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Case Report

Perioperative Management of a Patient with a Pheochromocytoma, Severe Mitral Regurgitation, and Prior Right Pneumonectomy Undergoing Robotic Adrenalectomy: A Case Report

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Abstract

This case highlights the perioperative considerations of a patient with a pheochromocytoma, prior pneumonectomy, and severe mitral valve regurgitation presenting for adrenalectomy. Adrenalectomy for pheochromocytoma carries risks, particularly in patients with cardiopulmonary abnormalities. In this case, we discuss the rationale for proceeding with the adrenalectomy prior to mitral valve intervention. We illustrate the intraoperative surgical and pulmonary challenges in the setting of a prior pneumonectomy. Additionally, we outline the unique considerations during and after pheochromocytoma removal in the setting of mitral regurgitation. Implementation of these considerations is critical for management of a patient with severe cardiopulmonary disease undergoing adrenalectomy.

ABBREVIATIONS

CT: Computer Tomography; CTA: Computed Tomography Angiography; FRC: Functional Residual Capacity; LVEF: Left Ventricular Ejection Fraction; MR: Mitral Valve Regurgitation; RVEF: Right Ventricular Ejection Fraction; SBP: Systolic Blood Pressure; TEE: Transesophageal Echocardiogram; TTE: Transthoracic Echocardiogram

INTRODUCTION

Pheochromocytomas are catecholamine-producing neuroendocrine tumors of the adrenal medulla. Severe cardiovascular complications can arise during adrenalectomy due to the exaggerated release of norepinephrine and epinephrine.[1] In the perioperative setting, excessive catecholamine release occurs most frequently during laryngoscopy, endotracheal intubation, surgical incision, abdominal insufflation, and tumor manipulation [1]. Cardiovascular complications include hypertension, arrhythmias, myocardial infarction, heart failure, pulmonary edema, and cardiovascular collapse. Perioperative management during pheochromocytoma removal in patients with pre-existing cardiopulmonary disease offers additional challenges. Case reports describing this rare occurrence include patients with hypertrophic obstructive cardiomyopathy [2], heart failure with reduced ejection fraction and mitral regurgitation [3], heart failure with preserved ejection fraction [4], and coronary artery disease [5,6], but to the author's knowledge no case reports have described adrenalectomy in patients with a prior pneumonectomy and severe mitral regurgitation, compounding the complexity of the patient's cardiopulmonary disease.

We present the case of a 62-year-old male with severe mitral valve regurgitation (MR) and prior right pneumonectomy with a diagnosis of a pheochromocytoma presenting for adrenalectomy. Specifically, we describe the management of severe MR in the presence of catecholamine release during tumor manipulation. Further, we address the notable physiologic and anatomic considerations in such a patient during robotic adrenalectomy.

The presented case highlights the unique perioperative management of pheochromocytoma removal in a patient with severe pre-existing cardiac and pulmonary disease. Written HIPAA authorization was obtained from the patient.

CASE PRESENTATION

A 62-year-old man presented to our hospital for evaluation of an incidental adrenal mass. His symptoms included hypertension with systolic blood pressure (SBP) exceeding 180 mmHg, headaches, and episodic profuse sweating. Other medical history included severe structural MR, lung cancer status post right pneumonectomy 25 years ago and hypertension. The adrenal mass was discovered after the patient presented for management of fever and dyspnea that prompted chest computed tomography angiography (CTA). Subsequent biochemical work up revealed elevated urine and plasma metanephrines, consistent with pheochromocytoma. After multidisciplinary discussion with cardiothoracic surgery and anesthesiology, the recommendation was made to proceed with right adrenalectomy before mitral valve surgical intervention.

In preparation for adrenalectomy, the patient was alpha-blocked with doxazosin. During preoperative testing, an electrocardiogram revealed normal sinus rhythm. Transthoracic echocardiogram was difficult to interpret due to poor acoustic windows from prior pneumonectomy. Preoperative labs were unremarkable. A cardiac MRI revealed left ventricular ejection fraction (LVEF) 66%, right ventricular ejection fraction (RVEF) 48%, and a structurally abnormal mitral valve with resultant severe, eccentric MR (regurgitant volume 41 mL, regurgitant fraction 52%). Computer tomography (CT), confirmed right pneumonectomy and resultant severe right mediastinal shift (Figure 1,2). Pre-operative pulmonary function testing demonstrated an FVC of 2.8L (51%), FEV1 of 1.8L (42%), and DLCO of 52%.

