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Pheochromocytoma In A Patient With Severe Prosthetic Valve Aortic Stenosis, HFrEF, And Turner Syndrome

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INTRODUCTION

Patents presenting for pheochromocytoma resection require significant peri-operative management. Further challenges arise in the setting of severe prosthetic aortic valve stenosis (AS) with Turner Syndrome. This case report discusses the anesthetic management of a patient with a pheochromocytoma, AS, and Turner Syndrome.

CASE REPORT

A 66-year-old female with severe prosthetic aortic valve stenosis with a mean gradient of 54mmHg and a valve area of 0.7cm^2 presented for pheochromocytoma resection. Her past medical history included Turner's syndrome, biventricular systolic heart failure with a LV EF of 45%, bicuspid aortic valve s/p aortic valve replacement, congenital coarctation of the aorta which had been stented in infancy, atrial fibrillation, CAD s/p PCI to the RCA, and hypertension. During workup for aortic valve replacement, review of systems was positive for episodic headaches, blurry vision, and palpitations. An MRI scan revealed a right adrenal mass, and further testing confirmed a pheochromocytoma. After a multidisciplinary team discussion, the plan was made to pursue a right adrenalectomy prior to AVR. Ten days prior to surgery, the patient was admitted to the ICU for alpha blockade and fluid resuscitation. However, she suffered from severe orthostatic hypotension with phenoxybenzamine treatment. Instead, her home metyrosine dose was administered prior to surgery.

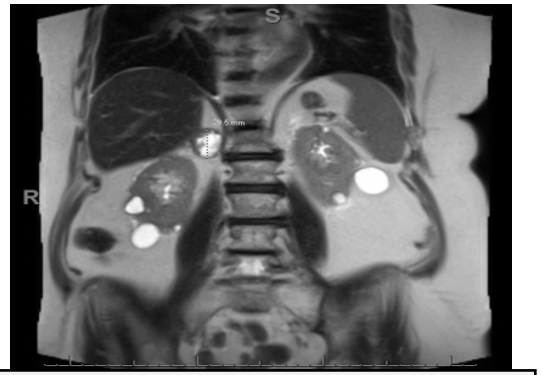
The scheduled adrenalectomy was performed under general anesthesia with arterial line, TEE, and pulmonary artery catheter monitoring. A cardiac surgeon and perfusion team were available. To facilitate potential rapid conversion to cardiopulmonary bypass in the event of hemodynamic instability, the right femoral artery and vein were cannulated with 5 french sheaths.

Intraoperatively, a Milrinone infusion was initiated due to elevated PA pressures. Octreotide, phentolamine, phenoxybenzamine, clevidipine, vasopressin, epinephrine, nitroprusside, and nitroglycerin were available. Upon surgical stimulation of the mass, the blood pressure increased to 180s/100s. Clevidipine and nitroglycerine were administered, and the blood pressure stabilized.

The specimen was removed, and the case proceeded uneventfully. Milrinone was weaned off, femoral catheters were removed, and the patient was extubated and transported to the ICU on 0.03mcg/kg/min of norepinephrine. Surgical pathology confirmed a pheochromocytoma and the patient was discharged without post-operative complications. A staged TAVR was performed three weeks later under deep sedation which proceeded uneventfully.

CASE REPORT

FIGURE 1



MRI image of the right adrenal mass measuring 29.5mm

DISCUSSION

This case demonstrates the difficult management of two rarely co-existing pathologies. In prior case reports of severe AS coincident with pheochromocytoma, AVR was performed prior to adrenalectomy, or AVR was aborted. This is a unique case in which adrenalectomy was performed prior to surgical management of the AS. This patient's care was further complicated by biventricular HF, CAD, and repaired coarctation of the aorta.

Preoperative recommendations for adrenalectomy include pharmacologic alpha blockade, but this patient did not tolerate phenoxybenzamine due to her AS. Metyrosine, a competitive inhibitor of tyrosine 3-monoxygenase, was used to impair synthesis of catecholamines.

Intraoperatively, we considered that decreased afterload might reduce coronary perfusion leading to myocardial ischemia and diminished cardiac output in the setting of severe AS. Our team was prepared to perform an urgent AVR under CPB if pharmacologic supports were not adequate. Further, peritoneal insufflation with attendant hypercarbia and diminished venous return can increase PA pressure and exacerbate RV dysfunction. We maintained continuous PA pressure monitoring and supported the RV with milrinone. Following successful adrenalectomy, a TAVR was performed three weeks after initial presentation to avoid the increased risk associated with redo sternotomy.

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