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CASO CLÍNICO

Anaesthetic Approach for Mitral Valve Replacement in a Patient with a Pheochromocytoma

Abordagem Anestésica para Substituição de Válvula Mitral num Doente com um Feocromocitoma

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Afiliação
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Palavras-chave
Anestesia; Feocromocitoma; Prótese de Válvula Cardíaca; Válvula Mitral

ABSTRACT

Pheochromocytomas are rare neuroendocrine tumours that produce and store catecholamines. Intraoperatively, the release of excessive amounts of catecholamines can produce life-threatening cardiovascular complications.

This is a case report of a woman in her 60s recently diagnosed with a pheochromocytoma in need of a mitral valve replacement due to anterior leaflet prolapse and ruptured chord.

Due to the possibility of worsening mitral regurgitation and cardiovascular collapse after a hypertensive crisis during adrenalectomy we prioritised the cardiac surgery.

Intraoperatively, we performed a multimodal total intravenous anaesthesia with propofol and low-dose infusions of remifentanil and dexmedetomidine. Hemodynamic stability was maintained throughout the procedure and two months later the patient underwent a laparoscopic adrenalectomy uneventfully.

Although pheochromocytoma removal is usually a priority surgery, when facing serious cardiovascular pathology, such as severe mitral regurgitation, it may be a better option to prioritise the cardiac surgery over the pheochromocytoma removal.

RESUMO

Feocromocitomas são tumores neuroendócrinos raros que produzem e armazenam catecolaminas. No período intra-operatório, a libertação de quantidades excessivas de catecolaminas está associada a um elevado risco de complicações cardiovasculares potencialmente graves. Neste caso clínico, reportamos a abordagem peri-operatória de uma mulher de 68 anos, recentemente diagnosticada com um feocromocitoma, proposta para cirurgia de substituição da válvula mitral por prolapso do folheto anterior e ruptura de corda tendinosa. Devido à possibilidade de agravamento da insuficiência mitral e colapso cardiovascular na eventualidade de uma crise hipertensiva durante uma adrenalectomia, a cirurgia cardíaca foi priorizada. No intra-operatório, procedemos a uma anestesia endovenosa total multimodal com propofol e perfusões de remifentanil e dexmedetomidina em baixas doses. A cirurgia decorreu sem complicações, mantendo estabilidade hemodinâmica durante todo o procedimento. Posteriormente, dois meses mais tarde, a doente foi submetida a uma adrenalectomia laparoscópica sem intercorrências. Embora a remoção do feocromocitoma seja geralmente considerada uma cirurgia prioritária, perante uma patologia cardiovascular grave, como a regurgitação mitral severa, a inversão desta prioridade pode ser uma melhor opção.

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INTRODUCTION

Pheochromocytomas are catecholamine-secreting rare neuroendocrine tumours of the adrenal medulla.[1] Intraoperatively, the release of excessive amounts of catecholamines can produce life-threatening cardiovascular complications.[2] In most cases, this diagnosis is considered a surgical priority but there are some case reports in which cardiac surgery was prioritised due to the severity of the cardiovascular pathology.[3,7] In these situations, anaesthesiologists must be prepared to manage the perioperative implications of a pheochromocytoma in major surgery. Multimodal anaesthesia may play a role for success.

CASE REPORT

This is a case report of a woman in her 60s, American Society of Anesthesiologists (ASA) IV, recently diagnosed with a pheochromocytoma in need of a mitral valve replacement (MVR) due to anterior leaflet prolapse and ruptured chord.

The pheochromocytoma was diagnosed previously during hospitalization for the diagnostic workup of a deep vein

thrombosis. Abdominal computed tomography (CT) showed a hypervascular tumour with marked enhancement after IV administration of contrast material. The initial biochemical tests included plasma and 24-hour urine fractionated metanephrines and catecholamines. The plasma fractionated levels were 0.438 nmol/L of metanephrines (normal value < 0.5 nmol/L) and 2.003 nmol/L of normetanephrines (normal value < 0.9 nmol/L) and 24-hour urine levels were 602 µg of metanephrine (normal value < 180 µg), 4166 µg of normetanephrine (normal value < 390 µg) and 1135.7 µg of noradrenaline (normal value < 80 µg). A PET/CT and a metaiodobenzylguanidine (MIBG) scan showed increased uptake of a lesion in the left adrenal gland (Fig. 1).

