

## A Case of Pheochromocytoma during Pregnancy Missed Diagnosis as Preeclampsia

Medhat M Fadel<sup>1</sup> Khine M M Lwin<sup>2</sup> M.A. Badawey<sup>3</sup> A. Moustafa<sup>4</sup>

<sup>1</sup> Department of Obstetrics and Gynaecology, Ormskirk and Southport District Hospital, UK

<sup>2</sup> Wythenshawe Hospital, Manchester Foundation Trust, Manchester, UK

<sup>3</sup> Bronglais general hospital, General medicine, Aberystwyth, UK

<sup>4</sup> Dudley Group NHS Foundation Trust, Russells Hall Hospital, Obstetrics and gynaecology, Dudley, UK

\*Corresponding author: Mahmoud M. Metwally, Mobile: (+20) 01279520640, Email: [magicyya74@gmail.com](mailto:magicyya74@gmail.com)

### ABSTRACT

**Background:** Adrenal pheochromocytomas and extra-adrenal paragangliomas are rare tumors, with an incidence of 2-8 cases per million per year, often presenting with non-specific symptoms such as palpitations, headaches, and sweating.

**Objective:** This study aimed to diagnose a case of pheochromocytoma during pregnancy.

**Material and methods:** It is challenging due to the limitations of radiological imaging and the risks associated with invasive procedures. Biochemical testing for urinary and plasma metanephrines is essential.

**Management:** It requires a multidisciplinary approach. The primary treatment is surgical removal, but this poses risks during pregnancy, including preterm labor. Control of blood pressure is crucial before considering surgery. Alpha-adrenergic blockers like phenoxybenzamine are typically used in non-pregnant patients, but their use in pregnancy is limited due to potential risks. Beta-blockers and labetalol are not recommended as standalone treatments. **Conclusion:** In this case, the plan was to manage blood pressure with calcium channel blockers, with surgical treatment deferred until after delivery.

**Keywords:** Pheochromocytoma, Pregnancy, Hypertension, Blood pressure, Calcium channel blockers, multidisciplinary approach, Adrenal tumors, Alpha-adrenergic blockers, Symptoms, Maternal and fetal safety.

### INTRODUCTION

This case report highlights the clinical presentation, diagnosis, and management challenges of a pregnant woman with a suspected pheochromocytoma.

Symptoms of pheochromocytoma could be mixed with many symptoms of pregnancy and many of common pregnancy problems like preeclampsia, diagnosis and management could be challenging during pregnancy due to physiological limitations.

### CASE REPORT

A 22-week pregnant woman presented to The Emergency Department with severe headache and blurred vision. Her blood pressure was significantly elevated, with readings of 180/110 mmHg and 190/120 mmHg. Urine analysis revealed no proteinuria, and liver and kidney function tests were within normal limits. She had a history of hypertension and was under investigation for a suspected pheochromocytoma, with an MRI scan planned post-delivery (Figure 1,2,3,4). Initial treatment with labetalol that was ineffective, causing further elevation in blood pressure.

