

# Management of Pheochromocytoma in Pregnancy: Case Report and Review of the Literature

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**Abstract:** Pheochromocytoma during pregnancy is rare and lethal to the mother and fetus. Its diagnosis is often missed during pregnancy and is usually made during labour or immediately postpartum with increased mortality rates for both the mother and fetus. Early diagnosis and timely, appropriate management reduce possible maternal and fetal complications. A case of pheochromocytoma recognized at early pregnancy is described.

**Keywords:** Pheochromocytoma, hypertensive crisis, pregnancy, maternal mortality, fetal mortality.

## INTRODUCTION

Pheochromocytoma is a neuroendocrine tumor arising from chromaffin cells of the adrenal medulla or the sympathetic ganglia. Its presence is usually suspected in hypertensive patients with the appearance of symptoms such as headache, sweating or palpitations. Although it is an unusual cause of hypertension accounting for about 0.2-0.4% of cases, it may lead to severe and potentially lethal hypertensive crises due to the effects of the released catecholamines [1,2]. During pregnancy pheochromocytoma occurs with a frequency of 0.002% of all pregnancies [3,4]. Confusion with the much more prevalent forms of pregnancy-related hypertension is the main cause of overlooking the diagnosis. Consequently, a timely diagnosis and proper treatment are crucial because they may improve maternal and fetal morbidity and mortality [5-8]. If it remains undiagnosed the maternal and fetal mortality are estimated at around 50%, whereas the timely diagnosis improves the rates to <5% and <15% respectively [9,10]. We report a pregnant woman with pheochromocytoma which was diagnosed at the ninth week of gestation. Additionally, we analyze both the importance of early diagnosis and proper treatment for a successful outcome to take place.

## CASE REPORT

A 34-year-old woman was admitted to our department at 9 weeks gestation for management of hypertension. A history of paroxysmal episodes of hypertensive crises accompanied with headaches, sweating and palpitations was present from 8 months. Actually, in three episodes she was severely hypertensive with a blood pressure (BP) of 240/140 mmHg. Her previous medical history raised the suspicion of

pheochromocytoma; therefore, imaging and biochemical testing was performed to confirm or exclude the diagnosis. Ultrasound and magnetic resonance imaging demonstrated a 7,5 cm x 9,2 cm right adrenal mass (Fig. 1, 2).

Specialized biochemical examinations revealed increase in urinary catecholamines (889mg/24h; reference values: <150mg/24h) and urinary vanillylmandelic acid (28mg/24h; reference values: 1.8-6.7mg/24h), which confirmed the diagnosis of pheochromocytoma.

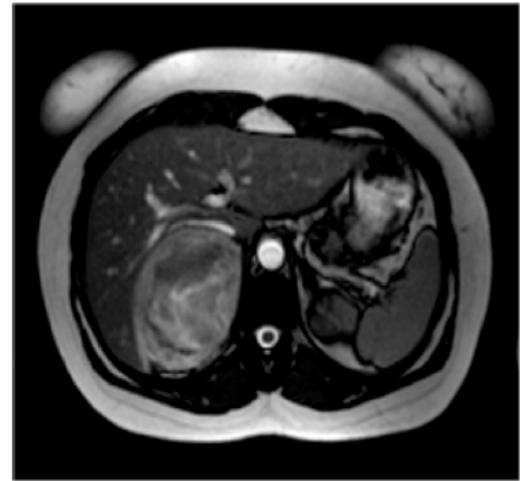
The patient was initially treated with alpha-blockers (terazosin) with a gradual increase in dose up to 5mg two times per day. Subsequently  $\beta$ -blockers (atenolol 25mg once daily) were added to the treatment 3 days later to account for tachycardia. This combination therapy was appropriate for the preoperative medical preparation so as to block the effects of released catecholamines. A multidisciplinary team of hypertension specialists, surgeons, anesthesiologists, pediatricians and obstetricians decided to perform tumor excision at the 13th week of gestation, in order to protect the fetus organogenesis and allow for proper preoperative management. The patient received the medication at home until the 13<sup>th</sup> week of gestation with a maximal pre-surgical BP of <130/80mmHg and complete disappearance of clinical manifestations. The surgery was conducted without any complications for the mother and fetus. BP remained remarkably stable during operation (even during tumor excision) due to proper preoperative management and the experience of the anesthesiology team. Figs. (3 and 4) depict the excised tumor, which had all the macroscopic features of pheochromocytoma with the characteristic color and within tumor hemorrhagic lesions and fusions.

The histological examination confirmed the preoperative diagnosis of pheochromocytoma. Postoperatively, the patient was in excellent clinical condition with a normal BP. The obstetrician confirmed the normal development of the fetus. The new sample of urinary catecholamines was within

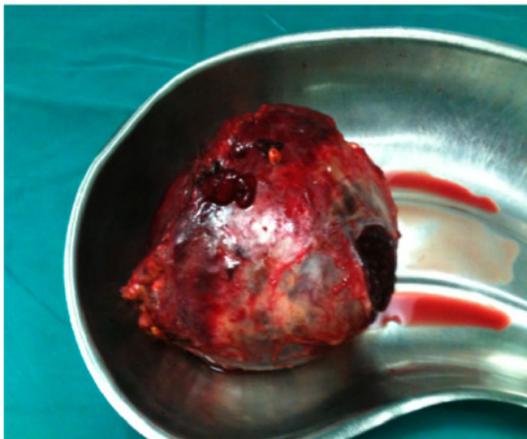
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**Fig. (1).** Ultrasound of the right adrenal showing a large mass.



**Fig. (2).** Abdominal magnetic resonance imaging revealing a large mass originating from the right adrenal gland.



**Fig. (3).** Photo depicting the mass right after surgical excision.



**Fig. (4).** Characteristic features of pheochromocytoma in the tumour.

reference values. The patient was discharged five days after the operation. Six months later (at the 39<sup>th</sup> week of gestation) a healthy infant was born with vaginal delivery.

## DISCUSSION

This case illustrates that the timely antenatal diagnosis of pheochromocytoma along with a proper preoperative adrenergic blockade and an experienced multi-specialist team result in the successful management of this highly risky clinical condition. The key factor of the successful management is the early detection of the tumor during pregnancy and its appropriate treatment [11,12]. Diagnosis in the antenatal period and resection during pregnancy were combined with better outcomes compared with the diagnosis in the immediate postnatal period or during delivery [13]. On the other hand, when the detection of the tumor was made before 23 weeks' gestation, slightly more successful results were observed with the excision in the second trimester than in the third trimester, but without statistical significance [13].

In the case of antenatal diagnosis the resection is recommended to be undertaken with laparoscopic procedures com-

pared to the conventional open surgery. With laparoscopic surgery better outcomes are achieved because of the timely control of catecholamine release and therefore hemodynamic stability and safety [14,15].

Caesarean section with epidural, general or combined anesthesia is the recommended mode of delivery and is related with lower maternal mortality (19%) than vaginal delivery (31 %) [6,15,16]. When the tumor is removed before 24 weeks of gestation, delivery can be conducted either way.

In conclusion, timely diagnosis of pheochromocytoma in a pregnant hypertensive patient and appropriate medical preoperative management followed by subsequent laparoscopic resection during pregnancy lead to reduction of maternal and fetal mortality. Most successful outcomes can be achieved with the contribution of a multidisciplinary team with high-level expertise in this specific field.

## CONFLICT OF INTEREST

The author(s) confirm that this article content has no conflicts of interest.

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