

# Pheochromocytoma Presenting as Hypertension in Pregnancy

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## Abstract

Pheochromocytoma is a curable, rare cause of hypertension, characterized by symptoms and signs related to increased catecholamine secretion. Pregnancy and labour increase the risk of hypertensive crisis. However, antepartum diagnosis reduces both maternal and foetal mortality, allowing for safe cesarean section and resection of tumor. Control of hypertension with  $\alpha$  blockers and  $\beta$  blockers is the medical treatment. Surgical removal of the tumour is the definitive treatment. Hypertensive crisis needs to be treated aggressively. We report this case for the rare presentation of pheochromocytoma presenting as hypertension in pregnancy.

## Introduction

Pheochromocytomas are neuroendocrine tumors that typically present with paroxysms of hypertension. Sporadic pheochromocytomas are usually unicentric and unilateral while familial pheochromocytomas are often multicentric and bilateral. The triad of symptoms including headache, palpitations and sweating in patients with hypertension should arouse immediate suspicion of pheochromocytoma. Other common complaints are dyspnoea, weight loss despite normal appetite (caused by catecholamine-induced glycogenolysis and lipolysis), visual problems during an attack, and profound tiredness and polyuria, most commonly experienced after an attack. Unusual symptoms related to paroxysmal blood pressure elevation or sudden arrhythmia during diagnostic procedures (e.g. endoscopy, catheterization), anesthesia or food ingestion (e.g. tyramine in chocolate) should promptly arouse a suspicion of pheochromocytoma.

## Case History

A 26 year old female, housewife, gravida 2 (with 1 male child 5 years old alive and well), with 14 weeks of amenorrhoea, came with complaints of vomiting since 7 days to the Gynaecology OPD. She was detected to have increased blood pressure on antenatal check-up and was referred to us for the evaluation and management of hypertension. The gynaecologist had started her on tablet  $\alpha$  Methyldopa 250 mg thrice a day and capsule Nifedipine 5 mg thrice a day. There was no history of hypertension in the previous pregnancy or anytime in the past. She did not complain of breathlessness, headache, diplopia or blurring of vision. On direct questioning, she gave history of palpitations which were irregular and occurred after exertion and subsided with rest. There was no history of bouts of excessive sweating, headache, tremors or anxiety.

On examination, she was afebrile, pulse- 88/min/regular all peripheral pulses well felt. Blood pressure was 144/90 mm Hg in right arm supine position. There was no thyroid swelling. Systemic examination did not reveal any abnormality. Her blood investigations like complete blood count, liver, renal and thyroid functions, blood glucose and serum electrolytes were normal. The ECG and funduscopy were normal. The ultrasound of the abdomen showed a right adrenal mass measuring 4 cm x 2.9 cm. The ultrasound of the pelvis showed a single viable foetus of 12.5 weeks showing generalized skin edema and was reported to be hydrops fetalis. In view of the abnormal ultrasound pelvis

report, the gynecologist advised termination of pregnancy, we estimated 24 hours urinary metanephrines which were elevated 2.20 mg (Normal range: 0.00-1.00 mg/ 24 hours) and the urinary vanillyl mandelic acid (VMA) was elevated; 33.60 mg (Normal range: 0.00-13 mg/ 24 hours). Magnetic Resonance Imaging (MRI) of the abdomen with contrast showed a well defined, heterogeneously enhancing mass in the right adrenal gland with hemorrhagic focus and small calcification within it (Fig.1). The imaging features were suggestive of adrenal neoplasm, likely to be pheochromocytoma.

The treatment was changed to Phenoxybenzamine 10mg twice daily and Atenolol 25 mg twice daily. When adequate control of blood pressure was achieved, the patient underwent medical termination of pregnancy (MTP) as advised by the gynaecologist. In the post procedure period (within 30 mins), there was an episode of paroxysmal hypertension; blood pressure was 220/120 mm Hg and the patient was shifted to the intensive care unit (ICU) for the control of hypertension. She was treated with intravenous Sodium Nitroprusside for a period of 24 hours. Once the blood pressure was controlled, she was transferred to the general ward.

She was referred to the Urologist for removal of the adrenal mass. She was operated laparoscopically and tolerated the procedure well, without any intra-operative and post-operative complications. Post-operatively she became normotensive and was off anti-hypertensives. Till date the patient follows up regularly and has remained normotensive since then.

## Discussion

Pheochromocytoma is a curable rare cause of hypertension, characterized by symptoms and signs related to increased catecholamine secretion.<sup>1</sup> Pheochromocytomas are neuroendocrine tumors that typically present with paroxysms of hypertension, but occasionally can lead to marked hemodynamic instability, left ventricular dysfunction, and cardiovascular collapse. Pheochromocytomas occur in about 0.05% to 0.1% of patients with sustained hypertension.

