Resection of the Inferior Vena Cava for Retroperitoneal Sarcoma: Six Cases and a Review of Literature

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Abstract

Resection of the inferior vena cava (IVC) for malignancy is a technically demanding procedure. We present a series of six cases of resection of the IVC for retroperitoneal sarcomas, four of which were primary caval tumors. We outline the technical difficulties faced in these complex procedures and discuss the oncological outcomes of these rare tumors. We performed a retrospective review of six patients operated for retroperitoneal masses involving the inferior vena cava between April 2015 and July 2016 at our tertiary care institute. Six patients underwent resection of the IVC, three of which required a multivisceral resection. An artificial prosthesis was used to reconstruct the IVC in three patients, whereas two patients underwent primary repair of the vein wall. One patient did not require any reconstruction. Margins were microscopically positive in two out of six patients. All patients received radiotherapy, either in the neo-adjuvant or adjuvant setting. Two patients developed local recurrences with a median follow-up of 24.5 months. Resection of the IVC for extirpation of retroperitoneal sarcomas is a technically complex and difficult procedure. The availability of a multidisciplinary team of surgeons and state-of-the-art intensive care support is essential for good outcomes.

Keywords Inferior vena cava · Leiomyosarcoma · Retroperitoneal sarcoma · Prosthetic graft · Reconstruction

Abbreviations

IVC Inferior vena cava
PTFE Polytetrafluoroethylene
ePTFE Expanded PTFE

Introduction

Resection of the inferior vena cava (IVC) for malignancy is a technically demanding procedure which may be required for retroperitoneal tumors involving the vein, or tumors arising primarily from the caval wall. In the latter setting, the tumor is usually a leiomyosarcoma, arising from the smooth muscle in the vein wall [1]. There are multiple methods of reconstruction

that have been described, including primary repair, patch grafting, prosthetic graft replacement, and replacement with an allograft [2]. Resection of the infra-renal segment without reconstruction has also been reported in literature [3]. We present our report of six inferior IVC resections performed for retroperitoneal sarcomas, four of which were primary caval tumors.

Materials and Methods

A total of 37 patients underwent resection of retroperitoneal sarcomas between April 2015 and July 2016 at our institute. We performed a retrospective review of six patients operated for retroperitoneal masses involving the IVC and requiring a partial or circumferential resection of the IVC in this period of time. There were four female patients and two males with a median age of 35.5 years (age range from 21 to 57 years). Demographic details, symptoms at presentation, and surgical details are summarized in Table 1.

The data was obtained from a prospectively maintained database and the histological and radiological details were obtained from the electronic medical records of the patients.

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Table 1 Patient and surgical characteristics

Patient	Age	Gender	Symptoms	IVC segment involved	Veno-venous bypass used	IVC reconstruction	Multivisceral resection
1	43	Female	Pain in abdomen, pedal edema	Inter-renal	No	Yes. PTFE graft	No
2	36	Female	Pain in abdomen	Inter-renal	Yes	Yes. PTFE graft	Yes. Right nephrectomy
3	35	Female	Palpable abdominal lump, abdominal pain	Inter-renal	No	No	No
4	57	Male	Pain in abdomen	Inter- and infra-renal	No	No	Yes. Right nephrectomy
5	35	Female	Incidentally detected	Retro- and infra-hepatic	No	No	Yes. Right nephrectomy, cholecystectomy with liver wedge
6	21	Male	Palpable abdominal lump	Infra-renal up to bifurcation	No	Yes. Dacron Y-graft	No

IVC inferior vena cava, PTFE polytetrafluoroethylene

Results

All patients except one were symptomatic at the time of presentation to the hospital (Table 1). The most common symptom was vague abdominal pain for which the patient had been investigated elsewhere and been detected to have a retroperitoneal mass. Two of these patients had received some form of neo-adjuvant treatment elsewhere before presenting to our institute. The indications for this were not known.

