Case Report

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Rupture Of A Giant Solid Pseudopapillary Neoplasm Of The Pancreas During Intraoperative Caesarean Delivery: A Case Report And Literature Review

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1. Abstract

Nonspecific presentations during pregnancy can mask early signs and symptoms of upper abdominal tumours, making preoperative diagnosis of upper abdominal tumours difficult. Solid pseudopapillary neoplasm of the pancreas (SPN) is a rare exocrine tumour of the pancreas, and SPN in combination with preeclampsia during pregnancy is even rarer. In this paper, we report a case of SPN combined with preeclampsia during pregnancy and sudden rupture of a giant retroperitoneal SPN during caesarean section, resulting in life-threatening intra-abdominal haemorrhage in the patient. After exclusion of obstetric factors, a rapid response team was activated, a multidisciplinary treatment (MDT) was carried out, and the patient was treated promptly and appropriately by resection of the giant retroperitoneal tumour, partial resection of the body and tail of the pancreas, and abdominal drainage. To our knowledge, this is the first reported case of SPN combined with preeclampsia during pregnancy, and a rapid and timely MDT could have ensured the patient's life.

2. Keywords:
Preeclampsia, Caesarean section, Solid pseudopapillary neoplasm of the pancreas, Rupture

3. Introduction

SPN is a rare exocrine tumour of the pancreas, classified pathologically as a low-grade malignant tumour of epithelial origin of the pancreas, with a low incidence and an unknown pathogenesis. SPN is particularly prevalent among young women in their twenties and thirties. The clinical manifestations of SPN are often nonspecific, with no clinical symptoms when the tumour is small. SPN is rare, and the combination of SPN and pregnancy is even rarer and is often difficult to diagnose preoperatively. This paper is the first to report an extremely rare case of SPN in pregnancy combined with sudden rupture of a large retroperitoneal pancreatic tumour encountered during caesarean section, resulting in life-threatening intra-abdominal haemorrhage. This case suggests that the diagnosis of epigastric tumours in pregnancy is difficult and that their nonspecific presentation can mask early signs and symptoms. The presence of clinical signs and symptoms in pregnant women that cannot be explained by pregnancy alone should be taken seriously by clinicians. It is also hoped that the report of this rare case will serve as a wake-up call to all clinicians to improve perinatal care, strengthen relevant investigations and detect abnormalities as early as possible. In the case of sudden intraoperative abdominal haemorrhage, the possibility of a ruptured surgical tumour should be considered after excluding obstetric factors. It is important to be calm, activate the rapid response team and seek MDT to ensure the life of the patient.

4. Case report

A 22-year-old woman was admitted to the hospital as an emergency case due to "gravidity 2, parity 0, 38 weeks of pregnancy, oedema for half a month and sudden increase in blood pressure for 1 day". The patient had regular menstruation, a urine HCG (+) test more than 30 days after menopause, and no early pregnancy reaction. She denied having a history of birth control or viral infection. The initial blood pressure during pregnancy was 120/80 mmHg. There was no abnormal NT, and there was spontaneous foetal movement at 4+ months of pregnancy and normal prenatal examinations. Non-invasive prenatal examinations revealed a low-risk pregnancy, and 4D ultrasound showed no abnormality. The OGTT result was 4.26-9.1-8.62 mmol/L, and she was diagnosed with gestational diabetes mellitus, which was controlled with diet and exercise. A routine pregnancy blood test revealed a HGB level of 94 g/L. The patient was treated with oral ferrous succinate to correct the anaemia, and the patients HGB level in her last pregnancy blood test was 102 g/L. At 14 weeks' pregnancy, the patient presented with bilateral foot swelling, which

gradually progressed to bilateral lower-limb oedema. Tests showed that the patient's blood pressure was in the normal range, her urine protein was negative, and she had no other discomfort.

