Case Report

Solid Pseudopapillary Neoplasm of Pancreas in Pregnancy Treated with Tumor Enucleation: Case Report and Review of the Literature

TT Huang, J Zhu, H Zhou[1], AM Zhao

Departments of Obstetrics and Gynecology and [1]General Surgery, Ren Ji Hospital, School of Medicine, Shanghai Jiao Tong University, Shanghai, China

Abstract

Solid pseudopapillary neoplasm of pancreas (SPN) during pregnancy is rare and presents a threat both to the mother and the fetus. We report a case of SPN in a 26-year-old woman diagnosed at 21 weeks of gestation. Tumor enucleation was successfully performed by a general surgeon. A healthy female infant was delivered at 39 weeks and 5 days of gestation vaginally without complications. Our report provides an example that tumor enucleation of SPN during the second trimester could be successfully performed during pregnancy. A multidisciplinary approach with respect to the pregnant patient's diagnosis, indications, and timing of surgery is necessary in ensuring the best possible outcomes for both the mother and the child.

Keywords: *Pancreas, pregnancy, solid pseudopapillary neoplasm, tumor enucleation*

Date of Acceptance:
30-Apr-2018

Introduction

Solid pseudopapillary neoplasm of pancreas (SPN) is a rare, benign or low-grade neoplasm. SPN is most often diagnosed with young women. It is particularly rare during pregnancy, yet the accelerated tumor growth of SPN during pregnancy can be life-threatening.[1-3] There are only a few reports of SPN discovered and resected during pregnancy.[4-6] In all cases, the goal is to minimize both maternal and fetal risk. Therefore, treatment of SPN for a pregnant patient remain a clinical challenge.[7-9] Here, we report a case of SPN in a 26-year-old woman diagnosed at 21 weeks of gestation on routine prenatal examination in Shanghai, China.

Case Report

A 26-year-old G1P0 woman undergoing routine prenatal examination was discovered to have an epigastric mass at 21 weeks of gestation. The patient had no experience of abdominal pain, nausea, or vomiting. There was no weight loss, constitutional symptoms, hematemesis, or changes in genitourinary or bowel habits. The patient's medical history was unremarkable and with no family history of similar growth. Physical examination revealed a large, firm mass in the left side of the epigastrium. Ultrasound scan showed a well-circumscribed nonhomogeneous tumor about 12.4 cm in diameter in the mid-abdomen [Figure 1a]. Magnetic resonance imaging (MRI) depicted a heterogeneous mass related to the pancreatic tail [Figure 1b]. There was mild splenomegaly. No ascites or hepatic lesions was identified, and no biliary ductal dilation was present. Laboratory tests such as blood count, liver function tests, and serum amylase were within normal range. Tumor markers including carcinoembryonic antigen (CEA = 1.57 ng/ml), cancer antigen 125 (CA125 = 34.83 U/ml), and carbohydrate antigen 199 (CA199 = 4.01 U/ml) were within normal ranges. The tentative differential diagnosis includes nonfunctional islet cell tumor, pancreatic pseudocyst, serous cystadenoma, serous cystadenocarcinoma, mucinous cystadenoma, mucinous cystadenocarcinoma, and intraductal papillary mucinous neoplasm.

At 22 weeks of gestation, the patient underwent exploratory laparotomy. The neoplasm, measured to be

Access this article online

Quick Response Code:

Website: www.njcponline.com

DOI: 10.4103/njcp.njcp_39_18

Address for correspondence: Prof. AM Zhao, Department of Obstetrics and Gynecology, Ren Ji Hospital, School of Medicine, Shanghai Jiao Tong University, No. 160, Pujian Road, Pudong New District, Shanghai, China.
E-mail: zamzkh0526@126.com

How to cite this article: Huang TT, Zhu J, Zhou H, Zhao AM. Solid pseudopapillary neoplasm of pancreas in pregnancy treated with tumor enucleation: Case report and review of the literature. Niger J Clin Pract 2018;21:1234-7.

