INTRODUCTION

Pheochromocytomas are catecholamine-secreting tumors that arise from chromaffin cells in the adrenal medulla.

Paragangliomas are tumors of the extra-adrenal sympathetic (catecholamine-producing) and parasympathetic (rarely catecholamine-producing) paraganglia, and can be located anywhere from the base of the skull to the pelvis.

These tumors are rare, occurring in 1:100,000 people, with only 10-20% diagnosed in childhood.

Though clinical presentation may vary, hypertension is the most common finding. Presenting symptoms may include headaches, sweating, and palpitations. Cardiac manifestations include tachycardia, arrhythmias, and catecholamine-induced cardiomyopathy.

CASE REPORT

Our patient is a 12 year old male who initially presented to an outside facility in acute heart failure of unknown etiology.

An echocardiogram revealed a markedly dilated left ventricle with very poor left ventricular function. The patient’s shortening fraction was 11%.

He did not respond to vasopressor therapy, and following a bradycardic arrest he was transferred to Vanderbilt Children’s Hospital and emergently placed on extracorporeal membrane oxygenation (ECMO), while being evaluated for possible heart transplant.

He subsequently had a left ventricular assist device (LVAD) placed, followed by right ventricular assist device placement one week later.

Close reevaluation of all prior medical records revealed a catecholamine-secreting tumor immediately adjacent to, or within the left adrenal gland, suspicious for a pheochromocytoma or paraganglioma.

After adequate alpha-blockade, the patient underwent successful laparoscopic left adrenalectomy and paraganglioma excision while on biventricular assist device support.

Approximately one month later, the patient received an orthotopic heart transplant and was discharged home approximately 80 days following his initial admission.

DISCUSSION

Pheochromocytomas & paragangliomas are rare neuroendocrine tumors that can cause drastic cardiovascular compromise due to excessive catecholamine secretion.

In the pediatric population, these tumors are more likely to have a genetic component and be a part of a hereditary syndrome such as von Hippel-Lindau disease, multiple endocrine neoplasia 2A & 2B, familial paraganglioma syndrome, and neurofibromatosis type 1.

Catecholamine-induced cardiomyopathy is usually improved with adreno-receptor blockade, and almost always resolves following excision of the tumor.

The diagnosis of these tumors remains difficult and a high index of suspicion is necessary. Plasma & urine metanephrine levels are the initial diagnostic tests of choice, followed by imaging studies to localize and look for multiple tumors.

Preoperative preparation is paramount. Goals include the control of blood pressure, heart rate, and rhythm, the restoration of blood volume, and ultimately, the prevention of a catecholamine crisis during surgical intervention. Adequate alpha- and potentially, beta-blockade are critical in this process.

Critical intraoperative events include periods of increased catecholamine secretion (intubation, incision, insufflation, & tumor manipulation), as well as the time following the ligation of venous drainage from the tumor.

The function of the BiVAD may be affected by the position of the patient and by the creation of the pneumoperitoneum.

REFERENCES