The role of Oncovascular Surgery in Primary Retroperitoneal Sarcomas

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Abstract:

Retroperitoneal soft tissue sarcomas are a rare clinical entity, with an annual incidence of less than 0,5 in 100.000. Surgical resection is the main therapeutic option for patient with favourable characteristics, with radiotherapy and neoadjuvant chemotherapy being utilised pre-operatively to optimise the tumour characteristics and are not often employed as stand-alone treatment. R0 resection is the most essential factor for achieving a long-term recurrence-free survival. Due to their close anatomic relations and their infiltrative character towards the abdominal vessels, en bloc resection of these tumors often requires a complex vascular reconstruction. Lack of standardised techniques or guidelines only adds to the complexity. After review of the recent literature, we summarize important information about the pathogenesis, diagnosis and treatment of retroperitoneal soft tissue sarcomas. We additionally analyse strategies employed by oncovascular surgeons that both preserve the vascular supply to the intraabdominal organs and lower extremities, after en block resection of the tumour along with infiltrated vessels. These include vessel ligation, or repair after partial or total resection. These strategies aid in the achievement of an optimal oncological outcome.

INTRODUCTION

Retroperitoneal soft tissue sarcomas (RSTSs) are rare malignant tumours arising from mesenchymal cells. RSTS account for 15% of all soft tissue sarcomas, with those affecting the extremities being most common. RSTSs are characterized by large variability in histologic type, with liposarcomas being the most prominent subtype, making up 1% of all malignant cancers in the adult population, followed by leiomyosarcomas[1,2]. According to the fifth edition of the WHO Classification of Tumours of Soft Tissue and Bone, the histological subtypes of liposarcomas include the atypical lipomatous tumour (ALT), also known as well-differentiated liposarcoma (WDLPS), dedifferentiated liposarcomas (DDLPS), myxoid liposarcomas (MLPS), pleomorphic liposarcomas (PLPS), and myxoid pleomorphic liposarcomas[3], with each subtype having its distinct diagnostic and biologic features. Mean patient age at diagnosis ranges between 59-61 years[4] and can either be an incidental finding or have symptoms due to extrinsic compression to other structures. Surgical resection is the only option offering the possibility of cure and recurrence-free survival. However, there seems to be distinct lack of official recommendations regarding the surgical management of retroperitoneal tumours involving the intraabdominal vessels, partly due to their heterogeneity, and partly due to vascular infiltration, which has been a criterion of non-resectability.

We performed a review study encountering the PubMed medical Database. The search strategy incorporated the following keywords: “primary retroperitoneal sarcomas”, “retroperitoneal sarcomas”, “oncovascular” ,“arterial”, “venous”, “vascular”, “reconstruction”, “repair” and combinations of them. Retrieved records were subsequently screened for relevance to the topic of the study. Cross reference was carried out to retrieve relevant studies. Our literature review (summarized in the Table) underscores the rarity yet clinical significance of retroperitoneal sarcomas resections along with the infiltrated vessels and combined with vascular reconstructions. We retrieved 9 case reports and 1 case series, including a total of 17 patients.

PATHOGENESIS

Mesenchymal tumours account for about 50% of all primary retroperitoneal tumours[5]. Due to their anatomical position, RSTSs often have no accompanying symptoms, thus remaining undetected and having grown to considerable size when diagnosed. In cases of sarcomas with large diameters, an increase in abdominal circumference and a palpable mass can be the first signs of RSTS. Other symptomatology can affect the digestive tract, the urinary tract (renal impairment, ureter obstruction due to extrinsic compression or infiltration), the vascular network (extrinsic compression, infiltration or malignant thrombus formation leading to phlebitis, edema secondary to partial or total occlusion), as well as nerve compression[6,7].

Another consideration is the tendency to arise in narrow proximity to vascular structures, making RSTSs historically

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Table. Demographic, surgical and follow-up data of 17 patients with retroperitoneal sarcomas, who underwent resection and simultaneous vascular reconstruction

