Solid pseudopapillary tumor of the pancreas in a pregnant woman

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Abstract

Solid pseudopapillary tumor (SPT) of the pancreas is a rare neoplasm of low malignant potential that mostly affects young women in the second or third decade of life. The number of such patients reported in the literature has increased in recent years, while SPT in pregnancy is extremely rare. To the best of our knowledge, only one case of SPT in pregnancy has been reported in the English-language literature. We herein report a case of asymptomatic SPT in a 26-year-old Chinese female in the 14th week of pregnancy, and present our experience of the surgical management of SPT in pregnancy. (Acta gastroenterol. belg., **2011**, 74, **560-563**).

Key words : solid pseudopapillary tumor (SPT), pancreatic neoplasm, pregnancy.

Introduction

Solid pseudopapillary tumor (SPT) of the pancreas, first reported by Frantz *et al* (1) in 1959, is an uncommon but distinct pancreatic neoplasm with a low malignancy and accounts for about 1-2% of exocrine pancreatic tumors (2,3). The tumor has been given several different names according to its macroscopic and microscopic pathological character until this name, solid pseudopapillary tumor of the pancreas, was defined by the World Health Organization (WHO) as a unique tumor in 1996 (4). With the widespread availability of high-quality imaging and a better understanding of its pathology, the number of such patients reported in the literature has steadily increased. SPT predominantly affects young women in their second or third decade of life (5), while SPT in pregnancy is extremely rare. To the best of our knowledge, only one case of SPT in pregnancy has been reported in the English-language literature (6).

We herein report a case of asymptomatic SPT in a 26-year-old Chinese female in the 14th week of pregnancy, and present our experience of the surgical management of SPT in pregnancy.

Case report

A 26-year-old Chinese female in the 14th week of pregnancy was admitted to our department with an abdominal mass, which was accidently detected by ultrasonography (US) in prenatal care. Prior to this presentation, she had been healthy, with no previous surgery, weight loss, abnormal pap smears, sexually transmitted diseases or medical illnesses. She denied any history of blood transfusion, alcohol abuse, or medications. Her family history was normal. On physical examination, abdominal palpation revealed tenderness in the middle quadrant, and a complex mass about 10 cm × 8 cm in the middle upper quadrant with minimal mobility.

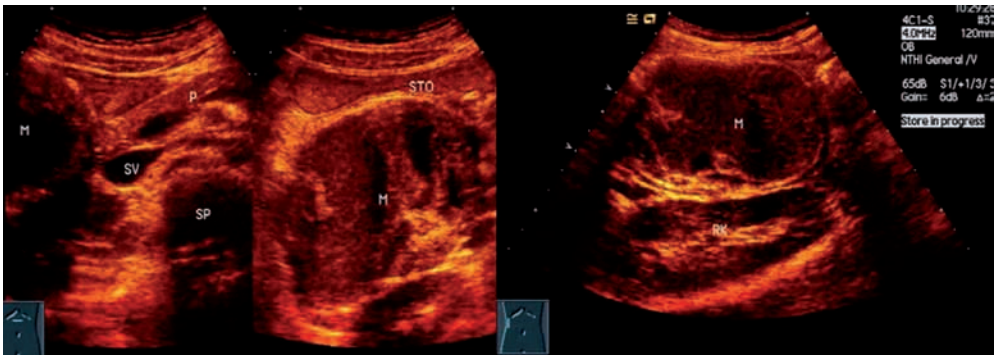

Fig. 1. — US revealed a well-circumscribed inhomogeneous mass with intact fibrous capsule, measuring about 9.5 cm × 6.2 cm × 9.0 cm, in the right of pancreatic head.
Abbreviation : M-mass ; SV-superior mesenteric vein ; SP-spleen ; P-pancreas ; STO-stomach ; RK-right kidney.

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Laboratory data as well as tumor markers, such as CEA and CA19-9, were normal. US revealed a well-circumscribed inhomogeneous mass with an intact fibrous capsule, measuring about 9.5 cm × 6.2 cm × 9.0 cm, in the right of pancreatic head (Fig. 1). Although computed tomography (CT) is usually the next imaging technology following US in the diagnosis of pancreatic neoplasms, in view of the pregnancy, magnetic resonance imaging (MRI) was performed to verify a giant solid-cystic mass with T1- and T2- weighted images in the right of pancreatic head (Fig. 2).

