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When Should Pheochromocytoma Be Suspected In Pediatric Patients? - A Case Report

José Antonio Coba Lacle
Pediatrician
Subspecialist in Rare Diseases, Hospital Pequeno Príncipe (HPP)
Master's Degree in Genetics, Universidad Simón Bolívar, Colombia
PhD Candidate in Health Sciences, Pontifical Catholic University of Paraná (PUC-PR)
Professor, Universidad Alexander von Humboldt, Armenia, Colombia
Lattes: https://lattes.cnpq.br/8374765335390438
ORCID: 0000-0002-3955-6183

Eirini Valakitsis Giraldo
Physician
Corporación Universitaria Empresarial Alexander von Humboldt, Colombia
ORCID: 0009-0008-6042-2228

Melisa Castaño Renteria
Physician
Unidad Central del Valle, Colombia
ORCID: 0009-0006-5394-5528

Jorge Luis Paez Corral
Medical Intern
Universidad Libre, Cali, Colombia
ORCID: 0009-0007-6900-0948

Jhon Alejandro Holguín López
Physician
Unidad Central del Valle, Colombia
ORCID: 0009-0009-7624-8049

Laura Andrea Suaza Cruz
Physician
Unidad Central del Valle del Cauca, Colombia
ORCID: 0009-0000-9778-4144

Helmer Andres Zuñiga Amu
Physician
Unidad Central del Valle del Cauca, Colombia
ORCID: 0009-0001-1244-2856

Cristian Gerardo Rendon Sánchez
Physician
Unidad Central del Valle del Cauca, Colombia
ORCID: 0009-0003-4663-9272

Jhonier Adrian Ussa Muñoz
Medical Intern
Unidad Central del Valle del Cauca, Colombia

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Abstract: Introduction: Pheochromocytoma is a rare neuroendocrine neoplasm in the pediatric population, arising from chromaffin cells of the adrenal medulla and potentially associated with excessive catecholamine secretion. Although hypertension is among its most common manifestations, presentations involving hypertensive emergencies and simultaneous damage to multiple target organs pose substantial diagnostic and therapeutic challenges.

Case Presentation: A 15-year-old adolescent girl with no previous diagnosis of hypertension presented with headache, abdominal pain, vomiting, and decreased visual acuity, progressing to a blood pressure of 261/196 mmHg. She was admitted to the pediatric intensive care unit and required intravenous sodium nitroprusside and the progressive addition of antihypertensive agents. Evaluation revealed severe hypertensive retinopathy with optic disc edema, retinal hemorrhages, and visual acuity of 20/400 in the right eye. Echocardiography demonstrated marked concentric left ventricular hypertrophy, intraventricular obstruction, and hypercontractility. Abdominal computed tomography angiography identified a 23-mm right adrenal nodule with heterogeneous arterial enhancement. Elevated plasma norepinephrine and epinephrine levels were documented, along with total urinary metanephrines of 1,809.70 µg/24 hours, approximately 3.5 times the reported upper reference limit. Following clinical stabilization and pharmacologic preparation, the patient underwent right retroperitoneoscopic adrenalectomy on March 30, 2023, without documented surgical complications. Histopathologic examination confirmed pheochromocytoma, demonstrating a characteristic *zellballen* architecture, no capsular or vascular invasion in the examined tissue sections, and a Pheochromocytoma of the Adrenal Gland Scaled Score (PASS) below 5. She was discharged in clinically stable condition.

Discussion: This case illustrates that a small adrenal lesion may be associated with severe systemic manifestations resulting from catecholamine excess. The coexistence of hypertensive retinopathy, substantial cardiac remodeling, proteinuria, and neurologic manifestations highlights the importance of promptly investigating secondary hypertension in adolescents.

Conclusion: Pheochromocytoma should be considered in the differential diagnosis of pediatric hypertensive emergencies, particularly when accompanied by adrenergic symptoms and target-organ damage. Accurate diagnosis, appropriate pharmacologic preparation, and surgical treatment are essential, as are genetic evaluation and long-term follow-up.

Keywords: Pheochromocytoma; Hypertensive Emergencies; Pediatric Hypertension; Hypertensive Retinopathy; Left Ventricular Hypertrophy; Adrenalectomy.

