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Case Report

The Transplant Surgeon: An Unlikely, Yet Suitably Qualified, Member of The Complex Neuro Endocrine Multi-Disciplinary Team

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ABSTRACT

Curative surgery for retro-peritoneal tumours involving vascular structures is challenging and multi-visceral resection is often required to obtain clear resection margins. Abdominal transplant surgeons have considerable experience in all aspects of visceral, vascular and retro-peritoneal surgery. Application of these skills to resect tumours involving vascular structures, and re-implant organs to preserve function is unique. We present the case of a 15-year old girl with a complex retro-peritoneal tumour which was resected *en-bloc* with the kidneys and vena-cava followed by auto-transplantation of the left kidney. Seven years later, the patient represented with a recurrent tumour which was successfully excised in its entirety. We discuss how innovative surgical strategies can be performed safely on an individualized basis. We highlight the importance of balancing the benefits of the 'technically possible procedure' with its risks, along with consideration of the outcomes of treatment and non-treatment alike.

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Introduction

Retroperitoneal tumours are uncommon and complex pathologies which often involve adjacent viscera and vascular structures. Attainment of clear margins during surgical resection is a key determinant of outcomes [1]. Surgical resection with curative intent therefore presents significant technical challenges which may require multi-visceral resection and, on occasions, complex vascular resection and reconstruction. Such procedures often require multi-specialty teams of surgeons for whom knowledge of, and operative experience within, the retroperitoneum is a routine part of clinical practice. The skill set of a transplant surgeon is unique amongst the contemporary general surgical sub-specialties as it inherently requires both competence and confidence in dealing with the gastro-intestinal, vascular and urological systems. Organ retrieval and transplantation provides unparalleled training and caseload in complex procedures which require extensive exposure of the entire retroperitoneum. The aim of this case report is to review the role of the transplant surgeon as part of the multi-disciplinary team (MDT) managing complex retroperitoneal tumours.

Case Report

A 15-year-old girl presented to the paediatric department with a one month history of headache, nausea, vomiting, and weight loss. She also experienced intermittent episodes of hypertension which caused fluctuations in her renal function. Ultrasound revealed a significantly smaller right kidney compared with the left. Computed Tomography identified a dumbbell shaped tumour in the retroperitoneal space which appeared to involve the renal arteries bilaterally and inferior vena cava (IVC) (Figure 1). Subsequent angiography demonstrated significant stenosis of the IVC at the level of the tumour and significant stenosis of both the left and right renal arteries, with poor arterial perfusion of the right kidney (Figure 2). This was further confirmed on DMSA scan which revealed a differential uptake of 1% in the right kidney and 99% on the left. Urinary catecholamines were marginally elevated. These findings, together with marginally elevated urinary catecholamine levels, were initially suggestive of a ganglioneuroma or ganglioneuroblastoma as the likely pathology.

A MDT panel, including a transplant surgeon, was convened to consider the operative options. A standard approach towards curative resection

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of the tumour would in all likelihood have rendered this young patient dialysis dependent. This was due to the tumour encroaching and stenosing the origin and most proximal two centimetres of the left renal artery. However, an *en bloc* resection followed by auto-transplantation of the left kidney provided the best option which balanced the risks and benefits of curative treatment and preservation of native renal function. An initial attempt at surgery was abandoned due to a hypertensive crisis upon induction of anaesthesia. A second successful attempt at surgery was made after further investigation and complete alpha blockade as in preparation for pheochromocytoma surgery. The tumour was excised along with the both kidneys and a section of the IVC. The left kidney was carefully removed, dividing the renal artery beyond the stenotic artery and perfused with cold perfusion fluid. It was then preserved at 4°C while the IVC was reconstructed and the retroperitoneum fully cleared of tumour and hemostasis achieved. The left kidney was then anastomosed end-to-side to the external iliac vessels on the right side without a carrel patch as in a standard kidney transplant. She made an excellent post-operative recovery with normalization of her renal function. Histological examination confirmed a paraganglioma.

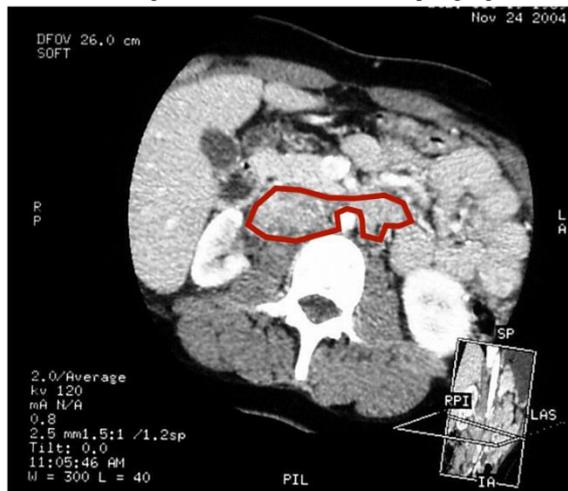


Figure 1: Abdominal CT scan showing a dumbbell shaped tumour (marked) in the retroperitoneal space.

