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CASE REPORT

Diagnosis and Management of Pheochromocytoma in Pregnancy: A Multidisciplinary Approach to Treatment and Outcomes at Mbarara Regional Referral Hospital: A Case Report

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Introduction: Pheochromocytoma in pregnancy is extremely rare but can be life-threatening for both mother and fetus. Its clinical manifestations often mimic common obstetric complications, such as preeclampsia, resulting in delayed or missed diagnosis with potentially catastrophic consequences. Prompt recognition and careful management are critical to improving maternal and neonatal outcomes.

Case Presentation: We report a 27-year-old Black African multiparous refugee woman at 26 weeks of gestation who presented with palpitations, shortness of breath, headaches, and markedly fluctuating blood pressure. Ultrasonography revealed a large right adrenal mass, and 24-hour urinary metanephrines confirmed catecholamine excess. Despite antihypertensive therapy, she continued to experience alternating hypertensive and hypotensive crises, necessitating an emergency cesarean delivery at 31 weeks of gestation. A live preterm neonate weighing 1.8 kg was delivered in good condition. Ten weeks postpartum, she underwent laparoscopic adrenalectomy. Histopathological examination and immunohistochemistry evaluation confirmed the diagnosis of pheochromocytoma.

Conclusion: This case highlights the diagnostic challenge of pheochromocytoma in pregnancy, which can masquerade as preeclampsia and other hypertensive disorders. It underscores the importance of considering pheochromocytoma in the differential diagnosis of labile blood pressure during pregnancy. Multidisciplinary coordination, vigilant hemodynamic stabilization, and timely postpartum surgery can achieve excellent maternal and neonatal outcomes.

Keywords: pheochromocytoma, pregnancy, adrenalectomy, hypertensive crisis, hypotensive crisis, preeclampsia mimic, catecholamine storm, multidisciplinary management, immunohistochemistry

Introduction

Pheochromocytoma (PHEO) is a rare metastatic potential neuroendocrine tumor that arises from neural crest-derived tissue and belongs to the broader group of paragangliomas (PGLs).[1] Paragangliomas are hormonally active tumors that secrete excess catecholamines such as norepinephrine, epinephrine, and dopamine, resulting in a spectrum of clinical symptoms.[2] Although uncommon, PHEO can present during pregnancy, posing significant diagnostic and therapeutic challenges. Its prevalence among patients with hypertension is estimated at 0.3%, while in pregnancy, it is even rarer, occurring in roughly 1 in 54,000 pregnancies (0.007%).[1,3] The clinical presentation is often nonspecific, with symptoms that may be subtle or easily mistaken for more common conditions such as preeclampsia. This can lead to delays in diagnosis until a hypertensive crisis occurs, sometimes with catastrophic maternal or fetal consequences.[4]

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A PHEO crisis can trigger placental abruption, fetal hypoxia, and even intrauterine fetal death, highlighting the life-threatening potential of this condition. Maternal mortality in pregnancies complicated by PHEO has been reported at 8%, with fetal and neonatal mortality reaching 17%.[5]

We present this case to raise awareness of PHEO as a rare but potentially life-threatening cause of labile hypertension in pregnancy, particularly when symptoms mimic preeclampsia or recur across multiple pregnancies. This case emphasizes the importance of careful assessment of labile hypertension, sequential imaging, biochemical confirmation, and a coordinated multidisciplinary approach, highlighting practical strategies that obstetricians can implement to optimize both maternal and fetal outcomes. Despite sporadic case reports, evidence-based guidelines for the diagnosis and management of PHEO in pregnancy remain limited, underscoring the value of sharing individual experiences. By presenting this case, we aim to improve the timely recognition and management of this rare but serious condition.

Case Presentation

A 27-year-old multiparous refugee woman (G3P2) with no notable past medical history presented at 26 weeks of gestation with worsening shortness of breath, palpitations, chest tightness, headaches, and markedly labile blood pressure, including inter-arm differences. Her obstetric history was significant for two prior adverse pregnancy outcomes. In 2022, she delivered at 36 weeks by spontaneous vaginal delivery at a Health Centre III near the refugee camp, resulting in a stillbirth weighing 2.0 kg with no external congenital abnormalities; the cause of death was unknown. In 2023, her second pregnancy was complicated by severe preeclampsia at 33 weeks, necessitating an emergency cesarean section at a Health Centre IV and resulting in a stillbirth weighing 2.4 kg. There was no known family history of endocrine tumors or hereditary syndromes. During her current pregnancy, she reported similar symptoms, raising concern that prior hypertensive crises may have been misattributed to preeclampsia when they could have represented PHEO related episodes.