Introduction

Pheochromocytoma is a rare neuroendocrine neoplasm arising from chromaffin cells of the adrenal medulla and characterized by variable catecholamine production. Similar tumors arising from extra-adrenal paraganglionic structures are classified as paragangliomas. Together, these neoplasms exhibit heterogeneous clinical manifestations related to both their secretory activity and their anatomic and biologic characteristics (1,2).

In the pediatric population, pheochromocytomas and paragangliomas have an estimated annual incidence of 0.5 to 2 cases per million children. Despite their rarity, they are particularly important because of their potential to cause severe hypertension, arrhythmias, cardiovascular complications, neurologic abnormalities, and other potentially life-threatening conditions (1).

The classic presentation includes headache, diaphoresis, and palpitations, often accompanied by sustained or paroxysmal hypertension. However, children and adolescents may present with nonspecific symptoms, including abdominal pain, nausea, vomiting, weight loss, visual disturbances, and behavioral or autonomic manifestations that may delay recognition of the disease (1,2).

Secondary hypertension is an important concern in pediatric practice, particularly in the presence of markedly elevated blood pressure, abrupt symptom onset, and evidence of target-organ damage. Under these circumstances, the diagnostic evaluation should include renal, renovascular, cardiovascular, and endocrine causes, including catecholamine-secreting tumors (2,3).

Excessive catecholamine activity may produce intense systemic vasoconstriction, increased peripheral vascular resistance, and hemodynamic overload. When sustained or particularly severe, these effects may lead to cardiac remodeling, myocardial ischemia, hypertensive retinopathy, and impaired cerebral and renal perfusion.

Identification of an adrenal mass in a patient with severe hypertension and systemic manifestations warrants targeted biochemical investigation. Plasma free metanephrines and fractionated urinary metanephrines are recommended diagnostic tests, and their results should be interpreted according to reference ranges, collection conditions, and potential interfering factors (1,2).

The treatment of choice for localized, resectable pheochromocytomas is surgical removal, preceded by clinical preparation aimed at reducing the cardiovascular effects of catecholamine excess. Alpha-adrenergic

blockade is a fundamental component of this preparation, whereas beta-adrenergic blockade may subsequently be added when indicated (1,2).

Another distinguishing feature of pediatric pheochromocytomas is their strong association with hereditary predisposition. International recommendations published in 2024 indicate that approximately 70% to 80% of pheochromocytomas and paragangliomas diagnosed during childhood and adolescence have a hereditary component, emphasizing the need for genetic evaluation and long-term surveillance (1).

We report the case of an adolescent girl with a 23-mm right adrenal pheochromocytoma who initially presented with an extreme hypertensive emergency accompanied by cardiovascular, retinal, and renal abnormalities. The objective is to discuss the importance of early recognition of this etiology in pediatric patients, the therapeutic challenges encountered during clinical stabilization, and the need for continued follow-up after tumor resection.

Case Presentation

A 15-year-old adolescent girl weighing 40 kg, with no previous diagnosis of hypertension or other known chronic diseases, was referred to a tertiary university hospital on February 15, 2023, after presenting with severe hypertension, headache, abdominal pain, vomiting, and visual disturbances.

According to her mother, the patient had experienced headache and abdominal discomfort since approximately February 11. On February 13, she developed episodes of abdominal pain accompanied by vomiting, prompting evaluation at an urgent care facility, where she received symptomatic treatment with dipyrone and was discharged.

The following day, she attended school without significant symptoms during the morning. However, after returning home, she remained asleep for much of the afternoon and awoke that evening with severe abdominal pain and recurrent vomiting.

Because of persistent symptoms, she returned to the emergency department, where her blood pressure was 261/196 mmHg. Intravenous sodium nitroprusside was initiated, resulting in a partial reduction in blood pressure to approximately 225/125 mmHg. Given the severity of hypertension and the insufficient therapeutic response, she was transferred to a specialized hospital for further management.

During the initial evaluation, the adolescent also reported persistent headache and a visual disturbance affecting the right eye, described as blurred vision accompanied by a grayish visual perception. She reported recent ibuprofen use for pain relief.

