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Pheochromocytoma in a Pregnant Woman with VHL leading to First Trimester Fetal Demise

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Introduction: Pheochromocytomas (PCCs) are rare neuroendocrine tumors that secrete catecholamines. They may be associated with familial syndromes such as Multiple Endocrine Neoplasia type 2 (MEN 2), von Hippel-Lindau disease (VHL), and Neurofibromatosis type 1 (NF 1). PCCs are a common cause of malignant hypertension in pregnancy and can lead to adverse maternal and fetal outcomes if left undiagnosed or untreated.

Case Description: A 39-year-old Latin American primigravid woman at 8 weeks 4 days gestation was referred to the endocrinology clinic for labile blood pressure (160/90 mmHg at recent ER visit) and a lab finding of elevated catecholamines (plasma norepinephrine 4720 pg/ml; normal 0-874 pg/ml). She reported chronic headaches and 2-lb unintentional weight loss during pregnancy. Past medical history was significant for right adrenal PCC diagnosed at age 13 (currently status post adrenalectomy), and a cerebrovascular accident at age 29. Family history was relevant for PCC in the patient's sister and mother. The patient's home medications were nifedipine 30 mg daily and prenatal vitamins. On physical exam, her blood pressure was 126/76 mm Hg, heart rate was 67/min and a scar of previous right adrenalectomy was noted. Doxazosin 1 mg daily was added to her regimen and nifedipine was discontinued. The patient was initially planned for an elective cesarean section at term. However, at 10 weeks 5 days of gestation, she had a missed abortion. Further genetic testing of syndromic PCC revealed a heterozygous mutation in the VHL gene. Abdominal MRI showed a left adrenal mass (3.6×2.6×3.6 cm^3) and retroperitoneal lymphadenopathy. The patient underwent left adrenalectomy and is currently hemodynamically stable on maintenance hydrocortisone and fludrocortisone.

Discussion: We report a case of PCC complicating pregnancy in a 39-year-old primigravida who had a family history of PCC. In females of reproductive age with a known PCC diagnosis, we suggest early genetic testing as it can further advise family planning, antepartum monitoring, and management. Medically, these patients can be managed with α-adrenergic blockers and calcium-channel blockers, followed by β-blockers. This can improve maternal and fetal outcomes primarily by minimizing paroxysmal elevations in catecholamines. Definitive treatment of PCC is surgical excision, usually performed during the second trimester or postpartum. Management of patients with VHL requires lifelong clinical, laboratory, and imaging surveillance for other manifestations of the disease.

References Bancos, Irina et al. Maternal and fetal outcomes in phaeochromocytoma and pregnancy: a multicentre retrospective cohort study and systematic review of literature. Lancet Diabetes Endocrinol 2021; 9: 13–21

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