In the current pregnancy, she first presented at 11 weeks with shortness of breath and chest pain. On assessment, she was found to have tachycardia and tachypnea, but no hemoptysis, and was suspected to have pulmonary embolism based on clinical findings. She was initially treated with low-molecular-weight heparin but was later maintained on warfarin after the first trimester due to financial constraints, attending the high-risk clinic every two weeks with close INR monitoring.

In the first trimester, she underwent an ultrasound at a peripheral clinic, where a large mass was noted and reported as a right ovarian mass. She did not raise any concerns at the time. Upon admission, review of her prior imaging confirmed this finding, though subsequent ultrasounds did not mention it. Later, during an ultrasound for fetal well-being, a right suprarenal mass was identified, revealing that the initial sonographer had visualized the mass but misinterpreted it as an ovarian lesion, resulting in delayed recognition of the true pathology.

At 26 weeks' gestation, during her third hospital admission for worsening symptoms, the initial diagnosis of pulmonary embolism was reconsidered. She was started on nifedipine and methyldopa for presumed preeclampsia without proteinuria. However, her blood pressure readings were intermittently low, leading to discontinuation of these medications. Despite this, her symptoms, including palpitations, headaches, and labile hypertension, gradually worsened. The complexity of her presentation prompted a multidisciplinary review. Differential diagnoses considered included hyperthyroidism, which was excluded by normal thyroid hormone levels, and Takayasu arteritis, though no definitive diagnosis was established.

At 28 weeks' gestation, a fetal well-being ultrasound revealed a right suprarenal mass 9.47×8.09 x 8.46 cm. The obstetric assessment confirmed a viable fetus with appropriate growth, normal amniotic fluid volume and no structural anomalies. Fetal Doppler assessment of cerebroplacental perfusion, including the middle cerebral and umbilical arteries, revealed normal pulsatility indices, consistent with preserved fetal perfusion.

Subsequently, a 24-hour urinary metanephrine test was performed, which showed a markedly elevated level of 11.9 ng/24 hours compared with the normal reference range shown in Table 1, supporting the diagnosis of a catecholamine-secreting adrenal tumor. Medical therapy was initiated with doxazosin (an alpha-1 adrenergic receptor blocker) and propranolol (a non-selective beta-adrenergic blocker) for symptom control. Despite pharmacologic management, her symptoms persisted.

Further investigations revealed no sinus tachycardia, no S1Q3T3 pattern on electrocardiography as shown in Figure 1, normal findings on echocardiography, and routine laboratory results, including complete blood count, liver function test, renal function test, urine protein, activated partial thromboplastin time (aPTT), prothrombin time (PT), and international

Table 1 Catecholamine Metabolite Testing

Catecholamine Metabolite	Patient Value	Reference Range	Interpretation
24-hour urinary metanephrine	11.9 ng/24 h	2-10ng/24hrs	Reported as markedly elevated
Normetanephrine	Not available	—	Not measured separately
3-methoxytyramine	Not available	—	Not measured

normalized ratio (INR) within normal limits. Daily obstetric surveillance included fetal heart-rate monitoring every four hours, while weekly ultrasound assessments consistently confirmed ongoing fetal viability.

In view of persistent maternal symptoms, a prior cesarean scar, and non-reassuring fetal status on continuous fetal monitoring, an emergency cesarean section was undertaken at 31 weeks' gestation after receiving dexamethasone 6 mg intramuscularly every 12 hours for four doses over 48 hours to promote fetal lung maturation, and a single 4 g intravenous dose of magnesium sulfate in 20% solution for fetal neuroprotection, followed by emergency cesarean delivery. A preterm female infant weighing 1.8 kg was delivered, attended by the neonatologist, and subsequently transferred to the neonatal intensive care unit for specialized care. The mother was transferred to the intensive care unit, where she remained for three days for ongoing crisis management.