Her mother denied fever, diarrhea, or urinary symptoms. There was no history of previous hospitalization, surgical procedures, known medication allergies, or chronic medication use before the current episode.

The pregnancy, birth, and neonatal history were described as unremarkable. Regarding family history, the paternal grandfather had hypertension and diabetes mellitus and had died suddenly at 50 years of age. Other family history was incompletely characterized, with no confirmed hereditary syndrome.

Upon admission to the referral hospital, intravenous hydralazine was administered without a satisfactory blood pressure response. Given the severity of hypertension and manifestations suggestive of target-organ damage, admission to the pediatric intensive care unit (PICU) was indicated, and sodium nitroprusside was continued at an initially documented dose of 1 µg/kg/min.

Initial Evaluation and Target-Organ Involvement

Laboratory testing performed on February 15 revealed a serum sodium concentration of 135 mEq/L, potassium of 3.03 mEq/L, blood urea of 26.8 mg/dL, and creatinine of 0.88 mg/dL. Arterial blood gas analysis showed a pH of 7.53, a partial pressure of carbon dioxide of 27 mmHg, and a bicarbonate concentration of 22.6 mmol/L, consistent with predominantly respiratory alkalemia at that time.

Urinalysis demonstrated 3+ proteinuria, 2+ ketonuria, and a positive reaction for blood, accompanied by mildly elevated leukocyte and erythrocyte counts in the urinary sediment. Urine culture showed no bacterial growth.

Assessment of urinary albumin excretion revealed an albumin-to-creatinine ratio of approximately 1,232 mg/g, suggesting marked albuminuria in the setting of severe hypertension. Insufficient data were available to determine whether this abnormality persisted after blood pressure control.

Renal ultrasonography and renal artery Doppler ultrasonography, performed on February 15, showed no structural renal abnormalities or hemodynamically significant stenoses. Both kidneys had preserved dimensions and morphologic characteristics.

The vascular evaluation was expanded to include Doppler ultrasonography of the carotid and vertebral arteries and the arterial systems of the upper and lower extremities. No hemodynamically significant abnormalities or ultrasonographic findings suggestive of vascular inflammation were identified.

Ophthalmologic examination revealed significant funduscopic abnormalities, including optic disc edema, retinal hemorrhages and microhemorrhages, vascular engorgement, and perimacular exudates, consistent with severe hypertensive retinopathy.

During a subsequent ophthalmologic assessment, uncorrected visual acuity was documented as 20/400 in the right eye and 20/30 in the left eye. Intraocular pressure was 12 mmHg bilaterally. Funduscopic examination demonstrated persistent optic disc edema, retinal hemorrhages, and macular abnormalities, prompting referral for specialized retinal follow-up.

Echocardiography performed on February 16 revealed marked concentric left ventricular hypertrophy, with an interventricular septal thickness of 13 mm and a posterior wall thickness of 12 mm. The examination described a reduced left ventricular cavity, marked papillary muscle hypertrophy, and an intracavitary systolic gradient between the ventricular apex and the left ventricular outflow tract.

Left ventricular hypercontractility was identified, with an estimated ejection fraction of 84%, along with moderate pulmonary infundibular stenosis and a reported systolic gradient of 60 mmHg.

The echocardiographic report raised the possibility of hypertrophic cardiomyopathy without establishing a definitive etiologic diagnosis. Given the extreme hypertension, pressure overload and catecholamine-mediated effects were considered relevant to the interpretation of these abnormalities, although the available data did not allow the relative contribution of each mechanism to be determined.

Chest radiography showed no significant pulmonary abnormalities or enlargement of the cardiac silhouette.

Head computed tomography (CT), performed on February 15, revealed nonspecific hypodense foci involving the white matter of the cerebral hemispheres, without evidence of intracranial hemorrhage, hydrocephalus, or mass effect. The radiology report suggested correlation with magnetic resonance imaging (MRI), according to clinical judgment.

Etiologic Evaluation of Hypertension

The severity of hypertension, the absence of renovascular abnormalities, and the presence of cardiac involvement prompted an expanded evaluation for endocrine and vascular causes of secondary hypertension.