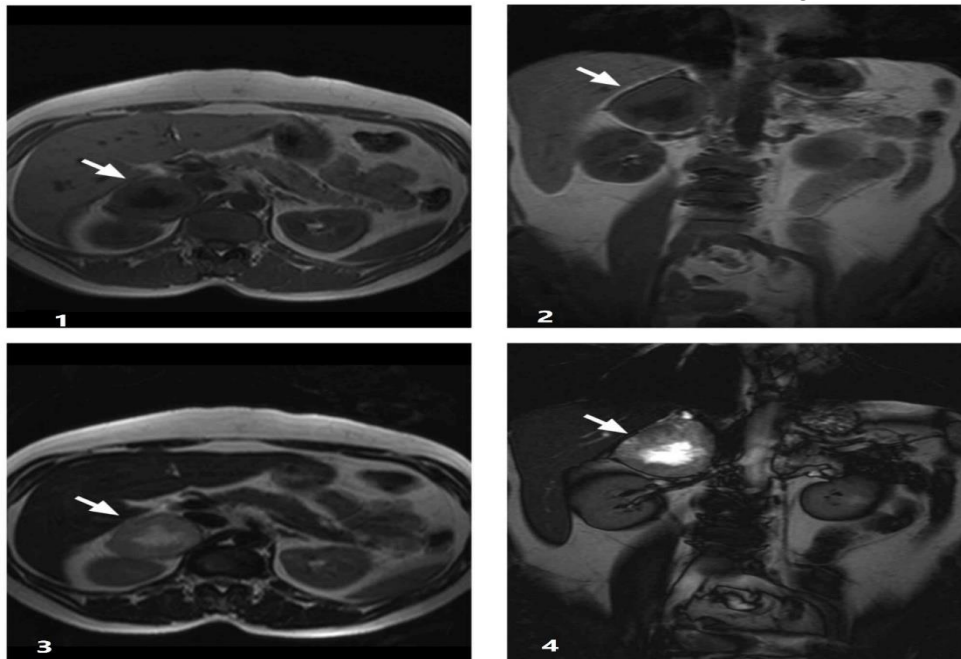


Figure (1,2,3,4): Showed consecutive sections in MRI scan for Pheochromocytoma on the right side.

## DISCUSSION

Adrenal pheochromocytomas and extra-adrenal paragangliomas, while rare, present a significant diagnostic and therapeutic challenge, especially during pregnancy. These tumors are characterized by the production of catecholamines, leading to symptoms like palpitations, headache, and sweating, which are non-specific and can overlap with other conditions common in pregnancy. The incidence of these tumors is relatively low, with only 2-8 cases per million per year, but their impact can be profound <sup>[1]</sup>.

Diagnosing pheochromocytoma during pregnancy is particularly challenging. The use of standard radiological imaging techniques, such as CT scans, is limited due to the potential risks of radiation exposure to the fetus. Similarly, invasive diagnostic procedures are avoided to prevent complications. Consequently, biochemical testing, including measurements of urinary and plasma metanephrines, becomes essential for a reliable diagnosis. These tests help in confirming the presence of pheochromocytomas and paragangliomas without exposing the fetus to unnecessary risks <sup>[2]</sup>.

Management of pheochromocytomas during pregnancy necessitates a multidisciplinary approach, involving endocrinologists, obstetricians, surgeons, and anesthesiologists to ensure both maternal and fetal safety. The definitive treatment for pheochromocytoma is surgical removal of the tumor. However, performing surgery during pregnancy carries significant risks, including the potential for preterm labor and other surgical complications. Thus, the timing of surgery is critical and is typically deferred until after delivery unless the mother's life is at imminent risk <sup>[2]</sup>.

Before surgery can be considered, achieving stable control of blood pressure is paramount. In non-pregnant patients, alpha-adrenergic blockers like phenoxybenzamine are commonly used to manage hypertension associated with pheochromocytoma. However, their use during pregnancy is constrained due to the potential risks to the fetus, making their application a case-by-case decision. Beta-blockers and labetalol, though often are used for hypertension, are not recommended as standalone treatments in this context because they can lead to unopposed alpha-receptor stimulation, potentially exacerbating hypertension <sup>[3]</sup>.

In the presented case, the decision was made to manage the patient's blood pressure with calcium channel blockers, which are considered safer during pregnancy. This strategy aimed to control the symptoms and stabilize the patient until delivery, at which point definitive surgical treatment can be performed. The goal was to balance the immediate need to manage severe hypertension with the long-term objective of tumor

removal, thereby minimizing risks to both mother and fetus <sup>[3]</sup>.

## Management of Pheochromocytoma during Pregnancy

Managing pheochromocytoma during pregnancy is challenging due to the physiological changes in pregnancy, the risks associated with diagnostic procedures, and the need to balance maternal and fetal health. The initial assessment involved a detailed clinical evaluation, focusing on symptoms such as severe hypertension, headaches, palpitations, and sweating. Biochemical testing included the measurement of urinary and plasma metanephrines and catecholamines, which is essential to confirm the diagnosis while avoiding the risks associated with ionizing radiation <sup>[4, 5]</sup>.

Diagnostic imaging plays a crucial role in identifying adrenal masses. Ultrasound is a non-invasive method that helps detect adrenal tumors without radiation exposure. MRI is preferred over CT scans during pregnancy because it does not involve ionizing radiation, making it safer for the fetus while providing detailed images to locate the tumor <sup>[5, 6]</sup>.

Effective blood pressure management is critical before considering surgical intervention. Alpha-adrenergic blockers, such as phenoxybenzamine, are typically used in non-pregnant patients to manage hypertension caused by pheochromocytoma. However, their use during pregnancy is cautious due to potential risks, as they are classified as class C drugs. Calcium channel blockers, like nifedipine, are considered safer alternatives during pregnancy for controlling blood pressure. Beta-blockers or labetalol are not recommended as standalone treatments because they can lead to unopposed alpha-adrenergic receptor stimulation, potentially exacerbating hypertension <sup>[4, 5]</sup>.