Date of Publication	Authors	Age, gender	Histological identity, involved structures	Pre-operative treatments	Resection plan	Reconstruction strategy	Recovery
April 2025	Kotsis et al. [24]	63yo (M)	Myxofibrosarcoma involving the IVC	Pre-op radioRX and chemotherapy (doxorubicin - ifosfamide) à partial radiographic response	R0 resection (En bloc resection of the sarcoma, the IVC, the right kidney and psoas-quadriceps muscle portions)	IVC replacement with PTFE tube graft	
Jan 2024	Bael et al. [30]	42yo (F)	Leiomyosarcoma involving the IVC in close proximity with the left kidney		R0 resection with nephrectomy	IVC reconstruction with a Dacron graft	Old hematoma drainage on 3rd post-op week. RT from 4th post-op month on
Jul 2023	Castro et al. [31]	48yo (F)	Well-differentiated liposarcoma		En bloc resection, along with the entire retrohepatic IVC	No reconstruction	Freer of disease, attending follow-ups
Dec 2021	Hasegawa et al. [22]	72yo (M)	Soft tissue high-grade sarcoma occluding the celiac trunk and the origin of the SMA		Radical tumour resection after SMA reconstruction	Saphenous vein graft to reconstruct the SMA, aortic defect reconstructed with Dacron patch, arterial bypass for the common hepatic artery with ringed PTFE graft (intraoperatively without pulse)	Local recurrence at patch site on 6th post-op month. Received chemotherapy, patient alive w/o recurrence at 6th post-op year
Nov 2021	Tirnavean et al. [32]	74yo	Leiomyosarcoma arising in the IVC near the renal ostia		En bloc resection	Reconstruction of the IVC with autologous graft harvested from the right superficial femoral vein	2 years of recurrence-free survival
March 2021	Kontopodis N et al. [35]	62yo (M)	Locally recurrent myxoid liposarcoma involving the CIV common iliac vein and distal IVC		Resection with removal of both CIVs and infrarenal IVC	reconstruction with a bifurcated Dacron graft	No post-op complication, patient discharged on 11th post-op day
Nov 2020	Quildrian et al. [29]	54yo (F)	Low-grade sarcoma with infrarenal aorta involvement		En-bloc resection with resection of left psoas muscle, left nephrectomy, partial aorta resection	Reconstruction with bi-iliac PTFE graft	Urgent thrombectomy and re-anastomosis on post-op day 0, patient alive and w/o recurrence 2 years post-op
Aug 2020	Grieff et al. [27]	63yo (M)	Leiomyosarcoma	Neoadjuvant chemotherapy with doxorubicin and dacarbazine	En-bloc resection of aorta, IVC, tumour and right kidney	Combined aortocaval reconstruction with Dacron Gelsoft tube graft for the aorta and temporary veno-venous (femoral vein to internal jugular) bypass, followed by a ringed GORE tex graft for the IVC reconstruction	1 year recurrence-free survival

2020	Homsy et al.[7]	57yo (F)	Leiomyosarcoma	No	Marginal resection with resection of the infrarenal aorta and IVC	Reconstruction of aorta with polyethylene prosthesis, IVC with bovine pericardium patch and temporary axillobirenal bypass	IVC thrombosis at 33 months
		62yo (F)	Metastatic Leiomyosarcoma without vascular invasion		Marginal resection with resection of aorta, IVC and left renal vein, as well as resection of the right kidney	Reconstruction of the aorta with polyethylene Y graft, IVC and left renal vein with homograft vein	IVC compressed by recurrent tumour at 13 months and eventually patient died to the neoplasm at post-operative month 15
		56yo (F)	Leiomyosarcoma	Pre-operative embolization of tumor-feeding arteries	Marginal resection with resection of the left iliac vein, sigmoid colon, and left nephrectomy	Reconstruction of the left iliac vein with autologous vein graft	Thrombectomy and stent at post-op day 5, patient died at post-op month 22
		26yo (M)	Leiomyosarcoma , involving the aorta, SMA, hepatic artery and right renal artery		Marginal resection of tumour with pancreatectomy, splenectomy and left nephrectomy	Reconstruction with polyethylene	Disease-free at 25th month post-op
		58yo (F)	Leiomyosarcoma involving the IVC		Marginal resection of tumour	Homograft vein reconstruction	Disease-free at 6th post-op year
		61yo (F)	Leiomyosarcoma involving the IVC and left renal vein		Wide resection of tumour and right nephrectomy	IVC reconstruction with homograft vein and renal vein with autologous vein graft	Living with sarcoma at 6th post-op year
		59yo (F)	Sclerosing liposarcoma involving the IIA, EIA, IIV, EIV		Marginal resection of tumour with hysterectomy	Reconstruction using homograft vein	Local recurrent tumour compressing the IVC at 4 months. Dead 1 year after her operation
		32yo (M)	Angiosarcoma epithelioeides arising from the aorta, involving the IVC and the celiac trunk		Resection with right nephrectomy	Reconstruction of the aorta, SMA, celiac trunk, left renal vein and both internal iliac arteries with polyethylene prosthesis, IVC with bovine pericardium. Additionally temporary axillo-renal bypass	Disease-free 2 years after
May 2020	Yokohama et al.[11]	59yo (F)	Dedifferentiated liposarcoma	Neoadjuvant chemotherapy with doxorubicin and ifosfamide. Femoro-femoral arterial bypass before laparotomy	Resection with right common iliac artery and vein resection, right nephrectomy, and resection of right psoas muscle and femoral nerve	Femoro-femoral bypass	Patient free of recurrence at 1 year post-op

