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Pheochromocytoma in Pregnancy

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Editorial

Pheochromocytoma during pregnancy remains a rare condition, occurring in 0.007% in one 22-year review of 30,426 pregnancies [1]. However, pheochromocytomas in pregnancy are associated with high maternal and fetal mortality (8% and 17%, respectively) in addition to fetal growth restriction and prematurity [2,3]. Antenatal diagnosis has been shown to reduce maternal and fetal mortality with early pharmacologic and surgical intervention [4].

We present a case of a 29 year-old G2P1011 female was referred to endocrine clinic for elevated plasma metanephrines that were checked in the setting of hypertension associated with headaches and hyperhidrosis that persisted following a spontaneous abortion at 30 weeks gestation.

The patient had a history of gestational hypertension during her first pregnancy, but had not routinely checked her blood pressure since her previous delivery 2 years prior. Early in her second pregnancy she began to develop episodes of paroxysmal headaches, palpitations, and heat intolerance associated with elevated blood pressure. She was diagnosed with gestational hypertension and diabetes in the first trimester and placed on bed rest. Despite the start of labetalol in pregnancy, she continued to have ongoing hypertension with episodic adrenergic symptoms. These symptoms persisted after the pregnancy loss. She remained on labetalol therapy after the pregnancy.

Two months after abortion, labs showed elevated plasma free metanephrines to 4.6 nmol/L (ref <0.50), plasma free normetanephrines elevated to 35 nmol/L (ref <0.90), 24 h metanephrines 6766 mg (ref 94-604), 24 h urine normetanephrines 5120 mg (ref 40-412). CT adrenal protocol showed a 3.7 cm mass replacing the left adrenal with poor washout on delayed imaging. Labetalol was discontinued and she was switched to phenoxybenzamine with left laparoscopic adrenalectomy two months after initial laboratory diagnosis.

Pheochromocytoma is often misdiagnosed as pregnancy induced hypertension (PIH), which remains the most common complication of pregnancy. However, there are some key differences that can aid in distinguishing the two entities. PIH rarely occurs prior to 20 weeks gestation, while symptomatic pheochromocytoma can present at any point during pregnancy. Pheochromocytoma often presents with paroxysmal hypertension associated with headache, palpitations (40%), hyperhidrosis (35%), and anxiety (18%) [5]. Idiopathic hypertension and PIH can often progress to preeclampsia, which is associated with proteinuria, leg edema, and extra weight gain during pregnancy. High uric acid, liver function abnormalities, and coagulopathies are more common in PIH and hyperglycemia can be seen in either condition [6]. Cardiomyopathy during pregnancy or in early post-partum should also raise the index of suspicion for pheochromocytoma [7]. One difficulty in distinguishing the two entities is that both plasma and urine metanephrines and catecholamines can be elevated in PIH, as much as 1.6 to 2.6 fold in one study [8]. However, other studies have reported that catecholamines are not elevated in normotensive pregnant women or preeclamsia [9]. It should be noted that some medications used in pregnancy (methyldopa, labetalol) can mildly increase metanephrine levels. However, these elevations are not to the same degree as is seen in pheochromocytoma. Plasma free metanephrines and 24 h urinary fractionated metanephrines remain the screening tests of choice, with sensitivity close to 100% [10].

Both adrenal ultrasound and MRI can be used as diagnostic modalities for identifying adrenal masses in pregnancy once biochemical testing has shown metanephrine excess. In one review, adrenal ultrasound had a sensitivity of 54% and MRI had a sensitivity of 93% [3]. The sensitivity of ultrasound declines in the third trimester with enlargement of the uterus.

The first step in management of pheochromocytoma in pregnancy is similar to that of non-pregnant adults. Alpha blockade, typically with phenoxybenzamine 10 mg twice daily should be started, with up-titration every 2-3 days to reach orthostatic blood pressures. Beta-blockade can then be added to address reflex tachycardia that develops in the setting of orthostasis. If antenatal

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diagnosis is made prior to 24 weeks gestation, improved maternal and fetal outcomes have been shown with laproscopic adrenalectomy between 12-24 weeks once adequate alpha blockade is achieved, particularly if tumor is less than 7 cm. Bilateral pheochromocytoma is not a contraindication to laparoscopic intervention. If diagnosis made after 24 weeks, cesarean section should be performed at term combined with tumor removal [11].

While pheochromocytoma in pregnancy remains rare, clinicians should have a low threshold for testing in several clinical scenarios: 1) women with labile or difficult to control blood pressure early in pregnancy 2) women with concurrent symptoms of paroxysmal headaches, palpitations, and/or hyperhidrosis 3) women with persistent symptoms after childbirth, or 4) finding of elevated blood glucose early in pregnancy along with elevated blood pressure and inability to gain weight as gestational age advances. No guidelines are available due to the rarity of this condition but early recognition and intervention are extremely important. Maternal and fetal outcomes are markedly improved with early antenatal diagnosis and rates of antenatal diagnosis have improved dramatically over the years with a higher index of suspicion and improved biochemical testing.

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