Case Report

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Pregnancy Complicated With a Giant Pancreatic Tumor and Decompensation of Liver Cirrhosis: A Case Report and Literature Review

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Abstract

Pregnancy with solid pseudopapillary tumor of the pancreas (SPTP) is rare. Because pregnancy hormones may cause tumor progression, the management and treatment of SPTP need to balance the safety of pregnant women and fetuses with surgical treatment. We reported a case of a giant pancreatic tumor diagnosed during pregnancy that was considered to be SPTP. Examinations also showed hepatitis B virus infection and severe decompensation of liver cirrhosis. Medical termination of pregnancy was performed. The patient has lived with the tumor until now without surgery. We retrieved the published case reports, summarized the clinical characteristics of pregnancy with SPTP, and explored its management during the perinatal period. Most patients with SPTP have a good prognosis with good maternal and fetal outcomes, and it is important to choose an appropriate treatment method and timing. However, pregnancy combined with decompensated liver cirrhosis needs to be terminated in a timely manner because of its high-risk status.

Keywords: Pregnancy complications; Pancreatic tumor; Solid pseudopapillary tumor of the pancreas; Liver cirrhosis decompensation; Abnormal coagulation function

Introduction

Solid pseudopapillary tumor of the pancreas (SPTP) is a rare low-grade malignant tumor that is particularly rare during pregnancy, but pregnancy likely accelerates the growth of SPTPs, which can be life-threatening.[1] So, it is hard to balance both maternal and fetal risk. Therefore, the treatment of SPTP during pregnancy remains a clinical challenge, and there has no clinical consensus accomplished on it. The incidence of pregnancy with liver diseases is approximately 3%, including both pregnancy-related and preexisting liver diseases.[2] Cirrhosis-complicated pregnancy is a major condition that considerably increases the morbidity and death of mothers and babies.[3] It is important to monitor the disease and terminate pregnancy timely to avoid tragic outcomes. Herein, we present a case of pregnancy with a giant pancreatic tumor considered to be an SPTP and decompensation of liver cirrhosis. We expected to determine a more appropriate clinical management by evaluating the relevant literature. The institutional review board from Peking Union Medical College Hospital approved the study. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Case presentation

A 30-year-old woman, gravida 2 para 1, was admitted to the obstetrics and gynecology ward of our hospital because of "25 weeks and 6 days of gestation, gingival bleeding and abdominal mass found for 2 months" in 2018. She never had a hepatitis B virus test and had no clinical manifestations before pregnancy, nor did she get prenatal care during pregnancy. Since gestation week 18, she had progressive gingival bleeding, systemic edema, abdominal pain and distension. Hepatitis B virus DNA level was 5,897,000 IU/mL. The ultrasound image and computed tomographic (CT) scan showed cirrhosis liver, a well-circumscribed nonhomogeneous mass (12.2 cm × 9.3 cm × 8.0 cm) occupying the left lobe of the liver, portal hypertension, thrombus formation of the portal system, enlarged spleen, and ascites. Blood tests showed severe anemia (hemoglobin, 67 g/L) and thrombocytopenia (platelet, 20 × 10^9/L), abnormal coagulation function (prothrombin time, 18.5 seconds; activated partial thromboplastin time, 57.4 seconds; fibrinogen, 0.88 g/L; D-dimer, 2.82 mg/L fibrinogen equivalent unit), and abnormal biochemical indicators (total bilirubin, 43.3 μmol/L; direct bilirubin, 30.0 μmol/L). The tumor markers were elevated: CA125, 102.2 U/mL (normal range, 0-35.0 U/mL); CA199, 72.7 U/mL (normal range, 0-35.0 U/mL); and α-fetoprotein, 75.6 ng/mL (normal range, 0-25.0 ng/mL). Pathological biopsy was not performed because of coagulation dysfunction and thrombocytopenia secondary to liver cirrhosis, so the diagnosis of SPTP was made according to clinical

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manifestations and image examination. The multidisciplinary team discussed that surgery was not suitable due to poor coagulation function; considering the severity of liver cirrhosis and that the tumor might be stimulated by hormones during pregnancy, it was recommended to terminate pregnancy immediately. After infusion of blood products to improve coagulation function, subsequent misoprostol-induced vaginal labor was conducted and uterine gauze packing was used for postpartum bleeding. The patient gave up resuscitating the newborn for financial reasons and was discharged 5 days after delivery. A pancreatic thin-layer CT scan was performed postpartum and showed that the tumor's diameter exceeded 10 cm and that it invaded and obstructed the portal vein and superior mesenteric vein (Figs. 1 and 2). Although the tumor was unresectable, an annual CT scan confirmed that its diameter did not increase.