The mother continued to experience persistent symptoms postpartum and was referred to Mulago National Referral Hospital two weeks after delivery. A preoperative contrast-enhanced CT scan characterized the adrenal lesion as heterogeneous with central necrosis, measuring 9.6×8.6 × 6.6 cm, without invasion of adjacent structures or distant metastasis, although mild left renal hilar lymphadenopathy was noted, as shown in Figure 2A and B. Ten weeks postpartum, she underwent a right laparoscopic adrenalectomy performed by a multidisciplinary surgical team, including urology specialists. Intraoperatively, the tumor was largely confined to the adrenal capsule with clear margins. Histopathology and immunohistochemistry confirmed the diagnosis of pheochromocytoma, with histopathological findings shown in Figure 3 and immunohistochemical results demonstrating strong positivity for synaptophysin and chromogranin, as shown in Table 2. Follow-up CT scan demonstrated complete excision of the adrenal mass, persistent mild left renal hilar lymphadenopathy (N1), and no distant metastasis (M0) as shown in Figure 4A and B.

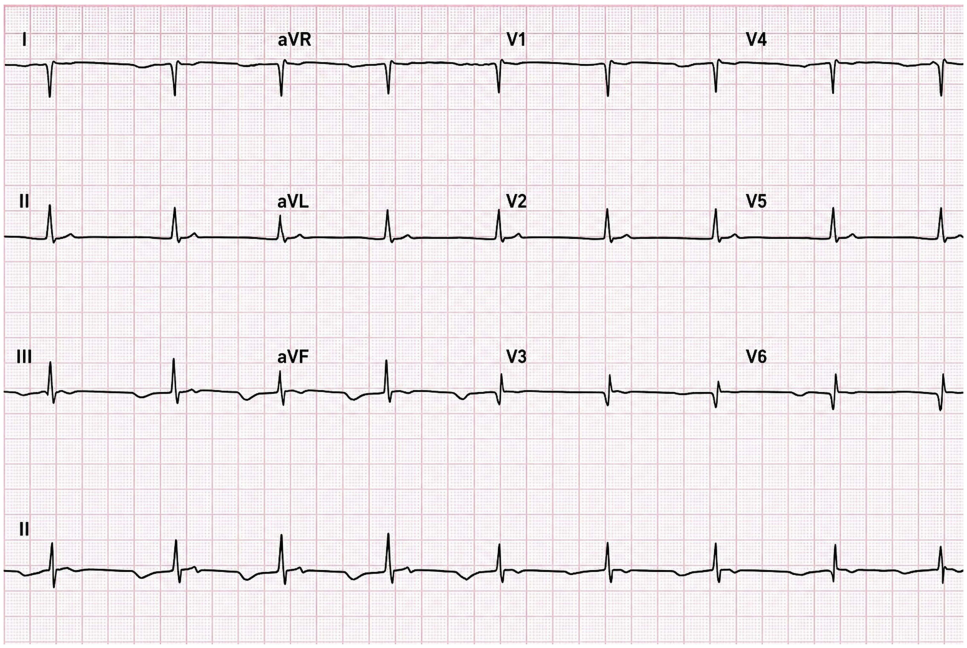


Figure 1 Normal Electrocardiogram (ECG).

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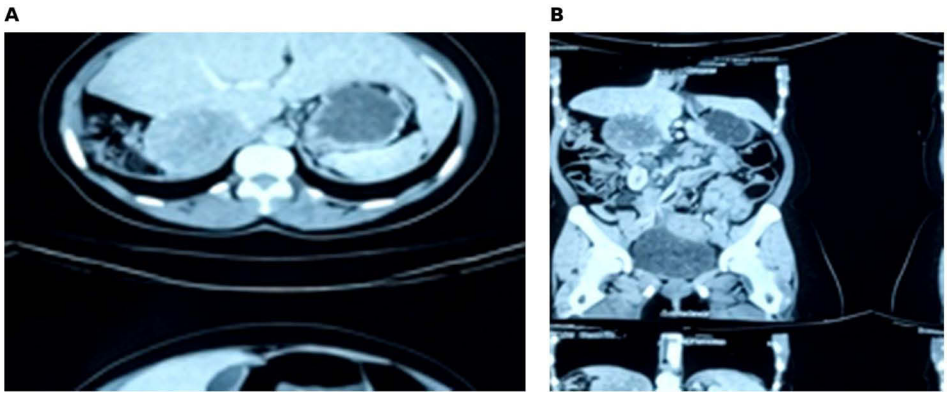


Figure 2 Contrast-enhanced computed tomography images showing a right adrenal mass. (**A**) Axial CT section demonstrating the adrenal lesion. (**B**) Coronal CT section showing the same lesion in relation to the right kidney and adjacent abdominal structures.

