CASE REPORT

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Adrenergic Storm with Obstructive Hydrocephalus: Atypical Neurological Presentation of Von Hippel-Lindau Disease with Bilateral Pheochromocytoma in an Adolescent

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Abstract

Pheochromocytoma is often not the first diagnosis considered for many neurological complaints, especially obstructive hydrocephalus. We report a case of Von Hippel-Lindau disease in an adolescent presenting with obstructive hydrocephalus due to adrenergic storm requiring emergency ventriculoperitoneal shunt. This vignette emphasizes the importance of an integral approach, particularly accurate diagnosis that would lead to targeted pharmacological therapy focusing on alpha-adrenergic receptor blockade, which was followed by total curative excision of pheochromocytoma in a timely fashion.

Key words: adolescent, germline mutation, obstructive hydrocephalus, Von Hippel-Lindau disease, pheochromocytoma, posterior reversible encephalopathy syndrome

INTRODUCTION

Pheochromocytoma (PCC) and paraganglioma (PGL) are rare chromaffin cell tumors that secrete catecholamine with an annual incidence of approximately 2-8 million person-years.[1,2] The classic triad of symptoms in patients with PCC consists of episodic headache, sweating and tachycardia. It has an atypical presentation in 9%-10% of patients which includes cerebrovascular events.[3] To date, cerebral infarction, intracranial hemorrhage, posterior reversible encephalopathy syndrome (PRES) and catecholamine-induced central nervous system (CNS) vasculitis have all been identified as unusual neurological manifestation of PCC.[4] Herein, we report a case of a PRES with atypical features in a 12-year-old male, newly diagnosed with bilateral PCC and VHL disease.

CASE

A 12-year-old male presented to the emergency department (ED) with a 3-day history of severe headache (pain score 8/10), accompanied by visual disturbances and an unintentional weight loss of 4 kg over three months. He denied fever, constitutional symptoms, or contact with tuberculosis. On examination, he was conscious but markedly hypertensive (BP: 180/120 mmHg) and tachycardic (HR: 155 bpm). Fundoscopic evaluation revealed bilateral grade 4 hypertensive retinopathy with papilledema. Electrocardiogram demonstrated sinus tachycardia with evidence of left ventricular hypertrophy. Laboratory parameters, including complete blood count, renal, hepatic and coagulation profiles, were within normal limits. CT scan with contrast of the brain revealed obstructive hydrocephalus with diffuse cerebellar edema and tonsillar herniation (Figure 1A). The provisional diagnosis at this juncture was obstructive hydrocephalus secondary to posterior fossa mass. Emergency ventriculoperitoneal (VP) shunt was performed after stabilization with intravenous labetalol, which was successfully tapered off within 4 hours postoperatively.

MRI of the brain performed on day 3 post-VP shunt showed diffuse cerebellitis without evidence of a posterior fossa mass. Given the abrupt neurological manifestation and typical neuroimaging findings of obstructive hydrocephalus, empirical treatment for tuberculous (TB) meningitis with anti-TB medications and intravenous dexamethasone were initiated after multidisciplinary discussion with intensivist and neurologist due to regional prevalence. Nonetheless, the cerebrospinal fluid (CSF) findings later did not support TB meningitis because the MTB GeneXpert was negative and CSF biochemistry was normal. Autoimmune screening, including anti-nuclear antibody and complement levels were within the normal range. Anti-TB therapy was discontinued on day 4. Despite initial improvement, the patient remained hypertensive

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(SBP: 160–190 mmHg, DBP: 90–110 mmHg) and tachycardic despite being treated with oral metoprolol 50 mg twice daily and oral prazosin 1mg thrice daily. He was referred to the endocrinology service for evaluation of secondary hypertension in a pediatric patient.