On the day of admission, the patient had a normal pregnancy test, a blood pressure of 151/94 mmHg, a urine protein result of 3+ and bilateral lower-limb oedema (++) and was referred to our hospital for treatment due to limited access to local hospitals. The patient's blood pressure was 167/109 mmHg, with no dizziness or headache, no blurred vision, no nausea or vomiting, and no significant upper abdominal discomfort, and the amniotic fluid sac had not broken. The patient had an abortion 1 year ago with clearance of 1 foetus. The patient had a history of being physically fit and denied having a specific medical history or family history of hereditary disease. Relevant tests on admission were performed. The results of routine blood testing were RBC 3.51×10^12/L↓, HGB 102 g/L↓, and PLT 114×10^9/L↓. The results of routine urine testing were urine protein 3+. The results of coagulation testing were PT-% 128.4%↑and D-Dimer 7.030 mg/L↑. The results of liver function testing were ALT 10 U/L, AST 21 U/L, and ALP 173 U/L↑. The results of renal function testing were CREA 57.9 mmol/L, UA 335 mmol/L, and BUN 3.73 mmol/L. The result of glucose and electrolyte testing was GLU 3.92 mmol/L↓, and those of the cardiac impairment evaluation were MYO myoglobin <21.00 ng/mL↓and NT-proB-type natriuretic peptide 895.90 pg/mL↑. The results of calcium, phosphorus and magnesium testing were calcium 2.10 mmol/L↓. ECG showed sinus tachycardia, and ST showed a 0.5-mm drop in V4-V6. The admission diagnosis was as follows:

1. Gravida 2, parity 0, 38 weeks of pregnancy, left occipitotransverse (LOT);
2. Hypertensive disorders of pregnancy, namely, severe preeclampsia;
3. Hypohydramnios;
4. Combined anaemia of pregnancy;
5. Umbilical cord encirclement; 6. Diabetes mellitus in pregnancy

In view of severe preeclampsia, 38 weeks of gestation, foetal maturity, the rapid progression of the condition over a short period, and the high risk of vaginal trial of labour, it was proposed that the pregnancy be terminated by emergency caesarean section after hospitalisation with completion of surgical ultrasound and cardiac ultrasound. Therefore, intravenous magnesium sulfate was used to relieve spasm, and oral labetalol (100 mg, TID) was given to lower blood pressure while presurgical preparation was carried out. On the night of admission, the patient complained of decreased foetal movement, a blood pressure of 172/105 mmHg, dizziness and breath-holding discomfort. Considering that the patient's blood pressure was not well controlled, the night duty doctor decided to terminate the pregnancy immediately to ensure the safety of the mother and baby. The patient was admitted to the operating room that evening, and combined lumbar and epidural anaesthesia was administered successfully. Intraoperative access to the abdominal cavity was made, and approximately 500 ml of dark bloody ascites was seen on exploration. The unusual dark bloody ascites alerted the operator to consider the possibility of severe preeclampsia complicated by rupture of a subperitoneal hematoma of the liver, rupture of the uterus, or rupture of other organs or tumors in the pelvic or abdominal cavity. In the operator's judgment, the fetus should be delivered without delay. The woman delivered a live female infant rapidly with no postnatal asphyxia and a 1-minute Apgar score of 9 (skin colour-1). The membranes and placenta are delivered intact, the uterine cavity is rapidly cleared, and the myometrium is closed with continuous full-layer sutures. During suturing, persistent active bleeding from the pelvis was noted. On immediate exploration, the uterus and bilateral adnexa were normal in appearance, and there was no bleeding from other parts of the pelvis. This patient had a history of severe preeclampsia, and a serious preeclampsia complication with sub liver capsule haematoma formation and possible liver rupture could not be excluded. At this point, the woman's blood pressure suddenly dropped, and her heart rate increased to 130-140 beats per minute, so the critical maternal resuscitation process was started immediately, general anaesthesia was requested, subclavian vein catheterization and tracheal intubation were established, the blood bank was urgently contacted, a green blood supply channel was opened, the abdominal cavity was filled with multiple blood pads to stop the bleeding, and the gastrointestinal surgeon was immediately contacted for consultation on the stage. The surgeon extended the incision from the caesarean section upwards to remove the old blood clot and blood from the upper abdomen by approximately 1500 mL. On exploration, the surface of the greater omentum, liver and spleen was smooth with no ruptured bleeding.

