Primary Leiomyosarcoma of the Juxtarenal Inferior Vena Cava: A Case Report

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Abstract Primary juxtarenal leiomyosarcoma of the inferior vena cava (IVC) is a rare disease and is invariably malignant. Prognosis depends on early patient diagnosis and management. We report a case of a primary IVC tumor in a 66-year-old woman which was managed successfully by surgical resection of involved IVC along with right nephrectomy. IVC reconstruction was done using a Dacron interposition graft, and the left renal vein was anastomosed end to side to the Dacron graft.

Keywords Leiomyosarcoma · Inferior vena cava · Dacron graft

Introduction

Leiomyosarcoma of the inferior vena cava (IVC) is rare tumors, with a few cases reported in literature. Invariably

these tumors are malignant with a slow growth pattern. Symptoms and signs are usually nonspecific and may precede diagnosis by several years making prognosis worse. The recommended treatment includes complete surgical resection with the role of adjuvant chemotherapy and radiotherapy unclear.

Case Report

A 66-year-old woman presented to us with complaints of pain in the right loin region for past 6 months, which was continuous and radiating to both lower limbs. She also complained loss of appetite and loss of weight for the past 2 months. There was no history of fever, hematuria, or dysuria. Her medical history revealed bilateral knee joint replacement for osteoarthritis in 2008 and laparoscopic cholecystectomy in 2009. On examination we found a few dilated veins in the lower abdominal wall and the upper part of both thighs. No abdominal lump was palpable as such on palpation. Hemogram, kidney function tests, and liver function tests were within normal limit. An abdominal ultrasound revealed a hypodense lesion in the retroperitoneum invading the IVC. Venous Doppler USG revealed compressed juxtarenal IVC with reduced upward venous flow. Contrast CT scan of the abdomen along with CT angiogram revealed either a primary tumor arising from the juxtarenal IVC and extending into the retroperitoneum or a retroperitoneal tumor invading the IVC. A CT-guided biopsy revealed a malignant spindle cell tumor.

The patient underwent exploratory laparotomy which revealed a firm tumor arising from the IVC extending 3 cm above and 4 cm below the right renal vein. The right

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Fig. 1 (a) & (b) Demonstrating involvement of IVC

renal vein and artery were invaded by the tumor. An on table frozen section, biopsy was done which came out to be a malignant mesenchymal tumor. The liver was subsequently mobilized to expose the subhepatic part of the IVC to achieve proximal clearance of tumor. The aorta was slung at the infradiaphragmatic level to control inflow. The patient was heparinized and the tumor was resected en block along with a right nephrectomy. An 18 mm Dacron interposition graft was used to reconstruct the IVC. The left renal vein was reimplanted in an end-to-side manner into the Dacron graft.

The postoperative recovery was uneventful. The patient had an adequate urine output and was put on anticoagulant drugs and was discharged on the 15th postoperative day. Her histopathology report revealed grossly a large tumor in the IVC lumen, infiltrating the wall measuring 7.5 cm × 7.4 cm. Microscopically, features were compatible with leiomyosarcoma. The tumor cells were positive for smooth muscle actin (SMA) and desmin, focally positive for CD34 and negative for S100. Resected margins were free of tumors. She has now been on our follow-up for the past 5 months and is doing well. She is currently on doxorubicin-based chemotherapy.

Discussion

IVC tumor may be primary or secondary in origin. A primary tumor arises directly from smooth muscle of the vessel wall, while a secondary tumor originates in the adjacent organs of the retroperitoneum and invades IVC by direct spread [1, 2]. Primary leiomyosarcoma of IVC is extremely rare tumor with a few cases reported in literature. These lesions are invariably malignant [3]. It is more common in females and usually presents in the sixth to seventh decades [4]. Patients are usually asymptomatic initially, thus delaying diagnosis. Symptoms and signs include abdominal pain

(66 %), abdominal mass (48 %), lower limb edema (39 %), weight loss (30.6 %), and Budd–Chiari syndrome (22.2 %). Other nonspecific symptoms are fever, weakness, anorexia, nocturnal sweating, and dyspnea [2, 4, 5].

Leiomyosarcoma of the IVC may arise in the lower (below the renal vessels), middle (renal vessels to retrohepatic IVC) and upper (suprahepatic) portions of the IVC. Typically these tumors are large solid masses with extensive area of necrosis. They may be primarily exophytic, or primarily intraluminal or combination of both [6]. Those with both intraluminal and exophytic components are difficult to differentiate from the secondary tumor arising from retroperitoneum and invading the IVC. Spiral computed tomography (CT scan) is a useful screening test to diagnose IVC tumors. However, on CT scan sometime it becomes very difficult to differentiate between the primary and secondary IVC tumors. Multiplaner magnetic resonance imaging (MRI) allows accurate assessment of relationship of tumor mass to surrounding structures. Fat suppression and Gadolinium enhancement increases quality of MRI image and helps in assessing retroperitoneal tumors [7].

Primary leiomyosarcoma of the IVC are invariably malignant and are slow-growing tumors [1]. Mingolia et al. reported 218 cases of primary leiomyosarcoma of IVC.

Fig. 2 Operative photograph showing reimplanted renal vein

Fig. 3 (a) & (b) Photograph and photomicrograph of tumor

Out of this, 134 (61.5 %) patients underwent radical resection, and 26 (11.9 %) patients had palliative resection while 58 (26.6 %) patients were inoperable. Radical tumor resection was found to be associated with better 5- and 10-year survival rates (i.e., 49.4 and 29.5 %, respectively) [3]. Resectability of the tumor is the prime factor affecting the prognosis. Late presentation and involvement of suprarenal IVC worsen the prognosis. Complete surgical resection with a 1 cm of tumor-free margin is considered the treatment of choice [8]. Hollenbeck et al. reported 25 patients with primary IVC tumor treated between 1982 and 2002 and showed that complete resection of IVC could be managed either by primary repair or ligation of IVC [5]. However, radical resection of IVC tumors is often associated with disruption of collateral venous channel, and simple ligation of IVC is associated with severe limb edema in some cases. Hence, current recommendation is to use prosthetic graft for reconstruction of the IVC [9]. Ring-reinforced Polytetrafluoroethylene (PTFE) or Dacron graft is commonly used for reconstruction. Role of post-operative chemotherapy and radiotherapy is controversial. Some studies have shown that patients receiving combined radiotherapy and chemotherapy have longer survival rates than those receiving only radiotherapy [10].

Conclusion

Primary leiomyosarcoma of the IVC is a rare tumor and is often difficult to differentiate from the secondary retroperitoneal tumor involving IVC. It is usually a very slow-growing tumor and is invariably malignant. Tumors involving the lower IVC are most amenable to complete removal, whereas upper IVC tumor are least amenable to complete

removal. Aggressive management of tumors by surgical resection and reconstruction of IVC by synthetic graft gives excellent results and is the treatment of choice (Figs. 1, 2 and 3).

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