A detailed history uncovered intermittent episodes of headache, diaphoresis and visual blurring for the past three months. Physical examination revealed a thin male with growth parameters below the 25th percentile. There were no cutaneous stigmata of neurofibromatosis or features suggestive of multiple endocrine neoplasia (MEN) syndromes. Birth and developmental histories were unremarkable. Family history, including a detailed three-generation pedigree, was negative for endocrinopathies, pheochromocytoma, renal tumors, or central nervous system (CNS) neoplasms. Both parents and a 16-year-old sister were healthy. The maternal side of the family, including six siblings of the mother, had no significant medical conditions. The paternal side, including four siblings of the father, were similarly unaffected. The paternal grandmother had end-stage renal disease in her 60s of unclear etiology, but there was no documented history of PCC, renal cell carcinoma, or CNS tumors.

Endocrinological evaluation revealed markedly elevated urinary normetanephrine (101.8 μmol/day; reference range: 2.46–23.0) and 3-methoxytyramine (4.9 μmol/day; reference range: 0.10–1.79). The aldosterone-renin ratio (ARR) was low, with a direct renin level of 106.3 mU/L (reference range: 3–66), plasma aldosterone of 582.3 pmol/L (reference range: 102.5–1196) and ARR of 5 (Table 1). Thyroid function, serum calcium and phosphate levels were within normal limits, excluding other common endocrine causes of secondary hypertension. A suppressed morning cortisol level (<13.8 mmol/L) was noted, though interpretation was limited due to prior dexamethasone therapy. Cross-sectional imaging via contrast-enhanced CT of the thorax, abdomen and pelvis revealed bilateral adrenal masses (right: 3.2 × 2.6 × 4.2 cm; left: 3.1 × 4.6 × 6.3 cm) with associated abdominal lymphadenopathy (Figure 2). Functional imaging with Ga-68 DOTATATE PET/CT confirmed intense somatostatin receptor expression in both adrenal lesions, while the lymph nodes remained non-avid (Figure 3).

Initial alpha-blockade with prazosin was transitioned to terazosin and subsequently to oral phenoxybenzamine, which was titrated up to 10 mg three times daily based on body weight, symptomatology and blood pressure targets

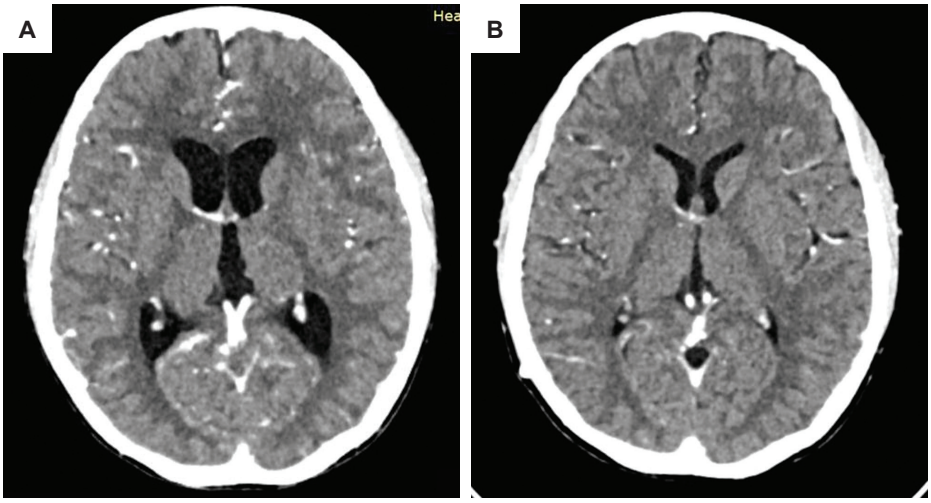

Figure 1. (A) Contrast CT of the brain on admission demonstrating diffused bilateral cerebellar oedema with obstructive hydrocephalus. **(B)** Repeat Contrast CT of the brain showed resolved cerebellar edema and obstructive hydrocephalus.

