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Case report

A solid pseudo-papillary tumor of the pancreas

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SUMMARY

Solid pseudo-papillary tumors (SPTs) of the pancreas are rare and typically present in young female patients. These are slowly growing masses that may attain large size, and are of low malignant potential. Surgical resection is curative in most of the cases. We presented the case of a young female referred to us with a large palpable epigastric mass. Preoperative diagnosis revealed a large SPT of the head of the pancreas. Surgical intervention, clinicopathological features, immunohistochemical findings and follow up are analyzed.

Key words: Solid pseudo-papillary pancreatic tumors, pancreatic tumors, cystic tumors

INTRODUCTION

Solid pseudopapillary tumors (SPTs) of the pancreas are rare. They have the form of an enlarging abdominal mass, and predominantly affect young women. These tumors are of unclear pathogenesis, low malignancy, and surgical resection offers an excellent chance for long-term survival.[1-5]

SPTs of the pancreas are uncommon neoplasms, accounting for 0.17% to 2.7% of the primary tumors of the organ[1-5]. This study reports a case of SPT of the head of the pancreas, in which the definitive diagnosis was confirmed by pathology

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CASE REPORT

A 29-year-old woman was admitted to our hospital with a large palpable mass in the epigastric region, which had become prominent the last two months, without any other accompanying symptoms. Barium swallow examination showed widening of the duodenal sweep and narrowing of the duodenal lumen (Figure 1). Ultrasound revealed a large pancreatic cystic lesion, while computed tomography (CT) (Figure 2) confirmed the existence of a cystic lesion having a diameter of about 18cm. CT strong-

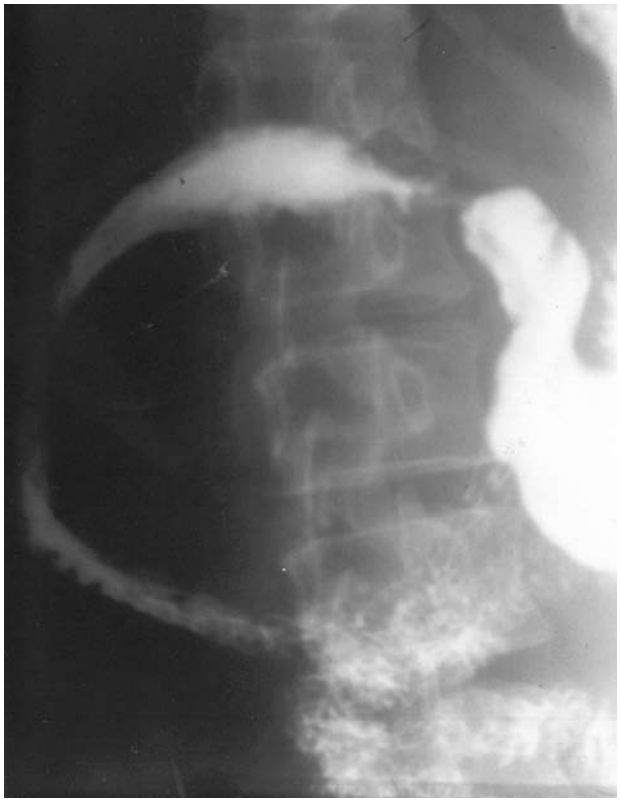

Figure 1. Barium swallow examination showing widening of the duodenal sweep and narrowing of the duodenal lumen.

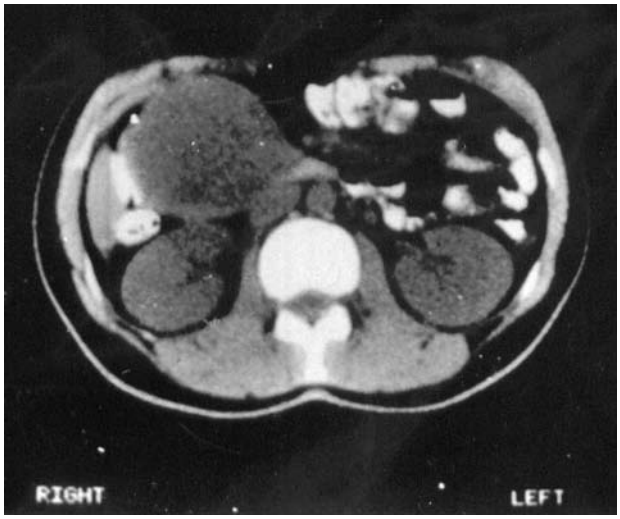

Figure 2. Computed tomography showing a cystic lesion of the head of the pancreas, 18cm in diameter.

ly supported the diagnosis of a pancreatic cystic neoplasm without any site of infiltration of the neighboring structures or diffusion of other intraabdominal organs.

At laparotomy, a cystic tumor of the head of the pancreas having the size of the head of a child and with hemorrhagic components was found. Pancreatoduodenectomy with preservation of the pylorus was performed (Figure 3). The postoperative course was uneventful and the patient was discharged on the 13th postoperative day.

Gross examination of the specimen showed a large encapsulated cystic neoplasm, with hemorrhagic and necrotic areas on cross sections. Microscopically, the most distinctive feature was the presence of pseudopapilla covered by several layers of epithelial cells. Their nuclei were avoided with few mitoses. The thick fibrovascular core often showed prominent mucinous change (Figure 4). In the central areas, the tumor cells were arranged around fibrovascular ribbons displaying papillary formations. These distinctive structures protruded into degenerated tissue including hemorrhagic cystic spaces. The neoplastic cells were found to infiltrate the cystic wall in some sites, without any invasion into the pancreatic tissue.

