MEDDOCS Open Access Publisher

ISSN: 2639-9237

Journal of Case Reports and Medical Images

Open Access | Case Report

Pheochromocytoma in a Young Female with Episodic Hypertension and Paroxysmal Symptoms - A Case Report

Prabin Duwadee[1]; Reechashree Dhungana[2]; Ashok Chhatkuli[3]; Abinash Baniya[4]; Abhash Kumar Jha[5]; Pratick Shrestha[6]; Prabin Kumar Bam[7]; Suraj Sharma[8]

[1]Chitwan Medical College and Teaching Hospital (CMCTH), Nepal.

[2]Institute of Medicine (IOM), Nepal.

[3]Universal College of Medical Sciences and Teaching Hospital (UCMS), Nepal.

[4]Chitwan Medical College and Teaching Hospital (CMCTH), Nepal.

[5]Chitwan Medical College and Teaching Hospital (CMCTH), Nepal.

[6]Vayodha Hospital, Nepal.

[7]Chitwan Medical College and Teaching Hospital (CMCTH), Nepal.

[8]National Academy of Medical Sciences (NAMS), Nepal.

***Corresponding Author(s): Prabin Duwadee**

Chitwan Medical College and Teaching Hospital (CMCTH), Nepal.

Email: dr.duwadeeprabin@gmail.com

Received: Apr 09, 2025

Accepted: Apr 29, 2025

Published Online: May 06, 2025

Journal: Journal of Case Reports and Medical Images

Publisher: MedDocs Publishers LLC

Online edition: http://meddocsonline.org/

Keywords: Pheochromocytoma; Hypertension; Paroxysmal symptoms; Young woman.

Abstract

Background: Pheochromocytoma is a rare, catecholamine-secreting tumor, typically presenting with episodic hypertension, headaches, diaphoresis, and palpitations. Early diagnosis and prompt management can lead to favorable prognosis, though challenging, particularly in younger individuals.

Case Presentation: A 28-year-old female with a history of episodic hypertension, headache, diaphoresis, and palpitations was diagnosed with pheochromocytoma. Elevated urinary metanephrines and imaging revealed a right adrenal mass. The patient underwent preoperative α- and β-blockade followed by laparoscopic adrenalectomy. Histopathology confirmed pheochromocytoma, and post-surgery, she remained symptom-free with normal biochemical markers.

Conclusion: This case emphasizes the need to consider pheochromocytoma in young patients presenting with typical symptoms of episodic hypertension. Timely diagnosis, preoperative management, and surgery lead to favorable outcomes. Although rare, pheochromocytoma should be on the differential diagnosis list for patients presenting with unexplained hypertension and related symptoms.

Introduction

Pheochromocytoma is a rare tumor that is derived from chromaffin cells situated within the adrenal medulla or extra-adrenal paraganglia. It is characterized by the excessive secretion of catecholamines, leading to a wide range of clinical manifestations [1]. Pheochromocytoma classically presents with a triad of paroxysmal headache, diaphoresis, and palpitations along with hypertension. The triad is present in around 40-80%

Cite this article: Prabin D, Reechashree D, Ashok C, Abinash B, Abhash KJ, et al. Pheochromocytoma in a Young Female with Episodic Hypertension and Paroxysmal Symptoms - A Case Report. J Case Rep Clin Images. 2025; 8(1): 1162.

of patients with pheochromocytoma and is highly sensitive and specific for the presumptive diagnosis of the disease [2]. Hypertension is the most common manifestation of pheochromocytoma and can manifest as sustained or paroxysmal episodes of elevated blood pressure. Pheochromocytoma equally affects both genders and its incidence are commonly observed during the fourth and fifth decades of life. Histologically, pheochromocytoma is derived from chromaffin cells which arise from neural crest cells; therefore, it is a neuroendocrine tumor [3]. Early diagnosis and management of pheochromocytoma is crucial to help prevent serious complications like cardiomyopathy and hypertensive crisis [4]. In this article, we present a case of pheochromocytoma observed in our center, highlighting the typical clinical features and course of treatment in a 28-year-old female diagnosed with this disease.