Table 1: Summary of previously published solid pseudopapillary neoplasm during pregnancy

Authors	Year	Patient age	Gestational age at surgery	Tumor location and size (cm)	Surgical procedure	Pregnancy outcome
Duff and Greene[1]	1985	35	14	Head; N/A	Needle biopsy, embolectomy, and Whipple	Spontaneous abortion after embolectomy
Bondeson *et al*.[2]	1990	19	4-5	Head; 8	EL, biopsy, and Whipple	Postoperative pregnancy termination
Morales *et al*.[3]	1998	21	6	Head; 8.2	EL; Whipple	SVD at 39 weeks
Ganepola *et al*.[4]	1999	37	23	Tail; 12	DP, splenectomy, and cholecystectomy	SVD at term
Levy *et al*.[5]	2004	27	16	Head; 6	Whipple	Labor induction at 34 weeks, SVD
Hajdu *et al*.[6]	2009	29	13	Tail; 16	DP	C-section at 38 weeks
Feng *et al*.[7]	2011	26	14	Head; 9.5	EL tumor enucleation	Labor at 38 weeks, C-section
Huang *et al*.[8]	2013	29	19	Body and tail; 17	Emergent exploratory reverse-T laparotomy; subtotal pancreatectomy and splenectomy	SVD at 29 weeks
MacDonald *et al*.[9]	2014	23	18	Body and tail; 16.3	Exploratory laparotomy and distal pancreatectomy, splenectomy and cholecystectomy	SVD at term
Sharanappa *et al*.[10]	2015	22	16	Head; 12	Pylorus preserving Whipple's pancreaticoduodenectomy	Medical termination of pregnancy

N/A=Not available; EL=Exploratory laparotomy; SVD=Spontaneous vaginal delivery; DP=Distal pancreatectomy; C-section=Cesarian section

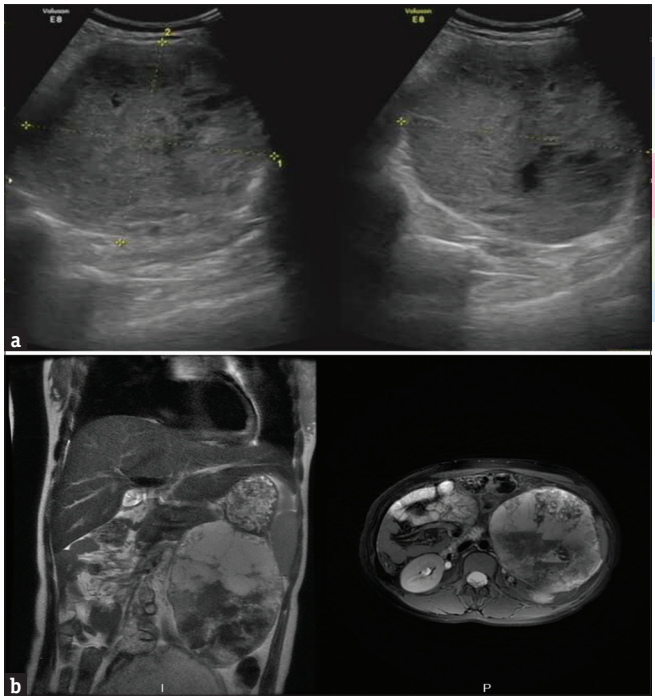

Figure 1: (a) Ultrasound image demonstrating a well-circumscribed non-homogeneous mass (12.4 cm × 11.8 cm × 12.4 cm) in the mid-abdomen. (b) Magnetic resonance imaging demonstrating a heterogeneous mass related to the pancreatic tail

13 cm × 12 cm × 12 cm [Figure 2a], was connected to the tail of the pancreas with a pedicle, but the common bile duct and main pancreatic duct were not dilated. Tumor enucleation was performed by a general surgeon, with the preservation of the splenic vessels and the spleen. The tumor is round and cystic-solid with the complete capsule. There was no metastasis or any

