed the tumor was very cellular and well-circumscribed without involvement of adjacent pancreas. Tumor cells were arranged in solid sheets and pseudopapillae, they were uniform, small to medium sized, polygonal with acidophilic cytoplasm and bland vesicular ovoid nuclei. The tumor separated from the normal pancreas with a well defined fibrous capsule without transgression of the capsule was seen (figure 3).
The patient regained oral intake after 24hours, the drain removed after 72hours, and discharged well on the 5th postoperative day.

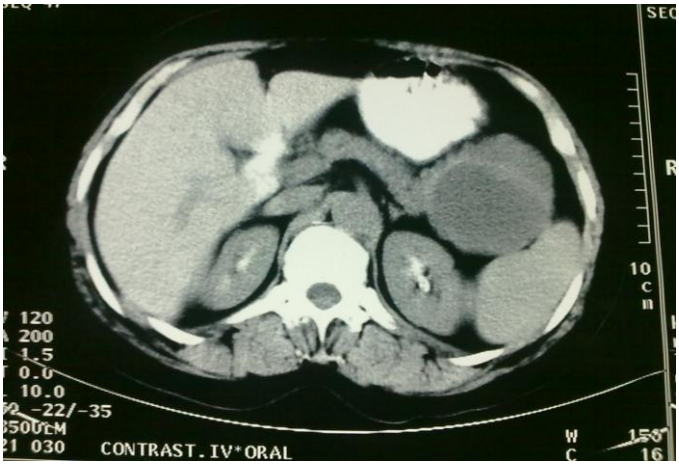

Figure 1 : Abdominal CT scan revealed cystic tumor at the tail of pancreas.

DISCUSSION:

A solid pseudopapillary tumor of the pancreas is a rare neoplasm of low malignant potential [7,8] . It accounts of 1.8% of patients underwent pancreatic resection [9], from 0.2% to 2.7% of all primary pancreatic neoplasms [8], and 9 % of patients with peripancreatic malignancy [7].

It occurs predominantly in females, that is about 90% of patients found in case series were females [9,10,11,12]. The age of presentation ranging from 21-39 years old [9,10] .

The clinical presentation of SPT can be clustered in three categories: nonspecific abdominal complaints, abdominal mass or fullness, and incidental radiological findings [7,10]. Abdominal pain was the predominant presenting symptoms [7,9,10] . Solid pseudopapillary tumor can develop in the entire gland, but it is slightly predominant in the pancreatic corpus and tail [9,13,14]. Robert et al [7] reported 33.3% in the head, 25% in the body and 42% in the tail, while de Castro et al [9] reported 42% in the head, 50% in the corpus/tail and central in 8.3%. In children, however, it can be more frequently found in the head of the pancreas [15,16].

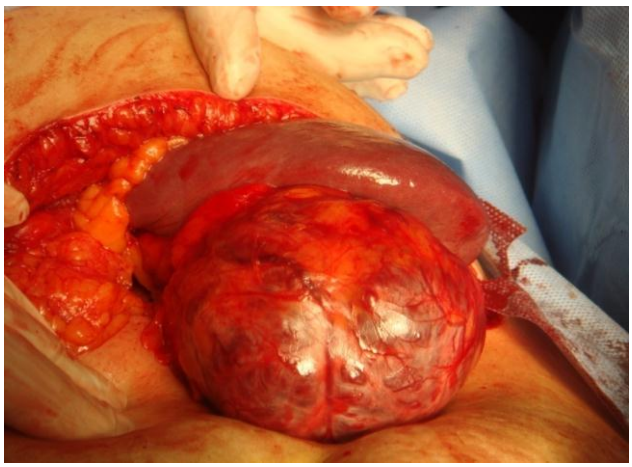

Figure 2: Operative photo showed the mass at tail of pancreas intimately in contact to the hilum of spleen after its mobilization