Case presentation

A 28-year-old female presented to the endocrinology clinic with a history of episodic hypertension (maximum documented BP was 210/120 mm of Hg), palpitation, headache (mostly left temporal), diaphoresis, and shortness of breath for one year. The symptoms were sometimes accompanied by vomiting, photophobia, and dizziness. She used to experience such episodes two to three times a week with each episode lasting about 10 to 15 minutes. The frequency had increased over the last two months occurring almost four to five times a week and sometimes two to three episodes in a single day which had incapacitated her daily activities. At the time of personation, laboratory studies showed Hemoglobin of 12.6 gm/dL, Total leucocytes of 9,500/mm3 (Neutrophil 75%, Lymphocytes 20%, Monocytes 3%, Eosinophils 2%), Fasting blood sugar of 108 mg/ dL, Urea of 37 mg/dL, Creatinine of 1.1 mg/dL, sodium of 134 meq/dL, potassium of 5.2 meq/dL, Calcium of 8.1 mg/dL, Phosphorus of 3.4 mg/dL, Total protein of 7.6 gm/L , albumin of 3.4 gm/L. ECG showed normal sinus rhythm at the time of presentation.

CECT of the abdomen and pelvis was done which revealed a well-defined mass (40 HU) measuring 62×50×59 mm arising from the right adrenal gland showing moderate heterogeneous contrast enhancement and poor contrast washout in 15 min delayed images as shown in figure 2. The features suggested possible pheochromocytoma. To confirm the diagnosis, a 24-hour urine sample was collected for evaluation of fractionated metanephrines (5309.28 mcg/24 hrs.) and nor-metanephrines (5565.60 mcg/24 hrs.). The results indicated elevated values consistent with secretory pheochromocytoma.

The patient was planned for surgery. She was initially managed with adequate salt loading (5 to 10 gm per day) and adequate fluids (3 to 5 liters per day). To achieve α and β adrenergic blockade, a 10-day course of Tab. Prazosin 5 mg PO QID and Tab. Metoprolol PO BD 50 mg after lunch and 25 mg after dinner, respectively, were prescribed and were continued till the day of surgery. After 10 days of pre-operative management, a laparoscopic right adrenalectomy was performed. The right adrenal mass, measuring 5×5 cm, was excised. Histopathological analysis of the removed mass revealed a well-circumscribed lesion with fibrous pseudocapsule showing proliferation of cells in Zellballen pattern, trabeculae and diffuse sheets as shown in figure 1. The individual cells were large and polygonal, exhibiting mild pleomorphism. They had round to oval nuclei with high nucleocytoplasmic ratio, irregular nuclear membrane, vesicular to coarse chromatin, prominent nucleoli, and abundant amount of finely granular cytoplasm. These histopathological features confirmed the diagnosis of pheochromocytoma. The post-operative period was uneventful and the patient was discharged on the sixth post-operative day being hemodynamically stable with blood pressure within normal range. The patient was followed up after two months of surgery. During the follow up, ambulatory blood pressure monitoring was done, which was normal. Additionally, the patient had no further complaints of headache, diaphoresis, and dizziness. After 6 months of surgery, 24 hour-urine metanephrine and nor-metanephrine levels were within normal range suggesting the absence of tumor elsewhere in the body as shown in Table 1. The patient was advised for a regular follow-up every three months to evaluate long term prognosis.

