

En-bloc resection of IVC and abdominal aorta as a part of the surgical management of a symptomatic myoepithelial tumour

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Background: Retroperitoneal sarcomas present significant technical challenges to surgical treatment. Close proximity to the major vascular structures is a common cause of incomplete resection and subsequent recurrence/continued symptoms. We describe a patient who presented such a challenge, in whom we undertook successful treatment.

Clinical problem and management: A young male presented with back pain in 2007 to the spinal surgeons at a University teaching hospital. Cross-sectional imaging revealed extensive tumour arising from L2/3 intervertebral foramen and L1/2 foramen, extensive tumour lying anterior to the vertebral bodies in the retroperitoneal space in the upper half of lumbar spine, encasing both the aorta and IVC, compressing but not occluding them. A working diagnosis of paraganglioma was made and functional studies were undertaken and the lesion was declared non-secretory (urine VMA, plasma metanephrines and metadrenaline were normal). A combined team of vascular, GI, orthopaedic surgeons with a spinal interest and neurosurgeons embarked on a resection (in 2007) at which the spinal component of the lesion was excised and a firm posterior instrumented stabilisation was achieved. The tumour was found to be arising from the region of the L2 nerve root ganglion. An anterior approach was made thru a midline laparotomy and the tumour was found adherent to the IVC, renal veins and aorta. After preliminary dissection and subsequent massive blood loss due to an IVC tear the procedure was abandoned. Frozen sections from the tumour were inconclusive. The patient made a good post-operative recovery and went home 2 weeks later. Post-op histological and immunological assessment (Prof Fletcher, Harvard) suggested a diagnosis of a soft tissue myoepithelioma, with no features of malignancy.

In view of continued disabling symptoms he was referred to our unit for consideration of surgical resection. Restaging with cross-sectional imaging confirmed some increase in size of the neoplasm. Split renal function assessment was performed in addition. Our operative strategy was to resect the tumour en-bloc along with the major vessels. A left nephrectomy was performed in a planned manner during the initial approach to the tumour. In conjunction with the vascular team, the aorta was resected from the level of the R renal artery to the common femoral arteries bi-laterally, and replaced with a Dacron trouser graft. The IVC was resected from the level of the R renal vein to the common iliac veins bi-laterally and reconstructed with a 'panel' graft, using bovine pericardium. Operating time was 6hrs and blood loss was 750mls. ICU stay was 3days and his post-operative recovery was uneventful. He was discharged following a hospital stay of 3 weeks. Histopathological examination confirmed complete resection of the lesion which was again confirmed to be a soft-tissue myoepithelial tumour.

Conclusion: Extreme vascular resection and reconstruction in the treatment of retroperitoneal sarcomas is feasible when undertaken by experienced surgical teams. Early referral to specialized units is to be encouraged and will benefit patients and improve outcome for them.