Leiomyosarcoma of Infrarenal Inferior Vena Cava: A Single Institution Experience and Review of Literature

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Abstract

We report three cases of primary leiomyosarcoma (LMS) of inferior vena cava (IVC). Vascular LMSs are rare tumors, arising most frequently from IVC. These tumors have a female predominance. Their diagnosis is often challenging, as patients may present with nonspecific complaints such as dyspnea, malaise, weight loss, abdominal pain, or back pain, preceding the diagnosis by several years. LMS of the IVC most frequently occurs in the middle segment. The final diagnosis can be made by an ultrasound or computed tomography-guided biopsy. Due to limited experience with this disease, optimal management of IVC LMS is unknown. Curative surgical resection remains the current treatment of choice for primary LMS of IVC. Neoadjuvant therapy may be given to downsize the tumor and increase resectability rates. Nonetheless, there is no proven role for adjuvant therapy, and recurrence is common. We, hereby, report three cases of this rare entity with emphasis on management.

Keywords: Inferior vena cava, leiomyosarcoma, PTFE

INTRODUCTION

Vascular leiomyosarcoma are rare malignant tumours of smooth muscle cells of media. It is now known that these tumours are most common malignant tumour of IVC. In this case series, we report reconstruction of ivc without immunosuppression.

CASE REPORTS

Case 1
Case 1 is a 60-year-old female with right-sided abdominal pain for 1 year, with no other known comorbidities. Physical examination revealed tenderness in the right upper quadrant and mild edema of lower extremities. Computed tomography (CT) revealed a heterogeneously enhancing mass with questionable involvement of inferior vena cava (IVC) and renal vessels [Figure 1]. The patient was diagnosed with a retroperitoneal sarcoma by a general surgeon who scheduled surgical excision. A midline incision was performed, and the vascular surgery service was consulted intraoperatively when it became evident that the tumor involved the IVC and was adherent to duodenum and right ureter and cava reconstruction would be mandatory for curative resection [Figure 2].

The tumour involved infrarenal IVC, which looped and controlled and after which intravenous heparin was administered. There was enough normal IVC below renal veins for proximal control, which was taken just below their origins. Distal control was taken just above the iliac bifurcation. The tumor was resected in en bloc with affected IVC. The tumor was found adherent to ureter. The repair was performed using 18 mm Dacron graft in end-to-end fashion. Patient underwent right ureter Double J stenting intraoperatively [Figure 3]. Specimen was sent for pathological examination [Figure 4] which revealed features suggestive of leiomyosarcoma (LMS) with intraluminal growth and negative margins positive for smooth muscle actin (SMA) and S-100. Is SMA which are tumor markers found positive in soft-tissue sarcomas [Figure 5]. The patient was placed on low-dose heparin and warfarin therapy. Adjuvant therapy was not administered and the patient was discharged to go home on postoperative day 5 and was doing

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well 16 months postoperatively. Follow-up duplex ultrasound has shown chronic laminar thrombus in the posterior wall of the graft causing mild (<30%) stenosis. The patient remains on warfarin therapy to help maintain graft patency and is followed up every 6 months with serial duplex ultrasounds, as well as CT scans to monitor for recurrence.

**Case 2**
Case 2 is a 61-year-old female with a history of vague abdominal pain in the epigastric and right hypochondrium for last 3 months. On physical examination, there was no tenderness and palpable mass. Patient underwent CT venography, which showed 8.8 cm × 5.5 cm well-defined heterogeneous mass arising from the infrarenal IVC with preservation of surrounding fat planes [Figures 5 and 6].

The patient underwent exploratory laparotomy with excision of tumor and primary repair of the IVC was done without reconstruction as patient had well-developed venous collaterals and primary repair looked feasible. The patient did not develop swelling during the postoperative period.

The pathological examination showed high-grade LMS with tumor cells positive for SMA.

The patient was placed on low-dose heparin and warfarin therapy. Patient has been followed for 11 months and without any recurrences.

**Case 3**
A 35-year-old female was admitted in general surgery ward with complaints of upper abdominal pain for the past 6 months and edema of bilateral lower limbs for 20 days. On physical examination, there was tenderness in the right hypochondrium and epigastrium.

The patient underwent ultrasonography abdomen which showed retroperitoneal mass. On CT abdomen, we observed well-enhancing retroperitoneal mass inferior to pancreas and in close relation to third part of duodenum [Figure 7]. The patient was diagnosed as retroperitoneal sarcoma, and was planned for excision by general surgeon, but on table the tumour was found arising from ivc extending into both common iliac veins, vascular team was
called and excision with reconstruction was planned [Figure 8]. The tumor was isolated and intravenous heparin was given. Proximal control of infrarenal IVC was taken and distally bilateral common iliac control was taken and clamped. The tumor was resected and sent for pathological examination, and the infrarenal IVC was reconstructed using Dacron 16 mm × 8 mm × 8 mm graft [Figures 9 and 10]. The pathological margins were negative, and tumor was primary leiomyoma positive S-100 tumor cells. The patient has been put on low-dose oral anticoagulant and is being followed with Doppler for graft surveillance.