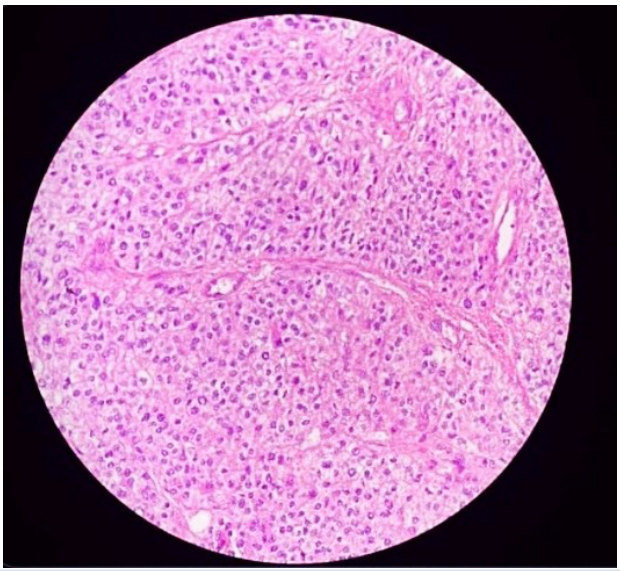

Figure 1: Histological appearance of pheochromocytoma using hematoxylin and eosin stain showing proliferation of cells in Zellballen pattern, trabeculae and diffuse sheets.

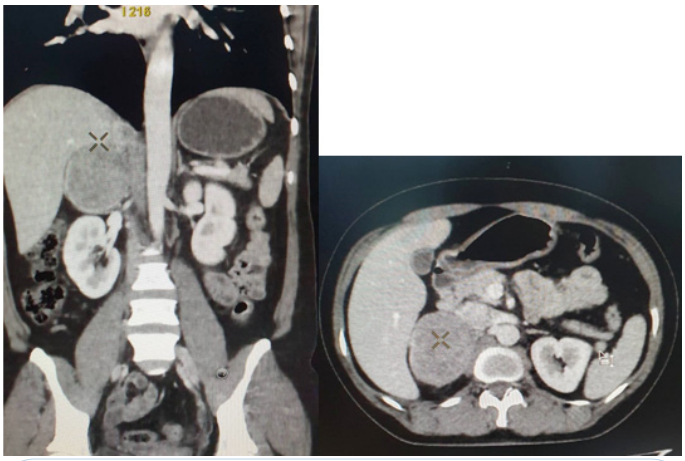

Figure 2: CECT abdomen and pelvis axial section and coronal section show heterogeneously enhancing mass lesion arising from the right adrenal gland at the suprarenal location.

Table 1: Showing normal values of urinary metanephrines and normetanephrines over 24 hours and values of urinary metanephrines and normetanephrines over 24 hours after 6 months of surgery.

Urinary levels of metanephrines and normetanephrines in 24 hours	Metanephrines	Normetanephrines
Normal range	24-96 mcg/24h	75-375 mcg/24h
Initial values	5309.28 mcg/24h	5565.60 mcg/24h
Values after 6 months surgery	29.27 mcg/24h	227.58 mcg/24h

Discussion

This case highlights the classical presentation of pheochromocytoma in a young female, with episodic hypertension, paroxysmal symptoms, and markedly elevated urinary metanephrines. Imaging revealed a right adrenal mass, and the subsequent histopathology confirmed the diagnosis of pheochromocytoma. Pheochromocytoma is typically diagnosed in fourth to fifth decade of life, though it can be diagnosed at any time from childhood to adulthood [1]. Our patient is a 28-year-old female, depicting an instance of its occurrence in a younger female. It is a rare catecholamine-secreting tumor, derived from chromaffin cells situated within the adrenal medulla or extra-adrenal paraganglia, also known as chromaffinoma. Pheochromocytoma is a rare cause of hypertension, accounting for approximately 0.1 to 1% of all cases of hypertension [1,2]. Pheochromocytoma is clinically manifested with signs and symptoms like paroxysmal hypertension, headache, diaphoresis, palpitations, pallor, dyspnea, and anxiety attacks from excessive catecholamines (i.e., mainly norepinephrine and epinephrine) secreted either intermittently or continuously by chromaffin cells of the tumor [2,3]. The clinical features observed in patients are influenced by the type and quantity of catecholamines released by the tumor which can produce epinephrine (E), Norepinephrine (NE), or dopamine continuously (resulting in persistent hypertension) or intermittently (causing paroxysms of symptoms) [3].