On February 16, CT angiography of the chest, abdomen, and pelvis was performed. The examination showed no relevant abnormalities of the aorta or the major vascular structures evaluated.

However, abdominal imaging identified a nodule in the region of the right adrenal gland measuring approximately 23 mm, characterized by intense heterogeneous enhancement during the arterial phase following intravenous contrast administration. These findings raised suspicion for pheochromocytoma.

Hormonal testing demonstrated elevated plasma catecholamine concentrations, including norepinephrine of 988.6, compared with a reported upper reference limit of 420, and epinephrine of 292.7, compared with an upper reference limit of 84. Dopamine was below 30, within the laboratory reference range. The measurement units for these determinations should be confirmed against the original laboratory reports.

Total urinary metanephrines measured in a 24-hour collection were 1,809.70 µg/24 hours, compared with a reported upper reference limit of 520 µg/24 hours for the patient's age group. This value was approximately 3.5 times the upper reference limit.

Urinary vanillylmandelic acid was 4.6 mg/24 hours, within the reference range provided by the laboratory for individuals older than 10 years.

Additional endocrine testing included cortisol of 19.9, aldosterone of 21.10, dehydroepiandrosterone sulfate (DHEA-S) of 89, and thyroid hormone measurements. Thyroid-stimulating hormone (TSH) was 5.87 µIU/mL, slightly above the adult reference range provided by the laboratory, without sufficient evidence to establish clinically significant thyroid dysfunction at that time.

The combination of clinical findings, the adrenal nodule, and biochemical abnormalities further supported the suspicion of a catecholamine-secreting pheochromocytoma.

Clinical Course in the Pediatric Intensive Care Unit

The patient remained in the PICU from February 15 through February 24, 2023, because of severe hypertension, the need for intravenous antihypertensive infusion, and investigation of the associated systemic manifestations.

During this period, she exhibited marked blood pressure variability, requiring successive adjustments to pharmacologic treatment.

Sodium nitroprusside was administered from February 15 through February 21. Intravenous hydralazine was also used on admission, clonidine was administered between February 17 and 18, and oral antihypertensive medications were progressively introduced.

Propranolol was initially prescribed before doxazosin was introduced. Subsequently, doxazosin, captopril, and amlodipine were administered, with dose adjustments based on blood pressure response and clinical course.

Doxazosin was introduced on February 18 and gradually titrated to 4 mg/day. Captopril was initiated on February 20 at 25 mg every eight hours, whereas amlodipine was introduced on February 22.

During hospitalization, recurrent hypokalemia was documented, with potassium concentrations of 2.4 mmol/L on February 17, 2.8 mmol/L on February 18, and 2.7 mmol/L on February 19. Reduced magnesium concentrations, ranging from approximately 1.5 to 1.66 mg/dL, were also observed, prompting electrolyte replacement.

On February 21, during an episode of hypotension temporally associated with manipulation of a vascular access device, an event described as a seizure was documented.

Subsequent head CT showed no significant acute abnormalities. Electroencephalography performed during the same period demonstrated infrequent epileptiform activity in the left posterior region.

The available documentation does not allow the mechanism of the seizure-like event to be definitively established, nor does it demonstrate an exclusive causal relationship with blood pressure changes, primary epileptic activity, or possible hypertension-related cerebral involvement.

Following progressive stabilization of blood pressure, sodium nitroprusside was discontinued on February 21.

The patient's clinical condition improved, and she was transferred from the PICU to the pediatric ward on February 24, where blood pressure monitoring and multidisciplinary management continued.

Hospital Ward Course and Preoperative Preparation

During her stay on the pediatric ward, the adolescent remained clinically stable, although visual disturbances persisted, particularly in the right eye.

She continued to be followed by the pediatrics, urology, ophthalmology, and anesthesiology teams while hormonal test results were reviewed and the surgical treatment strategy was determined.

An anesthesiology assessment performed on March 1 classified the patient as American Society of Anesthesiologists Physical Status III (ASA III) and recommended continuing the previously initiated adrenergic preparation before surgery.

On March 3, the patient was alert, active, afebrile, and free of headache, abdominal pain, nausea, or vomiting. She reported persistent blurred vision in the right eye, although gradual improvement was noted.