Abbreviations: M: Male, F: Female, EIA: external iliac artery, EIV: external iliac vein, IIA: internal iliac artery, IIV: internal iliac vein, IVC: inferior vena cava, pre-op: preoperative, post-op: post-operative, PTFE: polytetrafluoroethylene, RT: radiotherapy, SMA: superior mesenteric artery, yo: years old.

challenging to resect. A R0 excision requires wide dissection and removal of portions from the surrounding organs or muscles, and that often includes the large intra-abdominal vessels. Additionally, local recurrence is more frequent in patients who have undergone surgical resection of a primary RSTP compared to patients with extremity STS[8]. Thus, the need for wide resection while simmultaneusly keeping the vascular integrity of the abdominal organs has made the vascular surgeon an integral part of the multidisciplinary team for RSTS patients, giving rise to the field of oncovascular surgery.

DIAGNOSIS

As stated earlier, RSTSs can be found incidentally during imaging for other complaints, otherwise they can grow into a large size before being diagnosed. Experts agree that computational tomography (CT) is the gold standard for the diagnosis of RSTSs (Figure 1,2). This is especially true for tumours invading the Inferior Vena Cava (IVC)[9]. Magnetic resonance imaging (MRI) can be supplementary and is thought to better detect vessel wall invasion. Due to the infiltrative nature of RSTSs, some researchers mention the MRI tail sign as a guide

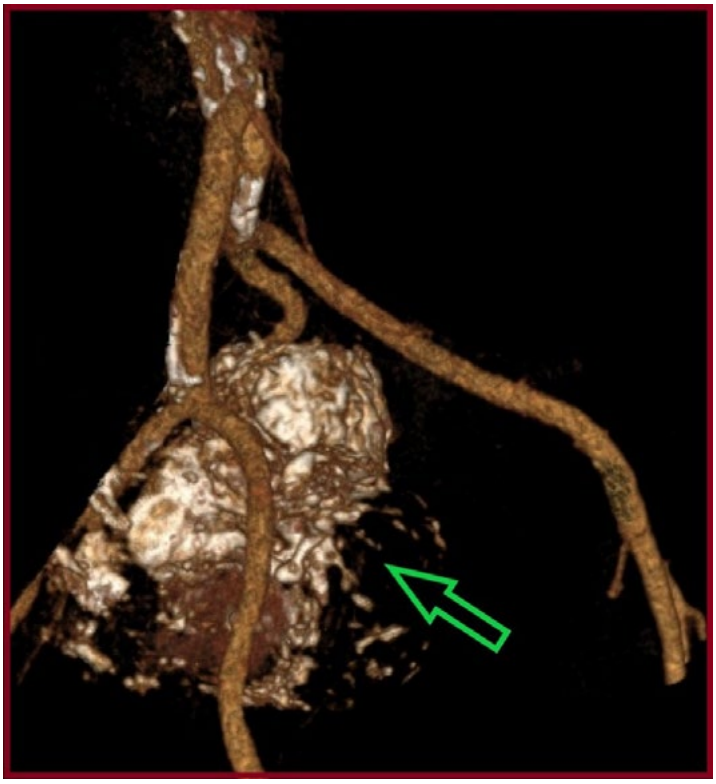

Figure 1: A retroperitoneal liposarcoma (arrow) infiltrating the right external iliac vein, surgically resected along with the vein.