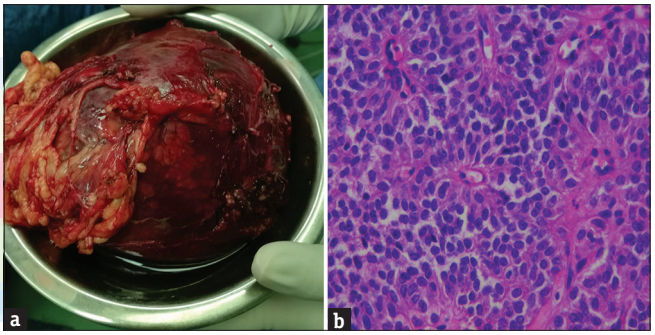

Figure 2: (a) Intraoperative picture of the solid pseudopapillary tumor of the pancreas. (b) Histopathology of solid pseudopapillary neoplasm of the pancreas

visible tumor residue at the operation site. Intraoperative histological examination confirmed the diagnosis of SPN [Figure 2b]. Positive staining for vimentin, pan-CK, CD56, β-catenin, progesterone receptor (PR), and alpha-methylacyl-CoA racemase (AMACR) were found by immunohistochemical analysis; CgA, Syn, and E-cadherin were negative.

There were no postoperative complications. Fetal evaluation including ultrasound scan on postoperative day 9 revealed a normal fetus with normal anatomy. The patient was discharged on postoperative day 10 in stable condition. After surgery, the patient's pregnancy proceeded uneventfully. At 39 weeks and 5 days of gestation, a healthy female infant was delivered vaginally. Her Apgar scores were 10 and 10, at 1 and 5 min, respectively. Six months after the delivery, there was no detectable evidence of residual or recurrent disease by MRI.

Discussion

SPN was first described by Frantz in 1959 and is rare in pregnancy. To the best of our knowledge, there are only ten cases of SPN found and treated during pregnancy [Table 1].[1-10] The presentation of SPN is usually nonspecific. Abdominal pain or mass is the most common presenting clinical symptom or sign. Nonspecific symptoms such as nausea, vomiting, fever, weight loss, and jaundice are other clinical symptoms; most of them are caused by the tumor compressing against the otherwise normal pancreas. However, a considerable percentage of the patient population is asymptomatic, and the neoplasm can only be incidentally detected. SPN can be detected by ultrasonography, computed tomography, or MRI. When making imaging decisions during pregnancy, one must take into account the risk to the fetus. Imaging procedures without ionizing radiation are preferable and should be considered. Ultrasonography and MRI are not associated with known adverse fetal effects.

Immunohistochemically, SPN cells strongly and diffusely express vimentin, α-1 antitrypsin, α-1 antichymotrypsin, neuron-specific enolase, PR, and β form of estrogen receptor. CD10, CD56, CD 1, FLI-1, and epithelial markers such as CK, AE1/AE3, and CAM 5.2 can also be focally positive. β-Catenin is expressed in all SPNs. In our case, we observed strong immunoreactivity for vimentin, CK, CD56, β-catenin, PR, and AMACR. PR seems to be responsible for the regulatoffing tumor cells' replication rate. This might be one of the explanations for the high prevalence of SPNs among young women, and for the increased speed of growth observed when serum progesterone levels are elevated such as in pregnant women. Ganepola *et al*. reported a 5.5 cm SPN discovered during the 4th week of pregnancy; the tumor was found to grow to >12 cm within 4 months.[4] In another case, the tumor had been slow growing, close monitoring for tumor growth was a possibility during pregnancy, and a successful pancreaticoduodenectomy was performed after term delivery.[11] Hence, it might be expected that more cases of SPNs would be observed in young pregnant women due to the effect of PR.