Literature review and discussion

Using “Pregnancy” and “Solid pseudopapillary tumor of the pancreas” as keywords, we searched the PubMed and China National Knowledge Infrastructure databases from construction to January 13, 2022. A total of 17 related case reports were retrieved. The clinical characteristics of pregnancy combined with SPTP were summarized. The patients' median age was 26 years (range, 19-39 years). Except for one instance in which the tumor was discovered before pregnancy, the median gestational weeks of onset was 14 (range, 4-28). The most prevalent clinical symptoms were stomach pain, abdominal mass, nausea, and vomiting. Only one case was asymptomatic, and the tumor was found on a routine prenatal ultrasound. Four cases had artificial or spontaneous abortions during early or middle pregnancy, with one undergoing surgery 1 week before the abortion and three undergoing surgery after the abortion. Whereas 13 cases had a cesarean section or vaginal delivery between 34 and 41 weeks of pregnancy, one had surgery early in pregnancy, seven had surgery between 13 and 23 weeks of pregnancy, and five had surgery after delivery. Twelve patients did not have tumor recurrence 3 to 18 months after the operation. One patient had distant metastasis 3 months after the surgery.

SPTP is a rare pancreatic mucus tumor that accounts for 1% to 3% of pancreatic tumors and is mainly seen in young women.[1] There are no specific clinical manifestations, which makes it hard to be recognized.[4] However, it has some unique manifestations on CT imaging, which makes CT the most

