

# Management and Long-Term Monitoring of a Young Patient with Pheochromocytoma and RET Mutation: A Case Report

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**Background:** Pheochromocytoma is a rare catecholamine-secreting tumor that can present with severe hypertensive episodes and other symptoms due to excessive catecholamine release. Approximately 30% of pheochromocytomas are associated with hereditary syndromes, including multiple endocrine neoplasia type 2A (MEN2A), an autosomal dominant disorder caused by mutations in the RET proto-oncogene. MEN2A is characterized by the presence of medullary thyroid carcinoma, pheochromocytoma, and primary hyperparathyroidism.

**Case Presentation:** We report the case of a 19-year-old female who presented with pheochromocytoma without experiencing a crisis, despite having a significant adrenal mass and undergoing high-dose glucocorticoid treatment. Genetic testing revealed a heterozygous missense mutation in the RET gene (c.1900T > C: p. Cys634Arg), associated with MEN2A. Further endocrine evaluation identified a thyroid nodule with mildly elevated calcitonin levels, but normal electrolyte and parathyroid hormone levels. Over a 15-month postoperative follow-up, the patient exhibited persistently mild hypercalcitoninemia with stable thyroid nodule size, while PTH and serum calcium levels showed a progressive increase. Further parathyroid scintigraphy using <sup>99m</sup>Tc-MIBI was performed, yielding a negative result for parathyroid adenoma.

**Conclusion:** Patients with MEN2A require comprehensive, long-term follow-up to monitor for recurrence of pheochromocytoma and the development of additional endocrine neoplasms. This case highlights the role of genetic testing in guiding the management of hereditary pheochromocytoma and supports the importance of personalized monitoring strategies in patients with MEN2A.

**Keywords:** pheochromocytoma, glucocorticoids, RET mutation, case report

## Introduction

Pheochromocytoma is a rare catecholamine-secreting tumor arising from chromaffin cells of the adrenal medulla, accounting for approximately 0.1% to 0.6% of cases in hypertensive patients.<sup>1</sup> Clinically, it often presents with paroxysmal or sustained hypertension, headaches, palpitations, and diaphoresis due to excessive catecholamine release.<sup>2</sup> While most pheochromocytomas are sporadic, about 30% are associated with hereditary syndromes, notably multiple endocrine neoplasia type 2A (MEN2A).<sup>3</sup> MEN2A is an autosomal dominant disorder characterized by the triad of medullary thyroid carcinoma, pheochromocytoma, and primary hyperparathyroidism, primarily linked to mutations in the RET proto-oncogene.<sup>4</sup> Among these, the Cys634Arg mutation is particularly associated with a higher penetrance of endocrine tumors. Early identification and management of MEN2A are crucial due to the potential for aggressive tumor behavior and the risk of recurrence.<sup>5</sup>

This case report discusses a 19-year-old female with a RET Cys634Arg mutation, presenting with pheochromocytoma without experiencing a crisis, even under high-dose glucocorticoid treatment. The case underscores the importance of genetic evaluation and long-term follow-up in patients with hereditary pheochromocytoma.

## Case Presentation

A 19-year-old female presented to our hospital's emergency room with a chief complaint of a two-day history of headache, accompanied by recurrent nausea, vomiting, and a one-day fever. On admission, her physical examination revealed a high fever of 39.1°C, elevated blood pressure at 189/120 mmHg, and a pulse rate of 148 beats per minute. Laboratory results indicated an elevated white blood cell count of  $14.77 \times 10^9/L$  and a neutrophil count of  $13.55 \times 10^9/L$ , suggesting a possible infection or inflammatory response. Initial empirical treatment with antibiotics was administered due to suspected infection, but her symptoms persisted. Given her abnormal vital signs, elevated inflammatory markers, and lack of symptom improvement, the patient was admitted for further diagnostic evaluation and transferred to the intensive care unit for close monitoring. A year prior, the patient had presented with similar symptoms and was diagnosed with myocarditis at a local hospital based on clinical findings at that time. During that hospitalization, she was also diagnosed with hypertension and prescribed antihypertensive medications. However, after discharge, the patient did not adhere to the prescribed antihypertensive therapy and did not regularly monitor her blood pressure. Additionally, it is notable that her father had a history of sudden, unexplained death.

The initial laboratory findings are presented in Table 1, with key laboratory data throughout the treatment course detailed in Figure 1. To investigate the underlying etiology of the patient's symptoms, a chest computed tomography (CT) scan was performed. Incidentally, this scan revealed a left adrenal mass with soft tissue density, measuring 43 mm × 36 mm (Figure 2). No pathological findings were observed in the head and chest CT scans. The electrocardiogram demonstrated sinus tachycardia with a shortened PR interval and tall, peaked P-waves in leads II, III, and aVF. Transthoracic echocardiography did not reveal any significant abnormalities.