Complete surgical resection is the mainstay treatment for all patients with SPN even in the presence of local invasion or distant metastasis. Surgical procedure should be selected according to the tumor position, size, operation times, and intraoperative histological examination. There is no treatment guideline for pregnant women with SPNs, and therefore, decisions on the optimal timing for surgical management can be challenging. Surgical intervention during the first trimester may be associated with spontaneous abortion or poor fetal outcome, including congenital anomalies. The second trimester is the preferred time window for surgical intervention for resectable SPN. The American College of Obstetricians and Gynecologists' current recommendations are for operation during the second trimester for any nonurgent abdominal surgical procedures during pregnancy. Many case reports agreed that the second trimester is favorable for surgical resection, as fetal organogenesis is complete and the size of the fetus may allow for an easier surgical procedure in comparison to third-trimester operations. In addition, the risk of spontaneous abortion is low during the second trimester. Surgical resection during the third trimester may be associated with an increased risk of premature induction of labor. The malignant potential of SPN is reported to be 10%–15%. Considering SPNs have such low malignant potential, SPNs could be managed after term delivery.[11,12] Several cases of ruptures of SPN have been reported causing maternal instability during pregnancy. In this situation, emergency surgical intervention is performed, regardless of the gestational age.

Conclusion

Our reported case provides an example that tumor enucleation of SPN during the second trimester could be successfully performed during pregnancy with the preservation of the fetus's life. We conclude that a multidisciplinary approach with respect to a pregnant patient's diagnosis, indications, and timing of surgery is necessary in ensuring the best possible outcomes for both the mother and the child.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Duff P, Greene VP. Pregnancy complicated by solid-papillary epithelial tumor of the pancreas, pulmonary embolism, and pulmonary embolectomy. Am J Obstet Gynecol 1985;152:80-1.
2. Bondeson AG, Bondeson L, Thompson NW. Early pregnancy masquerading as a marker for malignancy in a young woman

with curable neoplasm of the pancreas. Br J Surg 1990;77:108-9.
3. Morales A, Ruíz Molina JM, Estéves HO, Robles-Díaz G, Díaz-Sánchez V. Papillary-cystic neoplasm of the pancreas. A sex-steroid dependent tumor. Int J Pancreatol 1998;24:219-25.
4. Ganepola GA, Gritsman AY, Asimakopulos N, Yiengpruksawan A. Are pancreatic tumors hormone dependent? A case report of unusual, rapidly growing pancreatic tumor during pregnancy, its possible relationship to female sex hormones, and review of the literature. Am Surg 1999;65:105-11.
5. Levy C, Pereira L, Dardarian T, Cardonick E. Solid pseudopapillary pancreatic tumor in pregnancy. A case report. J Reprod Med 2004;49:61-4.
6. Hajdu N, Poharnok Z, Olah A. Successfully resected pancreatic tumor in pregnancy. Eur Surg 2009;41:48-50.
7. Feng JF, Chen W, Guo Y, Liu J. Solid pseudopapillary tumor of the pancreas in a pregnant woman. Acta Gastroenterol Belg 2011;74:560-3.
8. Huang SC, Wu TH, Chen CC, Chen TC. Spontaneous rupture of solid pseudopapillary neoplasm of the pancreas during pregnancy. Obstet Gynecol 2013;121:486-8.
9. MacDonald F, Keough V, Huang WY, Molinari M. Surgical therapy of a large pancreatic solid-pseudopapillary neoplasm during pregnancy. BMJ Case Rep 2014;2014. pii: bcr2013202259.
10. Sharanappa V, Tambat RM, Srinivas NM, Razack A. Solid pseudopapillary tumour of pancreas in pregnancy: Case report. Sch J Med Case Rep 2015;3:40-6.
11. Yee AM, Kelly BG, Gonzalez-Velez JM, Nakakura EK. Solid pseudopapillary neoplasm of the pancreas head in a pregnant woman: Safe pancreaticoduodenectomy postpartum. J Surg Case Rep 2015;2015. pii: rjv108.
12. Chhabra M, Daver RG. Solid pseudopapillary epithelial neoplasm of pancreas in pregnancy: Case report of a rare co-occurrence. JMSCR 2017;8:26978-83.