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Anaesthetic management of cardiac phaeochromocytoma: A case series

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ABSTRACT

INTRODUCTION: Primary cardiac phaeochromocytoma is uncommon, with few anaesthetists encountering this rare pathology in clinical practice. Further, there is little information available on the detailed intraoperative and postoperative haemodynamics and principles of the anaesthetic management of this condition.

PRESENTATION OF CASE: We present a retrospective, single-centre case series of four patients with cardiac phaeochromocytoma who presented for surgical excision. We describe the perioperative evaluation and management of these patients, consideration of the requirements for cardiopulmonary bypass, and the analgesic and pharmacologic interventions needed to maintain stable perioperative and intraoperative haemodynamics.

DISCUSSION: Octreotide scintigraphy, in addition to echocardiography, cardiac MRI and coronary angiography proved vital in the preoperative evaluation of these patients. Preoperative anaesthetic management of cardiac phaeochromocytoma involved alpha-adrenergic blockade, judicious beta-adrenergic blockade and hydration. Intraoperatively, the administration of vasodilatory agents prior to, and vasoconstricting agents with volume therapy after tumour excision, were the key elements of anaesthetic management. Furthermore, we believe that cardiopulmonary bypass plays a pertinent role in cardiac phaeochromocytoma excision and that the risks and benefits of pulmonary artery catheters should be considered before use in these patients.

CONCLUSION: Management of cardiac phaeochromocytoma is complex and demands careful perioperative planning and management. Perioperative morbidity is common and anaethetists play an important role in achieving a successful outcome for patients who present for excision of cardiac phaeochromocytoma.

1. Introduction

Phaeochromocytoma is a catecholamine-producing tumour arising from chromaffin-positive cells of the sympathetic nervous system. This tumour can produce considerable amounts of catecholamines - primarily noradrenaline, resulting in an extreme hyper-sympathetic state, with deleterious haemodynamic consequences. Whilst the majority of phaeochromocytomas are found in the adrenal glands, aberrant collections of chromaffin tissue may be the site for the development of phaeochromocytoma [1]. Less than 2% occur within the chest, mostly in the posterior mediastinum, whereas cardiac phaeochromocytoma of the middle mediastinum are rare [2–4].

Primary cardiac phaeochromocytoma is uncommon and many anaesthetists will never encounter this pathology. Besterman et al. reported the first case of primary cardiac phaeochromocytoma in 1974, with subsequent cases reported over the following years [5–8]. However, there is little information available on the specific anaesthetic considerations when managing these complex cases. Therefore, we present a case series of cardiac phaeochromocytoma describing the perioperative management, requirements for cardiopulmonary bypass and the pharmacological interventions used to maintain stable intraoperative haemodynamics. This work has been reported in line with the PROCESS criteria [9].

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Table 1
Anatomical and coronary angiography findings in four patients diagnosed with primary phaeochromocytoma.

	Case 1	Case 2	Case 3	Case 4
Age(years), gender	17, Male	35, Female	19, Male	44, Male
Location and size of tumour(s)	Anterior wall of the aorta and outflow tract of the right ventricle (4.5 x 3.0 cm)	Left atrium (4.6 x 4.9 cm)	Lateral aspect of the aortic root (6.0 x 3.0 cm)	Exterior surface of right atrioventricular groove (8.0 x 5.5 cm), and the inferior border of right pulmonary artery (6.0 x 4.0 cm)
Tumour vascular supply on coronary angiography	Right coronary artery, which was completely obstructed by the tumour at its root	Circumflex branch of left coronary artery	Left internal mammary artery and sinoatrial nodal artery (branch of the right coronary artery)	Right coronary artery and circumflex branch of left coronary artery. Incidental finding of 80% stenosis to the mid left anterior descending coronary artery