On the second day of admission, the patient exhibited rising levels of brain natriuretic peptide (BNP) and Troponin I (TnI). The cardiologist provisionally diagnosed the patient with myocarditis of uncertain etiology, based on clinical presentation, elevated cardiac biomarkers (BNP and TnI), and supportive electrocardiogram findings. Treatment was initiated with methylprednisolone (0.25 g daily) to address potential myocardial inflammation due to suspected myocarditis. Furosemide (20 mg every 12 hours) and spironolactone (20 mg every 12 hours) were administered as diuretics to manage fluid retention and reduce cardiac workload. Perindopril amlodipine (10 mg: 5 mg daily) was prescribed as an angiotensin-converting enzyme inhibitor and calcium channel blocker combination to control blood pressure and reduce afterload. Metoprolol tartrate (25 mg every 12 hours) was used to manage heart rate and decrease myocardial oxygen demand, while esmolol (0.2 g/hour intravenous infusion), a short-acting beta-blocker, was administered for additional

**Table 1** Laboratory Tests Results

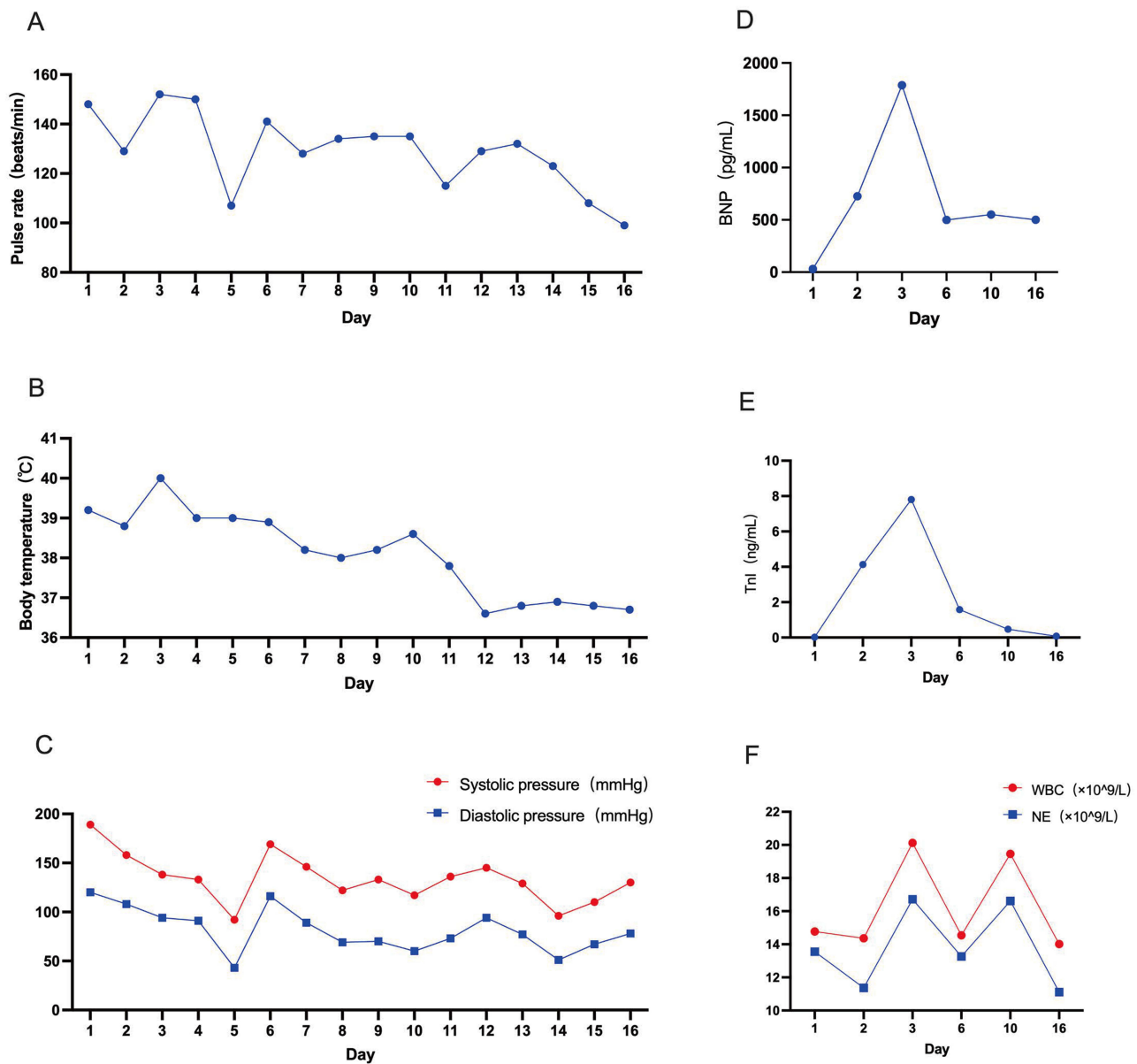
Laboratory Tests	Result		Reference Value
	Result in Admission	Result Before Discharge	
<b>Urine</b>			
Urinary free norepinephrine (nmol/24h)	>12,697.60		60.00–352
Urinary metanephrines (nmol/24h)	4078		< 216
Urinary normetanephrines (nmol/24h)	8312		< 312
Urinary vanillylmandelic acid (mg/24h)	58.1		≤ 12
Urinary free adrenaline (nmol/24h)	4368.89		4.31–61.60
Urinary dopamine (nmol/24h)	1802.74		750–2088