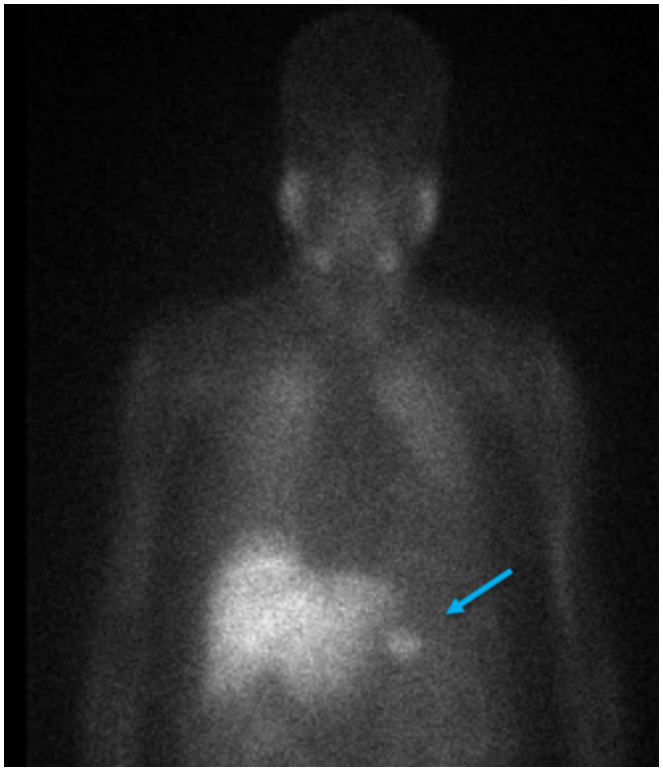

Figure 1. MIBG scan. The arrow shows the increased uptake in the left adrenal gland

Her past medical history included degenerative mitral valve disease with moderate mitral insufficiency and preserved left ventricular ejection fraction. She also suffered from systemic arterial hypertension, pulmonary hypertension, multinodular goiter with hypothyroidism and asthma. She had no known allergies and no coronary lesions diagnosed in a recent cardiac catheterization.

During medical preparation for adrenalectomy, she presented with an acute chord rupture of the mitral valve anterior leaflet with severe mitral regurgitation and NYHA IV heart failure with preserved ejection fraction and pulmonary artery systolic pressure (PSAP) of 50 mmHg. This posed a discussion regarding which surgery should be prioritised since the pheochromocytoma removal would now present as a much riskier procedure due to acute heart failure. For this reason, a multidisciplinary meeting with anaesthesiology, cardiology, cardiac surgery, general surgery and nephrology was held and the adrenalectomy was postponed for after the cardiac surgery. By this time, α blockade with phenoxybenzamine 10 mg twice a day was initiated.

Other than the previous imaging exams, the preoperative assessment included an electrocardiogram (ECG) which showed normal sinus rhythm and a chest radiography with no pathologic findings.

After 18 days of α blockade, during which the arterial pressure remained under normal values (systolic pressure under 120 mmHg), MVR with a mechanical valve via median sternotomy was planned under cardiopulmonary bypass and general anaesthesia.

Intraoperatively, standard monitoring included a five-lead ECG, non-invasive blood pressure, end-tidal partial pressure of carbon dioxide and peripheral pulse oximetry. In addition, we measured invasive arterial pressure in the right radial artery and central venous pressure through a deep venous catheter in the right jugular vein. To monitor depth of anaesthesia we used the Bispectral Index™ (BIS™) monitoring system and for cerebral oxygenation near-infrared spectroscopy (NIRS). We used intraoperative transesophageal echocardiography (TEE) throughout the procedure for real-time assessment of cardiac structure and function.

