

Excision of phaeochromocytoma and paraganglioma involving the great vessels

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Objective

- Phaeochromocytomas and paragangliomas occur across a wide distribution from the base of the skull to the pelvis, and may lie in close proximity to or involve the great vessels including the aorta and vena cava.
- The aim of the study was to describe the surgical management of phaeochromocytomas and paragangliomas involving the great vessels.

Design

- Retrospective case-series.

Patients

- 5 subjects undergoing surgical excision of either a phaeochromocytoma or paraganglioma involving the great vessels.
- Subjects were seen at St Bartholomew's Hospital, London between 2004 and 2013.

Results

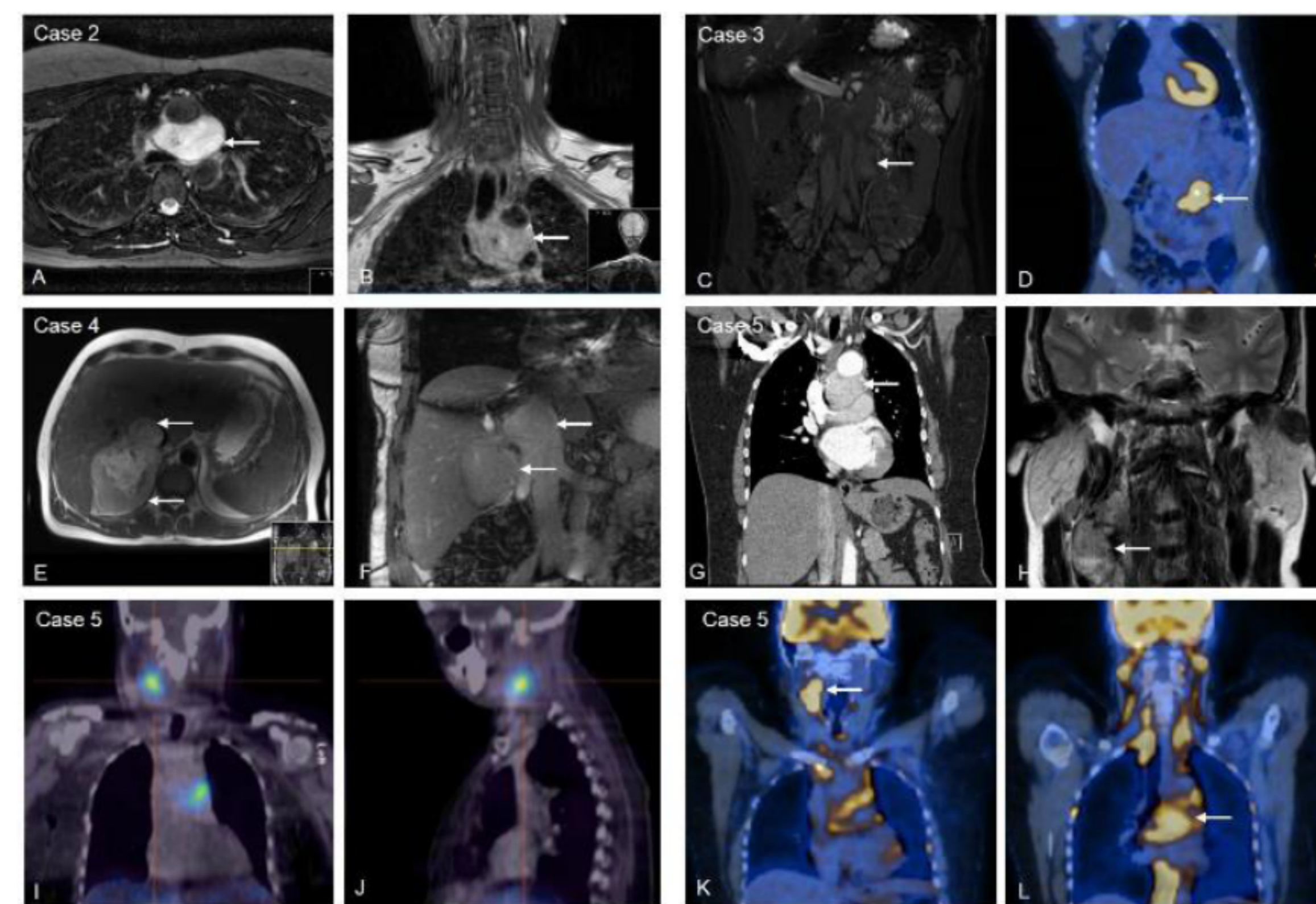
- Five subjects (age range 16-60 years)
- Three subjects had thoracic paragangliomas located under the arch of the aorta.
- 1 subject had an abdominal paraganglioma in which preoperative imaging was unable to delineate that the tumour was invading the aorta.
- 1 subject had a massive phaeochromocytoma invading the IVC.
- Three of the 4 subjects tested had predisposing germline mutations
 - 2 subjects with *SDHB* mutations
 - one subject with an *SDHA* mutation