A contrast-enhanced computed tomography (CT) scan was performed for all the patients to characterize the retroperitoneal mass and determine the extent and relation of the tumor to adjacent organs and blood vessels (Fig. 1). A CT thorax was performed for all patients to rule out distant metastases. Reformatted images were utilized to determine the exact extent of involvement of the inferior vena cava and involvement of adjacent organs in order to plan for likely multivisceral resections. In four of six patients, the inter-renal segment of the IVC was involved and a transplant team was kept on standby for possible renal auto-transplantation. A functional renogram was obtained as part of the pre-operative assessment when a nephrectomy was contemplated.

On exploratory laparotomy, the tumor was resected with en bloc resection of adjacent abdominal organs in three of six patients. Two of these patients required a nephrectomy with one patient additionally requiring a liver wedge excision with a cholecystectomy. Tumor resection was performed with adequate vascular margins which were macroscopically free from tumor. A thoraco-abdominal approach was utilized for one patient, with the remaining undergoing exploration via a generous midline incision. Radical resection of the IVC was performed for four of six patients and two patients underwent partial excision of the wall of the IVC to achieve a tumor-free cuff (Fig. 2a, b). Veno-venous bypass was required to maintain circulation only in one of the three patients that required a

prosthetic replacement of the IVC. Hemodynamic instability after clamping of the IVC was considered to be an indication for performing a veno-venous bypass to maintain venous return to the heart and preventing significant hypotension. However, two patients tolerated IVC clamping well without hemodynamic instability, and the resection and prosthetic replacement could be performed without the use of a bypass.

A prosthetic reconstruction with polytetrafluoroethylene (PTFE) or Dacron graft was performed in three patients. Prosthetic grafts were used in two of these patients as significant collaterals were not formed to allow simple ligation of the IVC. One of these patients additionally required a re-implantation of the left renal vein into the native IVC to preserve venous drainage of the left kidney after undergoing a right nephrectomy (Fig. 2c, d). In the third patient, a Dacron Y graft was utilized as the defect spanned across the bifurcation of the IVC into the common iliac veins which required to be reconstructed to maintain venous drainage from the lower limbs (Fig. 2e). Two patients did not require reconstruction as primary closure of the defect was feasible. One patient had multiple well-developed collaterals with pre-existing IVC thrombosis and reconstruction was deemed unnecessary in this case (Fig. 2f, Table 1).

The complexity of these procedures is underlined by the median operative time of 450 min (range 300–600 min). The median intra-operative blood loss was 1925 ml (range 1200–6000 ml) with blood replacement given as required (mean 2 units, range 0–12 units). Most patients required a post-operative ICU stay of 2 days. One patient developed a post-operative hemothorax and required a prolonged ICU stay of 9 days. The recovery of all patients except one was uneventful with a median hospital stay of 11 days (range 6–26 days) (Table 2).

Four of six tumors were primary leiomyosarcomas arising from the IVC wall, whereas two patients had tumors arising from the retroperitoneum with secondary encasement and

Fig. 1 CT scan showing pre- and post-operative images of a retroperitoneal mass involving the anterior wall of the inferior vena cava (IVC) with indistinct fat planes with the pancreas and duodenum and involvement of the ostium of the left renal vein

involvement of the IVC, one of which was a leiomyosarcoma and the other a dedifferentiated liposarcoma of the retroperitoneal tissues (Table 3). Five of six of these tumors were high-grade sarcomas and one patient had an intermediate grade

tumor. Macroscopically, negative margins could be achieved in all patients with microscopically focally positive margins in two patients at the transected edge of the vena cava. In both instances, the proximal margin of the IVC was involved

Fig. 2 Steps of resection of a tumor involving the anterior wall of the inferior vena cava (**a**) with proximal and distal vascular control (**b**) with anastomosis of the left renal vein to the distal segment of the IVC (**c**) and outcomes of the inferior vena cava replaced with prosthetic grafts (**d, e**) and unreconstructed IVC (**f**)

microscopically, which was seen to be macroscopically free during surgery. In both cases, the proximal margin was just below the confluence of the hepatic veins, and further excision of the IVC would entail complete hepatic vascular isolation. One patient with microscopic positive margins developed a local recurrence after an interval of 8 months. The other patient remains disease free at this time. Five patients received adjuvant radiation to the tumor bed, whereas one patient had received pre-operative chemoradiotherapy.