We performed a multimodal total intravenous anaesthesia with propofol, remifentanil and dexmedetomidine. Before induction we administered 2 mg of midazolam and an arterial line was placed in the right radial artery for invasive arterial pressure monitoring. The total intravenous anesthesia was then started with infusions of propofol at 6 mg/kg/h, remifentanil at 0.4 µg/kg/min and dexmedetomidine at 0.4 µg/kg/h. The trachea was easily intubated with a 7.0 orotracheal tube with resource to direct laryngoscopy after a bolus of rocuronium of 0.6 mg/kg.

After about half an hour of surgery, a noradrenalin infusion was started for maintenance of systolic arterial pressure above 100 mmHg, although there was no repercussion on cerebral oxygenation monitoring.

Cardiopulmonary bypass (CPB) was initiated one hour and a half following induction when the activated clotting time (ACT) was above 400 after administration of 20 000 units of unfractioned heparin.

According to the anesthetic depth monitoring, infusion rates were lowered during CPB to 3 mg/kg/h of propofol, 0.2 µg/kg/min of remifentanil and noradrenaline was also reduced to 0.1 µg/kg/min. An infusion of milrinone was initiated at 0.5 µg/kg/min three hours later. CPB lasted for three hours during which NIRS values did not drop below baseline and no complications were observed. During weaning of CPB an adrenalin infusion was started at 0.1 µg/kg/min

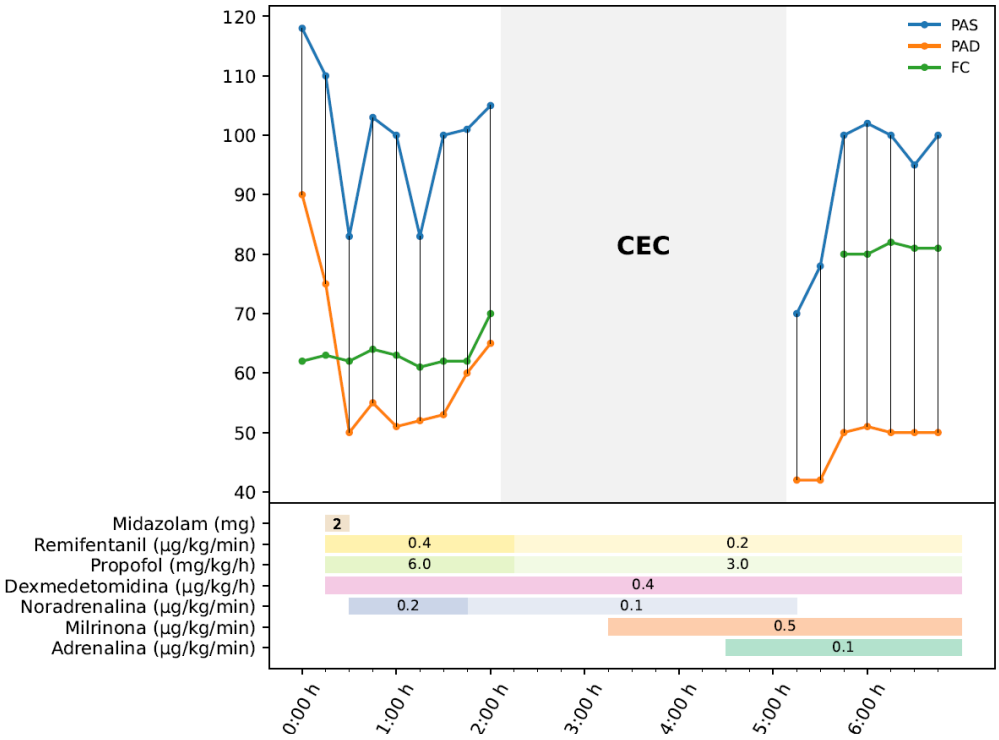

Figure 2. Part of the anesthesia record, focusing on the anesthetic agents and hemodynamic stability maintenance during the MVR

and noradrenalin was stopped (Fig. 2). Postoperatively, the patient was transferred to the intensive care unit (ICU) under mechanical ventilation, where she remained for two days. During the first day, inotropic and vasopressor support was maintained with noradrenalin at 0.05 µg/kg/min, adrenalin at 0.02 µg/kg/min and milrinone at 0.1 µg/kg/min, being progressively reduced and stopped during the second day.