Vital signs near discharge included a heart rate of 96 beats/min, respiratory rate of 24 breaths/min, peripheral oxygen saturation of 100%, and blood pressure of 120/90 mmHg.

She was discharged with prescriptions for captopril, amlodipine, doxazosin, and propranolol, along with instructions for home blood pressure monitoring, recognition of warning signs, and specialized follow-up.

The urinary metanephrine results, which became available subsequently, further supported the indication for surgical treatment of the adrenal lesion.

Surgical Treatment and Histopathologic Evaluation

On March 29, 2023, the patient was readmitted for surgical treatment of the right adrenal lesion.

On March 30, she underwent right retroperitoneoscopic adrenalectomy. The procedure was described as uneventful, and the surgical specimen was submitted for histopathologic examination.

During the immediate postoperative period, she remained under hospital observation, including monitoring in the PICU. The surgical team recommended discontinuation of the previously administered antihypertensive medications and continued clinical monitoring.

The histopathology results provided by the authors confirmed the diagnosis of an adrenal medullary neuroendocrine neoplasm consistent with pheochromocytoma.

Microscopic examination revealed a nested cellular architecture, known as the *zellballen* pattern, consisting of cells with abundant cytoplasm, round or oval nuclei, granular "salt-and-pepper" chromatin, and nuclear pleomorphism.

No capsular or vascular invasion was identified in the examined tissue sections. The Pheochromocytoma of the Adrenal Gland Scaled Score (PASS) was reported to be below 5.

Although this score represents a relatively favorable histologic finding, it does not allow the tumor to be definitively classified as biologically benign or exclude future recurrence or metastatic disease.

The macroscopic description provided did not allow the external characteristics, color, or presence of hemorrhagic and cystic changes in the specimen to be established with precision. Therefore, these features were not included as definitive findings.

The diagnosis of pheochromocytoma was established through integration of the clinical presentation, hormonal evaluation, imaging findings, and histopathologic confirmation following adrenalectomy.

Outcome and Follow-up

The immediate postoperative course was favorable, with no documented surgical complications.

On April 2, 2023, the patient was hemodynamically stable, had no significant clinical complaints, was voiding spontaneously, and had no signs of peritoneal irritation on abdominal examination.

She was discharged with instructions for outpatient urology follow-up.

The available records do not include serial postoperative metanephrine measurements, results of germline genetic testing, or sufficient information regarding long-term cardiovascular and ophthalmologic outcomes.

Consequently, although the immediate clinical outcome was favorable, definitive biochemical remission, complete regression of target-organ damage, and absence of tumor recurrence during follow-up cannot be established.

Discussion

This case describes a severe presentation of pediatric pheochromocytoma, initially identified through an extreme hypertensive emergency associated with cardiovascular, retinal, and renal involvement. Its distinctive clinical feature is the magnitude of systemic complications associated with an adrenal tumor measuring only 23 mm.

This presentation illustrates that the anatomic dimensions of a pheochromocytoma should not be used in isolation to estimate its hemodynamic impact. Although tumor size is relevant to prognostic assessment and surgical planning, clinical manifestations also depend on the secretory profile, intensity of catecholaminergic activity, duration of exposure, and individual cardiovascular response.

Hypertension is among the most important manifestations of pheochromocytoma. In children and adolescents, it may be sustained and frequently severe, contrary to the traditional perception that these tumors exclusively produce paroxysmal hypertensive episodes.

In this case, a blood pressure of 261/196 mmHg, accompanied by headache, vomiting, visual disturbances, and cardiac abnormalities, constituted a hypertensive emergency with systemic consequences.

Recognition of the severity of the condition was essential for the initiation of intravenous treatment and admission to intensive care. Management of pediatric hypertensive emergencies must account for the degree of blood pressure elevation, the presence of acute target-organ injury, and the risk of hypoperfusion associated with excessively rapid blood pressure reduction.

Identification of a catecholamine-secreting tumor also alters the therapeutic approach because sustained adrenergic stimulation may cause intense vasoconstriction, tachycardia, arrhythmias, and hemodynamic instability.

Retinal Involvement and Loss of Visual Acuity

One of the most clinically significant aspects of this case was the marked ophthalmologic involvement.