Pregnancy can elicit clinical manifestations of otherwise unrecognized pheochromocytoma. Pregnancy complicated by pheochromocytoma is a life-threatening situation for both mother and foetus. Pregnancy and labour increase the risk of hypertensive crisis as it may occur with the sudden release of catecholamine accompanying uterine contractility and straining. However, early antepartum diagnosis and proper management with medical treatment followed by surgical removal of the tumor reduces both maternal and fetal mortality, allowing for safe cesarean section and resection of tumor resulting in good maternal and fetal outcomes.<sup>2,3</sup>

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**Table 1 : Management of Gestational Hypertension.<sup>3</sup>**

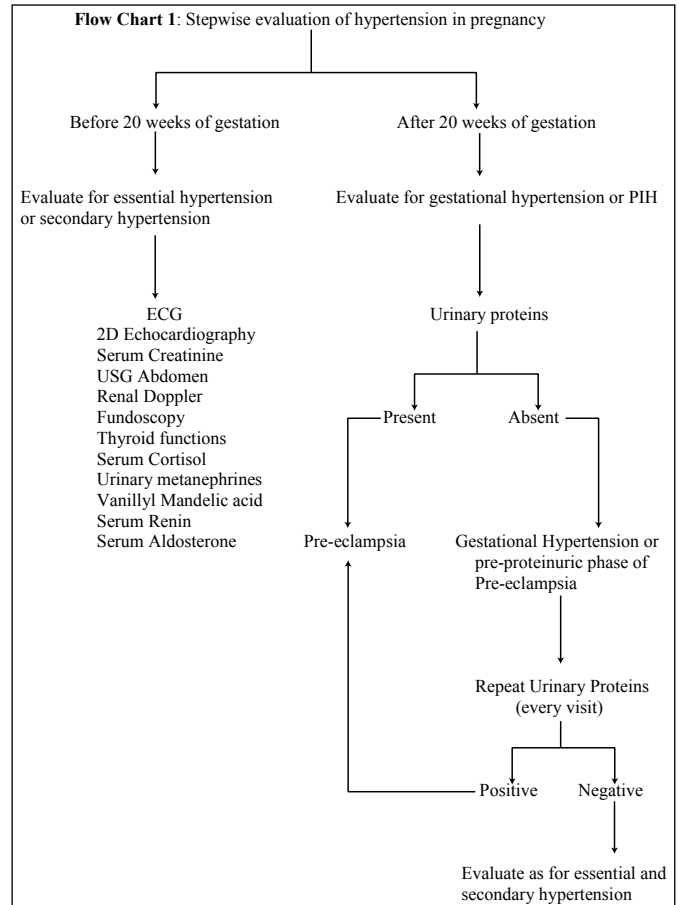
Stage I HT (BP > 140 mmHg Systolic or >90 mmHg Diastolic)	Methyldopa 250-500mg tds, Calcium channel blockers e.g. Amlodipin 5-10 mg od or Nifedipine 5-20 mg qds Diuretics like frusemide 20-40mg bd β blockers(Labetolol, atenolol). β blockers can cause IUGR(Labetolol is safer compared to atenolol);hence should be used with caution.
Stage II HT (BP > 160 mmHg Systolic or >100 mmHg Diastolic)	Combination of 2 or more drugs from the above(1 from each group) as required
Hypertensive crisis (BP > 180 mmHg Systolic or >110 mmHg Diastolic)	Nifedipine 10 mg orally can be repeated every 20 mins to a maximum of 30 mg. OR Nitroglycerine 5µg/min IV as a continuous drip with BP monitoring OR Hydralazine 5 mg IV bolus which can be repeated every 20-30 mins to a maximum of 25mg. OR Labetolol 20 mg IV bolus followed by 40 mg every 10 minutes till a maximum of 220 mg. OR Sodium nitroprusside 0.25µg/kg/min to a maximum of 5µg/kg/min (Used rarely due to fear of fetal cyanide poisoning)

Although pheochromocytoma in pregnancy is rare (estimated prevalence in full-term pregnancies of 1 in 54,000), factors specific to pregnancy can precipitate catecholamine crisis, making diagnosis and treatment challenging.<sup>4</sup> Pheochromocytoma should be searched for in the differential diagnosis of hypertension during pregnancy, especially in the young. The diagnosis can be made by elevated urinary catecholamines and by MRI scan as well as by ultrasonography. Surgical removal of the tumour is the definitive treatment. Pre-operatively, the patients are to be pre-treated with alpha-blockade followed by secondary beta-blockade. Post-operatively there is complete resolution of hypertension within a few hours.<sup>5</sup> The maternal mortality rate of undiagnosed pheochromocytoma is about 50%; this rate falls to 11% if diagnosis is made antepartum. The rate of foetal loss is similar even if the diagnosis is made during pregnancy.<sup>6</sup>

Surgery is indicated when tumors are functional or larger than 3 cm. Laparoscopy is the new gold-standard in treating adrenal pheochromocytomas. Nonfunctional and small tumors sized 3 cm or lesser may be closely followed up by imaging and hormonal investigations.<sup>7</sup>

The primary goals in the management of pheochromocytoma in pregnancy are early diagnosis, avoidance of a hypertensive crisis during delivery and definitive surgical treatment. Timing of surgical resection will depend on the gestational age at which diagnosis is made. Cesarean section is the preferred mode of delivery when the tumour is still present.<sup>8</sup>

We report this case for the rare presentation of pheochromocytoma presenting as hypertension in pregnancy, who had to undergo termination of pregnancy due to hydrops fetalis and became normotensive after surgical resection of the tumour.



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