The trachea was uneventfully extubated about 24 hours after the surgery.

After being discharged from the ICU, the patient stayed in the cardiothoracic ward for 9 days, during which time there were no complications to report.

This stay was somewhat prolonged for stabilization of the α-blockade which was continued postoperatively with phenoxybenzamine 10 mg per day. By the time of discharge, the patient's fatigue was getting better and her peripheral edema had significantly decreased.

The transthoracic ultrasonography revealed reduced right ventricular function related to a recent cardiopulmonary bypass with a PSAP of 30 mmHg and preserved left ventricular systolic function with a normally functioning mechanical mitral valve. Postoperative anticoagulation was achieved with warfarin.

Two months later the patient underwent a laparoscopic adrenalectomy uneventfully. During this surgery there was need of vasopressor support with noradrenalin after removal of the tumour which was maintained during the early postoperative period and suspended 48 hours later.

The patient was discharged one week later without complications.

DISCUSSION

Surgical resection is the only curative treatment for pheochromocytomas.[8] Surgery itself, however, has been found to result in measurable catecholamine release associated with the manipulation of the tumour. This leads to significant hemodynamic perturbations[1] and potentially lethal cardiovascular morbidity[6] with profound hypertension, bradycardia and tachyarrythmias.[9]

This already risky procedure will have additional considerations in the presence of acute mitral regurgitation and cardiac failure. On the other hand, few case studies in the literature address the anaesthetic management of cardiac surgery and CPB in patients with a diagnosed pheochromocytoma.

In our case, we held a multidisciplinary meeting in order to evaluate the risk of each intervention and decide which surgery to prioritise.

The possibility of a combined surgical procedure, with both surgeries done sequentially, was considered. Regarding a combined strategy with adrenalectomy followed by immediate MVR, the main concern was the risk of distributive and cardiogenic shock once the pheochromocytoma was removed. This option was considered too risky due to the expected myocardial stunning and vasoplegia after cardiopulmonary bypass (CPB).

On the contrary, as for a combined procedure with the MVR as the first surgery, CPB and systemic anticoagulation in the setting of a pheochromocytoma present serious considerations. The risk of catecholamine surge establishing bypass, potential retroperitoneal haemorrhage from the

tumour after the CPB-required anticoagulation and induced coagulopathy and severe hypertension and tachycardia after weaning from CPB[6] were considered to be too high.

Therefore, in order to avoid the additional risks of combining both surgeries, we decided to do them separately.

We considered the main risk to be the possibility of worsening mitral regurgitation and cardiovascular collapse after a hypertensive crisis during adrenalectomy and prioritised the MVR.

For prevention of cardiovascular complications, treatment with antihypertensive drugs before pheochromocytoma removal is recommended and α-adrenergic receptor blockers are considered the first choice.[8]

Although there are no guidelines on preoperative management of patients with pheochromocytoma who are going to be submitted to cardiac surgery, it seems prudent to manage them in a similar way. In our case, 18 days of presurgical administration of phenoxybenzamine were completed and no reflex tachycardia ensued, without need for a beta-adrenergic receptor blocker.

Regarding the intraoperative course of the MVR, we adopted a multimodal anaesthesia approach with different anaesthetic agents. The goal of this technique is to use the important synergistic effects of different drugs to reduce the dosing requirement for anaesthetics that affect hemodynamic stability such as propofol, and to spare consumption of opioids. This might be especially important during cardiac surgery, which by itself is a hemodynamic challenge due to mechanical impairment of cardiac function.[10]

In our case, this approach was successful and hemodynamic stability was maintained throughout the procedure with propofol, remifentanil and dexmedetomidine and only low-dose infusions of inotropic and vasopressor drugs.

CONTRIBUTORSHIP STATEMENT / *DECLARAÇÃO DE CONTRIBUIÇÃO*

FP e AR: Conceção, desenho do estudo; aquisição, análise e interpretação dos dados; redação.

Aprovação da versão final a ser publicada.

FP AND AR: Conception, study design; acquisition, analysis and interpretation of data; essay.

Approval of the final version to be published.

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