The patient was scheduled to undergo transabdominal robotic right adrenalectomy in the left lateral decubitus position. A preinduction radial arterial catheter was placed and anesthesia was induced with hydromorphone, lidocaine, propofol and rocuronium. The trachea was intubated and mechanical ventilation was initiated. Anesthesia was maintained with isoflurane. After intubation, a central venous catheter was placed followed by placement of a pulmonary artery catheter and transesophageal echocardiogram (TEE). TEE findings confirmed severe MR with bileaflet prolapse resulting in multiple eccentrically directed mitral regurgitation jets with Coanda effect, and normal biventricular function (Figure 3). Surgical exposure was difficult due to a shift of intra-abdominal contents into the right thoracic cavity. The adrenal mass was more retro-hepatic and retro-caval than usual. After controlling and dividing the adrenal vein the right adrenal gland with a single 4cm mass was completely removed. Pathology confirmed the diagnosis of pheochromocytoma. SBP's ranged from 100 to 225 mmHg and were managed with titrated doses of esmolol, nicardipine, propofol, and phentolamine. Intake was 2L of crystalloid, 500mL of 5% albumin, and output included an estimated blood loss of 10mL and urine output of 100mL.

At the end of the procedure, the patient was extubated and transferred to the surgical intensive care unit with invasive monitors and no vasoactive agents. He was discharged home from the hospital on post-operative day 2 after an uncomplicated recovery. He ultimately returned to the hospital for his elective mitral valve surgery at a later date.

DISCUSSION

The care of a patient with a medical history of severe MR, prior pneumonectomy, and pheochromocytoma undergoing robotic right adrenalectomy is described. To our knowledge, there is only one published case of a patient with severe MR undergoing adrenalectomy for pheochromocytoma [3]. There is no published literature regarding minimally invasive adrenalectomy in a patient with a prior pneumonectomy. The clinical decision making on whether to intervene upon the mitral valve or perform adrenalectomy first is important and not described elsewhere. The hemodynamic changes associated with a present pheochromocytoma could have serious consequences during cardiac surgery and potentially have a dramatic effect on bleeding, risk of aortic dissection, or stroke risk due to hypertension associated with catecholamine release. Additionally, the degree of MR may improve after adrenalectomy due to improvements in hemodynamics since MR is worsened by increases in afterload.

Numerous perioperative considerations exist for the patient diagnosed with pheochromocytoma undergoing adrenalectomy. Phenoxybenzamine or other α-adrenergic antagonists, such as doxazosin, are recommended by the 2014 Endocrine Society consensus guidelines as the initial agents for pre-operative blood pressure management [7]. Importantly, at least 7-14 days of α-blockade should always precede treatment with a β-adrenergic antagonist to avoid a hypertensive crisis due to unopposed α-receptor stimulation [7]. Of note, doxazosin poses less risk of post-operative hypotension compared with phenoxybenzamine once the active tumor is removed due to a shorter half-life [8]. In this case, doxazosin was prescribed pre-operatively and the patient had no vasopressor requirements post adrenalectomy.

Knowledge of the various surgical approaches to adrenalectomy and the accompanying patient positioning requirements aids in effective perioperative planning. Adrenalectomy can be performed via laparotomy or laparoscopy/robotic. The advantages of the laparoscopic or robotic over the open approach include reduced catecholamine levels and improved intraoperative hemodynamic stability [9]. Laparoscopy is performed through either a transabdominal or retroperitoneal approach with positioning varying by approach [7]. The retroperitoneal approach is performed in a modified knee-chest, prone jackknife, or lateral decubitus position. The knee-chest position is known to have significant associated hemodynamic perturbations, including decreased preload, cardiac output, and mean arterial pressure versus the prone jack knife position, which is not associated with positioning related hemodynamic alterations [10,11]. Retroperitoneal insufflation is often better tolerated hemodynamically than peritoneal insufflation, but may require a higher insufflation pressure to achieve similar operating space, leading to higher incidence of hypercapnia [12]. In our case, the robotic transabdominal approach in the lateral decubitus position was chosen, not only

Table 1: Distribution of the study population based on age, sex, religion and marital status.				
Age	**Frequency**	**Percent %**	**Valid %**	**Cumulative %**
≤ 20 years	7	3.4	3.4	3.4
21-40 years	182	87.5	87.5	90.9
41-60 years	19	9.1	9.1	100.0
Total	208	100.0	100.0	
Mean age – 30.5 ± 8.1**, range** 18 – 57 years				
Sex				
Female	4	1.9	1.9	1.9
Male	204	98.1	98.1	100.0
Total	208	100.0	100.0	
Religion				
Christianity	76	36.5	36.5	36.5
Islam	132	63.5	63.5	100.0
Total	208	100.0	100.0	
Marital status				
Single	107	51.4	51.4	51.4
Married	101	48.6	48.6	100.0
Total	208	100.0	100.0	