(Continued)

Table 1 (Continued).

Laboratory Tests	Result		Reference Value
	Result in Admission	Result Before Discharge	
<b>Blood</b>			
Cortisol (nmol/L)			
8:00	>1655.31	296.95	185–624
16:00	1095.63		< 276
24:00	961.98		< 50
ACTH (pg/mL)			
8:00	95.07	30.98	6–48
16:00	21.73		3–30
24:00	31.9		< 20
Dopamine (pmol/L)	524.5		≤ 195.7
Norepinephrine (pmol/L)	83975		414–4435.5
Epinephrine (pmol/L)	10579.3		≤ 605.4
TT3 (nmol/L)	0.94		1.01–2.48
TT4 (nmol/L)	141.67		69.97–152.52
FT3 (pmol/L)	4.28		3.28–6.47
FT4 (pmol/L)	8.35		7.64–16.03
TSH (mU/L)	0.469		0.56–5.91
Tn I (ng/mL)	0.023	0.081	0.02–0.06
BNP (pg/mL)	30.23	501.63	0–37.3
PCT (ng/mL)	0.683		0–0.05
ESR (mm/h)	39		0–34
CRP (mg/L)	3.08		0–10
IL-6 (pg/mL)	4.588		0–6.6
WBC ( $\times 10^9/L$ )	14.77	14.01	3.5–9.5
NE%	91.8	79.3	40–75
NE ( $\times 10^9/L$ )	13.55	11.11	1.8–6.3
FBG (mmol/L)	7.74		3.9–6.1
HbA1c (%)	6		4–6
AST (U/L)	22	70	13–35
ALT (U/L)	24	85	7–40
BUN	7.61	4.85	2.6–7.5
Cr	105	49	41–73
PTH (pg/mL)	83.4		12–88
Calcitonin (pg/mL)	19.21		0–18
K (mmol/L)	4.57	4.47	3.5–5.3
Na (mmol/L)	141.4	138.7	137–147
Ca (mmol/L)	2.76	2.26	2.11–2.52
<b>Venus blood gas analysis</b>			
pH	7.3		7.35–7.45
pCO <sub>2</sub> (mmHg)	39.1		35–45
pO <sub>2</sub> (mmHg)	123.8		80–100
Lac (mmol/L)	2.5		0.7–2.5
HCO <sub>3</sub> (mmol/L)	18.9		21–28

**Abbreviations:** ALT, alanine transaminase; AST, aspartate aminotransferase; BUN, blood urea nitrogen; BNP, brain natriuretic peptide; CRP, C-reactive protein; FBG, fibrinogen; Lac, lactic acid; NE, neutrophils; pO<sub>2</sub>, oxygen partial pressure; pCO<sub>2</sub>, partial pressure of carbon dioxide; pH, potential of hydrogen; Cr, serum creatinine; TP, total protein; TnI, troponin I; WBC, white blood cell; PTH, parathyroid Hormone; HbA1c, glycosylated hemoglobin; TSH, thyroid releasing hormone; FT4, free thyroxine; TT4, total thyroxine; FT3, free triiodothyronine; TT3, total triiodothyronine; ESR, erythrocyte sedimentation rate; PCT, procalcitonin; ESR, erythrocyte sedimentation rate; IL-6, interleukin-6.