Figure 1:

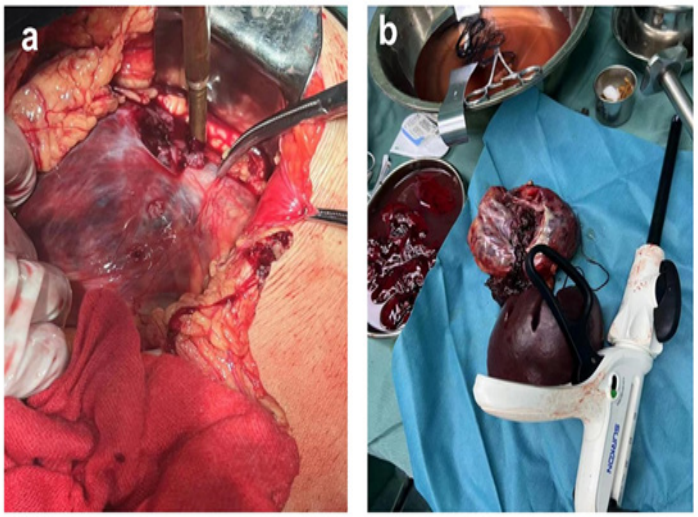

Figure 1: (a) Large retroperitoneal tumour detected intraoperatively; (b) Postoperative excised tumour specimen and spleen.

A large tumour, approximately 15 x 12 cm in diameter, with a surface rupture of approximately 4-5 cm, still with active bleeding, was visible after opening the gastrocolic ligament. The tumor was large and the spleen was torn during intraoperative dissection, so a parallel splenectomy was performed. After the superficial bleeding was controlled, the tumour was separated from the surrounding tissues, and finally, retroperitoneal giant tumour resection, partial resection of the body and tail of the pancreas, and intraperitoneal drainage were performed. The postoperative tumour

specimen was brittle and contained a large number of blood clots and some fish meat-like substance, which were sent for pathology. The total volume of intraoperative bleeding was approximately 3500 ml, with 1200 ml of blood and 800 ml of plasma transfused. After the operation, the patient was transferred to the ICU to continue transfusion of blood and plasma as well as coagulation factors to replenish blood volume and improve blood clotting, receive respiratory and circulatory support, and receive correction of electrolyte disorders and acid–base imbalance until stabilization of the patient's condition. A total of 2,400 ml of blood, 1,200 ml of plasma, 300 mL of platelets and 10 U of cold precipitation were transfused. The patient was awake two days after the surgery and successfully extubated. She was transferred to the general obstetric ward three days after the surgery and continued anti-inflammatory, hypotensive, rehydration support and thromboprophylaxis treatment and was discharged from the hospital two weeks after the surgery.

The postoperative pathology was as follows:

- (Retroperitoneal) solid pseudopapillary neoplasm of the pancreas, with immunohistochemical staining showing AACT (+), β-catenin (nuclear pulp +), Syn (+), CD10 (+), Vimentin (+), CyclinD1 (weak +), CD56 (partial +), PR (-), CgA (-), CKpan (-), E- cadherin (-), Ki67 (approx. 2% +);
- (Spleen) consistent with mild splenic stasis, 4 periportal lymph nodes with reactive hyperplasia and pancreatic tissue;
- In part of the tail of the body of the pancreas + large omentum, the examination material was consistent with omental tissue with 3 lymph nodes with reactive hyperplasia.