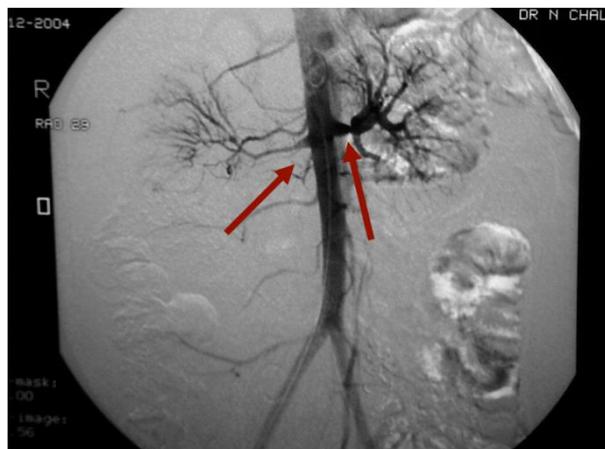


Figure 2: Angiography demonstrating bilateral stenosis of the renal arteries with poor perfusion to right kidney

Unfortunately, upon routine follow-up imaging after 5 Years, a 3cm left adrenal mass was demonstrated by a Magnetic Resonance scan. This was associated with elevated plasma normetadrenaline (3.77nmol/L) and

normal plasma metadrenaline (0.26nmol/L). Further consideration by the MDT panel agreed that, owing to her deteriorating renal function (eGFR 20mL/min) chemotherapy was not a tenable treatment option. Further surgery could render her dialysis dependent, with the need for an allogenic kidney transplant. However, surgical resection of the recurrent tumour offered the patient her only curative treatment option. At the third operation, by the transplant team, the use of a gamma probe ensured that the left adrenal mass and all residual tumour from the aorto-caval window was excised (Figure 3) with clear resection margins and no tumour recurrence to date. Her renal function also remains stable.

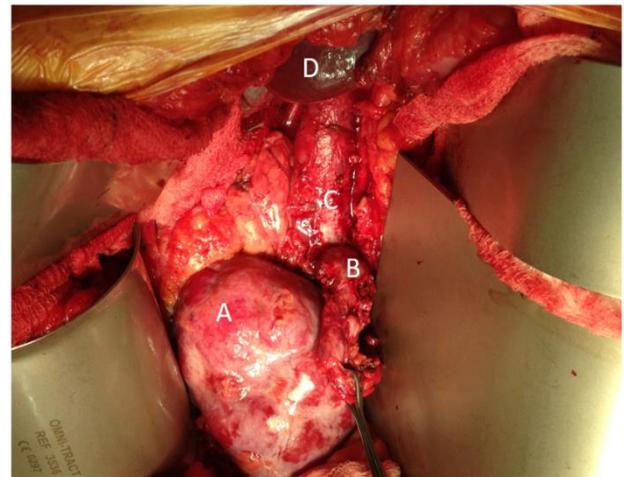


Figure 3: Appearance on second laparotomy. A) Auto-transplanted kidney; B) Recurrent tumour; C) Inferior Vena Cava; D) Liver

Discussion

Paragangliomas are rare tumours arising along the autonomic nervous system in the retroperitoneum and affect less than 1 in 2500 of the population [2]. Surgical resection offers the only curative option for retroperitoneal tumours such as paraganglioma [2]. Multi-visceral resection is frequently performed for complex retro-peritoneal tumours, and over 80% of cases require the resection of 3 or more organs [2]. Post-operative mortality is four times higher in patients requiring resection of 3 or more organs compared with 0 organs (HR 4.11 [1.88-9.00] p<0.001) [1].

However, despite the significant risks associated with procedures of this magnitude, vascular involvement and infiltration by retroperitoneal tumours need not represent a blanket contraindication to *en-bloc* oncological resection [3]. Survival in patients requiring vascular resection for oncological clearance exceeds 90% at 2 years after surgery [3]. Meanwhile, in non-malignant disease, resection and reconstruction of the IVC could result in up to 62% graft patency at 3 years [4].

A MDT panel review represents the minimum standard required of a contemporary surgical practice. The involvement of a transplant surgeon in the complex tumour MDT represents a willingness to consider innovative solutions for complex patients in an open and accountable environment. Moreover, the natural centralization of such cases in high volume transplant centres means that critical care facilities and tertiary support from associated specialties are immediately available.

The application of the transplant skill set to complex *unresectable* disease is not limited to tumours affecting the kidneys. *Ante-situm*, in-situ hypothermic perfusion and ex-vivo tumour resection are all familiar techniques to the liver transplant surgeon and which can be utilised as part of the surgeons' armamentarium to successfully excise hepatobiliary tumours involving the IVC [5]. Long-term patient survival following such complex surgery must be benchmarked against the known outcomes of advanced malignant hepatobiliary disease.

This case demonstrates the importance of flexibility when considering the most appropriate clinical therapies for complex patients. Membership of the MDT by individuals with specialist skills and interests outwith the *parent specialty* provides a unique opportunity to consider the best treatment options for selected patients. The role of the MDT is to contextualize treatment recommendations according to individual patient priorities. Potential benefits of the 'technically possible procedure' are balanced with its risks, along with consideration of the outcomes of treatment and non-treatment alike. Whilst the MDT has become, in recent years, a standard bearer of quality clinical practice, it is vitally important that patient receives personalized care in the process-driven pursuit of evidence based medicine.

Patient consent

Obtained

Conflicts of Interest

None

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