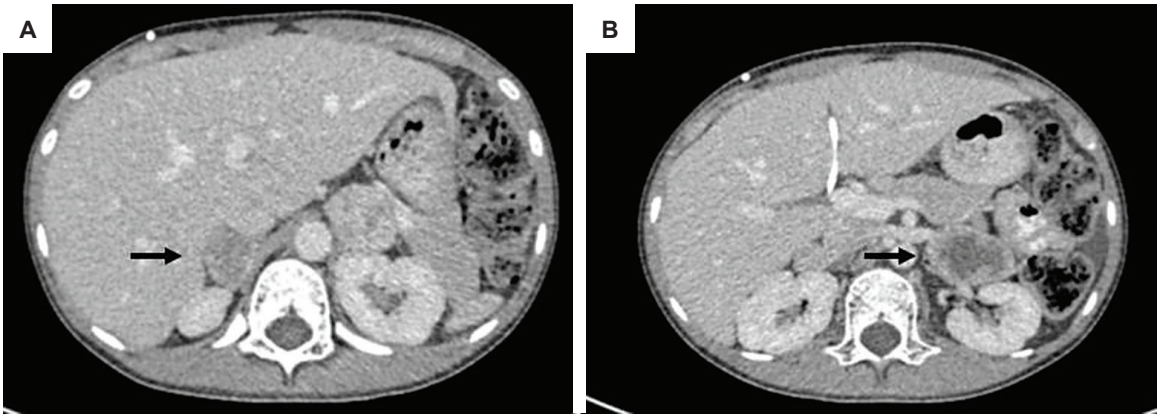

Figure 2. (A) Lobulated right suprarenal soft tissue lesion measuring 3.6 × 2.9 × 4.7 cm with central necrosis *(black arrow)*. No clear fat plane is seen between the lesion and segment VII/VI/V of liver; **(B)** Lobulated left suprarenal soft tissue lesion measuring 3.1 × 4.6 × 6.3 cm with central necrosis *(black arrow)*. Poor demarcation is seen between the lesion and body of the pancreas as well as the adjacent small bowel.

Table 1. Laboratory results

Investigations	Results	Normal range
24 hour urine metanephrine		
Normetanephrine	101.8 µmol/day	2.46 – 2.30
Metanephrine	0.6 µmol/day	0.30 – 0.95
3-methoxytyramine	4.9 µmol/day	0.10 – 1.79
Aldosterone renin ratio		
Direct renin level	106.3 mIU/L	3 – 66
Plasma aldosterone concentration	582.3 pmol/L	102.6 – 1196
ARR	5	
Thyroid function test		
TSH	1.1 mU/L	0.55 – 4.78
FT4	12.1 mmol/L	12.2 – 22
Cerebrospinal fluid analysis		
Appearance	Clear and colorless	
Cell count	0	
Acid Fast Bacilli direct smear	Negative	
Mycobacterium tuberculosis	Negative	
GeneXpert	Negative	
Protein	0.159 mmol/L	
Glucose	5 g/dL	

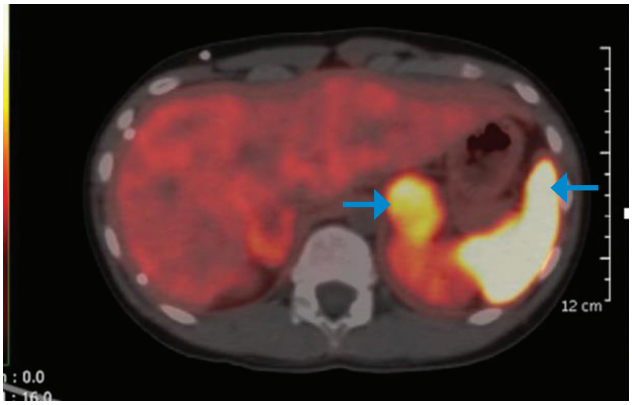

Figure 3. Ga-68 DOTATATE showing scan evidence of somatostatin receptor avid disease in bilateral suprarenal lesions with SUV_{max} 9.7 on the right suprarenal soft tissue and SUV_{max} 17.7 on the lobulated left suprarenal soft tissue *(blue arrows)*.