Table 2: Distribution of the study population based on education, tribe and employment status.				
Educational level	**Frequency**	**Percent (%)**	**Valid (%)**	**Cumulative (%)**
Primary	16	7.7	7.7	7.7
Secondary	60	28.8	28.8	36.5
Tertiary	128	61.5	61.5	98.1
None	4	1.9	1.9	100.0
Total	208	100.0	100.0	
Tribe				
Yoruba	184	88.5	88.5	88.5
Igbo	7	3.4	3.4	91.9
Hausa	2	1.0	1.0	92.9
Fulani	3	1.4	1.4	94.3
Nupe	2	1.0	1.0	95.3
Ebira	2	1.0	1.0	96.3
Igala	3	1.4	1.4	97.7
Others	5	2.4	2.4	100.1
Total	208	100.1	100.1	
Employment status				
Employed	138	66.3	66.3	66.3
Not employed	70	33.7	33.7	100.0
Total	208	100.0	100.0	

for improved hemodynamic stability, but also the ability to perform TEE throughout the case.

A laparoscopic or robotic adrenalectomy can be a challenging endeavor in a patient with a prior pneumonectomy. Significant anatomic changes post pneumonectomy include mediastinal deviation toward the side of the lung resection, compensatory hyperinflation of the remaining lung, and ipsilateral hemidiaphragm elevation causing a shift of intra-abdominal contents in a cephalad direction [13]. In this case, the patient's liver had partially migrated into the right chest, complicating the surgical anatomy. Pneumoperitoneum associated with laparoscopic abdominal surgery can have several deleterious effects, especially in the setting of prior pneumonectomy. Alterations in respiratory physiology are disadvantageous by compression of the remaining lung, leading to decreased diaphragmatic excursion, compliance, and functional residual capacity (FRC) [14]. Hypercarbia is common with both retroperitoneal and peritoneal CO_2 based insufflation and hypercarbia management can be particularly challenging with one lung ventilation, as minute ventilation increases are often limited by protective ventilation strategies (4-6mL/kg tidal volume) as was employed in this patient [15]. Endotracheal tube

Table 3: Prevalence of p24 antigen.

P24		Frequency	Percent	Valid Percent (%)	Cumulative Percent (%)
	Negative	194	93.3	93.3	93.3
	Positive	14	6.7	6.7	6.7
	Total	208	100.0	100.0	100.0

Table 4: Proportion of types of blood donors in the study population.

		Frequency	Percent (%)	Valid Percent (%)	Cumulative Percent (%)
Valid	FRP	164	78.8	78.8	78.8
	VOL	17	8.2	8.2	87.0
	PD	27	13.0	13.0	100.0
	Total	208	100.0	100.0	

**FRP – family replacement donor, PD – paid donor, VOL- voluntary donor*

Table 5: Comparison of p24 antigen positivity with sociodemographic variables and other parameters of the study population.

Variables	Number (n)	(n) positive for P24 antigen	(%) positive for P24 antigen	(n) negative for P24 antigen	X^2	P value
Age Group						
< 20yrs	7	0	0	7	2.645	0.067
21-40yrs	182	14	6.7	168		
41-60yrs	19	0	0	19		
Sex						
Male	204	14	6.7	190	0.271	0.603
Female	4	0	0	4		
Marital Status						
Single	107	9	8.4	98	0.404	0.956
Married	101	5	5.0	96		
Religion						
Christianity	76	2	2.6	98	2.849	0.61
Islam	132	12	9.1	120		
Tribe						
Yoruba	184	14	7.6	170	1.929	0.197
Igbo	7	0	0	7		
Hausa	2	0	0	2		
Fulani	3	0	0	3		
Nupe	2	0	0	2		
Others	10	0	0	10		
Occupation						
Not Employed	58	6	10.3	52	13.792	0.841
Artisan	41	3	7.3	38		
Students	12	2	16.6	10		
Teaching	11	2	18.0	9		
Military	3	1	33.3	2		
Others	93	0	0	93		
Types of donor						
FRP	164	11	6.7	153	1.866	0.002
PD	27	3	11.1	24		
VOL	17	0	0	17		
Level of Education						
Primary	16	3	18.8	13	3.639	0.303
Secondary	60	2	3.3	58		
Tertiary	128	9	7.0.	119		
None	4	0	0	4		

** FRP – Family replacement donor, PD – Paid donor, VOL – Voluntary donor, X^2 – Chi-square, level of significance – P value < 0.05*

migration is a known complication of peritoneal insufflation causing advancement into an endobronchial position. Caution should be given to avoid this occurrence on the side of the previous pneumonectomy as catastrophic respiratory complications may ensue including hypoxia, hypercarbia, and ultimately respiratory arrest if unrecognized.