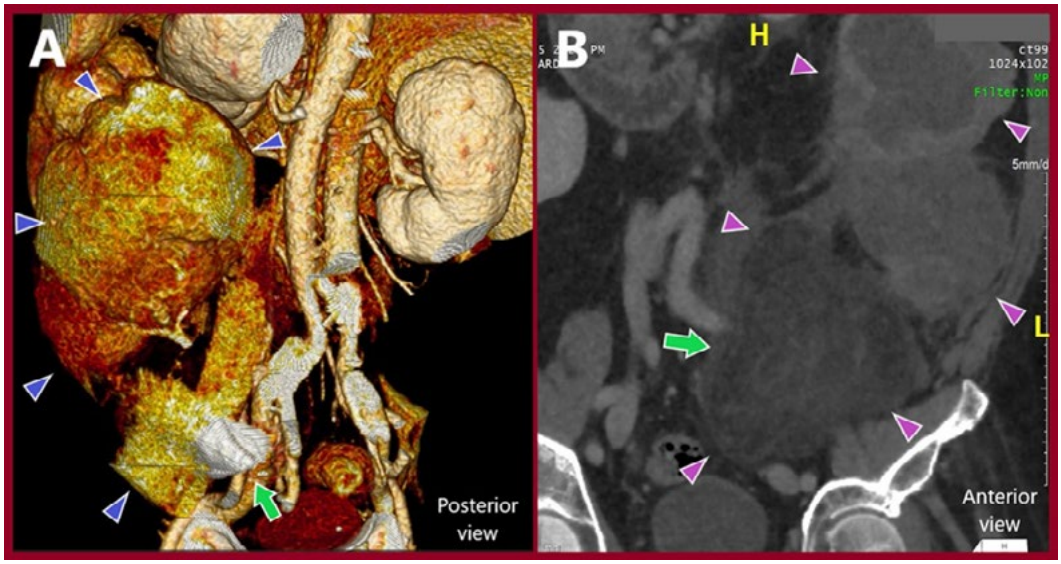

Figure 2: A large retroperitoneal sarcoma (arrowheads) encasing partially the iliac vessels (arrow), A) 3D-Reconstruction and B) Multiplanar Reconstruction of a Computed Tomography Angiography (Coronal view).

for planning the anatomical margins of resection[10]. Additionally, Magnetic Resonance Venography (MRV) can differentiate between ordinary or malignant thrombi and assist in mapping the extent of intraluminal thrombus, aiming to ultimately limit the intraoperative tumour dissemination, discussed later[8,9].

As with every oncologic surgery, a pre-operative biopsy should always be available, since a variety of tumours other than sarcomas can result in a retroperitoneal mass and which are optimally managed with modalities other than resection. A biopsy can only be omitted only in the rare case of a WDLPS or DDLPS[8.9]. For tumors invading the IVC, a CT-guided core needle biopsy should be preferred[9]. An MRI-guided biopsy can be obtained from the least well-differentiated area of the tumor, using specific advanced protocols[8]. A retroperitoneal approach is usually preferred to limit tumor dissemination and avoid damage to endoperitoneal organs. An open or minimally invasive surgical biopsy should be avoided for the same reason[8].

TREATMENT

A multidisciplinary team should summarize imaging and pathology results, as well as patient parameters and discuss the treatment plan. Neoadjuvant chemotherapy and radiotherapy (RT) are recommended for intermediate and high-grade lesions to mitigate the risk of local and distant recurrence and improve surgical outcomes. Recent guidelines advise for use of neoadjuvant systemic therapy in cases high risk for metastases or as a means of downstaging to allow resection[11]. However, it is not recommended for low-grade RSTSs and results are generally not hopeful[12, 13-17]. Radiotherapy has been shown to aid the control of local recurrence in patients undergoing en bloc resection with wide margins, but not in the overall survival of these patients. There is a tendency to extend overall survival but without conclusive results[12,18, 19]. Thus, neoadjuvant RT should be employed in patients with a high risk of local recurrence and should not be considered as a standard in the treatment in RSTS patients. RT toxicity is an additional factor that should be taken into account. Ultimately, the decision for administration of neoadjuvant therapy is made by the oncologist, depending on the performance status of the patient.

There are different strategies depending on the vascular and non-vascular structures involved when planning the surgical resection of RSTSs. Conversely, the involvement of the celiac trunk, the superior mesenteric artery, portal vein, invasive extension into the right atrium and infiltration of multiple organs such as pancreas and liver are criteria of non-resectability[20]. Nonetheless, recent advances in technology and technical skills have allowed progress to be made regarding tumour infiltrating the large intraabdominal vessels up to the level of the juxtarenal aorta.

Surgical therapy has two distinct goals: the first is to resect the tumour with a wide margin, macroscopically free of infiltrations, and the second is to minimize blood loss and avoid circulatory compromise. In cases of inferior vena cava (IVC) infiltration, IVC ligation can be considered in tumours lower than the renal ostia or tumours causing external obstruction of the IVC, providing that adequate collateral circulation has been established[9]. If less than 50% of the lumen is obstructed, or in the case that the suprarenal IVC is involved, then the IVC should be repaired. Repair by primary suture is not realisable, since the tumours often encase the IVC in a considerable length, also considering the need for a wide resection margin, creating distance between the two ends. There is also the possibility of partial vascular resection, with such an approach mainly utilised for veins due to their compliance. In this case, repair of venotomy can be achieved by primary suture, a biologic or a synthetic patch[21].

