Pheochromocytoma has long proven to be a diagnostic challenge for clinicians. This challenge is expanded when symptoms of this disease manifest during the peripartum period. Here we report the case of a 38-year-old woman with a history of migraine headaches that presented to the emergency department at the request of her obstetrician. During a routine office visit, one week before a scheduled cesarean section, the patient was discovered to have severe hypertension with a systolic pressure of 280/140. Tapping our differential for this peripartum patient with headaches and hypertension was pheochromocytoma. Subsequent workup revealed elevated urine metanephrines, followed by abdominal magnetic resonance imaging that revealed a left suprarenal mass consistent with pheochromocytoma, which was ultimately surgically removed.

Pheochromocytoma is a rare catecholamine-producing tumor that in order to diagnose requires a high index of suspicion from the clinician. This case emphasizes the importance of maintaining a broad differential before preeclampsia when diagnosing a patient with headaches and hypertension in the peripartum period.

**INTRODUCTION**

Pheochromocytoma is a rare neuroendocrine tumor of the adrenal medulla or extra-adrenal chromaffin tissue. This rare tumor may cause paroxysmal and occasionally sustained hypertension. If indiagnosed, pheochromocytoma may result in maternal and fetal morbidity and mortality. Most cases of pheochromocytoma are sporadic; however, according to current literature approximately 20% may be syndromic (1). The classic presentation is quite variable, ranging from an adrenal incidentaloma to a patient in hypertensive crisis with fever accompanied by hypertension. Cardiovascular consequences of pheochromocytoma are arrhythmias, heart failure, arrhythmias, visual disturbances, seizures, and sudden death (4). The classic features of pheochromocytoma in pregnant or postpartum patients are similar to those seen in other patients. Peripartum pheochromocytoma is challenging to diagnose because it shares many features with preeclampsia, which is much more common. There are several postulated reasons why pheochromocytoma may manifest during pregnancy and peripartum. Increased intra-abdominal pressure, fetal movement, uterine contraction, and the stress of delivery all contribute to the development of clinically relevant pheochromocytoma (5). Proper management of pheochromocytoma involves more than simply the excision of the tumor. Prior to surgery, adequate alpha blockade must be established. Current recommendations involve at least a seven day course of an increasing dose of the irreversible alpha blocker phenoxybenzamine up to 30 mg three times per day (6). Oral prazosin or intravenous phenolamine can be used to manage paroxysms. If a pheochromocytoma is discovered during pregnancy, current literature suggests the best outcomes for both the mother and fetus are realized if the tumor can be removed prior to 23 weeks gestation. Because of uterine obstruction, after 23 weeks, medical management with an alpha blockade bridge to post-partum surgery is recommended (1). It is crucial to diagnose pheochromocytoma during pregnancy to aid in the prevention of fetal and maternal morbidity and mortality.

**CASE PRESENTATION**

A 38-year-old female, G14 P11 with past medical history significant for two years of migraine headaches, presented to the emergency department at the request of her obstetrician for evaluation of headache and hypertension. At her obstetrician’s office, a blood pressure of 280/140 was discovered. In the emergency department evaluation revealed a gravid woman with normal vital signs. The patient was then admitted for observation of her blood pressure and scheduled cesarean section. Two days after admission the patient’s obstetrician delivered the fetus by cesarean hysterectomy rather than cesarean section due to placenta percreta being discovered intra-operatively. The patient reported worsening headaches postoperatively. Paroxysmal hypertension was noted both during the cesarean hysterectomy and postoperatively in the intensive care unit. Subsequent magnetic resonance imaging of her brain showed posterior reverse encephalopathy syndrome (PRE) with no infarctions to secondary severe hypertension. Pheochromocytoma was suspected as the cause of her paroxysmal hypertension and coinciding headache. She was discharged post-operative day six with 24-hour urine catecholamines pending. After discharge, headaches and paroxysmal hypertension continued.

24-hour urine metanephrine analysis results were completed showing 7342 mcg of metanephrine (reference range 36-190 mcg), total metanephrines 2613 mcg (range reference 35-482 mcg), and total normetanephrines of 9955 mcg (range reference 1195-695 mcg). Serum metanephrines were also elevated more than 10 times the upper normal limit. On post-operative day 15 she was again hospitalized due to transient episodes of expressive aphasia without other localizing features. Repeat brain magnetic resonance imaging then showed bilateral cortical infarcts and petechial hemorrhages throughout. Abdominal magnetic resonance imaging showed a left supra-renal mass consistent with pheochromocytoma. Alpha blockade was initiated with phenolamine and phenoxbenzamine. She was then discharged and continued alpha blockade with close follow-up. Beta blockade was initiated as well due to beta blockade with close follow-up. Beta blockade was initiated as well due to hypertension. She was discharged post-operative day six with 24-hour urine metanephrine analysis results were completed showing 7342 mcg of metanephrine (reference range 36-190 mcg), total metanephrines 2613 mcg (range reference 35-482 mcg), and total normetanephrines of 9955 mcg (range reference 1195-695 mcg).

On post-operative day 50 after the original cesarean hysterectomy and day 34 of alpha blockade. Highest blood pressure measured during surgery was 170/115 during manipulation of the left adrenal gland. After excision, the left adrenal pheochromocytoma measured 7.0 cm x 5.5 cm x 5.0 cm. Post-operatively, she reported resolution of her headaches and had no permanent neurologic sequelae.

**DISCUSSION**

The clinical features of pheochromocytoma are due to supraphysiologic catecholamine secretion. Hypertension is the most common symptom, either sustained or paroxysmal. Pheochromocytoma manifests most commonly as a hypertensive crisis. Other signs and symptoms can also occur including headaches, chest pain, dyspnea, abdominal pain, nausea, vomiting, dizziness, orthostatic hypotension, heart failure, arrhythmias, visual disturbances, seizures, and sudden death (4). The features of pheochromocytoma in pregnant or postpartum patients are similar to those seen in other patients. Peripartum pheochromocytoma is challenging to diagnose because it shares many features with preeclampsia, which is much more common. There are several postulated reasons why pheochromocytoma may manifest during pregnancy and peripartum. Increased intra-abdominal pressure, fetal movement, uterine contraction, and the stress of delivery all contribute to the development of clinically relevant pheochromocytoma (5). Proper management of pheochromocytoma involves more than simply the excision of the tumor. Prior to surgery, adequate alpha blockade must be established. Current recommendations involve at least a seven day course of an increasing dose of the irreversible alpha blocker phenoxybenzamine up to 30 mg three times per day (6). Oral prazosin or intravenous phenolamine can be used to manage paroxysms. If a pheochromocytoma is discovered during pregnancy, current literature suggests the best outcomes for both the mother and fetus are realized if the tumor can be removed prior to 23 weeks gestation. Because of uterine obstruction, after 23 weeks, medical management with an alpha blockade bridge to post-partum surgery is recommended (1). It is crucial to diagnose pheochromocytoma during pregnancy to aid in the prevention of fetal and maternal morbidity and mortality.

**REFERENCES**