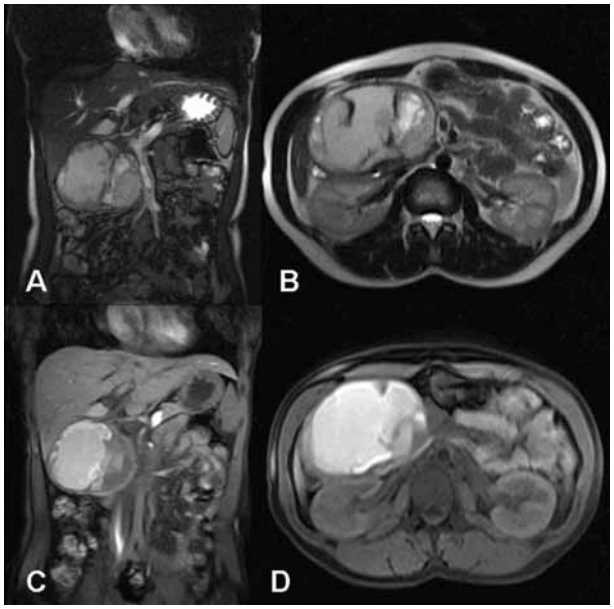

Fig. 2. — MRI was performed to verify the giant solid-cystic mass with coronal (A) and axial (B) T2- and coronal (C) and axial (D)T1- weighted images in the right of pancreatic head.

According to the imaging, it was assumed to be a pancreatic pseudocyst or pancreatic cystadenoma and a fine needle biopsy guided by US was performed (Fig. 3A). The puncture fluid was hemic, the smear showed 2-3 WBC per HPF and RBC all field/HPF (Fig. 3B). Amylase determination in the puncture fluid was negative (0 U/L). The needle biopsy showed tumor cells that were composed of papillary structures, with lots of neoplastic epithelial cells polygonal in shape (Fig. 3B). According to the needle biopsy and imaging, the diagnosis of SPT was suspected. Gynecological surgeons were consulted as the patient was in the 14th week of pregnancy. Surgery was considered as being relatively safe in the second trimester of pregnancy, and a peridural anesthesia was proposed as an ideal procedure for surgery. The surgery was performed in the 14th week of pregnancy. At laparotomy, a successful tumor enucleation was performed. There were no signs of metastases or any visible neoplastic tissues remaining at the operation site. Microscopically, the pancreatic tumor showed marked cellular proliferation in the solid areas that alternated with a pseudopapillary and cystic pattern (Fig. 4A-B).

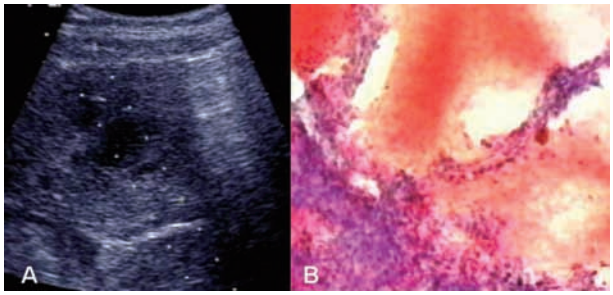

Fig. 3. — The needle biopsy guided by US was performed (Fig. 3A). The smear showed WBC 2-3/HPF and RBC all field/HPF. The tumor cells were composed of papillary structures, with lots of neoplastic epithelial cells polygonal in form (Fig. 3B).

Immunohistological results revealed that the tumor cells were positive for Vimentin (Vim), Cytokeratin (CK), CD56, CD10, Neuron specific enolase (NSE) and Synaptophysin (Syn) (Fig. 4 C-H). The Ki-67 proliferation index was less than 5% (data not shown). Based on these histological findings (7), the diagnosis of SPT was confirmed. According to the WHO criteria of malignancy, the tumor has low malignant potential.

In the early postoperative period, prophylactic tocolysis was carried out. Blood glucose monitoring during pregnancy and post partum showed normal carbohydrate metabolism. On the 3rd postoperative day, a pancreatic fistula occurred. The amylase in the liquid of abdominal drainage was 966 U/L. US detected a local opaque, dark area of fluid measuring about 52 mm × 32 mm in the upper middle abdomen (Fig. 5A), and the fetal position was good (Fig. 5B). Accordingly, the removal of abdominal drainage tube was delayed. Octreotide was not used in view of its side effect in pregnancy. On the 25th postoperative day, the abdominal drainage tube was removed as the liquid was less than 5 ml. After 28 days of hospital stay, the patient was discharged in a good general condition. On the 38th week, a healthy, mature girl with an Apgar score 9/10 was born by cesarean section.