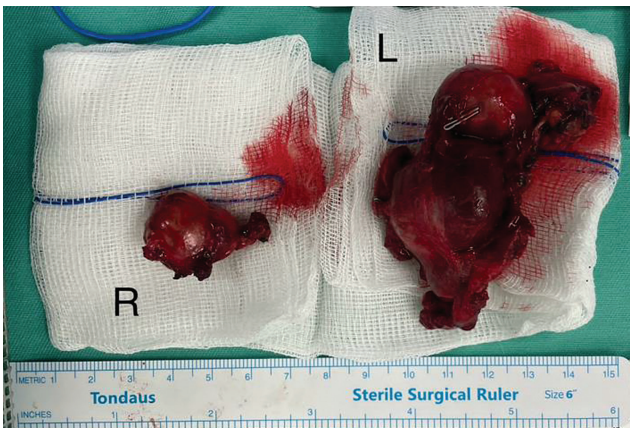

Figure 4. Gross pathology showing bilateral pheochromocytoma. The right adrenal tumor shows multilobulated with the main tissue mass measuring 6.0 × 2.5 × 3.0 cm with disrupted capsule. The left adrenal tumor shows nodular brownish tumor with smooth capsule, measuring 6.5 × 5.0 × 2.0 cm.

(<130/80 mmHg). The patient underwent successful bilateral adrenalectomy. Intraoperatively, both adrenal glands were found to contain well-encapsulated, lobulated tumors (right: 6 × 2.5 × 3 cm; left: 6.5 × 5 × 2 cm) (Figure 4). Histopathological analysis confirmed bilateral pheochromocytomas, exhibiting classical zellballen architecture with evidence of both capsular and vascular invasion. Immunohistochemistry was positive for chromogranin A and synaptophysin, consistent with neuroendocrine differentiation.

Given the bilateral presentation at a young age, genetic testing was conducted and identified a heterozygous pathogenic variant in the VHL gene (c.482G>A; p.Arg161Gln), confirming a diagnosis of von Hippel–Lindau (VHL) syndrome. Predictive genetic testing of both parents and the proband's sister yielded negative results, indicating a likely *de novo* mutation.

Follow-up contrast-enhanced CT of the brain performed three weeks after initiation of alpha-adrenergic blockade and effective blood pressure control showed complete resolution of cerebellar edema, hydrocephalus and no evidence of leptomeningeal enhancement (Figure 1B). Postoperatively, the patient's symptoms resolved entirely and he remained normotensive without the need for antihypertensive therapy. Electrolytes and glucose levels remained stable and steroid replacement was initiated as per protocol. Surveillance evaluations, including ophthalmologic and otolaryngologic assessments, revealed no evidence of other VHL-associated lesions such as retinal hemangioblastomas, renal cell carcinoma, pancreatic cysts or neuroendocrine tumors.

DISCUSSION

Acute neurological crises as the initial presentation of PCC often led to delay in the PCC work-up as the initial pursuit would be focusing on stabilizing the neurological emergency.[3,4] According to Anderson et al., who had retrospectively reviewed 93 patients with PCC aged 9-84 years, the common neurological crises were headache, seizures, strokes, delirium and subarachnoid haemorrhage.[5] To note, neurological manifestation with PRES is an exceedingly rare phenomenon among the pediatric population, albeit PRES occurs extensively in adults. The pathophysiology of PRES involve catecholamine-induced hypertensive surges overwhelming cerebral autoregulation, especially in the posterior circulation, leading to profound vasogenic oedema.[6,7] Furthermore, catecholamine excess could directly cause endothelial dysfunction and cerebral vasoconstriction, increasing the risk of both hemorrhagic and ischemic events.[3,7] In essence, this vignette adds to the limited literature of PCC with atypical neurological manifestations, reinforcing the need for early endocrine and pediatric evaluation in pediatric hypertensive crises.

PRES is a time-sensitive disease and the prognosis depends on a timely detection which would translate into delivery of appropriate therapy before the occurrence of irreversible

or even fatal neurological complications. The cornerstone of PRES management comprises of control of hypertension and treatment of the underlying cause. In another note, the outcomes of PRES associated with PCC varied with some patients experiencing significant neurological recovery, while others faced severe complications. Powell et al., reported a young man who presented with clinical features of PRES secondary to PCC crisis, diagnosed as MEN2A syndrome, which had a good outcome after prompt treatment.[8] Callao et al., reported a male patient who had left adrenalectomy done due to PCC who also presented with PRES but succumb due to complication of liver failure.[9] In our case, PRES induced by PCC had extended to the cerebellum and brainstem resulting cerebrospinal fluid outflow obstruction with ongoing tonsillar herniation due to cerebellar edema. In contradistinction to the typical parieto-occipital involvement in PRES, our case demonstrated an isolated posterior fossa involvement which is exceedingly rare among adolescent population and these findings mimicked posterior fossa mass on initial CT imaging.[10,11] This observation was also reported by Ogino et al., and Grossbach et al., who described acute obstructive hydrocephalus caused by PRES.[12,13] Despite in extremis presentation, the prompt neurosurgical intervention and intensive care in our case had successfully relieved the intracranial pressure and demonstrated a satisfactory neurological recovery.

