INTRODUCTION

• Pheochromocytoma is a rare condition, with an incidence of one per 100,000 per year. Approximately 10% of them occur in children.

• Anxiety and other psychiatric disturbances are common symptoms of pheochromocytoma, attributed to excess catecholamine surges.

• Post-adrenalectomy hypotension has been widely reported and is attributed to the acute drop in circulating catecholamines.

ABSTRACT

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

CASE REPORT

Presentation

• An 18 year old female had significant anxiety and depression since age 11.

• She began experiencing episodic symptoms of abdominal pain, vomiting, palpitations, headaches, anxiety and diaphoresis at age 12.

• She spent four weeks in an inpatient psychiatric hospital for depression and suicide attempt.

• A cardiologist, who saw her for hypertension, ultimately initiated the evaluation for pheochromocytoma.

Intraoperative Course

• After two weeks of medical management she underwent laparoscopic 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).

Postoperative Course

• 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, the patient underwent genetic testing. She was diagnosed with von Hippel-Lindau syndrome.

• Her psychiatric symptoms and hypertension largely resolved after surgery.

DISCUSSION

• Pheochromocytomas in children 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.

• Anxiety is a common symptom of pheochromocytoma. Lack of response to anxiolytic agents and hypertension should prompt consideration of other etiologies.

• The post-adrenalectomy hypotension resolved quite quickly postoperatively, suggesting that it may have been due to residual medication effects. The severity, however, indicates that there was likely a component of shock due to the sudden and abrupt discontinuation of endogenous catecholamine secretion from the tumors.

REFERENCES

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