Results

- All subjects had alpha and beta adrenergic blockade prior to surgery.
- Thoracic paragangliomas were excised following cardiopulmonary bypass and aortic transection to access the tumours.
- Abdominal paraganglioma invading the aorta was resected with part of the aorta to clear disease + insertion of an aortic Dacron graft.
- Phaeochromocytoma invading the IVC - resected en bloc with the right kidney. Intra-operatively it was possible to resect the tumour nodule invading the IVC excised via a venotomy and vena cava repair.

Outcomes

- All subjects made a good recovery from surgery.
- One subject had early post-operative complication managed conservatively.
- One subject required adjuvant radiolabelled MIBG therapy for residual disease.



Subject	Lesion	Surgery	Surgeon	Duration (*)	Max BP (systolic) mmHg	Minimum BP (systolic) mmHg	Anti-hypertensive agents	Vasopressor	Fluid input	Blood loss	Complications
1	Thoracic paraganglioma	Transsection aorta Cardio-pul bypass	Cardiothoracic	6h (2h)	250	61	GTN Esmolol Phentolamine SNP	-	Colloid 1.5 L RCC 2u FFP 2u Pit 1u	484ml	Atrial fibrillation, Pleural haemorrhage, Wound sepsis, Acute kidney injury
2	Thoracic paraganglioma	Transsection aorta Cardio-pul bypass	Cardiothoracic	4h 50 (1h 35min)	135	82	GTN Metoprolol Phentolamine	-	Colloid 1 L RCC 2u FFP 2u Pit 1u	270ml	-
3	Abdominal paraganglioma	Transsection + resection aorta Dacron graft	Endocrine Vascular	6h 10min (50 min)	165	88	SNP Labetolol	Adrenaline Noradrenaline	Colloid 7 L RCC 2u FFP 2u Pit 1u	450ml	-
4	Phaeochromocytoma	Venotomy, excision tumour, IVC repair Right nephrectomy	Hepatobiliary Vascular	3h 45min (22 min)	140	80	GTN Labetolol Metoprolol	Noradrenaline	Colloid 4 L RCC 4u (CS 720ml) FFP 2u Pit 1u	2200ml	-
5	Thoracic paraganglioma	Transsection aorta Cardio-pul bypass	Cardiothoracic	6h (2h 40min)	178	62	GTN Labetolol Phentolamine	Noradrenaline	Colloid 2.5 L RCC 2u FFP 2u Cryo 2u Pit 2u	664ml	-

Subject	Age	Germline mutation	Lesion	Size/cm	Great vessel	Great vessel involvement	MIBG imaging	PDS PET maging	Hypertension	Catecholamines 24h urinary/ nmol/24h	Metanephrines 24h urinary/ nmol/24h
										<560 <144 <3194	<2000 <4400 <2500
1	60	Not done	Thoracic	4.8 x 5.1	Aorta	Under arch	Avid	-	Yes	3677 <30 2501	- - -
2	40	SDHB	Thoracic	5.1 x 3.9	Aorta	Under arch	Non-avid	Avid	No	356 <30 2909	- - -
3	16	SDHB	Abdominal	6.5 x 5.4	Aorta	Invading	Avid	Avid	No	2863 57 3287	- - -
4	50	Nil	Phaeochromocytoma	12.6 x 7.2	IVC	Invading	Avid	-	No	- - -	41481 718 10011
5	49	SDHA	Thoracic	5.5 x 3.5	Aorta	Under arch	Avid	Avid	Yes	- - -	11626 509 15306

Conclusions

- Excision of phaeochromocytomas and paragangliomas involving the great vessels is high-risk surgery and should be undertaken in a tertiary referral centre within a multidisciplinary setting. Subjects require comprehensive radiological and biochemical assessment.
- Meticulous pre-operative preparation and appropriate intra- and post-operative back-up are essential.
- In some cases radiological imaging is unable to resolve the tumour anatomy and extent 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.