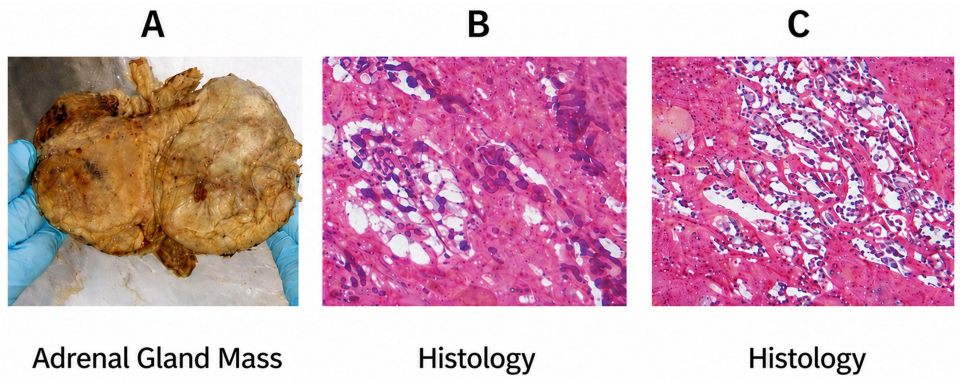


Figure 3 Surgical pathology findings of the adrenal gland mass. (**A**) Gross specimen showing the resected adrenal gland mass. (**B**) Histological section showing tumor cells arranged in nests/trabeculae. (**C**) Histological section showing vascular/stromal features consistent with pheochromocytoma.

Her care was coordinated by a multidisciplinary team comprising obstetricians, anesthesiologists, endocrinologists, internal medicine specialists, urologists, surgeons, and a neonatologist, which was pivotal in achieving a favorable outcome. Interventions were adjusted as needed: nifedipine and methyldopa were discontinued due to hypotension, low-molecular-weight heparin was switched to warfarin after the first trimester for feasibility, and doxazosin and propranolol were initiated after PHEO diagnosis to control labile hypertension.

During one year of follow-up, the patient remained asymptomatic with stable blood pressure and no biochemical or radiological evidence of recurrence. She demonstrated good adherence to prescribed medical therapy, perioperative

Table 2 Immunohistochemistry Results of the Adrenal Gland Mass

Immunohistochemical Marker	Result
Inhibin	Negative
Melan A	Negative
Synaptophysin	Positive
Chromogranin	Positive
Calretinin	Negative
Ki-67 proliferation index	Low (<10%)

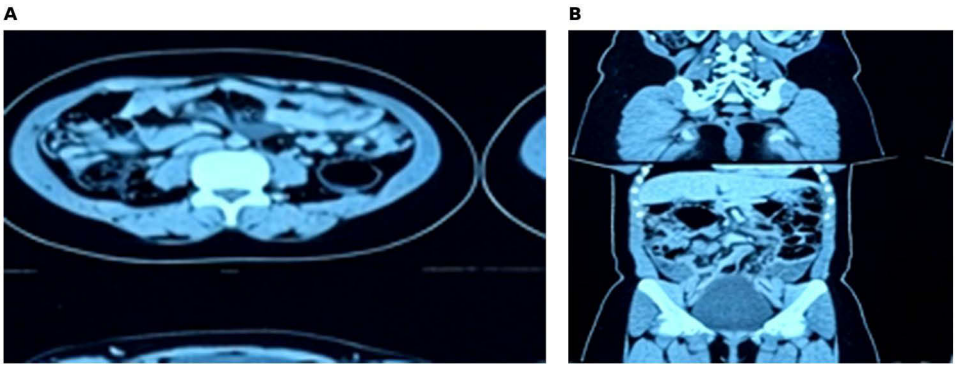


Figure 4 Contrast-enhanced abdominal CT images. (**A**) Axial CT section. (**B**) Coronal CT section.

management, and postoperative follow-up. All interventions were well tolerated, with no reported or observed treatment-related adverse events or surgical complications during the follow-up period. She was discharged in good condition, with both mother and infant thriving. As shown in Table 3, the detailed timeline of clinical presentation, investigations, management, and outcomes is summarized.

Discussion

PHEO is a rare catecholamine-secreting neuroendocrine tumor with metastatic potential that arises from chromaffin cells of the adrenal medulla, whereas paragangliomas (PGLs) are extra-adrenal tumors arising from sympathetic or parasympathetic paraganglia.[1] It accounts for approximately 0.2–0.4% of all cases of hypertension, and its occurrence during pregnancy is exceedingly rare, with an estimated prevalence of 1 in 54,000 pregnancies.[6]

Approximately 85% of pheochromocytomas and paragangliomas (PPGLs) arise in the adrenal medulla as pheochromocytomas, while the remaining 15% are extra-adrenal paragangliomas (PGLs). PGLs may occur at different anatomical sites, including retroperitoneal sympathetic paraganglia, mediastinal paraganglia, and parasympathetic head and neck paraganglia such as vagal paragangliomas.[7] In our patient, the tumor was located in the right adrenal medulla.