**DISCUSSION**

LMS of the (IVC) is a rare malignant tumor originating from the smooth muscle of the media.\(^1\) Symptoms and resectability
depend on the location and extension of the tumor as well as associated thrombosis.

More than half of all vascular LMSs occur at the IVC. Vascular LMS represents 2% of all LMSs and 0.5% of all soft-tissue sarcomas.[2,3]

IVC LMS is a malignant tumor of mesenchymal origin that develops from smooth muscular fibers of the tunica media.[4] Perl first described it in 1871, and the first surgical resection was in 1928 by Mechior.[5,6] LMS of the IVC is four times more common among women and is the most commonly diagnosed during the fifth to sixth decades of life.[1]

IVC LMS has a slow rate of growth and can remain asymptomatic for a long time, causing diagnosis to be delayed until advanced stages when prognosis is poor.

The presentation of LMS depends on its location along the IVC, which can be divided into three segments for this purpose. Segment I (lower) is below the renal veins and is involved in 36% of cases.[1,4] Typical presenting symptoms for IVC LMS in this segment may include lower-extremity edema, deep-venous thrombosis, abdominal pain, and palpable mass. Segment II (middle) is from the hepatic veins to the renal veins and is involved in 44% of cases. Symptoms in this region may include abdominal pain, nephrotic syndrome, and renal hypertension. Segment III (upper) is from the right atrium to the hepatic veins, is involved in 20% of cases, and can present with weight loss, nausea, Budd–Chiari syndrome, and cardiac arrhythmias.[7,8]

Diagnosis of IVC LMS is often incidental or at autopsy. When symptoms are present, CT or magnetic resonance imaging is useful to determine the extent of tumor involvement. The confirmation of diagnosis is always done by biopsy. Prognosis is better for tumors involving the middle segment of the IVC as compared with the upper segment. Surgical resection with negative margins, as was the case in all of our patients, has been shown to be the only treatment that improves survival.[1,2,4] Tumors involving the lower segment are best treated with total excision of the tumor and involved portion of the IVC and either primary IVC closure, patch angioplasty with autologous vein or prosthetic patch, or replacement with interposition polytetrafluoroethylene (PTFE) or Dacron or banked venous homograft.[9] The goal of cancer surgery is to resect for cure, which requires resecting all visible and microscopic tumors. Hence, this principle also applies for tumor invading IVC.

Undersizing of the PTFE or Dacron has been recommended with the rationale that the resulting increase in blood flow velocity within the graft might reduce thrombotic risk.[10] The use of ring PTFE is common in replacement of IVC.[11] The ring reinforcement, in theory, resists respiration compression better and thus prevents graft collapse that may be a factor in promotion of graft thrombosis. Others recommend a larger diameter graft, as PTFE tends to form a thick pseudointima that may result in obstruction.[12] We used (18 and 16 mm) grafts in our patients.

Creation of an arteriovenous fistula (AVF) either between the aorta and IVC or iliac vein, or between the femoral vessels, has been advocated. AVF creation is theorized to improve patency and prevent the need for anticoagulation by elevating blood flow velocity.[13] However, complications such as limb edema and congestive heart failure have been attributed to AVFs. We did not create AVF in our patients.

Finally, long-term postoperative anticoagulation is recommended by some authors to prevent thrombosis, while others report acceptable patency without warfarin therapy.[13,14] We used intravenous heparin bridging with oral warfarin therapy in our patients.

Following curative resection of the tumor, recurrence rates have been reported to be as high as 57% with a 5-year survival of 50% in a large series by Mingoli et al.[13] Adjunctive treatment with radiation or chemotherapy has not been proven to improve survival and was not used in our cases. Radical tumor resection has resulted in 5- and 10-year survival rates of 49.4% and 29.5%, respectively. Despite advances in treatment, perioperative mortality ranges as high as 15%.[16] The local tumor recurrence has been found as high as 57.3%. Although it was initially reported that high-grade tumors increased the risk of death, it has been found that tumor grade did not predict recurrence or survival. A retrospective analysis conducted at University of California, Los Angeles (UCLA) also found no difference in survival based on age, gender, tumor size, and lymph node status. Treating LMS of the IVC remains a challenge. Over the years, new techniques have resulted in decreased morbidity and mortality associated with operative management, yet local recurrence remains high. The rarity of this disease poses a great obstacle in its investigation.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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