Discussion

Solid pseudopapillary tumor (SPT) of the pancreas, first described by Frantz in 1959, forms about 1-2% of exocrine pancreatic tumors. The pathogenesis of SPT has not been determined yet, but accumulating evidence suggests a neuroendocrine origin (8,9). Progesterone has been proven to play an important role in the development of SPT (5,10). Only one case of SPT in pregnancy has been reported so far (6), in spite of the fact that pregnancy is the most common condition with high progesterone levels, which would suggest much higher incidence of this neoplasm.

The initial presentation of SPT is usually nonspecific. It is a non-functional, slow-growing neoplasm that very often reaches considerable size before the first symptoms appear (11,12). Papavramidis *et al* (5) summarized 718 SPT patients in English literature, showing that upper

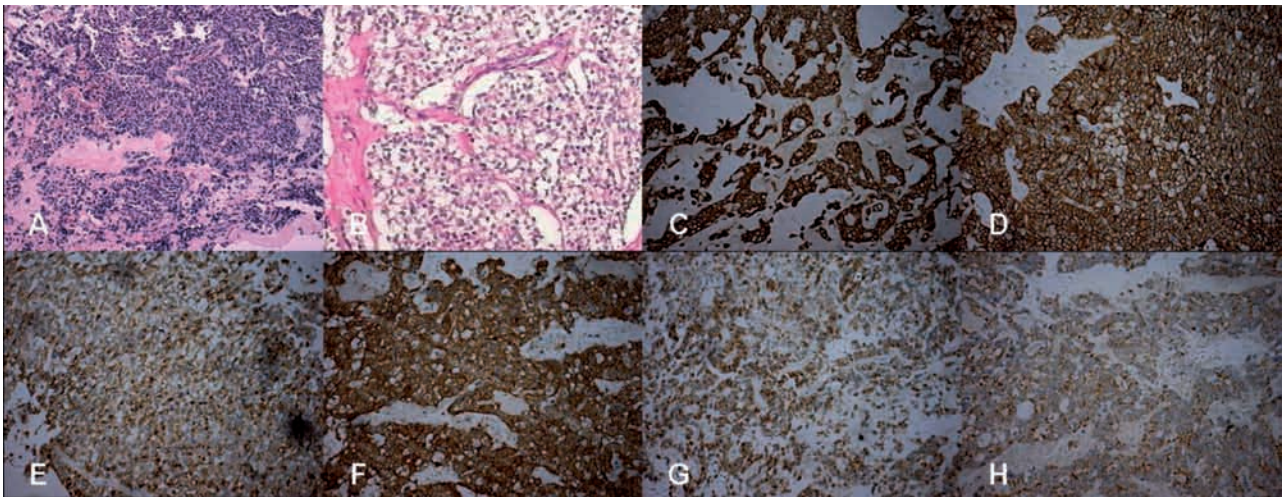

Fig. 4. — Microscopically, the pancreatic tumor showed marked cellular proliferation in the solid areas that alternated with a pseudopapillary and cystic pattern (Fig. 4A HE : 40×). The solid areas show sheets and cords of cells arranged around fibrovascular septa (Fig. 4B HE : 200×). Immunohistological results revealed that the tumor Vim (Fig. 4C), CD56 (Fig. 4D), Syn (Fig. 4E), NSE (Fig. 4F), CD10 (Fig. 4G), and CK (Fig. 4H) were positive (200×).