2. Presentation of case

Between 2006 and 2012, four patients with cardiac phaeochromocytoma presented to our university teaching hospital. All patients presented with symptoms of excessive sweating, palpitations and dizziness. The patient in Case 1 additionally reported intermittent chest discomfort. Clinical examination of all patients revealed resting tachycardia, hypertension and postural hypotension. Markedly elevated levels of 24-hour urinary total catecholamines, vanillylmandelic acid, and metanephrines were found. Octreotide scintigraphy (Tc-99m-Oct) diagnosed cardiac phaeochromocytoma in three of four cases, which was subsequently confirmed by echocardiography and cardiac magnetic resonance imaging (MRI). Cardiac MRI diagnosed cardiac phaeochromocytoma in Case 4. Preoperative coronary angiography was then performed to evaluate the vascular supply of the tumours. Anatomical and coronary angiography findings for all four patients are presented in Table 1. In Case 2, it was noted that vascularisation to the tumour was supplied from the circumflex branch of the left coronary artery. The need to ligate a portion of the circumflex branch of the left coronary artery distal to the tumour and the subsequent risk of postoperative myocardial ischaemia was anticipated, allowing for careful intraoperative planning. Coronary angiography in Case 3 verified that the internal mammary artery and sinoatrial nodal artery were supplying the tumour. Tumours supplied by the internal mammary artery may adhere to the pericardium and thus, during pericardial manipulation, haemodynamic fluctuation was also anticipated.

Preoperative management over 4-weeks involved alpha and beta-adrenergic blockade with phenoxybenzamine and metoprolol. Upon normalisation of blood pressure and heart rate, elective median sternotomy followed, performed under general anaesthesia with moderate hypothermic cardiopulmonary bypass (CPB) (Table 2). Monitoring included 5-lead electrocardiography, invasive central venous and blood pressure monitoring, pulse oximetry, bispectral index and bladder and nasopharyngeal temperatures. Anaesthesia was induced with midazolam (3–5 mg), fentanyl (5–10 u g/kg), propofol (1–2 mg/kg) and pancuronium (0.1 mg/kg). Anaesthesia was maintained with 1 MAC of isoflurane with a FiO_2 of 0.5. Intraoperative analgesia included boluses of fentanyl (200ug). Muscle relaxation was maintained with boluses of pancuronium.

A summary of the surgical approach, cardiopulmonary bypass (CPB) requirements and aortic cross-clamp times for each patient are summarised in Table 2. Endotracheal intubation, skin incision and sternotomy did not provoke significant haemodynamic fluctuation. However, in Case 1, during dissection of the tumour prior to aortic cannulation, blood pressure rose abruptly to 170/100 mmHg (Fig. 1). This was stabilised with esmolol (0.5 mg/kg) and fentanyl (300ug). In Case 3, blood pressure rose to 200/100 mmHg with a sudden sinus tachycardia of 110 bpm during manipulation of the pericardium. This was stabilised with phentolamine (4 mg) (Fig. 2). An image of the cardiac tumour visualised in Case 3 is shown in Fig. 3.

Modest hypothermia between 33–34 degrees Celsius was maintained for all patients during CPB. Each patient was uneventfully separated from CPB with continuous noradrenaline (1–10 ug/min) and dopamine infusions (1–3 u g/kg/min). Fluid intervention in Cases 1–3 included lactated Ringer's solution (1500–2500 ml), colloid (500–1500 ml), residual CPB blood (500–1000 ml), and additional packed red blood cells (560–1100 ml). Fresh frozen plasma (15 ml/kg) was required in all cases to correct a medical coagulopathy. In Case 4, significant and major active bleeding from the right pulmonary artery was identified, which was surgically managed with 4.0 prolene sutures, primary closure and packing. Haemostasis was achieved after almost four hours, necessitating 12 units of packed red blood cells and 1600 ml of cell saved blood. This large volume resuscitation with blood resulted in a subsequent dilutional coagulopathy, compounded by hypothermia, necessitating

Table 2
Summary of the surgical approach, cardiopulmonary bypass (CPB) requirements and aortic cross-clamp times.

	Case 1	Case 2	Case 3	Case 4
Surgical approach	Median sternotomy, extracapsular dissection of the tumour initiated at its junction with the muscle of the right ventricular outflow tract. The right coronary sinus of the aorta and part of the pulmonary artery was excised and reconstructed with bioprosthetic patches for complete tumour removal.	Median sternotomy, tumour on the roof of the left atrium dissected, supplying circumflex branch of the left coronary artery distal to tumour ligated.	Median sternotomy, tumour identified at aortic root extending into right atrioventricular groove and excised. The right coronary artery was unintentionally divided at its first segment during tumour excision and a vein graft was used to bypass the right coronary artery.	Median sternotomy, tumours to the exterior surface of the right atrioventricular groove and the inferior border of the right pulmonary artery were identified and excised. The left anterior descending coronary artery was bypassed with the left internal mammary artery.
CPB duration	143 min	80 min	93 min	134 min
Aortic cross clamping time	90 min	35 min	50 min	91 min
Total duration of surgery	280 min	200 min	310 min	600 min