Can the Right Kidney be Salvaged in the Surgical Management of Leiomyosarcoma of the Inferior Vena Cava—a Rare Case Report

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Abstract Primary leiomyosarcoma is rare malignant tumour arising from the smooth muscles and can appear at any site where smooth muscle is present. It is more common in the peripheral vessels and has predilection for the female sex. We here report a rare case of leiomyosarcoma involving the segment I & II of inferior vena cava with extension into the renal veins in a 70 year old male patient. Midline transperitoneal approach was taken to excise the tumour along with the vessel wall with anastomosis between the right renal vein and the infrarenal inferior vena cava with a reversed great saphenous vein graft thereby salvaging the right kidney.

Keywords Leiomyosarcoma · IVC tumor · Right kidney salvage

Introduction

Leiomyosarcomas are mesenchymal tumours most commonly seen in the retroperitoneum and the uterus [1]. In the vasculature it has predilection for the venous system. Most commonly seen in middle aged females in the mid segment of the IVC [2]. Only about 200 cases have been reported in literature since 1871 [3]. Symptoms are nonspecific and may precede the diagnosis by many years [4]. Diagnosis is challenging as patients present with non specific complaints such as dyspnoea, malaise, weight loss, abdominal or back pain [4]. Computer tomography (CT), Magnetic resonance imaging (MRI)

individually or in combination with cavography, ultrasound (US), and echocardiography, allow an early and accurate pre-operative diagnosis [4]. Curative surgical resection is the choice of treatment for primary leiomyosarcoma of the IVC. We describe an unusual case of primary leiomyosarcoma of the IVC with review of literature (Figs. 1, 2, 3 and 4).

Case Report

A 70 year old male patient presented with vague abdominal discomfort of 1 year duration. Physical examination was normal. Our patient had been taking symptomatic treatment regularly and underwent an ultrasound of the abdomen which detected a mass in the retroperitoneum with mixed echogenicity. A contrast CT scan of the abdomen was performed which revealed a 10.5×6×6 cm lobulated heterogeneously enhancing intraluminal mass with necrotic changes in the retrohepatic and supra-renal inferior vena cava (segment I & II) distending and obliterating the lumen with extension into the distal portion of left renal vein. The superior margin of the lesion extends into the proximal hepatic segment of the IVC beyond which the veins showed normal opacification with normal right sided cardiac chambers. Hepatic veins were patent with normal luminal opacification. The Right renal vein was patent with eccentric luminal patency of IVC at this level. The peri-caval fat planes appeared to be intact with medial displacement of pancreatic head and posterior displacement of right adrenal gland. No obvious retroperitoneal/mesenteric lymphadenopathy was noted. Mild diffuse luminal enhancement of infra-renal IVC was noted suggestive of patency of the lower IVC segment. Surgical excision of the tumour was planned. On exploration a 10×6 cm mass was palpated in the suprarenal IVC extending to the left renal vein ostium with development of collaterals. En bloc resection of the tumor

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Fig. 1 Abdominal computed tomography scan showing 10.5×6×6 cm mass in involving Segment I & II of inferior vena cava

with 1 cm cuff of IVC with both the renal veins and closure of the lower end of IVC was performed with excision of a cuff of the liver which was adherent to the IVC. Right renal vein appeared normal with no mass. Reversed saphenous vein graft was anastomosed between the right renal vein to lower end of IVC in an end to side fashion. The patient tolerated the procedure well and was discharged from the hospital. Histopathology revealed tumour mass composed of spindle cells arranged in sheets and fascicles. Tumour cells showed nuclear atypia with increased N:C ratio, hyperchromatic nuclei and significant mitotic activity (10–15 mitotic figure/ 10 hpf in cellular area). The tumour cells were immunopositive for smooth muscle actin, desmin, KI-67 (positive, 12–15 %), CD 34 (focal weak positive) and negative for S100, consistent with Leiomyosarcoma. On follow up of our patient for 1 month an intravenous pyelogram was performed which revealed bilateral normally functioning kidneys and urine analysis was within normal limits.

Discussion

Leiomyosarcoma of the inferior vena cava is a rare malignant tumour arising from the smooth muscle of the media [3]. Leiomyosarcomas of the IVC can arise in the lower (below the renal vessels), middle (renal vessels to retrohepatic IVC), and upper (supra-hepatic) portions of the IVC. Due to their location deep in the retroperitoneum these tumours have an

insidious presentation, with symptoms occurring at later stages of the disease [5]. Leiomyosarcoma of the IVC occurs more commonly in the middle segment [6]. Tumour may be extrinsic or intrinsic or may have both components [6]. Hematogenous metastasis has been reported, especially to liver, lung, bone, lymph node and brain. At advance stages lymphatic spread has been documented [7]. Primary leiomyosarcoma of the IVC are reported to occur more often in the fifth and sixth decade of life with a female predominance [3]. Infrarenal leiomyosarcoma are often dormant for a protracted period and may cause only venous obstruction at a later stage. Tumour involving the middle segment may involve the renal vein and if occlusion occurs present with nephrotic syndrome, which was not seen in our patient. Tumours in the upper segment of IVC gives rise to various degree of Budd Chiari syndrome due to hepatic vein thrombosis [6]. Until the introduction of USG and CT, IVC leiomyosarcoma was most commonly diagnosed during laparotomy or autopsy [6]. Leiomyosarcomas tend to enlarge via intraluminal or extraluminal growth rather than infiltration [8]. Modern imaging modalities using CT, MRI individually or in combination with cavography, ultrasound and echocardiography, allow for an early and accurate preoperative diagnosis resulting in high rate of surgical resection and improved survival [4]. Recent progress of diagnosed investigations such as spiral CT with reconstruction will be able to further define the tumour extent and aid in defining the most

Fig. 2 Gross specimen 10×6.5×6 cm IVC tumour

Fig. 3 Microphotograph showing spindle cells with atypia arranged in fascicles consistent with diagnosis of leiomyosarcoma

Fig. 4 Intravenous pyelogram taken 1 month after surgery showing bilateral normally functioning kidney

appropriate treatment approach. In the present case CT reconstruction was done prior to surgery. Despite advances in imaging technology histopathology with IHC confirmation is required for formal diagnosis. Leiomyosarcoma on histopathology reveals spindle cell tumours which are positive for smooth muscle activity including desmin, smooth muscle actin [1] which was positive in our case in addition to KI-67 and CD34. Aggressive surgical approach is recommended in the light of the tumour slow growth pattern and relatively low metastatic potential [9]. Complete surgical resection with tumour free margin of 1 cm is the treatment of choice [10] which was performed in our case. Complete resection of tumour is often possible in lower segment. If the tumour involves the upper segment complete excision is not possible due to frequent extension to hepatic vein and right side of heart. In middle segment more complicated en-bloc resection along with right kidney is required if the renal vein is involved [6]. Extensive collateral venous drainage preserves left kidney preserves renal function in middle caval tumours [9]. In our case the right kidney was preserved and reversed great saphenous vein graft from the right limb was put end to end with the right renal vein and end to side to the lower segment of IVC. To our knowledge this procedure has not been mentioned in literature of management of IVC tumours.

Conclusion

Leiomyosarcoma of IVC is an extremely rare entity. Patients often present with non-specific symptoms. Various imaging modalities have allowed early diagnosis, staging, treatment planning and subsequently better outcomes. Both the kidneys can be preserved with careful planning as was in our case wherein we avoided a right sided nephrectomy as we placed a venous graft between the right renal vein and the infrarenal inferior vena cava. This procedure has not been reported in literature till date.

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