Specific precautions should also be taken when managing patients with cardiac disease for adrenalectomy. Pheochromocytomas can lead to severe hypertension and tachycardia with associated cardiovascular complications during surgical removal. Careful selection of invasive monitoring can assist in the early detection and management of life- threatening hemodynamic changes. In this case, pre-induction arterial cannulation was performed to detect acute blood pressure changes and a central venous catheter was placed for administration of vasoactive agents. TEE was utilized to evaluate intravascular volume status, MR severity, and cardiac function. A pulmonary artery catheter was placed for additional cardiac monitoring and assessment of filling pressures both intra- and post-operatively. For hypertensive episodes we used short-acting vasodilator and beta blocking agents, including esmolol, nicardipine, phentolamine, propofol and increased volatile anesthetic depth. Short-acting vasodilatory agents are prudent since hypotension often occurs after tumor removal due to catecholamine deficiency, volume depletion, and residual effects of preoperative α-blockade [1]. A few strategies were employed to manage these acute hemodynamic disturbances in the setting of severe MR. Euvolemia was maintained with fluid status assessment based on overall hemodynamic conditions, pulmonary artery filling pressures, and frequent TEE evaluation. Intraoperative SBP was maintained between 100-120 mmHg in anticipation of an acute hypertensive episode and peritoneal insufflation was initiated incrementally with a maximum intraabdominal pressure of 15 mmHg. Despite these precautions, the SBP rapidly increased above 200 mmHg four separate times resulting in elevated mean pulmonary artery pressures of 60 mmHg. Despite the increased systemic afterload during tumor manipulation, there was not a notable increase in the amount of MR on TEE. Each hypertensive episode was treated expeditiously with vasodilators, avoiding cardiac or respiratory decompensation. Norepinephrine infusion was utilized to maintain SBP>100 and the patient only had one brief (<5 min) period of hypotension (SBP<100) throughout the case. Norepinephrine was selected for its balanced alpha and beta receptor stimulation and physiologic similarity to tumor secretory product. Overall, the patient tolerated the procedure well with no cardiopulmonary complications.

We discuss the perioperative considerations of a patient with a pheochromocytoma, prior right pneumonectomy, and severe mitral valve regurgitation undergoing robotic right adrenalectomy. The various surgical approaches to an adrenalectomy carry individual intraoperative anesthetic considerations, particularly in a patient with pre-existing severe cardiopulmonary dysfunction. Furthermore, surgical anatomy and ventilator management is often complicated in patients with prior pneumonectomy. Pre-existing cardiac disease offers additional challenges in maintaining safe hemodynamic control during pheochromocytoma removal. This case illustrates the implementation of each of these considerations into the successful management of a complex patient undergoing pheochromocytoma removal.

DECLARATION OF CONFLICTING INTERESTS

Andreas C. Plackis declares that there is no conflict of interest.

Kelly L. Mishra has an investigator initiated industry funded research grant with Baxter that is unrelated to this publication. Kelly Mishra declares that there is no conflict of interest.

Amber N. Wesoloski declares that there is no conflict of interest.

Kara K. Siegrist declares that there is no conflict interest.

Carmen C. Solorzano declares that there is no conflict of interest.

Susan S. Eagle has developed a minimally-invasive volume status device that has been licensed via Vanderbilt University to Baxter Healthcare. This declaration has no influence on this manuscript.

EACH AUTHOR'S INDIVIDUAL CONTRIBUTION TO THE MANUSCRIPT

Andreas C. Plackis: This author contributed to the outline and performed a preliminary literature search for this manuscript. He composed the bulk of the initial rough draft and was the resident physician who carried out the anesthetic plan during the adrenalectomy surgery.

Kelly L. Mishra: This author helped make substantial contributions with regards to manuscript draft and obtaining images. She was the attending cardiac anesthesiologist who participated in the care of the patient during the eventual mitral valve repair surgery.

Amber N. Wesoloski: This author helped with correspondence in the submission of this manuscript as well as manuscript edits.

Kara K. Siegrist: This author helped with formatting and editing of the manuscript.

Carmen C. Solórzano: This author helped contribute to manuscript writing with regards to the surgical perspective of the case and performed the adrenalectomy on this patient.

Susan S. Eagle: This author helped make substantial contributions with regards to writing the manuscript and editing the final draft. She was the attending anesthesiologist responsible for anesthetic plan during adrenalectomy surgery.

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