The presence of optic disc edema, retinal hemorrhages, perimacular exudates, and substantially reduced visual acuity in the right eye demonstrates microvascular injury associated with severe hypertension.

Persistently elevated blood pressure may impair the autoregulatory mechanisms of retinal circulation, resulting in arteriolar vasoconstriction, increased vascular permeability, ischemia, and extravasation of blood components into retinal tissues.

In severe cases, the combination of optic disc edema, hemorrhages, and exudates is consistent with advanced hypertensive retinopathy and represents important evidence of target-organ damage.

In the present case, visual acuity of 20/400 in the right eye, compared with 20/30 in the left eye, demonstrated substantial functional impairment that partially persisted even after initial blood pressure stabilization.

However, the available records do not establish the ultimate degree of visual recovery. This limitation highlights the importance of specialized ophthalmologic follow-up after control of the underlying hypertensive condition, particularly when macular and optic nerve abnormalities are present.

Cardiovascular Involvement and the Differential Diagnosis of Ventricular Hypertrophy

Echocardiography revealed marked concentric left ventricular hypertrophy, a reduced ventricular cavity, hypercontractility, and an intraventricular systolic gradient.

These findings require careful interpretation, particularly because the original echocardiographic report raised the possibility of hypertrophic cardiomyopathy.

Left ventricular hypertrophy may represent an adaptive response to sustained pressure overload. In addition, catecholamine excess may produce a range of cardiovascular manifestations, including abnormalities of myocardial contractility, ventricular dysfunction, arrhythmias, and cardiomyopathy associated with a hyperadrenergic state.

In this case, marked concentric hypertrophy associated with extremely elevated blood pressure supports the consideration of hemodynamic overload as a possible contributing mechanism.

Nevertheless, the available documentation does not allow primary cardiomyopathy or other associated structural conditions to be completely excluded.

The reported moderate pulmonary infundibular stenosis also warrants attention because it is not a typical or specific manifestation of catecholamine excess. Its presence further supports the need for cardiologic reassessment following hemodynamic stabilization.

The absence of echocardiographic follow-up after adrenalectomy prevents determination of whether left ventricular hypertrophy regressed, the intraventricular gradient normalized, or cardiac function changed.

Therefore, the most scientifically appropriate description is that of structural and functional cardiac involvement documented during the hypertensive emergency, without attributing its etiology exclusively to pheochromocytoma or establishing a definitive diagnosis of primary hypertrophic cardiomyopathy.

Renal and Electrolyte Abnormalities

Urinary evaluation revealed substantial proteinuria and an elevated albumin-to-creatinine ratio, consistent with renal involvement in the setting of severe hypertension.

Despite these abnormalities, no structural renal anomalies or significant renal artery stenoses were identified, and serum creatinine values did not show marked elevations during the initial assessment.

These findings suggest that proteinuria may have been associated with hemodynamic injury to the renal circulation. However, resolution following blood pressure stabilization was not documented, preventing determination of whether the abnormality was transient or persistent.

Another relevant finding was recurrent hypokalemia, with a documented nadir of 2.4 mmol/L, accompanied by acid-base abnormalities at certain points during hospitalization.

Although hypokalemia may occur in several clinical settings, including gastrointestinal losses, medication effects, and hormonal disturbances, its presence in a patient with severe hypertension justified a broader endocrine evaluation.

The aldosterone measurement alone was insufficient to establish primary aldosteronism, particularly in the absence of adequate information regarding renin levels, sampling conditions, and medication-related interference.

Potassium and magnesium replacement were important components of clinical stabilization, especially given the potential cardiovascular risks associated with electrolyte abnormalities.

Neurologic Manifestations During Hospitalization

The patient experienced an event described as a seizure that was temporally associated with hypotension during manipulation of a vascular access device.

Although the event occurred in the setting of circulatory instability, its etiology cannot be established solely from the available information.

The initial head CT demonstrated nonspecific white matter hypodensities, whereas the subsequent examination revealed no significant acute abnormalities.

Electroencephalography showed infrequent epileptiform activity in the left posterior region. This finding requires clinical interpretation but does not, by itself, establish the mechanism of the observed event.

