Excision of phaeochromocytoma and paraganglioma involving the great vessels

St Bartholomew’s Hospital, London, UK

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 germine mutations
  • 2 subjects with SDHB mutations
  • one subject with an SDHA mutation

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.