Although pheochromocytoma-paragangliomas (PPGLs) are rare in children accounting for 0.2–0.5 cases per million population annually, they remain the most common endocrine tumors in this age group and are implicated in approximately 0.5–1% of pediatric hypertensive cases.[14] Compared to adults in whom paroxysmal hypertension is more characteristic, adolescent presentations often feature persistent hypertension as the dominant symptom. Up to 91% of PCC patients present with classic symptoms such as episodic headache, diaphoresis, palpitations and hypertension.[15] However, atypical and severe neurological presentations, including PRES, cerebral infarction, CNS vasculitis and even hemorrhagic stroke, have been documented in children and adolescents with PPGL.[3,7]

PCC associated with VHL syndrome, categorized under Cluster 1B of PPGL classification, typically demonstrate a noradrenergic biochemical phenotype.[16,17] In this patient, 24-hour urinary catecholamine metabolite analysis showed markedly elevated normetanephrine (101.8 μmol/day; >40 times the upper limit of normal), a moderate increase in 3-methoxytyramine and normal metanephrine levels. This biochemical profile aligns with the characteristic pattern seen in VHL-related tumors. The underlying mechanism involves VHL gene mutations leading to stabilization of hypoxia-inducible factor 2-alpha (HIF-2α), which regulates genes involved in catecholamine biosynthesis. HIF-2α suppresses phenylethanolamine N-methyltransferase (PNMT), the enzyme responsible for converting norepinephrine to epinephrine, resulting in a predominant noradrenergic phenotype. Additionally, a modest elevation in 3-methoxytyramine, a dopamine metabolite, may reflect minor dopaminergic activity, consistent with the biochemical spectrum of VHL-associated pheochromocytomas.[17] Notably, the low aldosterone-to-renin ratio observed in our case can be attributed to the elevated plasma renin activity commonly seen in pheochromocytoma. This phenomenon is primarily driven by chronic catecholamine excess, which induces sustained vasoconstriction and leads to reduced circulating plasma volume. The resulting hypovolemia stimulates renin release via activation of β1-adrenergic receptors. Additionally, catecholamines directly enhance renin secretion through β1-adrenergic receptor-mediated pathways. These mechanisms collectively explain the increased plasma renin activity and consequently, the low aldosterone-to-renin ratio in patients with PCC.[18]

Cross-sectional imaging with contrast-enhanced CT of the thorax, abdomen and pelvis revealed bilateral adrenal masses and abdominal lymphadenopathy. The right adrenal mass measured 3.6 × 2.9 × 4.7 cm and the left measured 3.9 × 4.8 × 6.0 cm. Both lesions were heterogeneous, well-circumscribed and confined to the adrenal glands, without evidence of local invasion or distant metastases. Cluster 1B VHL/EPAS1-related PPGLs specifically PCC seem to show stronger expression of the L-type amino-acid transporter and less SSTR2 expression. Therefore, PET/CT imaging based on [18F]FDOPA is more sensitive than [68Ga]-DOTA-SSA PET/CT in VHL/ EPAS1-related PPGLs.[19,20] However, this modality was not available in our setting. Consequently, Ga-68 DOTATATE PET/CT was selected as the next best available molecular imaging tool, allowing functional assessment of the adrenal lesions. Ga-68 DOTATATE PET/CT has demonstrated sensitivity of 92–100% and specificity of 88–100% in various studies particularly for detecting extra-adrenal and metastatic disease in VHL-associated pheochromocytoma.[17]