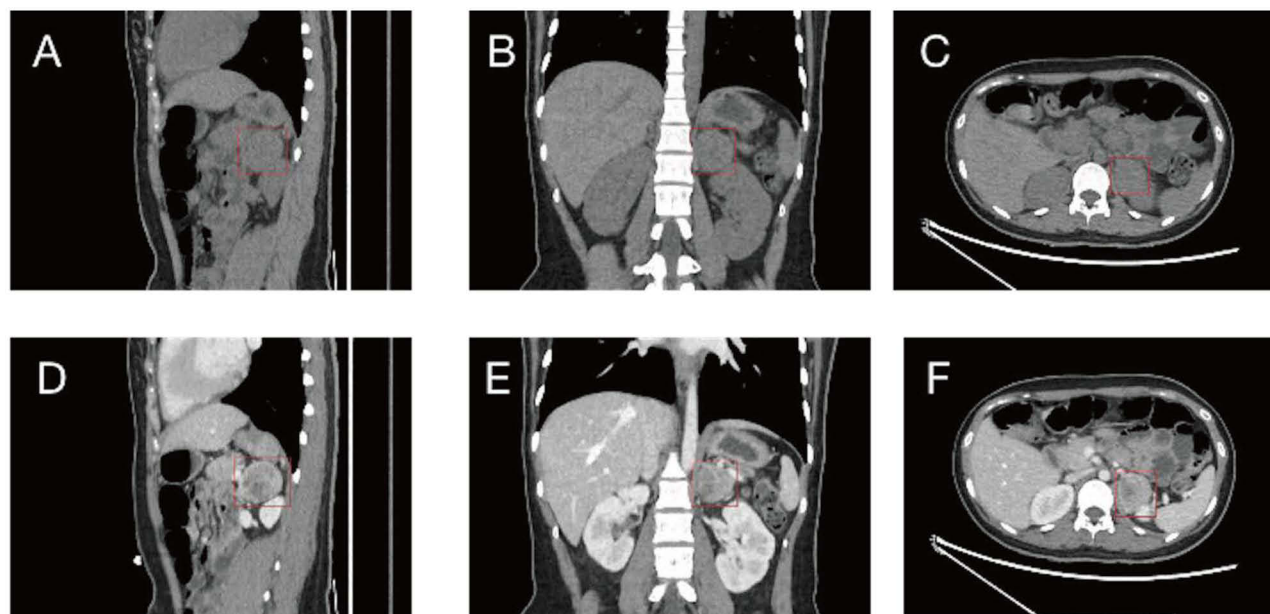


**Figure 1** The important laboratories throughout the entire treatment process. **(A)** Heart rate changes during hospitalization; **(B)** Body temperature changes during hospitalization; **(C)** Blood pressure changes during hospitalization, with the red line representing systolic blood pressure and the blue line representing diastolic blood pressure; **(D)** BNP level changes during hospitalization; **(E)** Troponin I level changes during hospitalization; **(F)** White blood cell and neutrophil level changes during hospitalization, with the red line representing white blood cells and the blue line representing neutrophils.

**Abbreviations:** NE, neutrophils; WBC, white blood cell; TnI, troponin I; BNP, brain natriuretic peptide.

acute heart rate control due to sinus tachycardia. Due to concerns about a potential infection, moxifloxacin was added as empiric antibiotic therapy.

Given the patient's presentation with an adrenal mass and hypertension, the endocrinologist recommended an evaluation of the aldosterone-to-renin ratio, plasma cortisol, plasma catecholamines, and 24-hour urinary catecholamines along with their metabolites. In the recumbent position, plasma and urinary catecholamine levels were markedly elevated (Table 1), including plasma dopamine at 524.5 pmol/L, norepinephrine at 83975 pmol/L, and epinephrine at 10579.3 pmol/L. Additionally, the 24-hour urinary levels showed free adrenaline at 4368.89 nmol/24 hours, free norepinephrine exceeding 12697.60 nmol/24 hours, normetanephrine at 8312 nmol/24 hours, metanephrines at 4078 nmol/24 hours, and vanillylmandelic acid at 58.1 mg/24 hours. These findings supported a clinical diagnosis of pheochromocytoma. On the



**Figure 2** Image showing a left adrenal tumor. (A–C) Unenhanced abdominal CT in transverse plane scan, sagittal plane scan and coronal plane scan which depicts a left adrenal gland nodule (red boxes, over 3 cm diameter). (D–F) Enhanced abdominal CT in transverse plane scan, sagittal plane scan and coronal plane scan which confirm the presence of a left adrenal mass.

fifth day post-admission, glucocorticoid therapy was discontinued, and perindopril amlodipine was substituted with terazosin for more targeted blood pressure management.