The median follow-up duration was 24.5 months (range 15 to 30 months). Two patients developed local/regional recurrences at a disease-free interval of 8 months. Of these, one patient had an operable recurrence in the right lumbar region and underwent excision of the same followed by adjuvant chemotherapy. She currently remains disease free at 30 months following initial surgery. The other patient developed an unresectable local recurrence at the operated site and has received palliative chemotherapy for the same. One patient developed distant metastasis to the right lung after 20 months from initial surgery. The metastatic lesions were deemed resectable on imaging and the patient underwent pulmonary

metastasectomy. She presently remains disease free and on regular follow-up. There has been no mortality to date.

Discussion

Resection and Reconstruction: Techniques

Resection of the distal segment of the IVC was first reported by Melchior in 1928 for a leiomyosarcoma arising from the vena cava [4]. Since then there have been numerous individual case reports and small series reporting various resections of the IVC for malignancy.

Kieffer et al. in their series of 22 patients, have described resections of sarcoma affecting different segments of the IVC, which they divided into the retro-hepatic or supra-renal, the inter-renal, and the infra-renal portions [5]. The authors report various techniques of resection including the use of bilateral subcostal or midline abdominal incisions with thoracic access via a median sternotomy or a phrenotomy when required. A similar description is provided by Mastoraki et al., who

Table 2 Peri-operative parameters

Patient	Operative time (min)	Blood loss (ml)	Blood replacement (units)	ICU stay (days)	Hospital stay (days)	Peri-op complications
1	450	1850	2	2	10	None
2	600	6000	12	9	26	Yes. Hemothorax
3	420	1800	2	2	12	None
4	480	5000	11	2	8	None
5	300	1200	0	2	6	None
6	450	2000	2	2	14	None

ICU intensive care unit

recommend using a thoraco-abdominal incision, a combination of median laparotomy with sternotomy, a thoracophreno-laparotomy, or a midline incision with a subcostal incision as the case demands [6]. We could achieve adequate exposure and access to all relevant intra-abdominal structures using a midline vertical incision in five of our patients. One patient required a thoraco-abdominal access since the tumor mainly involved the retro-hepatic and supra-renal segments of the IVC, and supra-hepatic control of the IVC was required.

Involvement of the inter-renal segment raises a separate concern regarding the venous drainage of the kidneys. Three patients in our series required a nephrectomy, whereas one patient required a re-implantation of the renal vein. It is important to keep in mind that both renal veins may be affected by the tumor, as in our first patient, and that a kidney auto-transplant may be required to salvage at least one of the kidneys if a vein re-implantation is not feasible [7]. In this situation, the kidney is transplanted in the iliac fossa and the blood supply is re-established by anastomosis to the iliac vessels. We planned for this eventuality in the pre-operative setting after careful review of the imaging and keeping the transplant team on standby. The necessary intra-operative protocols for renal transplant would then need to be followed for an optimal outcome. Intra-operative surprises could lead to disastrous results. Kraybill et al. have reported a similar series of four

patients, wherein the uninvolved kidney was auto-transplanted in two patients [7]. Multivisceral resections also may be required to achieve complete resection of the tumor [2, 5, 8]. Hardwigsen et al., in their series of 14 patients, report a variety of tumors which entailed a resection of the IVC along with multivisceral resections including renal, hepatic, bowel, and aortic resections [8]. Stauffer et al. report the resection of an IVC leiomyosarcoma along with removal of the adrenal gland, distal pancreas, spleen, and left kidney which were involved by the tumor [9]. Involvement of the retro-hepatic segment may entail a complete vascular exclusion of the liver for resection and may also require major liver resection for adequate disease clearance [2, 5, 10].