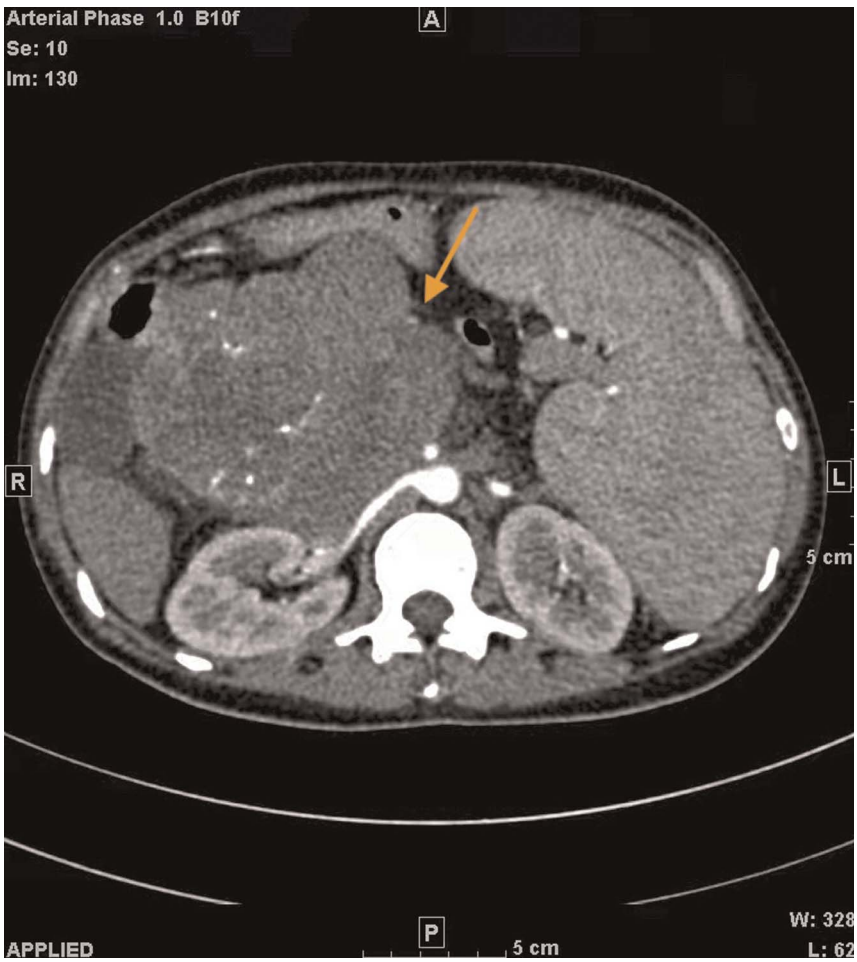

Figure 1. The transverse section of pancreatic thin-layer computed tomography reveals an irregular solid and cystic mass (12.3 cm × 9.3 cm × 8.0 cm) in the pancreas head with a lobulated appearance (arrow).

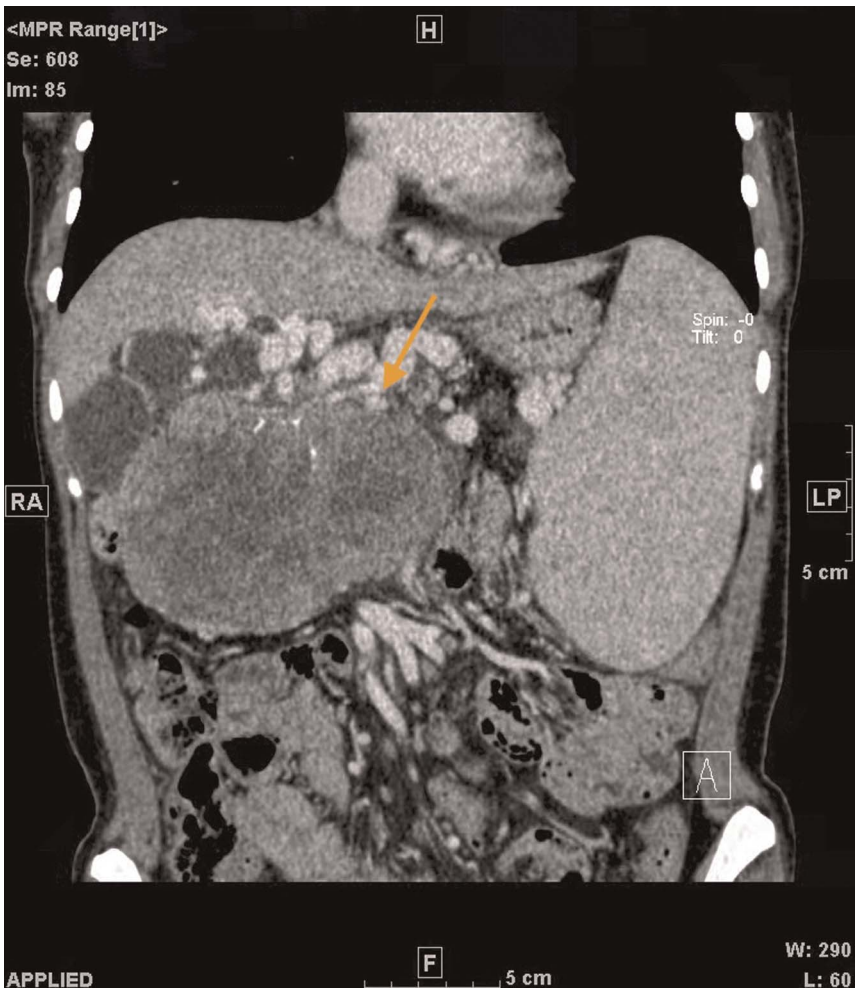

Figure 2. Contrast-enhanced images in coronal views reveals an irregular solid and cystic mass (arrow).