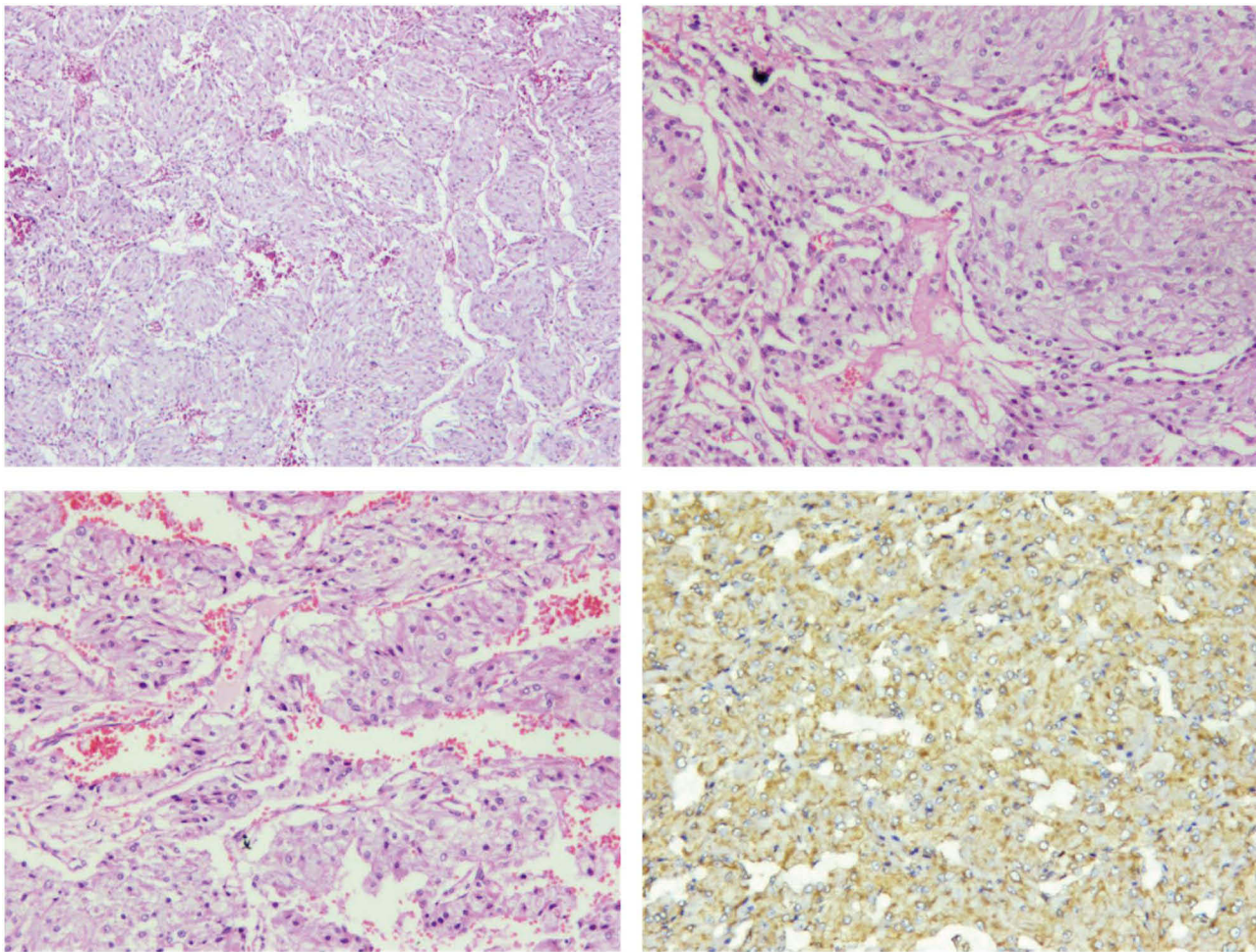
An enhanced abdominal CT scan (Figure 2) further confirmed a left adrenal mass, highly suggestive of pheochromocytoma. Additionally, after obtaining informed consent, whole-exome sequencing was performed, revealing a heterozygous missense mutation, c.1900T > C: p. Cys634Arg, in the RET gene, leading to a substitution of cysteine with arginine at codon 634. This mutation raised suspicion for multiple endocrine neoplasia syndrome, prompting further evaluation of the thyroid and parathyroid glands. Thyroid color Doppler ultrasound identified a hypoechoic mass measuring 6 mm × 4 mm in the left thyroid lobe, and a mild elevation in calcitonin levels was noted. No additional significant abnormalities were detected.

As the patient's condition gradually improved, plasma cortisol and ACTH levels returned to normal. The patient was subsequently discharged with a prescription for metoprolol tartrate (100 mg every 12 hours) and ivabradine hydrochloride (5 mg every 12 hours) for home management. Three months later, after achieving stable clinical status, the patient underwent resection of the left adrenal tumor, which measured 50 mm × 40 mm × 30 mm. Immunohistochemical analysis (Figure 3) confirmed positive staining for Vim, CD56, Syn, CgA, and NSE, with S-100 positive in Sertoli cells, while CKpan, CD10, MART-1/Melan-A, and Melan-A were negative. The Ki67 index was 1%, leading to a definitive diagnosis of adrenal pheochromocytoma. The patient was discharged without further medications and has since been regularly followed up postoperatively without recurrence of symptoms. Over a 15-month postoperative follow-up, the patient exhibited persistently mild hypercalcitoninemia with stable thyroid nodule size, while PTH and serum calcium levels showed a progressive increase (Table 2). Further parathyroid scintigraphy using 99mTc-MIBI was performed, and the conclusion was a negative result for parathyroid adenoma (Figure 4).

