☐ Case report ☐

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Early Detection and Successful Laparoscopic Adrenalectomy for Pheochromocytoma in Pregnancy; A Case Report

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Pheochromocytoma is an extremely rare tumor in pregnant women with potentially fatal consequences. We report a case of pregnant woman at 22 weeks of gestation with pheochromocytoma. A correct diagnosis on the basis of differential clues from severe preeclampsia was obtained and laparoscopic adrenalectomy was performed.

Key Words: Pheochromocytoma, Pregnancy, Laparoscopy, Adrenalectomy

Pheochromocytoma is a rare catecholamine producing tumor and its occurrence during pregnancy is extremely rare (reported incidence of one in 54,000 pregnancies).[1] Because of its rarity and confusion with the much more common forms of pregnancy-related hypertension, the diagnosis of pheochromocytoma in pregnancy may be overlooked. If unrecognized, maternal and fetal mortality amounts to 40-50%.[1] However, after antenatal diagnosis of pheochromocytoma, maternal mortality was reduced substantially to <2% and fetal mortality to <14%.[2] Early diagnosis of pheochromocytoma in pregnancy needs high index of suspicion due to the wide variability and non-specificity of its clinical signs and symptoms.

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We report a case of a 28-year-old woman who was diagnosed with pheochromocytoma early on the basis of detailed history taking and underwent laparoscopic adrenalectomy at advanced gestational age compared with those of previous reported cases.

Case report

A 28-year-old primigravida presented with a blood pressure of 173/104 mm Hg at 22 weeks of gestation. She complained of headache and was otherwise asymptomatic. A urine dipstick test showed 2+ for glucose and trace for protein. A presumptive diagnosis of severe preeclampsia and gestational diabetes were made. However, on physical examination, there was no evidence of peripheral edema. Fetal sonographic examination was unremarkable. Laboratory investigations were all within normal limits except for trace proteinuria and 4+ glycosuria. With these findings, she was considered to be inconsistent with a diagnosis of preeclampsia.

An evaluation of secondary hypertension was carried out. Biochemical investigation showed that

24-hour urinary excretion of vanillylmandelic acid (VMA) and metanephrine was 18.2 mg (normal range 1-8 mg/24 hours) and 2 mg (normal range 0-0.8 mg/24 hours), respectively. With a high suspicion of a pheochromocytoma, additional biochemical tests and magnetic resonance imaging (MRI) of the abdomen were performed. Urine norepinephrine was 1,011.6 μg/24 hours (normal range <80 μg/24 hours) and epinephrine was 21.9 μg/24 hours (normal range <20 μg/24 hours). Plasma norepinephrine was 3,053.8 pg/mL (normal range <80 pg/mL) and epinephrine was 10.5 pg/mL (normal range <20 pg/mL). MRI of the abdomen confirmed a right adrenal mass measuring 4 cm, with internal cystic and solid component (Fig. 1).

At 25^{+6} weeks of gestation after 14 days of preparation with the α-blocking drug doxazosin, a laparoscopic right adrenalectomy was successfully carried out. Histopathologic examination confirmed pheochromocytoma. The postoperative course was uneventful. At the 14^{th} day postoperation, 24-hour urinary excretion of VMA and metanephrine was normalized to 2.4 mg (normal range 1-8 mg/24 hours) and 0.3 mg (normal range 0-0.8 mg/24 hours), respectively. Plasma norepinephrine and epinephrine was decreased to 311.0 pg/mL (normal range <80 pg/mL) and 22.6 pg/mL (normal range <20 pg/mL), respectively. The remaining weeks of the pregnancy progressed normally and a normal baby of 3.2 kg weight was delivered at 39 weeks of gestation by vaginal delivery. Apgar scores were 9 at 1-minute and 10 at 5-minute. The mother and the newborn recovered well and were discharged on the 3^{rd} postpartum day.

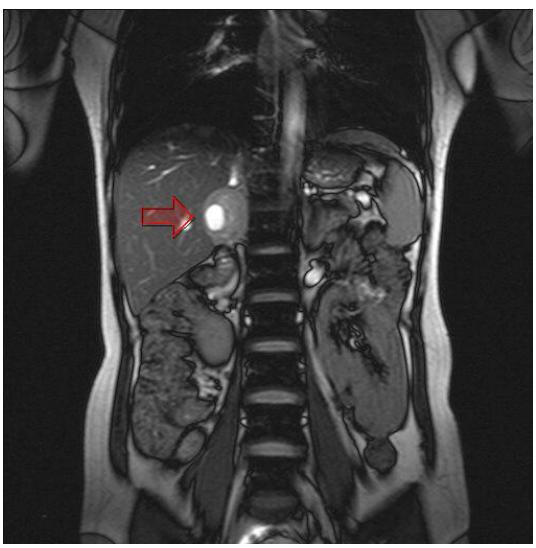

Fig. 1. Magnetic resonance imaging of the abdomen. Arrow indicates right adrenal tumor.

