CASE REPORT

Solid Pseudopapillary Tumor of the Pancreas in a Young Malay Woman

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Summary

Solid pseudopapillary tumors of the pancreas are very rare, low-grade malignant potentially curable neoplasms. Despite having non-specific symptomatology, they have typical features such as being more common in young women, and classically presenting as large abdominal masses. Accurate diagnosis is important because long-term survival hinges on complete resection of the tumor.

Key Words: Pancreas, Pancreatic neoplasms, Solid pseudopapillary tumor

Case Study

An 18-year-old previously healthy Malay woman was referred by her local doctor to a hospital in Singapore in 1998 for a deep painless lump located in the left hypochondriac region, roughly 5 x 5cm. Initially the patient refused treatment for non-medical reasons. During 2000, the patient became pregnant and gave birth to a healthy baby in early 2001. It was during 2002 when the lump continued to increase in size and cause occasional severe pain that the patient decided to seek treatment. The pain was intermittent over the left upper abdomen. Symptoms such as weight loss, lethargy, altered bowel habits and malaena were absent. There was also no history of abdominal trauma, alcohol abuse or pancreatitis.

On examination, the patient was comfortable and not jaundiced. A 10cm well defined non-tender, non-pulsatile mobile mass was palpable in the left hypochondriac region and extending to the left flank. No other masses or lymph nodes were palpable.

The patient was mildly anaemic (Hb 10.7g/dL), but the liver function tests, amylase, and tumor markers CEA, CA-125, CA-19-9, alpha-FP, were all within normal limits. A CT scan of the abdomen (Figure I.) revealed a mass 10.6 x 8.1cm of soft tissue density with a heterogeneous appearance. It appeared to arise from and lie laterally to the stomach. The hepatobiliary structures and spleen were normal, and there was no evidence of abdominal lymph node metastases. At this stage, the impression was that the lump appeared to be a leiomyosarcoma arising from the body of the stomach. However, a gastroscopy was performed, showing that the body of the stomach was compressed externally, thus making a leiomyosarcoma unlikely.

Despite having an uncertain preoperative diagnosis, it was decided to surgically resect the tumor. It was noted during laparotomy, that the tumor was attached to the posterior border of the tail of the pancreas and the transverse mesocolon, but not attached to the splenic vessels or omentum. The tumor was completely dissected off the pancreas and measured 13 x 10 x 11cm covered by an intact fibrous capsule. On cross section, it was composed of mixed cystic and solid components, with haemorrhagic areas. Histopathology revealed sheets of cells with moderate

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amounts of eosinophilic cytoplasm. Prominent fibrovascular septae with a pseudopapillary pattern were present. Small cystic spaces with eosinophilic material, areas of myxohyaline degeneration, and aggregates of foamy macrophages were noted. Normal pancreatic tissue surrounded the tumor. The histological appearance of the lump was suspicious of a solid pseudopapillary tumor of the pancreas. The diagnosis was then confirmed by immunohistochemical staining the tumor cells positive for alpha-1-antitrypsin, vimentin and NSE (Neuron-specific enolase).

The patient made an uneventful recovery, is presently well and being regularly followed up at the hospital outpatient clinics.

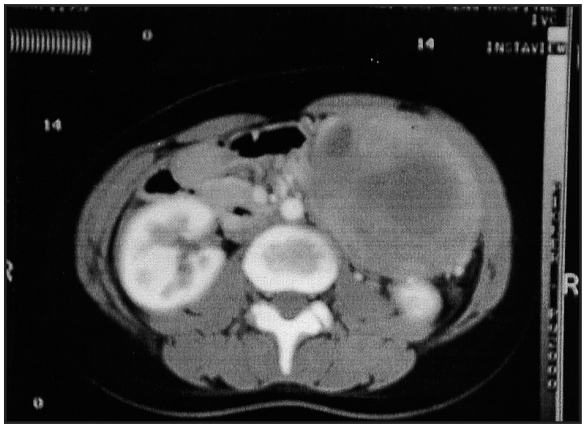

Fig. I: Abdominal CT scan showing a large well-circumscribed mass of soft tissue density with a heterogeneous appearance

Discussion

Since the first description of solid pseudopapillary tumor (SPT) of the pancreas by Virginia Frantz in 1959, just over 400 cases have been reported in the world literature. It is therefore a very uncommon tumor, representing about 1% of all pancreatic neoplasms, and is roughly 9.5 times more common in females, with the mean age of diagnosis being 27 years[1]. There is also an overrepresentation among blacks and east Asian racial groups.