A primary closure of the defect in the vein is acceptable when it results in luminal narrowing less than 50% [2]. Else, a patch closure of the defect has to be performed. Materials utilized for patch closure include pericardial patches, autologous venous patches using the saphenous vein or the internal jugular vein, synthetic patches composed of PTFE, and peritoneo-fascial patches [2, 11]. Various options of reconstruction after segmental resection have been described in literature [10]. The commonest form of reconstruction is with an expanded PTFE (ePTFE) graft. This allows for selection of length and diameter which can be individualized to each patient. The commonly used diameters of PTFE grafts range

Table 3 Oncologic outcomes

Patient	Tumor size (cm)	Histology	Grade	Margins	Adjuvant treatment	Disease-free interval	Recurrence
1	13 × 8.5	Leiomyosarcoma (IVC)	High	R1	Adjuvant RT	24 months	No
2	7.5 × 6.5	Leiomyosarcoma (IVC)	High	R0	Adjuvant RT	20 months	Yes. Distant (lung-operated)
3	20 × 15	Leiomyosarcoma (retroperitoneum)	Intermediate	R0	Adjuvant RT	8 months	Yes. Regional (operable)
4	29 × 19	Dedifferentiated liposarcoma (retroperitoneum)	High	R1	Adjuvant RT	8 months	Yes. Local (inoperable)
5	10 × 4.8	Leiomyosarcoma (IVC)	High	R0	None (pre-op CTRT received)	22 months	No
6	18 × 14	Leiomyosarcoma (IVC)	High	R0	Adjuvant RT	15 months	No

IVC inferior vena cava, RT radiotherapy, CTRT chemoradiation

from 12 to 20 mm. In our series, we utilized PTFE grafts with diameters of 12 and 14 mm to reconstruct the IVC. For selection of the graft size, we used a smaller graft size as compared to the native IVC. There are various opinions in this respect. Quinones-Baldrich et al. have described the use of narrower graft sizes to maintain a higher velocity of blood flow across the graft and avoid thrombosis [2]. Others recommend the use of a larger graft diameter up to 20 mm to match the native IVC diameter as well as the use of ringed ePTFE to prevent collapse of the graft and subsequent thrombosis [9]. A larger diameter of the graft has also been recommended since a thick layer of pseudo-intima can form and compromise the lumen [12]. The risk of thrombosis and occlusion of the graft can be reduced by performing an arterio-venous fistula to maintain a high rate of blood flow through the graft. Kieffer et al. have used this technique with success in their patients with good results [5]. However, Bower et al. state that creation of an arterio-venous fistula is not necessary for tumors involving the supra-renal IVC because of the large blood flow at this level, and that it may be performed for long segment prosthetic replacement of the IVC [10]. Long-term anti-coagulant treatment may also be avoided with this technique. We have not utilized an arterio-venous fistula for any of the patients requiring prosthetic reconstruction of the IVC and have not faced any problems related to graft thrombosis or occlusion. Our use of anti-coagulation is not routine, but case based.

Simple ligation of the IVC is also acceptable after resection of the segment involved by tumor [3]. This is especially true in the event of pre-operative thrombosis or occlusion of the IVC leading to the formation of collaterals and adequate venous drainage is maintained after resection of the affected segment. We could perform simple ligation and division of the IVC in one patient who had developed extensive retroperitoneal collaterals. Care was taken during the dissection to meticulously preserve these collaterals. However, one must be cautious when choosing not to reconstruct the IVC as poor collateral formation may result in debilitating lower limb edema and poor functional outcomes [9]. In tumors requiring extensive retroperitoneal dissection, these collaterals may be disrupted leading to an acute increase in venous pressures if the IVC is not reconstructed. Intra-operative fecal or biliary contamination or bowel resection may also preclude the use of a prosthetic material to reconstruct the IVC. The infra-renal segment of the IVC can usually be ligated without significant hemodynamic changes or deterioration of renal function. The commonest sequela of acute obstruction of the IVC is lower limb edema which can usually be managed conservatively [3, 13]. However, resection of the inter-renal segment usually requires a reconstruction with appropriate re-implantation of the renal veins to avoid renal decompensation and anuria. We chose to reconstruct the IVC with a graft in three of our patients since the inter-renal segment was affected in two patients and the resection of the bifurcation of the IVC with

division of the lumbar vessels was required in the third patient. All patients demonstrated good graft patency on follow-up imaging without lower limb edema or renal decompensation.