In cases of primary sarcomas of the aorta or of DDLPSs involving the infrarenal aorta, the involved segment is excised and replaced by a synthetic graft. If the juxta- or supra-renal aorta is involved, the complexity of repair increases due to the need to reimplant the renal arteries, the superior mesenteric artery, and the celiac trunk[21,22]. Homsy et al.[7] report several cases of aortic resection in which reconstruction was successful and who succumbed to causes other than graft failure, proving that, despite the high-risk of complications, the benefit from an R0 resection should be weighed against it [23].

When it comes to graft repair, polytetrafluoroethylene (PTFE) grafts are the first choice. Autologous grafts can be utilized when infection is a concern, such when interventions on the intestines are needed[7]. However, they are being abandoned as they seem to compromise the oncological outcome[24]. Ringed PTFE grafts of smaller diameter than the original IVC seem to yield the best results, offering more support and resisting collapse to extrinsic pressure, thus maintaining adequate flow. This also lowers the risk of in-graft thrombosis, as graft flow is increased[25,24,26].

Regarding in-graft flow and thrombus formation, there are several tools that several surgeons have used intraoperatively. The use of intraoperative doppler can confirm that in-graft flow is sufficient[24]. Likewise, transesophageal echocardiography (TEE) is used intraoperatively when it is suspected that an existing malignant thrombus extends to the supradiaphragmatic IVC[9]. Some authors have used an IVC filter to avoid pulmonary embolism due to malignant emboli or the creation of an arteriovenous fistula after IVC resection[6]. However, others do not recommend such practice, supporting that careful dissection of the IVC and good proximal and distal control are sufficient[9]. This requires mapping of thrombus extent, achieved via pre-operative MRI, intraoperative ultrasound and if necessary, TEE.

For tumors invading or surrounding the common, internal and/or external iliac arteries (CIAs, IIAs, EIAs respectively), often both arterial and venous reconstruction is required, owing to their close proximity[27]. The IIA can be ligated without limb compromise, while the CIA and EIA can be reconstructed with synthetic or biologic grafts[28,21,29,30,31,32,23]. In case of concomitant bowel resection, an extra-anatomical bypass might be considered[21]. The contralateral femoral artery or vein can be utilized as an autologous graft should biologic grafts be unavailable[28,21]. Should the graft pass over the inguinal ligament, a muscle flap can be constructed to protect it[33] Similar to the

IVC, the iliac veins can be reconstructed by biologic or PTFE graft or by an autologous contralateral femoral vein graft. Contrary to the IVC, smaller diameters yield a lower flow, thus lowering patency rates. Contralateral IIA transposition for reconstruction of the CIA is also described in patients with microscopic tumour infiltration[34]. Common and internal iliac veins can be safely resected, when infiltrated, accounting for the adequate collateral vessels draining the extremity. In any strategy it is important to avoid external compression of the grafts. Kontopodis N et al. [35] used a bifurcated graft to reconstruct the right common iliac vein, passing the left limb over the native right common iliac artery to avoid exactly that.

As extended resection of multiple organs is required to achieve R0 resection, timing of each one is crucial to the outcome. Kotsis et al. opted to first tackle the repair of the IVC and then followed the oncological resection of the tumour[24]. Although this logic allows for an ideal field for the tumour excision, it is not completely without risk of graft compromise.

Anticoagulation therapy is lifelong in the cases of graft repair. There are no established guidelines or a clear consensus. Pneumatic compression devices as well as prompt anticoagulation therapy in the immediate post-operative period is the optimal combination. There is the benefit of improving the flow rate in the reconstructed IVC by improving venous return[24]. Post-operative follow-up should include routine CT-angiography, starting in the 1st postoperative month to assess in-graft flow and thrombus formation, with the possibility to titrate the anticoagulation therapy further[9]. Upon unifocal recurrence, reintervention with complete excision is warranted, whereas in multifocal recurrence intervention is unlikely to be radical and has palliative character[37].

CONCLUSION

RSTSs pose a complex challenge to the oncologic surgeon, often requiring extended vascular resections and replacement of the large intra-abdominal vessels with grafts in order to achieve the R0 resection. In these techniques sometimes there is absence of specific training. To achieve best results for RSTS patients, the presence of an experienced vascular surgeon during such operations is of paramount importance to aid in prolonging the overall survival of patients.

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