The patient was instructed to undergo follow-up every 3 months for 1 year, every 6 months after 1 year, and once per year after 2 years and thereafter. At the present six-month follow-up, no abnormalities were found on ultrasound and CT.

Figure 2:

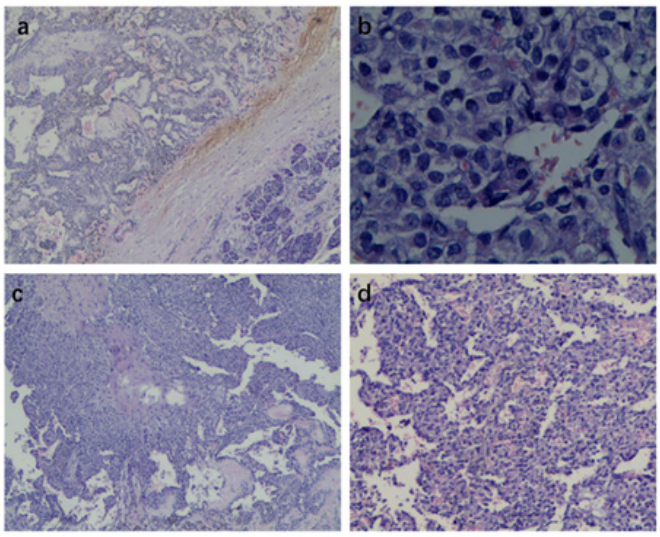

Figure 2: Postoperative pathological return images. (a) SPN tumor tissue, the upper left is the tumor area, the lower right is the normal pancreas, the boundary between the tumor and the normal pancreas is still clear. (b) Tumor cells with unclear boundaries, eosinophilic cytoplasm, some cells with slightly clear boundaries, foamy or transparent cytoplasm, round or oval nuclei located in the center, some nuclear grooves are seen, and nuclear schizophrenia is rare. (c) The cystic area is often seen as a collection of foamy tissue cells, and the cyst wall is fibrotic. (d) Pseudopapillary area with branching papillae, papillae covered with several layers of tumor cells, with abundant blood vessels or blood sinuses.

5. Discussion

Solid pseudopapillary neoplasm of the pancreas are rare exocrine tumours of the pancreas, also known as papillary and solid epithelial tumours of the pancreas, cystic solid tumours of the pancreas, papillary cystic epithelial tumours, solid papillary epithelial tumours of the pancreas, and Frantz tumour[1]. In the latest pathological classification, SPN of the pancreas is categorized as a low-grade malignant tumour of epithelial origin of the pancreas[2], with a low incidence of 0.17% to 3%[3-4]. Its pathogenesis is still unclear. Most scholars think that SPN originates from pancreatic tissue, but with the in-depth study of its immunohistochemistry and other studies, it was found that the tumour has multidirectional components, namely, endocrine, exocrine and focal epithelial components of the pancreas, suggesting that SPN most likely originates from pancreatic embryonic pluripotent stem cells[5]. SPN has a clear sex- and age-specific tendency, occurring mostly in young women, with over 90% of pancreatic pseudopapillary tumours occurring in young women in their twenties and thirties[6]. The clinical presentation is mostly nonspecific, and there are no clinical symptoms when the volume is small. Preoperative diagnosis is therefore difficult and easily misdiagnosed[7]. Some patients present with vague pain or discomfort in the upper abdomen, which is not clearly distinguishable from hepatobiliary disease. The tumour is usually found accidentally by ultrasound during physical examination, and the diameter of the SPN tumour is usually large. Some patients show symptoms of tumour compression, such as vague pain and discomfort in the upper abdomen, abdominal distension, nausea, and vomiting. In severe cases, the tumour may manifest as intestinal obstruction. Cases of acute pancreatitis and tumour rupture leading to acute abdomen have also been reported[8].