The left renal vein, if involved, can be resected without reconstruction in most cases since it communicates with the lumbo-hemiazygous collateral system [3, 13]. Left renal vein reconstruction or an auto-transplantation may be required if a longer segment needs to be resected along with the gonadal vein or the lumbar collaterals. However, if the right renal vein requires division, it almost always needs to be re-implanted into the abdominal vasculature due to the paucity of collaterals on the right side [2]. Three of the patients in our series required a right nephrectomy due to involvement of the right renal vessels by the tumor. One patient underwent re-implantation of the left renal vein into the native IVC for preservation of renal function following a right nephrectomy. The length of the left renal vein allowed for some migration of position of the ostium and a comfortable anastomosis to the native IVC. In case this is not feasible, the vein can be re-implanted into the PTFE graft and ringed PTFE is preferred in this scenario as it allows the ostium to remain open [2].

Guerrero et al. have reported the use of a fresh IVC allograft to reconstruct the IVC after resection without the use of immunosuppression [14]. The patient had a patent IVC at 14 months of follow-up without the requirement of anticoagulants. Pulitano et al. have described the use of biologic grafts composed of autologous peritoneo-fascial and bovine pericardial grafts in their series of 32 patients requiring either a patch repair or complete replacement of the IVC, with good results [15].

Resection of the IVC is a technically demanding procedure, requiring the collaboration of teamwork of surgical oncologists, vascular surgeons, and transplant specialists. This complexity is reflected in the median operative time of 450 min with some procedures taking longer as well. This is comparable to an average operative time of 355 min reported by Hines et al. and 420 min as reported by Mann et al. [16, 17]. The median blood loss was 1925 ml with an average transfusion requirement of 2 units. Quinones-Baldrich et al. report a mean blood loss of 1000 (80–3700) ml in their series of 47 patients undergoing vena caval resections [2]. Hines et al. report an average blood loss of 1115 ml in their series of 14 patients [16]. Similarly, a transfusion requirement of an average of 2–3 units has been reported by various authors [8, 17, 18]. The median hospital stay was 11 days. This, too, is comparable to a mean hospital stay of 9 (5–19) days reported by Quinones-Baldrich et al., 18 (14–20) days by Hardwigen et al., and 11 (4–55) days by Mann et al. [2, 8, 17].

Oncologic Outcomes (Tables 3 and 4)

Resection of tumors arising from the IVC is the only definitive treatment to offer a possible cure in these patients [19].

Adjuvant therapies such as chemotherapy and radiotherapy have been used; however, it is difficult to assess the benefit of these treatments due to the relatively small number of cases treated. In spite of the current standards of multimodality treatment, the prognosis in tumors of the IVC remains poor with local recurrences and distant metastases developing in many patients. Two of the six patients in our series have developed local recurrences within 1 year of follow-up and one patient developed lung metastases, underlining the aggressive nature of these tumors (Table 3).

The reported peri-operative mortality in various series ranges from 0 to 15% [13]. Few authors have reported intra-operative deaths in their series, seen in patients with large tumors requiring complex multivisceral resections. Kieffer et al. report a post-operative mortality of 4 patients of 20 that underwent a curative resection [5]. Hollenbeck et al. report a post-operative mortality of 8%, both patients in their series dying due to pulmonary thrombo-embolism [13]. Quinones-Baldrich et al. report a post-operative major morbidity of 10.6% including bowel obstruction, bleeding, renal failure, and graft thrombosis [2]. Only one of six patients in our series had a major morbidity which was managed conservatively. There was no peri-operative mortality in our series.