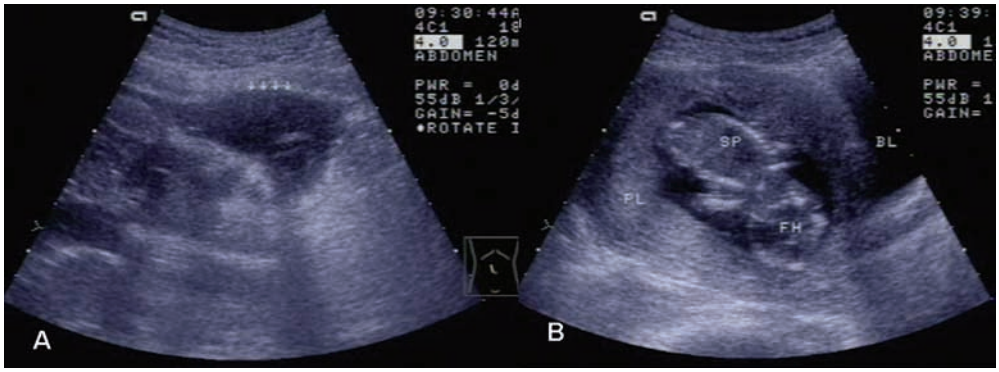

Fig. 5. — US detected a local opaque, dark area of fluid (small arrows) measuring about 52 mm × 32 mm in the upper middle abdomen (Fig. 5A), and the fetal position was good (Fig. 5B).
Abbreviation : PL-placenta ; SP-spine ; FH-forehead ; BL-bladder.

abdominal pain is the most common symptom (46.5%), followed by a slowly enlarged, palpable, non-tender upper abdominal mass (34.8%). Asymptomatic cases are reported in 15.5% of cases. The tumor size can be quite large (mean diameter of 6.08 cm) at diagnosis with more than 5 cm in diameter in 83% of cases. The most common location of the SPT is the tail (35.9%) and the head (34%) of the pancreas (5). Both, the previous reported case of SPT in pregnancy (7.2 × 8.0 × 5.4 cm) and our case (9.5 × 6.2 × 9.0 cm) had tumors larger than 5 cm located in the pancreatic head. In conclusion, a large cystic-solid mass in the pancreas of a young pregnant woman should raise suspicion for SPT.

Accurate preoperative diagnosis of SPT is difficult because of the similarity of the findings among cystic lesions of the pancreas (5,13). As a part of a general investigation, the first imaging technique is US showing a well-circumscribed inhomogeneous mass in the epigastrium. Following US, CT scan is the next most common imaging technique. In our case, CT scan, in view of the pregnancy, was not performed and thus MRI was preferred to verify a giant solid-cystic mass with T1- and T2- weighted images in the right of pancreatic head. If MRI reveals an encapsulated mass with solid and cystic components as well as hemorrhage without obvious internal septum, SPT should be highly suspected (14). In our case, the fine needle biopsy guided by US was performed. The puncture fluid was hemic, and a smear showed WBC 2-3/HPF and RBC all field/HPF. The needle biopsy showed tumor cells that were composed of papillary structures, with lots of neoplastic epithelial cells polygonal in shape. According to the needle biopsy and imaging, the preoperative diagnosis of SPT was highly suspected.

SPT exhibits pathological features and is diagnosable based on the classic histological appearance (15,16). Microscopically, the growth pattern of the tumor cells is remarkably uniform, with a combination of solid,

pseudopapillary, or hemorrhagic pseudocystic structures in various proportions (17). However, SPT and pancreatic endocrine tumor (PET) may be very similar and the results of immunohistochemistry showed that expression profiles of the two tumors overlapped, which sometimes results in difficulty in distinguishing the two entities. Recently, it was shown that ß-catenin and E-cadherin immunostaining can be applied to make definite differentiation between SPT and PET (18). Immunoreactivity for ß-catenin is found in the cytoplasm and the nuclei of almost all tumor cells in the majority of SPT (8,19). Loss of membrane staining and/or nuclear staining for E-cadherin is seen in 100% of cases of SPT of the pancreas (20).

At present, radical resection is the treatment of choice for SPT even with metastasis or local extension (5, 12,21). The surgical approach depends on the location, size, and nature of the neoplasm as well as time of surgery. On evaluation of international literature, a surgical procedure providing R0 resection with limited radicality seems to be an acceptable therapeutic strategy in SPT (22,23). In the previous case, although the patient underwent a successful Whipple procedure at 16 weeks of gestation, this did not ameliorate abdominal pain, nausea or vomiting. In our case, because the mass was surrounded by a dense fibrous capsule in a pregnant woman, a simple enucleation of the neoplasm was performed. There were no signs of metastases or any visible neoplastic tissues remaining at the operation site. Surgery is relatively safe in the second trimester of pregnancy. In conjunction with gynecologists, the large SPT was removed with the R0 resection of the head of the pancreas.

In conclusion, our case provides convincing evidence on the fact that SPT may present in pregnancy, and surgical resection could be successfully performed during pregnancy with the preservation of the foetus's life. The cooperation between gynecologists and surgeons, the appropriate timing of the operation, the precise surgical technique followed by careful postoperative care are all essential requirements of the successful therapy.

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