In patients with severe hypertension, neurologic manifestations may result from several processes, including impaired cerebral autoregulation, hypoperfusion, metabolic disturbances, and epileptic seizures.

Posterior reversible encephalopathy syndrome (PRES) should also be considered in the differential diagnosis of patients presenting with severe blood pressure elevation, headache, visual disturbances, and seizures. However, confirmation requires appropriate clinical evaluation and neuroimaging, particularly brain MRI, which was not available in the reviewed records.

Accordingly, PRES cannot be established as a diagnosis in this patient, although it remains a relevant differential diagnostic consideration.

Biochemical Diagnosis and Tumor Localization

Recognition of pheochromocytoma requires integration of clinical suspicion, biochemical evidence of catecholamine secretion, and anatomic localization.

International recommendations identify plasma free metanephrines or fractionated urinary metanephrines as the preferred initial diagnostic tests because of their greater sensitivity compared with isolated catecholamine or vanillylmandelic acid measurements (1,2).

In this case, elevated plasma norepinephrine and epinephrine supported the suspicion, whereas total urinary metanephrines of 1,809.70 µg/24 hours provided additional relevant biochemical evidence.

However, the available result refers to total metanephrines without differentiation between metanephrine and normetanephrine. This limitation prevents a complete biochemical characterization of the tumor's secretory phenotype.

Similarly, a urinary vanillylmandelic acid concentration within the reference range did not exclude pheochromocytoma, particularly in the presence of the clinical manifestations, other biochemical findings, and adrenal imaging abnormalities.

CT angiography identified a 23-mm right adrenal lesion with intense heterogeneous arterial enhancement.

The small size of the lesion is a notable feature of this case because it demonstrates that a relatively limited anatomic tumor burden may coexist with major systemic manifestations.

Nevertheless, this association does not establish a direct quantitative relationship between tumor size and hormonal production, since clinical severity may depend on multiple factors that were not measured in the present report.

Pharmacologic Management and Preoperative Preparation

The management of functional pheochromocytomas requires careful preparation to minimize the risks associated with catecholamine release, particularly during tumor manipulation and anesthesia.

Alpha-adrenergic blockade is recommended as an initial component of preoperative preparation. Beta-adrenergic agents may subsequently be added when necessary to control tachycardia or other cardiovascular manifestations, after adequate alpha-adrenergic blockade has been established (1,2,4).

This sequence is important because isolated beta-adrenergic blockade in the presence of intense catecholamine secretion may result in predominance of alpha-adrenergic receptor-mediated vasoconstriction.

In the present case, the medical records indicate that propranolol was introduced before doxazosin, while the underlying diagnosis was still under investigation.

This aspect warrants critical consideration in light of current recommendations; however, it does not establish that the sequence of medication administration caused a specific complication in this patient.

As the suspicion of pheochromocytoma became stronger, doxazosin was introduced and progressively titrated, together with other agents for blood pressure control.

Intravenous sodium nitroprusside was used during the critical phase to manage extreme hypertension, while the gradual introduction of oral therapy contributed to stabilization and preparation for definitive treatment.

Current recommendations also emphasize the importance of assessing intravascular volume expansion and reversing the contracted circulating volume associated with chronic catecholamine excess, particularly during the preoperative period.

The available documentation does not allow the specific volume optimization measures implemented before surgery to be fully characterized.

Surgical Treatment and Histopathologic Interpretation

Adrenalectomy is the treatment of choice for localized and resectable pheochromocytomas.

A minimally invasive approach may be used in selected patients, taking into consideration tumor location, size, radiologic characteristics, and the surgical team's experience (1,2).

In the present case, right retroperitoneoscopic adrenalectomy was performed without documented complications following clinical stabilization and preanesthetic evaluation.

Histopathologic confirmation demonstrated a *zellballen* architectural pattern, characteristic of chromaffin cell neoplasms, with abundant cytoplasm and granular nuclear chromatin.

No capsular or vascular invasion was identified in the examined tissue sections, and the PASS was below 5.