important imaging examination and also the reason we made the diagnosis without pathology.[5] SPTP is biologically borderline or low-grade malignancy. Therefore, active surgical radical resection should be performed as soon as possible.[6] The most tricky feature of SPTP is the presence of progesterone receptors, which makes the treatment of pregnancy complicated with SPTP in a dilemma. Kosmahl *et al.*[7] reported 59 cases of SPTP, and progesterone receptors were identified in tumor specimens in more than 90% of cases through immunohistochemistry. The progesterone level rises during pregnancy, which may lead to the progression of tumor lesions.[8] Surgical treatment in early and late pregnancy may increase the risk of early pregnancy abortion and preterm birth, respectively. The current recommendation is to perform elective surgery in the 2nd trimester.[9] However, there has no recognized treatment plan accomplished for pregnancy with SPTP. Of the 17 reported cases, all patients received surgical treatment, whereas eight patients underwent surgery during pregnancy. Yee *et al*[10] reported a tumor found at 18 gestational weeks and closely observed until delivery at 40 weeks, during which the tumor size did not increase significantly. The patient underwent surgery postpartum. Al-Umairi *et al.*[11] reported progression in the tumor size from 13.7 cm to 16.8 cm from 28 weeks to 38 weeks of pregnancy. The growth rates of the tumor during pregnancy were not described in three cases who underwent surgery postpartum[12–14] and one case[15] who underwent surgery 19 weeks after the tumor was found; however, it can be inferred that the tumor did not affect the safety of mothers and fetuses. The other seven cases were surgically treated within 8 weeks after the onset, and the tumors increased rapidly during pregnancy in three cases.[15–17] A patient reported by Morales *et al.*[16] had a tumor size of approximately 5 cm at 4 weeks of pregnancy; 7 days later, CT examination showed that the tumor size was approximately 8.2 cm, and 12 days later, the tumor size was approximately 12 cm during laparoscopic exploration. Huang *et al.*[17] reported spontaneous rupture and bleeding of pancreatic tail tumors in patients at the 19th week of pregnancy. During the operation, the diameter of the tumor reached 17 cm, and spontaneous rupture may have been caused by the rapid growth of the tumor. All three patients had a smooth pregnancy until 39 to 40 weeks of delivery. Balancing the safety of the mother and child with surgical treatment is very important. Because of the small number of cases, it is temporarily impossible to evaluate the impact of pregnancy on tumor growth. Established cases showed some pregnancies with tumors reaching full term

without significant acceleration of tumor growth, so it is necessary to closely monitor tumor growth during pregnancy in combination with the patient's surgical willingness. From previously reported cases, surgical operation in the 2nd trimester would be relatively safe. There are not any traceable case reports and reliable clinical evidence to know whether the tumor loses progesterone stimulation after the termination of pregnancy will shrink or even obtain surgical opportunities for patients who are unable to undergo surgery.

Pregnancies complicated with liver diseases, especially liver cirrhosis, are rare and dangerous.[18] A total of 10% to 25% of patients with preexisting liver cirrhosis will have their condition worsened during pregnancy.[3,18,19] The incidence of variceal hemorrhage, hepatic encephalopathy, liver and kidney syndrome, splenomegaly, and coagulation dysfunction caused by decompensated liver cirrhosis is significantly higher in pregnancy.[19] If there is evidence of pregnancy complicated with serious liver dysfunction, the pregnancy should be terminated in a timely manner.[20] The patient has no known history of hepatitis B and cirrhosis and even has not presented any clinical manifestations before 18 gestational weeks, while presented to the hospital with decompensated manifestations directly, including ascites, splenomegaly, portal hypertension, coagulation dysfunction, and thrombocytopenia, which may indicate a progression of liver cirrhosis caused by pregnancy. Continued pregnancy and pancreatic head occupancy may increase the burden on the liver and lead to further deterioration, so the pregnancy should be terminated as soon as possible.

Conclusion

In the absence of other complications and contraindications, surgical treatment during pregnancy and conservative management are both acceptable, because the majority of the pregnancy outcomes were good. There is a certain risk of accelerated tumor growth with continuing pregnancy, which may affect the mother and child. However, SPTP is not an absolute indication for termination of pregnancy according to the review of the above cases. If there are contraindications or unwillingness for surgery during pregnancy, appropriate treatment should be assessed to achieve the best outcomes for both the mother and the baby.

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Author Contributions

Juntao Liu and Xinyan Liu (corresponding authors) guaranteed all aspects of the reliability. Yi Yu collected the data and wrote this article. Ping Peng and Lirong Teng participated in reviewing the cases. Congcong Liu and Qian Zhou illustrated the table. All authors approved the final version of the article.

Conflicts of Interest

None.

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