中文題目：實性假乳突狀瘤併有組織學上侵犯：一種少見的胰臟腫瘤

英文題目：Malignant solid pseudopapillary neoplasm with histological lymph/vascular/ perineural invasion in a young female：A rare pancreatic tumor

作者：王宿鴻[1]，郭行道[2]

服務單位：財團法人奇美醫院永康總院肝膽胃腸科

Abstract

Solid pseudopapillary neoplasm (SPN) is a rare pancreatic tumor. It most affects female patients within their second or third decade. It has low potential to become malignancy. Surgery is the mainstay choice of treatment.

Case report

A 32-year-old female presented to the emergency department with complaint of sudden onset epigastric cramping pain since 3 hours ago. She had underlying disease of type 2 diabetes mellitus, essential hypertension and chronic epilepsy, following up at our clinics regularly. She denied vomiting, jaundice, body weight loss or bowel habit change. Physical examination revealed significant tenderness over the upper abdomen. Her Vitals, basic blood panels including liver function tests were all within normal limits. Enhanced-contrast computed tomography (CT) scan showed an encapsulated mass, about 4.7x3.7cm in size, with mild enhanced central portion at body of the pancreas. (Fig.1) In attempt to identify this lesion the patient had a magnetic resonance imaging (MRI) scan which confirmed a 4.2cm size solid, hypovascular tumor with peripheral enhancement and compression of splenic vein. (Fig.2) She also received endoscopic ultrasound (EUS) which showed hypoechoic and hypovascular mass without liquid part. (Fig.3) However, fine needle aspiration was not performed because of the nearby vessel. Tumor marker including CEA and CA199 were normal. According to above finding, solid pseudopapillary neoplasm was considered. The patient received distal partial pancreatectomy five weeks later. The specimen revealed a 4.5cm mass over pancreatic body and tail with thickened wall and >90% tumor necrosis. (Fig.4) During operation it was difficult to separate splenic vessel from pancreas, so splenectomy was performed before pancreatectomy. Histopathology showed ill-circumscribed tumor with microscopic

pseudopapillae and sheets of polygonal cells, extending beyond the pancreas but without involvement of celiac axis, the superior mesenteric artery, lymph node or spleen. However, it presented lymph, vascular and perineural invasion, representing its malignant nature. The neoplastic cells are immunoactive for vimentin, CD10, but not Chromogranin A, confirming the diagnosis of malignant SPN without distant metastasis. After surgery, there were no special events or additional treatments.

Discussion

Solid pseudopapillary neoplasms (SPNs) are rare pancreatic tumors, accounting for about 1-2% of pancreatic exocrine tumor.[1,2] It was first described by Virginia Kneeland Frantz in 1959.[3] At 1996, the World Health Organization gave it the official name: Solid pseudopapillary neoplasm, based on its pathological features.[4] In recent years, the incidence of SPNs increases with the improvement of imaging modalities. SPNs primarily affect 2nd to 3rd decade young female with a median age of 22 years.[2] About 90% of SPNs occur among female patients.[2] The majority of patients are asymptomatic and they are usually diagnosed by incidental finding on image performed for other unrelated reasons, although some patients may complain non-specific abdominal pain, nausea or vomiting, especially in the situation of solid tumor hemorrhage.[5] Unlike pancreatic adenocarcinoma, SPNs rarely cause jaundice even the tumors are large or locate at pancreatic head, maybe because of the soft nature of these tumors.[2] Rare complications include rupture and hemoperitoneum, coagulopathy, cholangitis, sepsis and death. Generally, they are slow growing tumors and have good prognosis. Because of the indolent course, they are often large with a mean diameter of 9 cm at diagnosis.[6] The most common site of the tumors is head and tail.[2] SPNs still have low potential to become malignant. Pathologically, SPNs were classified as malignant if it revealed perineural, vascular, extrapancreatic, pancreatic parenchyma invasion, or distant metastases. [7]