A multidisciplinary team approach is essential in managing pheochromocytoma during pregnancy. This involves coordination among endocrinologists, obstetricians, surgeons, and anesthesiologists to create an optimal treatment plan. Regular monitoring of maternal blood pressure and fetal well-being, including regular antenatal visits and serum catecholamine level assessments, is crucial <sup>[6]</sup>.

Surgical intervention remains the definitive treatment for pheochromocytoma, but the timing of surgery is critical. Surgery during pregnancy is risky and typically deferred until after delivery unless there is an immediate threat to the mother's life. If surgery is necessary during pregnancy, it is ideally performed in the second trimester after achieving adequate blood pressure control with alpha-blockade. Postoperative care includes continuous monitoring of blood pressure and serum catecholamine levels to prevent complications <sup>[6]</sup>.

Delivery planning must consider the mode and timing of delivery, which depends on obstetric indications and the patient's clinical stability. If possible, delivery is planned once blood pressure is well-controlled. Definitive surgical treatment is then planned post-delivery to remove the pheochromocytoma, minimizing risks to the fetus and allowing for optimal surgical conditions<sup>[7]</sup>.

Supportive care is also integral to the management of pheochromocytoma during pregnancy. This included hydration and electrolyte management through intravenous fluids and electrolyte replacement, which may be necessary to manage dehydration and electrolyte imbalances due to severe hypertension. Anti-emetics and nutritional support were provided to manage symptoms like vomiting and ensure adequate nutrition for both the mother and the fetus<sup>[7]</sup>.

Patient education and counseling are vital components of managing pheochromocytoma during pregnancy. Educating the patient about the nature of pheochromocytoma, the associated risks, and the treatment plan is crucial. Providing psychological support and counseling helps address any concerns related to the condition and its management during pregnancy. Also, ensuring the patient was well-informed and supported throughout the process<sup>[7]</sup>.

## CONCLUSION

The management of pheochromocytoma during pregnancy is a complex and multifaceted challenge that necessitates a thorough and coordinated approach to ensure both maternal and fetal well-being. This case underscored the importance of considering pheochromocytoma in the differential diagnosis of severe hypertension during pregnancy. A multidisciplinary team, comprising endocrinologists, obstetricians, surgeons, and anesthesiologists, is crucial for developing and implementing an effective treatment plan.

**Ethical Approval: This case study has been published after a written informed consent from the patient, all**

**related information and images have been kept strictly confidential and the publication is kept anonymous .**

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## REFERENCES

1. **Lenders J, Duh Q, Eisenhofer G et al. (2014):** Pheochromocytoma and paraganglioma: an endocrine society clinical practice guideline. *The Journal of Clinical Endocrinology & Metabolism*, 99 (6): 1915-1942. doi:10.1210/jc.2014-1498
2. **Manger W, Gifford R (2002):** Pheochromocytoma: A Multidisciplinary Approach. Springer Science & Business Media.
3. **Neumann H, Eng C (2009):** The approach to the patient with paraganglioma, pheochromocytoma, or familial syndromes associated with these tumors. *The Journal of Clinical Endocrinology & Metabolism*, 94 (8) : 2677-2683. doi:10.1210/jc.2009-0574
4. **Young W (2019):** Pheochromocytoma and paraganglioma in children and adolescents. *Journal of Internal Medicine*, 285 (1): 1-12. doi:10.1111/joim.12825
5. **Plouin P, Amar L, Dekkers O et al. (2016):** European Society of Endocrinology Clinical Practice Guideline for long-term follow-up of patients operated on for a phaeochromocytoma or a paraganglioma. *European Journal of Endocrinology*, 174 (5): G1-G10. doi:10.1530/EJE-16-0033
6. **Chen H, Sippel R, O'Dorisio M et al. (2010):** The North American Neuroendocrine Tumor Society consensus guideline for the diagnosis and management of neuroendocrine tumors: pheochromocytoma, paraganglioma, and medullary thyroid cancer. *Pancreas*, 39 (6): 775-783. doi:10.1097/MPA.0b013e3181ebb4f0.
7. **Biggar M, Lennard T (2013):** Systematic review of phaeochromocytoma in pregnancy. *British Journal of Surgery*, 100 (2): 182-190. doi:10.1002/bjs.8949.