This study was approved by the Ethics Committee of Taizhou People's Hospital (Approval Number: 2024-096-01). Written informed consent was obtained from the patient for the publication of their clinical details and any accompanying images.

## Discussion

In the discussion of this case, the complexity of diagnosing pheochromocytoma, especially when it presents with non-specific symptoms and coexisting conditions, is highlighted. This patient initially presented with symptoms of headache, fever, and hypertensive crisis, combined with elevated BNP and TnI, which led to a preliminary diagnosis of myocarditis. Given the



**Figure 3** Tumor cells are diffusely distributed with focal necrosis. Vim (+), CD56 (+), CgA (+), Syn (+), NSE (+), S-100 (part +), CKpan (-), CD10 (-), Ki67 (1% +), MRRT/MenlanA (-), supporting the diagnosis of pheochromocytoma.

patient's background of prior hypertension and family history of sudden death, these findings initially supported a cardiovascular focus in her treatment plan. However, her persistent symptoms despite conventional management prompted further investigation, ultimately revealing an adrenal mass and confirming a diagnosis of pheochromocytoma.

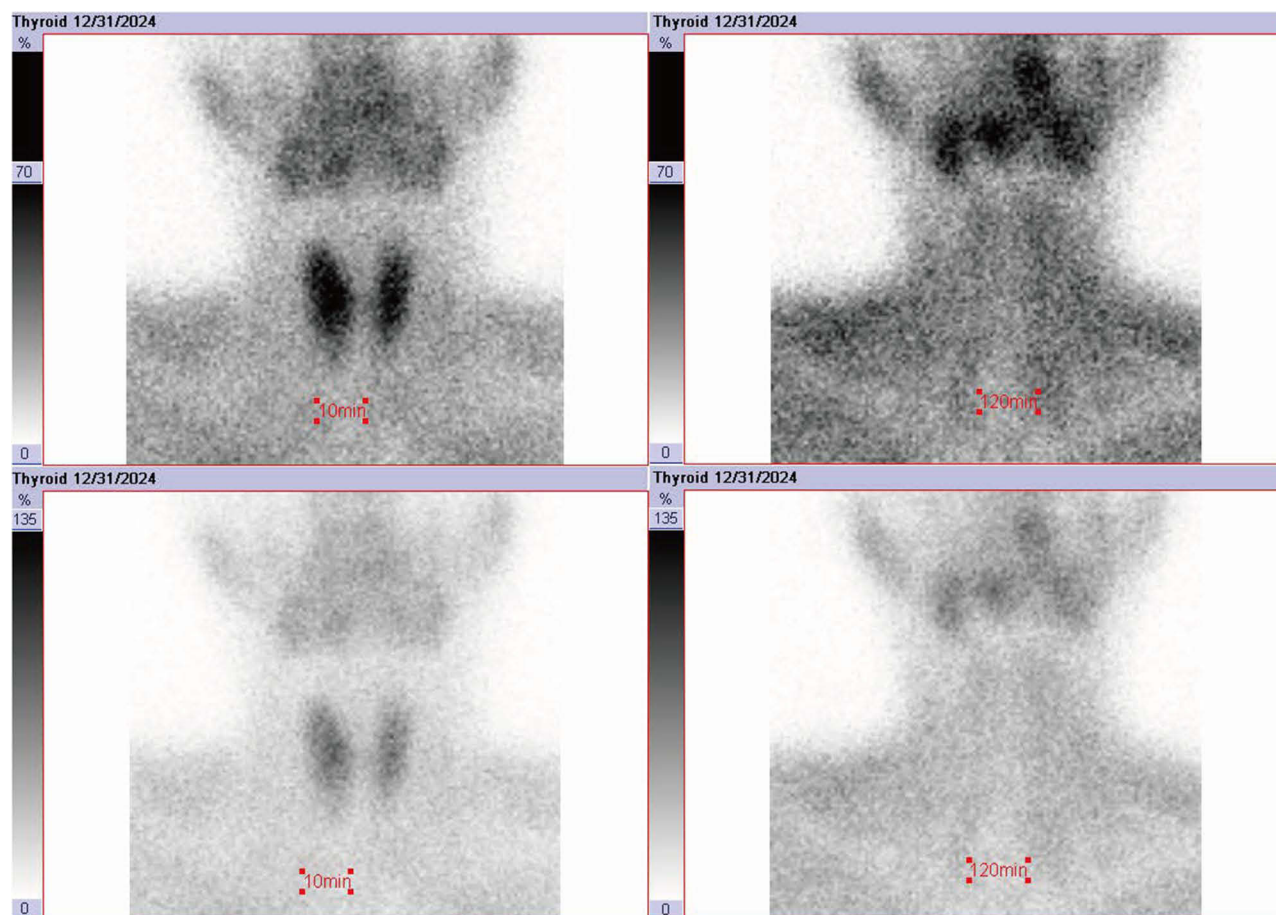
Pheochromocytoma is a rare catecholamine-secreting tumor, often challenging to diagnose due to its variable clinical presentation. Studies show that up to 50% of pheochromocytoma cases present with paroxysmal hypertension, while some cases, as highlighted in previous literature, may exhibit hypotension or normotension, depending on the catecholamine profile and the duration of catecholamine release.<sup>6,7</sup> In this case, the patient's hypertensive crisis, combined with sinus tachycardia, high fever, and elevated BNP and troponin I levels, underscored the adrenergic surge likely driven by excessive catecholamine release from the adrenal mass. Other cases have documented similar presentations where pheochromocytoma manifests as myocarditis or stress cardiomyopathy due to catecholamine toxicity, complicating initial diagnostic efforts.<sup>8</sup>