Table 3 Timeline of the Clinical Events

Time Point	Clinical Events
2022 (1st pregnancy)	Spontaneous vaginal delivery at 36 weeks; stillbirth (2.0 kg); cause unknown.
2023 (2nd pregnancy)	Severe preeclampsia at 33 weeks; emergency cesarean section; stillbirth (2.4 kg).
11 weeks gestation (current pregnancy)	Presented with shortness of breath and chest pain; suspected pulmonary embolism; anticoagulation initiated.
First trimester ultrasound	Large mass misinterpreted as right ovarian mass at peripheral clinic.
26 weeks gestation	Re-admission with palpitations, headaches, labile blood pressure; treated as preeclampsia without proteinuria.
28 weeks gestation	Ultrasound revealed right suprarenal mass (9.47 × 8.09×8.46 cm); suspicion of pheochromocytoma.
28–29 weeks gestation	24-hour urinary metanephrines markedly elevated; alpha- and beta-blockade initiated.
31 weeks gestation	Persistent maternal crises and non-reassuring fetal status; emergency cesarean delivery; live preterm neonate (1.8 kg).
2 weeks postpartum	Referral to Mulago National Referral Hospital; CT scan confirmed adrenal mass.
10 weeks postpartum	Right laparoscopic adrenalectomy performed; histopathology and immunohistochemistry confirmed pheochromocytoma.
1 year follow-up	Mother asymptomatic; stable blood pressure; no biochemical or radiological recurrence; infant thriving.

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The classic clinical triad of episodic hypertension, palpitations, and diaphoresis (Menard's triad) is characteristic[8] and was present in this case. However, as commonly reported, the patient experienced a prolonged diagnostic delay, having received multiple alternative diagnoses across several healthcare encounters. Her symptoms dated back to a previous pregnancy and were initially misattributed to more common obstetric conditions, particularly preeclampsia. This reflects both the rarity of PHEO and its non-specific presentation, which often overlaps with hypertensive disorders of pregnancy. Delayed diagnosis is associated with high maternal and fetal morbidity and mortality.[4]

Early diagnosis during pregnancy is challenging but critical. Progressive gestation increases the risk of hypertensive crises due to rising intra-abdominal pressure, tumor vascularity, fetal movements, and labor-related stress. These crises can result in uteroplacental insufficiency, fetal growth restriction, hypoxia, or intrauterine fetal demise. The risk is greatest in the third trimester, during delivery, and in severe cases, early delivery or pregnancy termination may be required because of hemodynamic instability.[9]

PHEO is rarely reported among Black African populations, although available evidence suggests that clinical presentation does not differ significantly across ethnic groups. In sub-Saharan Africa, diagnosis is further complicated by limited access to specialized biochemical assays and advanced imaging modalities. Most regional reports consist of retrospective case series, with very few cases describing pregnancy-associated presentations.[7]

The systemic effects of excess catecholamines can involve multiple organs, particularly the cardiovascular system. Sustained hypertension and prolonged catecholamine exposure may result in left ventricular hypertrophy, arrhythmias, dilated cardiomyopathy, cardiogenic shock, or acute coronary syndromes.[10] Pregnancy may exacerbate these complications due to increased hemodynamic demands and heightened catecholamine sensitivity.[5]

Diagnosis is established biochemically through measurement of catecholamines and their metabolites, including plasma free metanephrines and 24-hour urinary catecholamines, metanephrines, and vanillylmandelic acid (VMA), which are not affected by pregnancy. Tumor localization is achieved using imaging modalities such as ultrasound, CT, or MRI, while definitive confirmation relies on histopathological and immunohistochemical analysis. Cardiac evaluation with ECG and echocardiography is essential, and genetic testing should be considered where indicated.[11,12] In this patient, biochemical testing, imaging, and histological evaluation confirmed the diagnosis.

Functional nuclear medicine imaging has an important role in the evaluation of pheochromocytoma and paragangliomas, particularly for tumor localization, detection of metastatic or multifocal disease, treatment planning, and follow-up. SSTR2-targeted DOTATATE PET/CT using ^68Ga or ^64Cu tracers has been highlighted as a valuable approach in PPGL diagnosis and management.[13] However, nuclear medicine imaging was not performed in this case because it was not available in our setting, which represents a limitation of this report.