On image, SPNs usually have mixed solid and cystic components with regions of calcification, hemorrhage or cystic degeneration.[8] On computed tomography, SPNs typically revealed well-demarcated, encapsulated pancreatic lesion compromising both solid and cystic components. The cystic component usually locates at central area while the solid component at peripheral area

with calcifications.[9] In contrast to other pancreatic neoplasm, SPNs tend to have similar peripheral enhancement with the surrounding pancreatic parenchyma during both arterial and venous phases. Compared to CT, magnetic resonance imaging could demonstrate well-circumscribed lesions with a heterogeneous signal intensity on T1- and T2 weighted images, and could show the presence of hemorrhage within these tumors, which is a very characteristic feature of SPNs.[10] The clinical or imaging features for predicting turning malignant are still controversial. It is also unclear that which pathological or clinical factors could differentiate between benign and malignancy, or predict tumor recurrence/metastasis. According to one recent published data, high-grade malignancies, synchronous metastases, peripancreatic fat infiltration, and lymphovascular invasion correlated with the increasing recurrence rate.[11] The other published data said that focal discontinuity of capsule was more common in malignant tumors. There were no differences between benign and malignant tumors based on location of tumor, tumor margin, proportion of solid component, growth pattern, presence of calcification, presence of upstream pancreatic duct dilatation, or enhancement pattern.[12]

The mainstay of therapy is surgical resection. Pre-operative histologic examination could be done by EUS-guided fine needle aspiration. The sensitivity and specificity is up to 70%, respectively.[13,14] The retrieved cells are usually ovoid or polygonal in shape with typical small central nuclei and abundant cytoplasm. More than 90% of these tumors stain positive for Vimentin, enolase, alpha1 antitrypsin, alpha1 antichymotrypsin, beta-catenin and negative for E-cadherin, chromogranin, and CK19. [15]

As in our case, it was difficult to differentiate between benign or malignancy according to clinical presentation or imaging finding, so surgical en-bloc resection could be the only therapeutic option and a curative management for all patients, even in patients with distant metastasis, because long-term survival could be predicted. The overall survival rate is generally >95%.[11] Microscopic or distant metastasis is not contraindication for surgical resection. However, it is difficult to identify the predictive factors for evaluating survival because of the long survival rate of SPNs patients with local

recurrence or distant metastasis. The local recurrence rate is less than 10% within 5 years after surgery. Follow-up is essential, and surgery is still the treatment of choice in such situation, whereas radiotherapy or chemotherapy has no clear role in SPNs.

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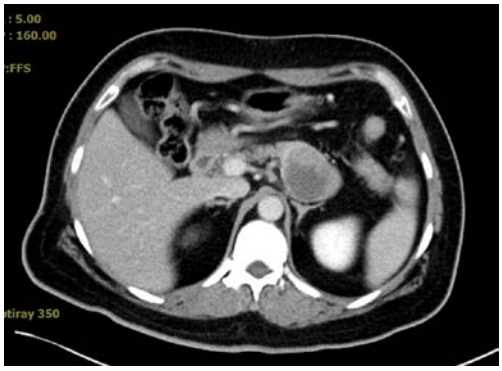

Fig 1

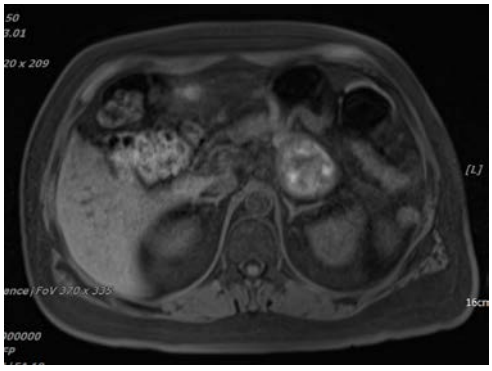

Fig 2

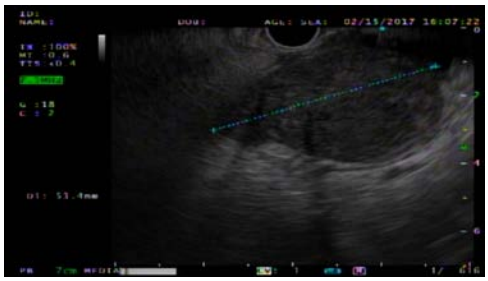

Fig 3

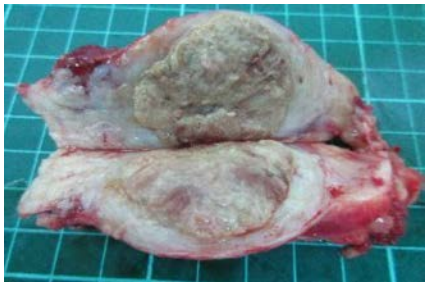

Fig 4