The role of glucocorticoid therapy in this case is particularly interesting. The patient received glucocorticoids for suspected myocarditis; however, glucocorticoids are known to potentially worsen catecholamine secretion in pheochromocytoma by stimulating adrenergic receptor sensitivity, posing a risk for crisis. Previous studies have shown that glucocorticoids can promote the synthesis and release of catecholamines. Steroids have been shown to stimulate the synthesis of catecholamines by inducing biosynthetic enzymes (phenylethanolamine-N-methyltransferase, tyrosine hydroxylase, and dopamine- $\beta$ -hydroxylase).<sup>9–11</sup> Previous study has shown that the release of catecholamines from perfused canine adrenal glands by corticosteroids.<sup>12</sup> Glucocorticoids have also been shown to increase catecholamine synthesis and storage in PC12 pheochromocytoma cell cultures.<sup>13</sup> A review highlighted the need for careful consideration

**Table 2** Postoperative Biochemical Trends Over 15 Months

	Urinary VMA (mg/24h, < 12)	Urinary Adrenaline	Urinary Norepinephrine	Urinary Dopamine	PTH (pg/mL, 12–88)	CT (pg/mL, 0–18)	Ca (mmol/L, 2.11–2.52)	P (mmol/L, 0.85–1.51)	Abdominal CT	Thyroid Ultrasound
14 days post-surgery	6				68.5	31.12	2.48	1.37	Post-left adrenal surgery; Dense lesion in the left adrenal gland.	Left lobe 6×4 mm, right lobe 2×1 mm.
3 months post-surgery	7.6	17.97 (nmol/24h, 4.31–61.6)	259.69 (nmol/24h, 60–352)	2691.86 (nmol/24h, 750–2088)	86.9	32.07	2.53	1.17	Post-left adrenal surgery.	Left lobe 6×4 mm, right lobe 2×1 mm.
9 months post-surgery	7.2				93.40	27.33	2.53	1.20	Post-left adrenal surgery.	Left lobe 6×4 mm, right lobe 2×1 mm.
15 months post-surgery	7.2	4.0 (ug/24h, 0–20)	53.7 (ug/24h, 0–90)	421.5 (ug/24h, 0–600)	112.8	32.67	2.56	1.33	Post-left adrenal surgery.	Left lobe 6×4 mm, right lobe 2×1 mm.

**Abbreviations:** CT, Computed Tomography; Ca, Calcium; PTH, Parathyroid Hormone; VMA, Vanillylmandelic Acid.



**Figure 4**  $^{99m}\text{Tc}$ -MIBI parathyroid scintigraphy showing no abnormal radiotracer retention.

of corticosteroid use in patients with undiagnosed adrenal masses to avoid exacerbating adrenergic symptoms.<sup>14</sup> In this case, glucocorticoids were discontinued upon suspicion of pheochromocytoma, after which alpha-blocker therapy with terazosin was initiated to provide more effective blood pressure control. This intervention underscores the importance of avoiding beta-blockers alone in pheochromocytoma due to the risk of unopposed alpha-receptor stimulation, which can precipitate a hypertensive crisis.