The effects of catecholamines on different organs may differ depending on their blood concentration and types of adrenergic receptors in the organs which may lead to highly variable clinical presentations and thereby possible delay in diagnosis [4]. Pheochromocytoma typically presents with a triad of paroxysmal headache, diaphoresis, and palpitations that occur with a frequency ranging from monthly to several times a day and usually lasts for a few seconds to a few hours and in approximately 90% of cases, the patient presents with sustained or paroxysmal hypertension [3]. The paroxysmal attacks are generally precipitated by exertion, trauma, stress, induction of anesthesia, drugs (metoclopramide, steroids, TCA's), and contrast dye [5]. The classic triad along with hypertension has a 90% sensitivity and 93% specificity in diagnosing pheochromocytoma [6]. Although hypertension is a common finding, Hao-Yu Wu et al. [7] reports a case of pheochromocytoma presenting with hypotension. The reason for hypotension and sometimes even shock, is due to factors that include tumor necrosis leading to a sudden decline in continuous catecholamine secretion, adrenergic receptor desensitization, and a decrease in vascular volume [8].

The conventional understanding that 10% of pheochromocytomas are malignant, 10% are bilateral, 10% are extra-adrenal (with 10% of those being extra-abdominal), 10% occur in non-hypertensive patients, and 10% are hereditary no longer accurately reflects the nature of these tumors. With growing recognition of germline mutations, it is now understood that up to 40% of pheochromocytoma and paraganglioma cases can be attributed to these genetic alterations [9].

The laboratory diagnosis of pheochromocytoma can be made by measuring catecholamines and their metabolites in the blood and urine. As secretion of catecholamines from pheochromocytoma is episodic, single sample estimation of catecholamines and their metabolites is not helpful for the diagnosis of the disease. Therefore, 24-hour urinary catecholamines and urinary metanephrine (i.e. catecholamines metabolite) levels are generally estimated for the diagnosis of the disease and elevated 24-hour urinary catecholamines and urinary metanephrine has a sensitivity of 97% and specificity of 98% for diagnosing pheochromocytoma [5]. Increased plasma fractionated metanephrine level has a sensitivity of 98% and specificity of 92% for diagnosing pheochromocytoma and hence is the gold standard method. If available, free plasma methoxytyramine can be measured to detect a rare dopamine-producing tumor [10]. Imaging modalities such as MRI and CT scan are employed once laboratory findings suggest pheochromocytoma for anatomical localization of the tumor. MRI and CT scans have around 95% sensitivity and 70% specificity for localizing adrenal pheochromocytoma [5]. Functional imaging techniques like 18F-fluorodeoxyglucose positron emission tomography (^{18}F-FDG PET), iodine-123 meta-iodobenzylguanadine (^{123}I-MIBG), 18F-flurodihydroxyphenylalanine (^{18}F-FDOPA), 68Ga-DOTA coupled somatostatin analogs (^{68}Ga-DOTA) are employed for tumor characterization, confirmation of metastasis and treatment planning of the pheochromocytoma and are usually done after anatomic localization by CT or MRI scan. Functional imaging is also employed if laboratory findings suggest pheochromocytoma but no definite abnormality is seen on CT or MRI scans [11].

Additional genetic testing is also recommended for the diagnosis of pheochromocytoma as various familial genetic disorders such as Multiple Endocrine Neoplasia type 2a (MEN 2a), Multiple Endocrine Neoplasia type 2b (MEN 2b), Von Hippel Lindau syndrome (VHL), Meurofibromatosis type 1 (NF 1), hereditary paraganglioma syndrome are associated with pheochromocytoma [12]. Hereditary paraganglioma syndrome is an autosomal dominant disorder characterized by multiple paragangliomas and pheochromocytomas due to mutation in mitochondrial enzyme succinate dehydrogenase that occurs with a frequency of 1/300000 [13]. About 10% of pheochromocytomas are malignant and have characteristic features like large tumor mass, local invasion of neighboring tissues or organs, and distant metastases on nuclear imaging. In CT scan tumor size of diameters ≥4 cm, ≥6 cm, and ≥8 cm has a chance of 20%, 65%, and 89% malignancy respectively [14].