Management involves careful medical optimization followed by definitive surgical resection. Preoperative alpha-adrenergic blockade, with or without beta-blockers, is essential to control hypertension, mitigate catecholamine-mediated effects, and restore intravascular volume, thereby reducing perioperative complications. Calcium channel blockers, such as nicardipine, may be used as adjuncts.[7,11] In our patient, medical therapy failed to adequately control hypertensive crises, likely due to a large tumor burden and physiological changes of late pregnancy. Symptomatic improvement occurred only after delivery.

Surgical resection remains the definitive treatment and may be performed via open, laparoscopic, or robot-assisted approaches, depending on tumor characteristics. In pregnancy, the second trimester is considered the optimal timing for adrenalectomy, balancing maternal safety with fetal viability.[11,14] In this case, late third-trimester diagnosis precluded immediate surgery. Medical management was continued to allow fetal lung maturation; however, escalating crises and poor fetal reassurance necessitated delivery. Definitive adrenalectomy was successfully performed 12 weeks postpartum.

Postoperative follow-up is guided by recurrence risk. High-risk patients—those with tumors larger than 5 cm, young age at diagnosis, genetic mutations, or paragangliomas—require lifelong annual biochemical surveillance with periodic imaging due to the risk of recurrence or metastasis. Low-risk patients may be followed with annual biochemical testing alone.[15] Our patient was classified as high risk based on tumor size and is currently on annual follow-up. At one year post-surgery, biochemical and imaging evaluations show no evidence of recurrence.

This case reinforces existing clinical practice guidelines that recommend early consideration of PHEO in pregnant women presenting with labile or refractory hypertension accompanied by adrenergic symptoms, particularly when

hypertension is atypical for preeclampsia. It underscores the importance of biochemical screening with plasma or urinary metanephrines and multidisciplinary management, as well as individualized timing of delivery and definitive surgical intervention. In low-resource settings, this report highlights the need for heightened clinical suspicion and careful interpretation of ultrasound findings to prevent diagnostic delay and improve maternal and fetal outcomes.

This case is hypothesis-generating, suggesting that targeted early biochemical screening for PHEO in pregnant women with atypical or recurrent hypertensive disorders may reduce diagnostic delay and improve maternal and fetal outcomes, a hypothesis that warrants evaluation in future prospective studies.

Strength and Limitations

This case report is strengthened by a detailed longitudinal obstetric history across multiple pregnancies, diagnostic confirmation using biochemical, radiological, histopathological, and immunohistochemical evidence, and effective multidisciplinary management in a low-resource setting. Adherence to CARE reporting guidelines and one-year follow-up showing favorable maternal and neonatal outcomes without recurrence further enhance its clinical relevance and educational value. However, the report is limited by its single-case design, which may restrict generalizability, and by resource constraints that limited access to advanced investigations, including nuclear medicine imaging, genetic testing, separate catecholamine metabolite profiling, objective imaging confirmation for suspected pulmonary embolism, and phase-specific CT Hounsfield unit assessment.

Conclusion

Pheochromocytoma in pregnancy, though extremely rare, poses significant risks for both mother and fetus due to its potential to cause hypertensive crises and multisystem complications. Early recognition, biochemical confirmation, and careful imaging are crucial for diagnosis, while a multidisciplinary approach involving obstetricians, endocrinologists, anesthesiologists, surgeons and neonatologists is essential for optimal maternal and fetal outcomes. Timely medical stabilization, appropriate timing of delivery, and definitive postpartum surgical management can result in favorable long-term outcomes, as demonstrated in this case.

Patient Perspective

The patient reported significant relief of symptoms and improvement in quality of life following definitive treatment.

Abbreviations

PHEO, pheochromocytoma; PGL, paraganglioma; ECG, electrocardiogram; CT, computed tomography; MRI, magnetic resonance imaging; NICU: Neonatal Intensive Care unit; INR, international normalized ratio; N1, regional lymph node involvement; HTN, hypertension; MEN II, multiple endocrine neoplasia type II; VMA, vanillylmandelic acid.

Data Sharing Statement

All data generated or analyzed during this case report are included in this published article. No additional datasets were generated or analyzed.

Ethics Approval

Ethical approval for the conduct and publication of this case report was granted by the Mbarara Research Ethics Committee (MUST-2025-293).

Informed Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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Disclosure

The authors declare no conflicts of interest related to this study.

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