

Ropp K, Lalwani K, Koh J
Oregon Health & Science University , Portland , OR, USA

Introduction

We describe a teenager who was diagnosed with bilateral pheochromocytomas six years after developing symptoms of anxiety, depression and rage, which were inaccurately attributed to psychiatric illness. This led to a delay in diagnosis and unnecessary treatment. Her operative course was complicated by severe and refractory hypotension after bilateral adrenalectomy.

Case Report

An 18 year old female (MH) presented for elective resection of bilateral adrenal pheochromocytomas. Her past medical history was significant for anxiety and depression since age 11. MH began experiencing episodic symptoms of abdominal pain, vomiting, palpitations, headaches, anxiety and diaphoresis at age 12 and was treated for hypertension at age 16. She spent four weeks in an inpatient psychiatric hospital for depression and suicide attempt. A cardiologist ultimately initiated the evaluation for pheochromocytoma.

After two weeks of medical management with phenoxybenzamine, she underwent bilateral adrenal gland resection. Induction, intubation and invasive line placement were uneventful. Upon manipulation of the first adrenal gland, moderate hypertension was managed with nicardipine and magnesium infusions. After the first adrenalectomy, hypotension was treated with a norepinephrine infusion.

After the second adrenalectomy, the hypotension was far more severe, prolonged and refractory, requiring continuous infusions of norepinephrine (0.3 mcg/kg/min), vasopressin (15 units/hr) and epinephrine (0.1 mcg/kg/min). Within hours of arrival to the ICU, most of the infusions were discontinued. She was extubated later that day.

Due to the bilateral tumors and the childhood onset, MH underwent genetic testing. She was diagnosed with von Hippel-Lindau syndrome. Her psychiatric symptoms resolved after surgery.

Discussion

Pheochromocytoma is rare, with a reported annual incidence of one in 100,000. Approximately 10% of pheochromocytomas occur in children, and they are more likely to be associated with other syndromes, such as neurofibromatosis, multiple endocrine neoplasia (MEN), or von-Hippel Lindau (VHL) syndrome. VHL, in particular, is associated with significantly elevated catecholamine levels.¹

The delay in diagnosis in this case appears to be unprecedented in published literature. Anxiety is a common symptom of pheochromocytoma.² However, “total lack of response to anxiolytic agents” and hypertension should prompt the provider to consider other diagnoses.³

Conclusion

This report describes a rare case of von Hippel-Lindau syndrome and bilateral pheochromocytomas in a teenage girl, masquerading as anxiety and depression for six years before the tumors were identified. Her postoperative course was complicated by severe and refractory, albeit brief, hypotension.

References

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