Case Report

Leiomyosarcoma of Inferior Vena Cava: Surgical Excision and Reconstruction of Inferior Vena Cava with Bifurcated Dacron Graft

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Abstract
Leiomyosarcomas of the inferior vena cava (IVC) are rare tumors that occur commonly in women of middle age group and usually present with nonspecific symptoms. Treatment consists of surgical excision with or without IVC reconstruction, combined with chemotherapy and radiotherapy. We present a case of leiomyosarcoma of infrarenal IVC extending into both iliac veins, which was resected with reconstruction of IVC and both iliacs with bifurcated Dacron graft. Although IVC reconstruction is mentioned in literature, reconstruction with bifurcated graft is rare.

Key Words: Bifurcated graft, inferior vena cava, leiomyosarcoma, reconstruction of inferior vena cava

Introduction
Vascular leiomyosarcoma is a rare entity, but it is the most common malignant primary tumor of inferior vena cava (IVC). The first report of IVC leiomyosarcoma was done by Peri and Virchow[1] in 1871. A majority of patients are women in their fifth or sixth decade.[2] The most common clinical presentation is chronic vague right-sided abdominal pain. In the management, pre- or post-operative adjuvant chemotherapy and radiotherapy are considered, but surgery remains the mainstay of the treatment. En masse resection of the tumor with involved IVC is the accepted treatment with or without establishment of continuity for venous return. Various materials are used for restoring continuity such as polytetrafluoroethylene (PTFE) graft and autogenous vein grafts. We present a case of IVC leiomyosarcoma, who was treated with complete resection of the tumor with IVC and both iliac veins and replaced with bifurcated Dacron graft. To our knowledge, in literature, there has been no mention of usage of bifurcated graft for IVC reconstruction.

Case Report
A 62-year-old female diabetic patient diagnosed to have retroperitoneal mass while being evaluated for mild leg edema in the lower extremities. She did not have any pain in abdomen, bowel or urinary symptoms, except for a positive history for breathing difficulty. Her laboratory investigations revealed nothing abnormal. Computed tomographic (CT) examination revealed a large mass in the retroperitoneum [Figure 1]. She underwent exploratory laparotomy and found to have a huge mass enclosing IVC and compressing right renal vein. Distal extension was up to external iliac vein in the right and common iliac vein in the left. The tumor was resected en bloc [Figure 2a and b] with IVC and both iliac veins. Venous reconstruction was done with Dacron bifurcated graft (18 × 9 × 9) [Figure 3].

Microscopic examination of the tumor revealed short, spindled out cells with open hyperchromatic nuclei showing increased mitosis and arranged in closely packed whorls/sheets and reticular pattern. The diagnosis was confirmed with tumor markers (positive for vimentin, desmin, and smooth muscle actin) [Figure 4]. Postoperatively, patient developed right limb of graft thrombosis, which was treated medically and patient recovered with no morbidity. She was advised adjuvant chemo- and radio-therapy. With 1 year follow-up, the patient is disease-free and functioning graft.

Discussion
Vascular leiomyosarcoma represents just 2% of all leiomyosarcomas with venous involvement more common than arterial sarcoma. Even in primary venous leiomyosarcoma, 60% originate from IVC. After the first documented case in 1871, only 197 cases had been
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In few cases of complete IVC obstruction preoperatively with development of adequate collaterals, resection of IVC without reconstruction may be considered. During reconstruction of infrarenal segment of IVC, simple clamping of vein is sufficient and does not cause proximal venous hypertension or arterial hypotension. Even if they occur, it can be managed by infrarenal aortic clamping. Various materials are used for the reconstructive material, such as PTFE, autogenous vein, but we used Dacron because of availability and cost factor.

Considering the development of recent advances in adjuvant chemotherapy and radiotherapy, adjuvant therapies may be considered in selected patients with some survival benefit, but surgical resection with microscopically negative margins is still the treatment of choice.
We recommend aggressive surgical team work by oncologist and vascular surgeon for better survival of the patient.

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**Conflicts of interest**

There are no conflicts of interest.

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