Follow-up consisted of CT scanning once a year, for the first five years, and thereafter once every two years. The patient, after a marriage and 3 successful pregnancies, is healthy and has no recurrence during this 15-year follow-up period.

DISCUSSION

SPT is a very uncommon pancreatic tumor that affects mainly women (F/M 10:1). The mean age of its appearance is 21.97 years (ranging from 2 to 85 years). Most of the patients are young (~22% are younger than 18 years), but a considerable 6% of the cases are older than 51 years.[5]

SPT generally becomes clinically evident as a palpable mass at the epigastrium, the left or the right hypochondrium. Furthermore, pain could also be one of the first signs of a SPT[5]. Rarely other symptoms can also exist, as a our recent case with hematemesis.[6]

The mean size of SPTs is around 6cm. Despite technical advances, pre-operative diagnosis is difficult. The differential diagnosis includes: disongogenetic cysts, retention cysts, pseudocysts and cystic tumors such as cys-

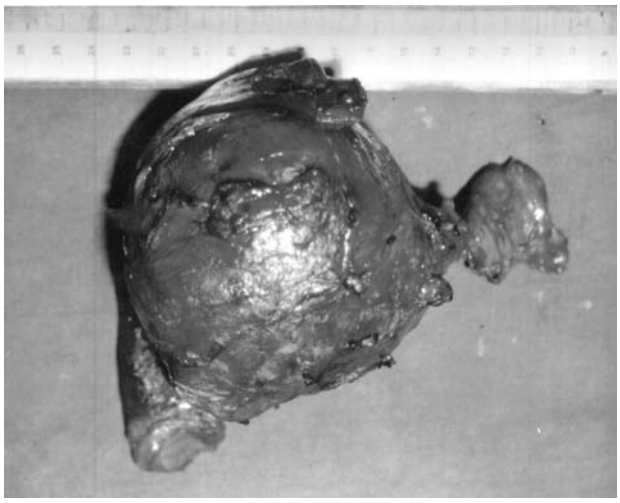

Figure 3. Surgical specimen of the tumor.

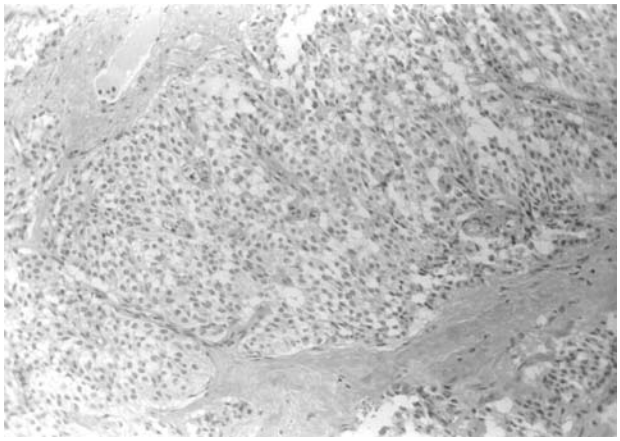

Figure 4. Papillary and solid epithelial neoplasm. Pseudopapillae covered by several layers of epithelial cells (H+E x 100).

tadenoma, cystadenocarcinoma, microcystic adenoma, lymphangioma, hemangioma, angiosarcoma and acinary cell cystadenocarcinoma. Percutaneous fine needle biopsy of the cystic wall and aspiration of the cystic component may be useful and may influence the subsequent management.[5]

Concerning the surgical approach, a great number of techniques are employed. The low grade of malignancy of this tumor has led some surgeons to perform simple enucleation of the neoplasm. However, distal pancreatectomy with splenic preservation or pancreatoduodenectomy, depending on the location of the tumor, represents the procedure of choice.[5]

In general, prognosis even in the case of a malignant SPT with metastasis is favorable. Some patients with "unresectable" tumors or those with hepatic metastasis have survived more than 10 years after the operation.[1,3-5]

Macroscopically, the tumor presents a well-circumscribed fibrous capsule and focal or widespread hemorrhage and cystic changes.[4] Microscopically, thin papillary structures are clearly made up of layers of cells on a fibrovascular axis along with solid areas of cellular laminae mixed with hemorrhagic zones of various sizes. Nuclei are avoided and folded with indistinent nucleoli and few mitosis. Hyaline globules and collections of foamy cells may be present. The thick fibrovascular core often shows prominent mucinous changes. It is not clear whether this tumor originates from ductal cells, acinar cells or primitive ones. Ultrastructural evidence of acinar, ductal and sometimes endocrine cell differentiation has been found. In addition, local positivity for neuron specific enolase, a1-antitrypsin and a1-antichymotrypsin was found. These findings suggest that papillary and solid epithelial cells predominantly have exocrine features but with the capacity for dual (exocrine and endocrine) differentiation.

In our case, the most noteworthy observations concern the large size of the tumor, the radical treatment by pancreatoduodenectomy with preservation of the pylorus, the immunohistochemical findings which support the opinion that these tumors could be derived from both the exocrine and endocrine pancreas, and the long survival with a good quality of life without any disease recurrence. Findings in accordance with the international literature support the concept that, despite the large size of these tumors and their ability to extend locally, complete surgical excision offers benefits in almost all cases. SPTs are of low-grade malignancy and potentially curable by extended resections of the primary tumor mass. Concerning the surgical approach, the international literature supports the that surgery must be as conservative as possible, with particular respect to the radical oncology, because of the relative low malignancy and the encapsulated form of the neoplasm.

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