Mingoli et al. created the International Registry of IVC leiomyosarcoma in 1992 and reported a 5-year survival of 49.4% in patients who underwent a curative resection with a disease-free survival of 31.4% [19]. Hines et al., in their series of 14 patients who underwent resection and adjuvant/neo-adjuvant radiation with or without chemotherapy, report a 5 year survival of 53% [16]. The median disease-free survival was 17.5 months in our series with a median overall survival of 24.5 months. There has been no mortality to date.

In the analysis by Hines et al., positive surgical margins were found to significantly affect the survival [16]. Addition of adjuvant therapy had a trend towards improved survival in this series. Two patients in our series had a microscopic positive margin (33%) and underwent adjuvant radiotherapy with a boost to the site of the positive margin, one of which developed a local recurrence which was deemed inoperable.

Microscopic margin positivity despite a macroscopic negative margin highlights the aggressive biology of these tumors, as well as the technical difficulties faced in the resection of tumors located in this region. Hollenbeck et al. have reported a mere 33% negative margin rate in their series of 21 patients [13]. Similar series of resections of these tumors have reported margin negativity in the range of 40 to 67% which is comparable to our series [3, 20]. An attempt at achieving a clear macroscopic margin of at least 1 cm should be made in all cases, as the survival for patients with margin positive resections has been seen to be dismal [16].

The histological grade of the tumor has been reported to have an influence on the prognosis and survival in the treatment of retroperitoneal sarcomas. Ferrario et al. reported a 5-year survival of 92, 54, and 48% for grades I, II, and III, respectively, in their retrospective study of 130 patients with retroperitoneal sarcomas [21]. Singer et al. performed a retrospective study of 183 patients with truncal and retroperitoneal sarcomas and reported an increased risk of death in patients with high-grade and intermediate-grade tumors as compared to low-grade tumors [22]. In our current series, there were five high-grade tumors, including all the tumors arising from the IVC primarily (Table 3). One patient had an intermediate grade retroperitoneal leiomyosarcoma, which was secondarily involving the IVC. The patient with an intermediate grade tumor developed a local recurrence, which was operable. One each of two patients in the high-grade tumor group developed local (inoperable) and distant (operable) recurrences respectively (Table 3). Sarcomas arising from the IVC are more likely to be high-grade tumors as reported by Quinones-Baldrich et al. and Daylami et al. [2, 3]. In a series of six retroperitoneal tumors requiring IVC resection, Wachtel et al. report four patients having high-grade tumors, one patient with a low-grade tumor, and one patient in whom the grade was unreported, which is similar to the distribution of grade in the current series [23].

In their series of 218 patients, Mingoli et al. report a poorer survival for patients presenting with upper segment tumors, lower limb edema, Budd-Chiari syndrome, intra-luminal

Table 4 Comparison of other series

Author	Year	Number of patients	Time period	R0 resection (%)	IVC reconstruction/repair	Recurrence (%)	5-year survival (%)
Hines et al. [16]	1999	12	19 years	83	7/12	25	53
Hollenbeck et al. [13]	2003	21	20 years	33	10/21	67	33
Kieffer et al. [5]	2006	20	25 years	NA	13/20	NA	34.8
Ito et al. [20]	2007	20	16 years	40	17/20	68	62
Daylami et al. [3]	2010	6	6 years	67	0/6	17	NA
Quinones-Baldrich et al. [2]	2012	47	21 years	NA	47/47	NA	45
Current study		6	14 months	66	5/6	33	NA

IVC inferior vena cava

growth of tumor, and occlusion of the IVC [19]. Whereas patients presenting with abdominal pain, a palpable abdominal mass, and with tumors affecting the middle segment of the IVC were seen to have a longer disease-free interval and a longer survival.