Since 1959, synonymous names used for this tumor have included 'Frantz tumor', 'solid and papillary tumor', 'papillary cystic tumor', 'solid cystic tumor', and 'solid, cystic, and papillary epithelial neoplasm', reflecting its variable macroscopic and histological appearance. Although it is classified as a pancreatic epithelial tumor, the cell of origin of this tumor is not well understood because normal adult pancreatic cell lines (ductal, acinar or endocrine) are not mimicked by the cells of SPT[1]. Hence their origin may arise from primitive multipotential pancreatic cells. The preponderance of cases in women and during the second or third decade of life, as in our patient, suggests that sex related/hormonal factors (e.g. steroid receptors) play a role in its pathogenesis[2]. To the best of our knowledge, there has only been one reported case of accelerated growth of a SPT during pregnancy[3]. The size of our patient's SPT roughly doubled in size during a period of 4 years, which included the duration of her pregnancy. Our case illustrates that pregnancy may have a role in accelerating the growth of SPT.

Macroscopically, SPT are well demarcated, spherical masses measuring 3-18cm, with an even distribution in the pancreas. Smaller SPT appear as soft, tan to red tumors with variable amounts of fibrosis, and often appear unencapsulated. Larger SPT often have a fibrous pseudocapsule around it, and possess large areas of haemorrhage and cystic degeneration inside with ragged fronds, although some may be entirely fibrotic[1]. The microscopy of SPT show two characteristic patterns. The first one consisting of solid sheets of polygonal eosinophilic cells, with prominent nucleoli, and the other of cyst like spaces with pseudopapillae. The diagnosis of SPT can be confirmed by immunohistochemical staining of tissue, as was the case in our patient. The most consistent markers with the strongest immunoreactivity are vimentin, alpha-1-antitrypsin, and NSE, each present in over 90% of SPT[4].

The usual presenting symptoms of SPT are vague and non-specific. Common presentations include upper abdominal discomfort caused by an enlarging mass, or a mass felt by the patient. The tendency for slow growth results in most SPT being asymptomatic until it has reached a considerable size, as in our case study. It is not uncommon for SPT to be detected incidentally, or cause symptoms after trauma inducing bleeding into the mass. Jaundice on presentation is exceedingly rare, and there are no known associations with any paraneoplastic syndromes[1].

Pancreatic function tests, liver functions tests and tumor markers such as CEA, CA19-9 are usually normal,

although anaemia may be due to bleeding from the cyst[2]. Imaging by ultrasound and CT scans remain the most useful preoperative investigations for SPT. Scans typically show a large (average diameter 8-10cm), well circumscribed mass located in the pancreas, and surrounded by a pseudocapsule. There are solid, cystic or mixed components, and if calcifications are present, it is highly specific for SPT[2]. The CT scan done in our case study showed a large encapsulated mass, but no calcification. Due to the lack of calcification and its intimate relation to the body of the stomach, a provisional diagnosis of leiomyosarcoma was made, and a pancreatic tumor was not considered as likely. In adults, an endoscopic ultrasound scan with fine needle aspiration (EUS-FNA) is commonly used in the evaluation of cystic pancreatic masses, and is suggested when a SPT is suspected[2]. Fine needle aspirates which show pseudopapillae, and bland tumor cells containing acidophilic, PAS positive and diastase resistant cytoplasmic granules are suggestive of SPT. However, an EUS-FNA was never attempted on our patient since neither a cystic pancreatic mass nor a SPT were suspected.

The treatment of choice for SPT is surgical resection irrespective of size or stage of the disease, unlike some other pancreatic tumors. A recent review done by Martin et. al.[5] concluded that SPT of the pancreas should be treated aggressively with attempts made at complete resection, even if it requires resection of proven metastases, especially hepatic metastases. Complete resection of the tumor is usually achieved by distal pancreatectomy or partial duodenopancreatectomy, depending on the location of the tumor. Tissue sparing surgery is possible if there is no spread to adjacent structures, which is often the case since a dense fibrous pseudocapsule often surrounds the mass. There is no clear role for adjuvant chemo-radiotherapy[1,5].

If left untreated, SPT runs a malignant, though indolent course. Very few patients have died from this tumor. Recurrence of SPT has not been reported with complete resection of local disease, therefore it is a tumor which generally carries a good prognosis[5]. There are few reports in the literature of long term results evaluating follow up management. However, we suggest that regular follow up of the patient is important since recurrent disease can still be responsive to treatment.

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