Discussion

The diagnosis of pheochromocytoma during pregnancy is often overlooked due to the similarity of usual signs and symptoms, such as nausea and hypertension in patients with pregnancy-related hypertension. In a review of the literature from 1980 to 1987, the overall maternal mortality was found to be 17% with a fetal loss of 26%, although antenatal diagnosis of pheochromocytoma has reduced the maternal mortality to <1% and the fetal loss to 15%.[3, 8] However, only 26% of pheochromocytomas were diagnosed antenatally.

Based on history and clinical exam, sustained episodes of hypertension occurring after the 20^{th} week of gestation, edema and weight gain are all important clues for preeclampsia, whereas paroxysmal episodes of hypertension occurring throughout the entire pregnancy, severe headaches, sweating, palpitation and orthostatic hypotension are clues for pheochromocytoma.[3] The presence of proteinuria, oliguria or anuria, hyperglycemia, elevated liver enzymes and thrombocytopenia are laboratory clues for preeclampsia.[4]

Paroxysmal attacks occurring in pheochromo-

cytoma during pregnancy as potentially fatal hypertensive crisis can be precipitated by general anesthesia, vaginal delivery, the mechanical effects of the gravid uterus, uterine contractions and increased fetal movements.[5] The fetal risks are mainly determined by the vasoconstrictive effects of catecholamines on the uteroplacental circulation and by the severity and duration of hypertensive spells.[1]

Urinary levels of catecholamines do not change during normal pregnancy, therefore, the diagnosis of pheochromocytoma can be confirmed by measurement of 24-hour urine VMA, metanephrines, or catecholamines. Following biochemical confirmation, the next step is to localize the pheochromocytoma.

Abdominal ultrasound as an initial imaging test is safe but less reliable, particularly with the gravid uterus in the third trimester.[1] Computed tomography or MRI can be used, but MRI is the preferred modality in pregnancy as it is free from ionizing radiation and produces high quality images.[6] If other imaging methods are unhelpful, the Iodine 123-metaiodoenzylguanidine scan is mainly indicated for detecting multiple tumors or extra-adrenal tumors and metastasis.[7]

Definitive treatment of pheochromocytoma is surgical removal. Appropriate preoperative medical management to block the effects of released catecholamine is essential, therefore, α-adrenergic blockade using phenoxybenzamine or doxazosin should be initiated about 10-14 days preoperatively.[1] β-blockade may be required to treat tachycardia, but it should never be prescribed before α-blockade, because β-blockade alone can precipitate a hypertensive crisis attributed to unopposed α-adrenergic effects by catecholamines.[4]

In pregnant patients, the optimal time of surgical tumor removal is a challenging and controversial issue depending on the gestational age, clinical response to treatment, the accessibility of the tumor, and the presence or absence of fetal distress.[4] Harper et al8 have recommended that tumor resection should be performed in the first and second trimesters, and elective cesarean delivery followed by tumor resection in later pregnancy. During the first trimester, surgical tumor removal may lead to increased risk of spontaneous abortion.[1, 8] Some authors have advocated that the tumor diagnosed before 24 weeks of gestation should be removed as soon as possible after hypertension is controlled medically, but after 24 weeks of gestation, tumor removal should be deferred until the fetus is viable and removed simultaneously at the time of cesarean section or shortly after delivery.[1, 4] However, a case who received conservative treatment until fetal maturity is attained, showed the difficulty in managing pheochromocytoma in pregnancy.[4]

Although mid-pregnancy laparoscopic surgery is difficult to perform this because it is difficult to secure visual field exposure, we undertook a laparoscopic surgery since our institution has had experience in laparoscopic adrenalectomy in the last decades and we expect to have a better prognosis in patients with fetal surgical intervention. In case of present case, compared to the most of previously presented cases, we made an early diagnosis and successfully undertook adrenalectomy through subsequent laparoscopic surgery. Keeping the well-being states of the mother and fetus to the full term and it was possible to have the childbirth through vaginal delivery. This could be the great significance this case has.

In conclusion, despite its low incidence, pheochromocytoma should be kept in mind as one of the causes of hypertension in pregnant patients, owing to its high

mortality. Here we present a hypertensive pregnant patient presenting with pheochromocytoma and illustrate the importance of careful history taking and physical examination in preventing such serious maternal and fetal complications.

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