Adjuvant Therapy

Adjuvant radiotherapy has been used after curative resection of these tumors [16, 20, 24]. These tumors are known to recur locally with a local recurrence rate up to 36%. Five patients received post-operative radiotherapy to the tumor bed as they were large, deep seated, high-grade tumors. Radiation may reduce the number of local recurrences by sterilizing the tumor bed [13]. However, delivery of radiation to the retroperitoneum has its pitfalls, due to the proximity of vital organs such as the kidney, liver, and spinal cord. Recent techniques of radiotherapy such as conformal, image-guided, and intensity-modulated radiation have improved radiation delivery to the target area with minimal toxicity to adjacent vital structures. Large, borderline resectable tumors can potentially be downsized by pre-operative radiotherapy and may become resectable [24].

In their retrospective study of 204 patients which compared the use of peri-operative radiotherapy compared to surgery alone, Kelly et al. reported an improved local recurrence-free survival and recommended that the use of peri-operative radiotherapy be investigated further in a randomized trial [25]. A Scandinavian study of 94 patients analyzing adjuvant radiotherapy in patients treated surgically for retroperitoneal sarcomas reported improved local recurrence (LR) rates of 77% with the used of adjuvant RT as compared to 39% when it was not utilized [26]. They also reported an overall survival (OS) benefit of 71 vs. 52% with the use of adjuvant radiation. Nussbaum et al. performed a case-control propensity score-matched analyses of US National Cancer Data Base which included 9068 patients [27]. They demonstrated that both pre-operative and post-operative radiotherapy were associated with significantly improved overall survival as compared to surgery alone. An ongoing European trial (Surgery With or Without Radiation Therapy in Untreated Non-metastatic Retroperitoneal Sarcoma [STRASS]) is currently evaluating the role of pre-operative radiotherapy in retroperitoneal sarcomas [28].

As we await the results of this trial, Baldini et al. have provided guidelines for the pre-operative radiotherapy for retroperitoneal sarcomas in their consensus report of an international expert panel [29]. It is unlikely that level I evidence will be generated in the adjuvant treatment of sarcomas requiring resection of the IVC due to their rare incidence and few reported cases. The treatment algorithms would likely have to be extrapolated from the treatment of a broader group of all retroperitoneal sarcomas.

The role of adjuvant chemotherapy is not established in the treatment of retroperitoneal sarcomas and most of the data are extrapolated from the treatment of extremity soft tissue sarcomas. However, when used, the standard regimen includes ifosfamide and doxorubicin. Kim et al. have utilized adjuvant chemotherapy with these agents and mention that time to recurrence and overall survival may be improved [30]. Neo-adjuvant chemotherapy, and occasionally radiotherapy, has been used to downsize the tumor and allow for safe resection, especially in histologies such as synovial sarcoma which are sensitive to chemotherapy [16, 20, 24]. Palliative chemotherapy has been used in the metastatic setting and can provide disease control and some prolongation of life [30]. One patient in our series developed an inoperable local recurrence and has received chemotherapy for the same. He remains alive with disease at the time of last follow-up. Miura et al. analyzed the US National Cancer Database and identified 1525 patients of 8653 patients who received chemotherapy, 10.6% of which received chemotherapy in the neo-adjuvant setting [31]. They reported that the median OS was worse for patients receiving chemotherapy as compared to surgery alone (40 vs. 68.2 months respectively) and concluded that the currently available chemotherapy regimens do not confer a survival benefit in the treatment of retroperitoneal sarcomas.

Conclusion

Resection of the IVC for extirpation of retroperitoneal sarcomas is a technically demanding and difficult procedure. The availability of a multidisciplinary team of surgeons and state-of-the-art intensive care support is essential for good outcomes. Adjuvant treatment in the form of radiation with or without chemotherapy is part of the multimodality treatment though the exact benefit is not known. The prognosis of these tumors remains dismal with a high incidence of local or distant relapses. Curative resection with adjuvant therapy remains the only option to offer long-term survival for these patients.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. As this is a retrospective study, for this type of study, formal consent is not required. However, informed consent was obtained from all individual participants included in the study.

Consent to Publish Consent has been obtained from the patients/guardians for publication of relevant medical data and operative photographs without revelation of patient identity in this study.

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