Nevertheless, interpretation of this score requires caution. Histologic risk stratification systems, including PASS and the Grading System for Adrenal Pheochromocytoma and Paraganglioma (GAPP), have limitations regarding reproducibility and prognostic performance. Therefore, the World Health Organization classification does not recommend their isolated use to predict metastatic behavior (5).

The absence of capsular or vascular invasion in the examined sections also does not guarantee freedom from future recurrence or metastasis.

Thus, although the histopathologic diagnosis confirms pheochromocytoma, assessment of clinical, molecular, and genetic risk factors remains necessary to determine the likelihood of future disease manifestations.

Hereditary Predisposition and Follow-up

Genetic evaluation is an essential component of care for children and adolescents diagnosed with pheochromocytoma.

The 2024 international consensus statement highlights that approximately 70% to 80% of pediatric pheochromocytomas and paragangliomas are associated with hereditary predisposition, a higher proportion than that observed in many adult populations (1).

Susceptibility genes include *VHL*, *RET*, *NF1*, and genes encoding succinate dehydrogenase subunits, among other molecular alterations associated with the development of these neoplasms.

The absence of a characteristic family history does not exclude genetic predisposition because variable penetrance, previously unrecognized familial manifestations, and de novo variants may occur.

In this case, no germline or somatic genetic testing results were available. Therefore, it was not possible to determine whether the tumor was associated with a specific hereditary syndrome.

The need for prolonged follow-up must also be emphasized.

International recommendations advise postoperative biochemical reassessment, preferably within two to eight weeks after surgical treatment, to document hormonal remission. In patients with confirmed genetic predisposition, lifelong follow-up is required, with surveillance tailored to the molecular and clinical risk profile.

Even when no hereditary predisposition has been identified, prolonged follow-up is recommended because of the potential for recurrence or development of additional tumors.

The patient was discharged in clinically stable condition after surgery, but the available records do not confirm normalization of metanephrine levels, regression of cardiac abnormalities, or complete visual recovery.

These outcomes would be particularly relevant for characterizing the reversibility of target-organ injury following removal of the catecholamine source.

Clinical Relevance and Limitations of the Report

This case presents several clinically relevant features: an initial presentation with extreme hypertension, nonspecific symptoms, significant ocular involvement, marked ventricular hypertrophy, urinary and electrolyte abnormalities, identification of a small adrenal tumor, and histopathologic confirmation following surgical resection.

The principal clinical message is that the severity of pheochromocytoma should not be underestimated based on tumor size alone.

Furthermore, the presence of target-organ damage in an adolescent without a previous diagnosis of hypertension reinforces the need for prompt etiologic investigation when severe blood pressure elevation is identified.

The limitations of this report include the absence of fractionated urinary metanephrine measurements, incomplete documentation of preoperative intravascular volume optimization, lack of available genetic testing results, absence of postoperative catecholamine or metanephrine measurements, and unavailable long-term cardiac and ophthalmologic follow-up.

The histopathologic documentation provided also lacks several macroscopic, immunohistochemical, and molecular parameters that could contribute to additional prognostic characterization.

Despite these limitations, the case consistently demonstrates the temporal association between histopathologically confirmed pheochromocytoma and a severe hypertensive emergency accompanied by multiple manifestations of target-organ damage.

Conclusion

Pheochromocytoma is a rare but potentially severe and treatable cause of secondary hypertension in children and adolescents.

This case demonstrates that an adrenal lesion measuring only 23 mm may coexist with an extreme hypertensive emergency, severe retinopathy, substantial cardiac involvement, and renal abnormalities, underscoring that tumor dimensions should not be interpreted in isolation as an indicator of clinical severity.

Systematic investigation of the causes of secondary hypertension, combined with target-organ assessment and appropriate biochemical and imaging studies, was essential for identifying the underlying etiology and guiding definitive treatment.

Right adrenalectomy enabled resection of the lesion, with histopathologic confirmation of pheochromocytoma and a favorable immediate postoperative course.

However, the absence of complete follow-up data prevents confirmation of definitive biochemical remission or complete reversibility of the cardiovascular and ophthalmologic manifestations.

This case reinforces the importance of multidisciplinary management, appropriate pharmacologic preparation, genetic evaluation, and prolonged follow-up in pediatric patients with pheochromocytoma.

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