Genetic analysis identified a *de novo* germline *VHL* mutation, with no pathogenic variants detected in either parent or sibling. While *VHL* syndrome is autosomal dominant, approximately 20% of cases result from *de novo* mutations.[21] Genetic testing in all pediatric PPGL cases is essential, regardless of family history, as germline mutations underlie 50–70% of pediatric cases.[1] Syndromes such as MEN2, *VHL*, *NF1*, and SDHx mutations remain the most common etiologies. In children, *VHL* is the most frequently implicated genetic syndrome in bilateral adrenal PCC, consistent with our patient's presentation.[14]

Management of hypertension in PCC requires careful and sequential pharmacologic intervention. Our patient initially received intravenous labetalol which is a combined α- and β-adrenergic blocker in the Emergency Department as part of standard care for hypertensive emergency with obstructive hydrocephalus. However, in undiagnosed PCC, beta-blockade without adequate prior alpha-blockade may exacerbate hypertension due to unopposed α-adrenergic receptor stimulation.[22] This phenomenon was observed in our case: despite treatment with oral metoprolol and low-

dose prazosin (1 mg TID), the patient remained significantly hypertensive. Blood pressure control was only achieved following the initiation of phenoxybenzamine, a non-selective and non-competitive α_1- and α_2-adrenergic receptor blocker, which remains the cornerstone of preoperative management in PPGL. Dosage must be individualized, accounting for body weight and hemodynamic targets. In this 36-kg patient, phenoxybenzamine at 10 mg TID effectively achieved normotension, targeting a preoperative blood pressure of <130/80 mmHg. This case highlights the essential principle that adequate alpha-blockade must precede beta-blockade in the management of PPGL to avoid life-threatening cardiovascular complications. Beta-blockers may be added subsequently to manage reflex tachycardia, but only after sufficient α-blockade has been established. Long-term follow-up is crucial in patients with *VHL*-associated PPGLs due to the risk of multisystem involvement, including retinal hemangioblastomas, renal cell carcinoma and pancreatic cysts or neuroendocrine tumors.[23] Multidisciplinary surveillance involving endocrinology, oncology, ophthalmology and genetics is essential for optimal patient management.

CONCLUSION

Adolescents with neurological manifestation warrant detailed and vigorous diagnostic evaluation, especially in the presence of *de novo* onset hypertension. Investigations should gravitate towards the exclusion of PCC because adrenergic storm could be its initial manifestation as exemplified in this vignette. Importantly, timely diagnosis establishment allows alignment with multi-disciplinary teams that ultimately lead to curative PCC excision. Bespoke treatment via genetic testing allows further management strategy, familial screening and prognostication, though this vignette represents a case of sporadic VHL disease.

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Ethical Approval
This case report has obtained approval from National Medical Research Register (NMRR), Ministry of Health Malaysia: NMRR-ID-23-03447-BEA

Statement of Authorship
All authors certified fulfillment of ICMJE authorship criteria.

CRediT Author Statement
JMC: Conceptualization, Methodology, Formal analysis, Investigation, Resources, Writing – original draft preparation, Writing – review and editing, Visualization, Project administration; **TLT:** Conceptualization, Methodology, Formal analysis, Investigation, Resources, Writing – original draft preparation, Writing – review and editing, Visualization, Project administration; **YHL:** Conceptualization, Methodology, Formal analysis, Investigation, Resources, Supervision, Project administration; **SKYH:** Conceptualization, Methodology, Formal analysis, Investigation, Resources, Supervision, Project administration; **FBY:** Conceptualization, Methodology, Formal analysis, Investigation, Resources, Supervision, Project administration; **SWL:** Conceptualization, Methodology, Formal analysis, Investigation, Resources, Supervision, Project administration; **XYO:** Conceptualization, Methodology, Formal analysis, Investigation, Resources, Supervision, Project administration; **HCW:** Conceptualization, Methodology, Formal analysis, Investigation, Resources, Supervision, Project administration; **SLY:** Conceptualization, Methodology, Formal analysis, Investigation, Resources, Supervision, Project administration.

Data Availability Statement
No datasets were generated or analyzed for this study.

Author Disclosure
The authors declared no conflict of interest.

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