Pheochromocytomas can be effectively treated and cured if diagnosed early. However, if the condition goes undetected, the excessive release of catecholamines can lead to severe and potentially life-threatening consequences. Over time, patients with pheochromocytoma may develop a dangerous form of dilated cardiomyopathy, which can be fatal. Chronic cardiomyopathy appears to be linked to consistently elevated levels of catecholamines in the bloodstream, as it often resolves rapidly following surgical removal of the tumor [15].

Surgical resection of the tumor is the mainstay of treatment. Given the accuracy of imaging methods for accurately localizing the tumor, the ability to conduct extensive visual exploration under laparoscopy, and the ease of intraoperative conversion to open surgery, the laparoscopic approach can be considered for the treatment of most pheochromocytomas, including bilateral tumors [16]. However, size, location and tumor characteristics help to determine the need of open surgery [17].

Adequate fluid and salt intake, along with alpha-blockers, are preoperative regimens recommended to patients undergoing surgical resection to reduce perioperative morbidity and mortality [10]. Selective alpha-1 blockers (prazosin, terazosin) or nonselective alpha-blockers (phenoxybenzamine) are the commonly used alpha-blockers. Beta-blockers should be given only after the complete alpha blockade to control tachycardia and maintain a pulse rate of 60-80 beats/min. complete alpha blockade can be clinically assessed by postural hypotension.

Beta-blockers should not be given until the patient is on alpha blockade because unopposed alpha-adrenergic receptor stimulation can precipitate a hypertensive crisis [5]. Generally, non-selective beta-blockers (Propranolol) or selective beta-blockers (Atenolol, Metoprolol) are used. Beta blockade after the complete alpha blockade is helpful in maintaining blood pressure in the pre-operative period [10]. Pre-operative fluid and salt intake are recommended to maintain hemodynamic instability during tumor manipulation [5,10]. Surgeons and anesthesiologists should pay extra attention during intraoperative period as the patient is prone to develop hypertensive crisis, hypotension, and arrhythmias due to the manipulation of the tumor and effects of anesthetic agents [5]. Hypertensive crisis during intraoperative period is treated with IV sodium nitroprusside or IV phentolamine. Laparoscopic adrenalectomy is the preferred method for surgical resection of tumors. Hypotension is a common problem during post-operative period which is treated with rapid infusion of IV fluids [5,10]. Due to the possibility of recurrence, long-term follow-up is required in all patients with pheochromocytoma. After surgery, plasma and/or urine metanephrines should be assessed after 2-6 weeks [18]. The patient was preoperatively managed with α- and β-adrenergic blockade, which stabilized her hemodynamics and ensured a smooth laparoscopic adrenalectomy. Post-surgery, the patient remained asymptomatic, with normalized biochemical markers, demonstrating the effectiveness of early diagnosis and appropriate surgical intervention in the management of pheochromocytoma.

Conclusion

This case emphasizes the importance of considering pheochromocytoma in the differential diagnosis of young patients presenting with episodic hypertension and associated symptoms. Early recognition through biochemical testing and imaging, followed by appropriate preoperative management and surgical intervention, ensures favorable outcomes. Long-term follow-up is recommended to monitor for recurrence or metastasis. Additionally, clinicians should be aware of potential mimics of pheochromocytoma to avoid misdiagnosis and to tailor management accordingly. This case underscores the necessity of a multidisciplinary approach in the management of pheochromocytoma, ensuring that both diagnostic and therapeutic strategies are optimally utilized.

Highlights

1. Pheochromocytoma is a rare catecholamine secreting tumor arising from chromaffin cells within the adrenal medulla or extra-adrenal paraganglia.
2. The patient usually presents with a triad of episodic headache, palpitation, diaphoresis and other associated symptoms.
3. CECT of abdomen revealed a well-defined mass arising from the right adrenal gland which was supplemented with a 24-hour urine sample that showed elevated levels of fractionated metanephrines and normetanephrines.
4. The patient underwent laparoscopic adrenalectomy after adequate preoperative alpha blockade followed by beta blockade resulting in resolution of symptoms.
5. Early diagnosis and management of pheochromcytoma is crucial to prevent life threatening cardiovascular complications.

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