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CASE REPORT

Phaeochromocytoma and Paraganglioma Excision Involving the Great Vessels

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Objective/background: Phaeochromocytomas and paragangliomas are vascular neuroendocrine tumours distributed between the neck and the pelvis and may be associated with catecholamine secretion. The aim of the study was to describe the complex surgical management required to excise these tumours when in close proximity to the great vessels (aorta and vena cava).

Methods: This was a retrospective case series. Patients included those undergoing surgical excision of a phaeochromocytoma or paraganglioma involving the great vessels. Data on clinical presentation; genetic mutations; tumour location; catecholamine/metanephrine secretion; surgical strategy; pre-, intra-, and post-operative course were collated.

Results: Five patients (age range 16–60 years) were identified; three had thoracic paragangliomas located under the arch of the aorta, one had an abdominal paraganglioma invading the aorta, and one had a massive phaeochromocytoma invading the inferior vena cava via the adrenal vein. Three patients had predisposing germline mutations. All patients had adrenergic blockade prior to surgery. A diverse range of complex surgical techniques were employed to excise tumours, including cardiopulmonary bypass, aortic resection, grafting and venotomy of the vena cava. Early post-operative complications were limited.

Conclusions: Excision of phaeochromocytomas and paragangliomas involving the great vessels is high risk surgery optimally undertaken within a multidisciplinary setting in a tertiary referral centre. Comprehensive radiological and biochemical assessment, meticulous pre-operative preparation and close intra- and post-operative monitoring are essential. Radiological imaging may be unable to resolve the tumour extent and anatomy pre-operatively and direct visualisation of the tumour may be the only way to clarify the surgical strategy. Pre-operative knowledge of the genetic predisposition may influence surgical management.

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INTRODUCTION

Phaeochromocytomas are chromaffin cell neuroendocrine tumours of the adrenal medulla. Paragangliomas (PGLs) are extra-adrenal tumours of sympathetic (secretory) or parasympathetic (mainly non-secretory) origin, located between the base of the skull and the pelvis. These highly vascular tumours are, on occasion, positioned in close proximity to the great vessels (aorta and vena cava). Tumour related morbidity and mortality results from associated catecholamine excess; hypertensive crises; cardiovascular sequelae; mass effect and metastatic disease. Approximately 30% of tumours are associated with germline mutations, 30% are associated with somatic mutations, and the remainder appear sporadic.[1,2] Malignancy rates for those with phaeochromocytoma or sympathetic PGLs are estimated to be 17%, although rates are higher with certain germline mutations, for example succinate dehydrogenase B (*SDHB*).[3]

The mainstay of therapy is surgical excision of tumours. When a tumour abuts or invades a great vessel, surgical decision making can be complex. The extent of tumour involvement in surrounding structures; symptoms; comorbidities; local surgical expertise; availability of alternative treatments; and patient preference all need consideration. Non-resectable disease is treated with radionucleotide therapy or chemotherapy.[4] Experience in managing five patients who underwent surgical excision of either a phaeochromocytoma or a PGL involving the great vessels is presented.

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CASE SERIES

Five patients over a 10 year period (2004—13), from a single institution, were identified as having surgical excision of either a phaeochromocytoma or a PGL involving the great vessels (aorta or vena cava). All five had pre-operative alpha- and beta-adrenergic blockade with phenoxybenzamine and propranolol. A summary of presentations and clinical characteristics is given in Table 1, intra-operative surgical data in Table 2, and radiological imaging from cases 2—5 is shown in Fig. 1.

Thoracic PGLs

Three patients had thoracic PGLs (patients 1, 2, and 5). Patient 2 had an *SDHB* mutation and patients 5 an *SDHA* mutation. Aortic transection and cardiopulmonary bypass was required to access the tumours for excision. A median sternotomy was followed by the establishment of cardiopulmonary bypass with aortic and bicaval cannulation and aortic cross-clamping (Fig. 2). Cardioplegia was then established. Following tumour excision, the aorta was re-anastomosed, unclamped, and the patients were taken off cardiopulmonary bypass.

Abdominal PGL invading aorta

Patient 3, an *SDHB* mutation carrier, required excision of the abdominal PGL with resection of part of the infrarenal abdominal aorta, which had been invaded by tumour. The aorta was cross-clamped and then transected. A 16 × 8 mm bifurcated Dacron graft was inserted. An additional nodule beneath but separate from the PGL was also excised and confirmed on histological examination to be a lymph node metastasis.

Phaeochromocytoma invading inferior vena cava

Patient 4, on computed tomographic imaging, was found to have a 8.3 × 9.5 cm ill defined heterogeneous right adrenal mass with a necrotic centre invading the inferior vena cava (IVC) superiorly and displacing the right kidney inferiorly (Fig. 1E, F). Twenty-four hour metanephrines (normetadrenaline and 3-methoxytyramine) were markedly elevated. The metaiodobenzylguanidine (MIBG) scan showed avidity in the tumour but no evidence of distant metastasis. A magnetic resonance scan was able to delineate some of the feeder vessels to the tumour and confirm the IVC invasion in more detail. A MAG 3 renogram demonstrated equal differential function of right and left kidney. It was unclear whether the tumour should be debulked or fully resected. Given the potential requirement for hepatic and IVC resections, surgery was carried out jointly by hepatobiliary and vascular surgeons. Pre-operative coiling was considered but not used as it was unlikely to significantly alter the magnitude of the proposed operations. Endovascular measures such as balloons or stents were precluded owing to the intracaval extension. Direct inspection of the tumour at the time of surgery revealed that the retroperitoneal phaeochromocytoma was infiltrating and displacing the right kidney downwards. It lay behind the right lobe of the liver but did not infiltrate it. A palpable nodule was felt within the IVC and a large number of

Table 1. Patient demographics and disease characteristics.

Patient	Age (y)	Reason for investigation	Germline mutation	Site of phaeo/PGL	Size (cm)	Great vessel involvement	MIBG imaging	FDG PET imaging	Hypertension	Catecholamines 24 h urinary/nmol/24 h			Metanephrines 24h urinary/nmol/24h		
										< 560	< 144	< 3,194	< 2,000	< 4,400	< 2,500
1	60	Hypertension, sweating, and tachycardia	Not done	Thoracic	4.8 × 5.1	Under aortic arch	Avid	—	Yes	3,677	< 30	2,501	—	—	—
2	40	Familial SDHB surveillance	SDHB	Thoracic	5.1 × 3.9	Under aortic arch	Non-avid	Avid	No	356	< 30	2,909	—	—	—
3	16	Familial SDHB surveillance	SDHB	Abdominal	6.5 × 5.4	Invading abdominal aorta	Avid	Avid	No	2,863	57	3,287	—	—	—
4	50	Anaemia investigations	Nil	Right adrenal	12.6 × 7.2	Invading IVC	Avid	—	No	—	—	—	41,481	718	10,011
5	49	Chest pain and exertional shortness of breath	SDHA	Thoracic	5.5 × 3.5	Under aortic arch	Avid	Avid	Yes	—	—	—	11,625	509	15,306

Note. PGL = paraganglioma; MIBG = metaiodobenzylguanidine; FDG PET = fludeoxyglucose positron emission tomography; SDHB = succinate dehydrogenase B; IVC = inferior vena cava; SDHA = succinate dehydrogenase A.