This patient experienced nausea and vomiting in early and mid-pregnancy, but this was considered a normal reaction to pregnancy and was not taken seriously. If the tumour ruptures and bleeds, she may even have symptoms such as gastrointestinal bleeding and anaemia. Pseudopapillary tumours of the pancreas are more commonly found in the caudal part of the body of the pancreas but can also occur in the head of the pancreas. They rarely cause dilatation of the pancreaticobiliary ducts and jaundice, as they tend to grow exophytically. Imaging is the main diagnostic tool for this condition. These include ultrasound, abdominal CT and MRI. Ultrasound of the abdomen is simple and inexpensive and can show the location, size and morphology of tumours. Colour Doppler (CDFI) can detect the internal blood flow of a tumour, but in practice, abdominal ultrasound is highly susceptible

to intra-abdominal gas interference, has relatively limited value for definitive diagnosis, and can be used as a primary screening tool. CT and MRI are more advantageous in showing the components of the lesion and can improve the accuracy of the diagnosis. Most of the laboratory tests for SPN are normal, and CA199, CA125, AFP, CEA and other tumour indicators are also mostly in the normal range, so they have no significant value for diagnosis. SPN is rarely diagnosed preoperatively and needs to be differentiated from nonfunctioning islet cell tumours, pancreatic adenocarcinoma and pancreatic cystadenoma. The treatment of SPN is mainly surgical resection[9]. Surgical resection provides a good prognosis with a cure rate of 95% [10], a five-year survival rate of 95-98% and a 10-year survival rate of 93% [11], but close follow-up is still required [12].

SPN is rare, and no clinical cases of SPN in pregnancy have been reported. This paper is the first report of an extremely rare case of SPN in pregnancy combined with a sudden intraoperative rupture of a large pancreatic tumour, resulting in life-threatening intra-abdominal haemorrhage. The tumour was found in time, and the multidisciplinary effort eventually resolved the problem. Many lessons were learned from the management of this case. Usually, if there are no special clinical symptoms, it is difficult to perform targeted ultrasound of the upper abdomen and more diagnostic CT and MRI, so it is difficult to detect abdominal tumors by means of conventional pregnancy tests, and it is difficult to achieve early diagnosis and treatment. This patient is a young woman who had no conscious discomfort except for nausea and vomiting in the early and mid-pregnancy period, so the doctor in charge did not improve the surgical ultrasound and other examinations before the emergency surgery, which led to a passive situation during the operation. As for intraoperative tumor rupture, we analyzed that it might be related to the following factors: the most basic pathophysiological changes of hypertensive disorders in pregnancy are spasm of small blood vessels and endothelial damage in the whole body, increase of vascular permeability, the huge pancreatic tumor is densely vascularized and rich in blood transport, and the rapid change of blood pressure before operation led to spontaneous rupture of tumor blood vessels and intratumor hemorrhage. This case is a wake-up call for clinicians and emphasizes the importance of perinatal care, strengthening investigations, paying attention to the complaints of pregnant women, and drawing sufficient attention from clinicians when pregnant women present with clinical symptoms and signs that cannot be explained by pregnancy alone. In pregnant women with acute abdomen combined with pregnancy, if ultrasound examination fails to reveal positive findings, CT or MRI of the abdomen should be considered for further diagnosis [13]. The American College of Obstetricians and Gynecologists (ACOG) states that, at present, radiation doses from diagnostic tests do not affect the developing embryo or foetus (but should be avoided at 8-15 weeks of gestation) [14].

6. Conclusion

When dealing with a pregnant woman with an acute abdomen, the obstetrician should fully communicate with her and recommend imaging to further clarify the diagnosis and to try to detect the lesion as early as possible to reduce complications for the mother and child and to obtain the best possible prognosis. Due to the unpredictable nature of obstetrics, if sudden intra-abdominal bleeding occurs intraoperatively, the possibility of surgical tumour rupture should be considered at this point after obstetric factors have been ruled out. It is important to remain calm, activate the rapid response team and seek MDT to ensure the preservation of maternal life.

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