The discovery of a RET gene mutation (c.1900T > C: p. Cys634Arg) in this patient raises the possibility of MEN2A, which is characterized by pheochromocytoma, medullary thyroid carcinoma, and primary hyperparathyroidism. This mutation prompted a thorough evaluation of other endocrine organs, leading to the identification of a thyroid nodule and mildly elevated calcitonin levels. A study with an average follow-up of 7 years found higher penetrance of medullary thyroid carcinoma, pheochromocytoma and hyperparathyroidism in Cys634Arg carriers than in Cys634Tyr carriers.<sup>15</sup> And the Cys634Arg mutation was an independent factor for persistent/recurrent of MEN2A.<sup>15</sup> MEN2A is associated with specific mutations in the RET proto-oncogene, with Cys634Arg being a common variant linked to this syndrome, necessitating genetic counseling and close monitoring for associated endocrine tumors.<sup>2</sup> A meta-analysis demonstrated that in MEN2, the initial presentation was MTC in 60% of patients, synchronous MTC and pheochromocytoma (PHEO) in 34%, and isolated PHEO in only 6%.<sup>16</sup> Among patients with PHEO, 72% had bilateral tumors, with the majority (82%) being synchronous.<sup>16</sup> In contrast, our patient presented with unilateral PHEO, and it remains unclear whether this represents the initial manifestation of MEN2. Further follow-up is required to determine whether MTC or other MEN2-associated endocrine abnormalities will develop over time.

The patient presents with a thyroid nodule and mildly elevated calcitonin levels but has declined fine-needle aspiration for further evaluation. According to existing studies, serum calcitonin measurement is useful for detecting

medullary thyroid carcinoma (MTC);<sup>4</sup> however, given the rarity of MTC among patients with thyroid nodules, calcitonin testing may yield false-positive results, potentially leading to unnecessary surgical intervention.<sup>17</sup> Therefore, in cases of mildly elevated calcitonin levels without additional malignant features, close follow-up may be a reasonable approach. Additionally, the patient demonstrated a progressive increase in parathyroid hormone (PTH) and serum calcium levels, yet 99mTc-MIBI parathyroid scintigraphy was negative. This raises the suspicion of primary hyperparathyroidism (PHPT), which is commonly caused by a parathyroid adenoma, hyperplasia, or, less frequently, parathyroid carcinoma.<sup>18</sup> The negative scintigraphy findings could be attributed to several factors, including small lesion size, parathyroid hyperplasia, ectopic parathyroid glands, and thyroid pathology.<sup>19</sup> Despite the negative nuclear imaging findings, PHPT remains a differential diagnosis based on the clinical and laboratory findings. Given the patient's test results and personal preference, continued follow-up is recommended to monitor for potential disease progression.

A literature review reported that most cases of glucocorticoid-induced pheochromocytoma crisis involved tumors  $\geq 30$  mm in diameter and glucocorticoid doses equivalent to  $\geq 60$  mg/day of hydrocortisone.<sup>20</sup> In our case, the tumor was 50 mm  $\times$  40 mm  $\times$  30 mm, and the glucocorticoid dosage was equivalent to 1250 mg/day of hydrocortisone, far exceeding this threshold. Despite these risk factors, our patient did not experience a pheochromocytoma crisis, which may be attributed to the influence of the MEN2A-associated RET mutation and individual variability in response to glucocorticoids. Studies suggest that MEN2A-associated pheochromocytomas may differ in their biochemical and clinical behavior from sporadic cases, potentially reducing the risk of acute catecholamine surges in response to glucocorticoids.<sup>21</sup> Additionally, there is significant individual variation in glucocorticoid sensitivity in pheochromocytoma patients; some individuals are more susceptible to catecholamine release, while others, like our patient, may show a lower risk of crisis even with high glucocorticoid doses.<sup>22</sup>

## Conclusion

This case exemplifies the diagnostic complexity of pheochromocytoma, particularly when it mimics cardiac or infectious conditions. It underscores the importance of comprehensive endocrine evaluation in patients with refractory hypertension and adrenal masses, as well as the critical role of genetic testing in patients with suspected hereditary syndromes. This case contributes to the body of literature that highlights the diverse presentations of pheochromocytoma and emphasizes best practices for management and follow-up, especially in cases associated with genetic mutations such as RET.

## Data Sharing Statement

All Data and material collected during this study are available from the corresponding author upon reasonable request.

## Ethics Approval and Consent to Participate

The Ethics Committee of Clinical Research, Taizhou People's Hospital approved this study and the publication of case details. In addition, we confirm that all processes were executed in accordance with all applicable rules and guidelines, and informed consent was obtained from all subjects.

## Consent for Publication

The written informed consent was obtained from subjects for publication of case details and images.

## Author Contributions

Qingqing Zhang and Xue Wei are equal contributors and co-first authors. 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 that they have no competing interests.

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