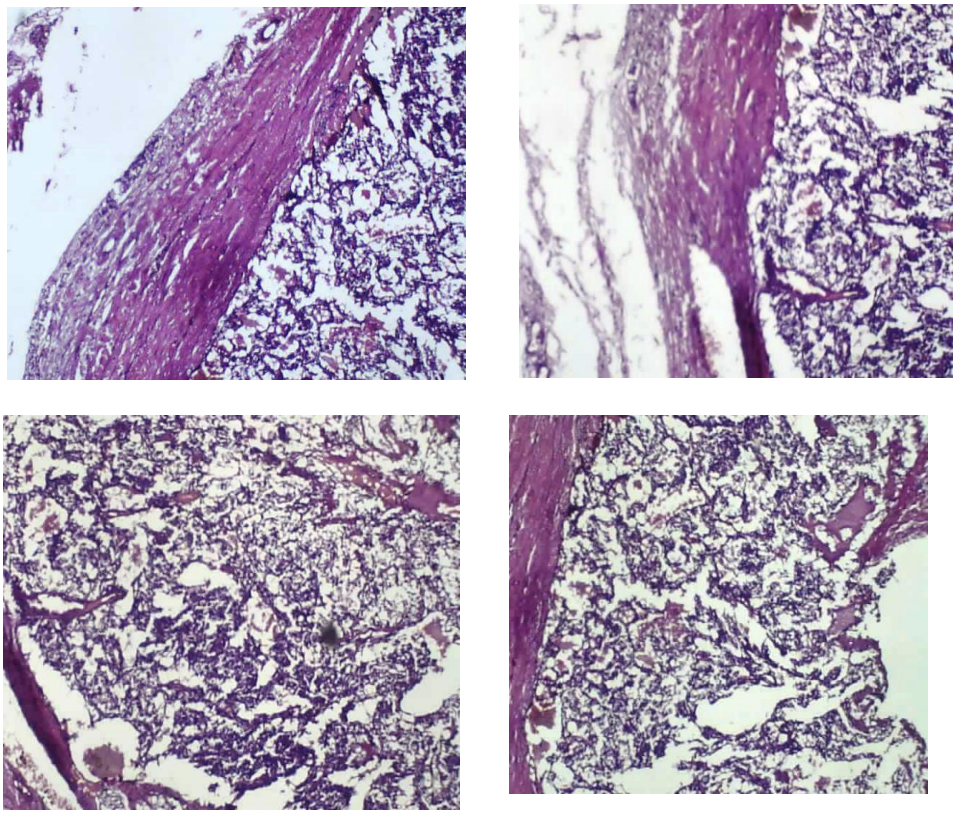

Figure 3: The tumor comprised branching pseudopapillary structures, solid sheets of fairly uniform cells with well defined capsule

The mean size of the tumor is variable, between 5.9 cm and 12.5 cm [7,9,10,17]. The typical ultrasonographic features of SPT includes a heterogeneous, encapsulated mass with solid echogenic and cystic hypoechoic components as well as peripheral calcifications, and displacement of surrounding structures[18,19,20]. The characteristic features of SPT on CT are a well-defined, low-attenuation mass with peripheral enhancement and complex cystic components with areas of necrosis and internal hemorrhage [9,21]. Differential diagnosis of SPT in young women with a solid and/or cystic pancreatic mass—based on the imaging modalities discussed above—would include serous cystic neoplasm, pancreatic endocrine tumors, cystadenoma, traumatic pseudocyst, acinar cell carcinoma, pancreatic lymphoma, papillary mucinous carcinoma, pancreatic pseudocyst, hydatid cyst, hepatoma, and hemangioma [9,21,22].

Metastatic disease does occur with SPTs with only 20 previously reported cases[9,19,23], the most common site of distant disease is the liver, very rare cases of peritoneal spread (n=4) and lymph nodes metastases (n=5) had reported [7], also there were patients found to have unresectable disease at initial presentation because of vascular invasion [7]

The mainstay of therapy is surgical resection, since this will achieve microscopic clear margins in most of the patients with more than 95% cure rate [12]. It is, however, recommended to make sure that the resection margins are tumor free, different types of operations done ; distal pancreatectomy with or without splenectomy[9,11,17], central pancreatectomy, central enucleation[20], and duodenopancreatectomy[24].

CONCLUSION :

SPTs should be put in differential diagnosis of cystic tumors of pancreas, and curative resection will be the best management.

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