Primary Leiomyosarcoma of inferior vena cava: a Case of Rare Malignancy

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ABSTRACT

Vascular leiomyosarcoma is a rare tumor, only about 200 cases have been reported worldwide. It commonly arises from inferior vena cava (IVC). There is a strong predilection for females and middle segment of IVC is most commonly affected. The diagnosis is often challenging as patients present with non-specific complaints such as dyspnoea, abdominal discomfort, or back pain. Computed tomography (CT) and Magnetic resonance imaging (MRI) individually or in combination with cavography, ultrasonography (US) and echocardiography, allow an early preoperative diagnosis. Here in, we present a rare case report of leiomyosarcoma of IVC in a 70 year old male patient who presented with vague abdominal discomfort. Ultrasound of abdomen detected a mass in retro peritoneum with mixed echogenicity. CT scan revealed a 10.5 x 6 x 6 cm lobulated, heterogeneously enhancing luminal mass with necrotic changes in supra-renal segment of IVC. Diagnosis of primary Vena caval tumor was made. En bloc resection of the tumor with 1 cm cuff of IVC with closure of the lower end of IVC was performed with excision of a cuff of the liver which was adherent to the IVC. Specimen grossly revealed a soft tissue mass adherent to vessel measuring 10 x 6.5 x 6 cm. External surface was smooth and bosselated. Cut section was nodular, grey white and firm in consistency. Microscopically diagnosis of Leiomyosarcoma was made. IHC study revealed positivity for Smooth muscle actin and desmin and Ki 67 positivity in 12-15 % of tumor cells and was consistent with diagnosis of Leiomyosarcoma.

Key words: Desmin, Inferior vena cava, Leiomyosarcoma, Smooth muscle actin, Vascular tumor.

INTRODUCTION

Leiomyosarcoma of vascular origin is a rare tumor, with its common origin from smooth muscle cells of inferior vena cava (IVC). Since 1871, only about 200 cases have been reported worldwide.[1] Leiomyosarcoma are most frequently encountered in sixth decade with a female predominance.[2] The diagnosis is often challenging as patients present with non-specific complaints such as dyspnoea, abdominal discomfort, or back pain.[3] Computed tomography (CT) and Magnetic resonance imaging (MRI) individually or in combination with cavography, ultrasonography (US) and echocardiography, allow an early preoperative diagnosis.[3] Histopathological and IHC examination further confirms the diagnosis of leiomyosarcoma. Herein, we present the clinical, radiological and histopathological features of an unusual entity of leiomyosarcoma of the IVC.

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CASE REPORT

A 70 year old male presented with history of vague abdominal discomfort since one year. He had no other significant medical and surgical history. Ultrasound of abdomen detected a mass in retro peritoneum with mixed

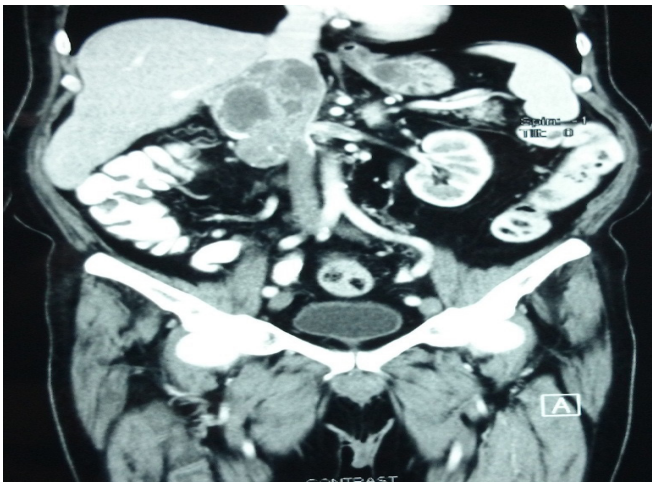

Figure 1: CT scan revealing a lobulated luminal mass in the supra-renal IVC

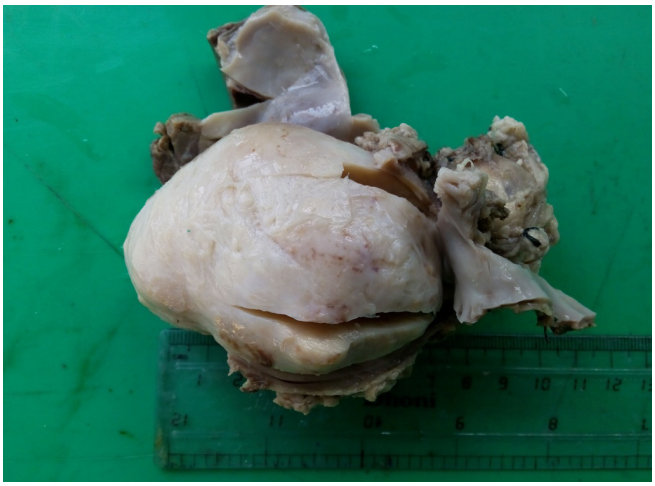

Figure 2: Gross photograph showing a nodular tumor arising from wall of IVC.

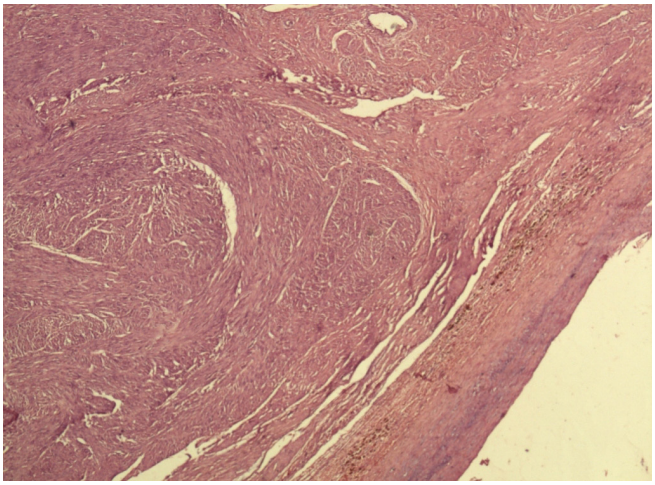

Figure 3: Scanner view showing spindle cell tumor arising from wall of IVC (H&E 40x)

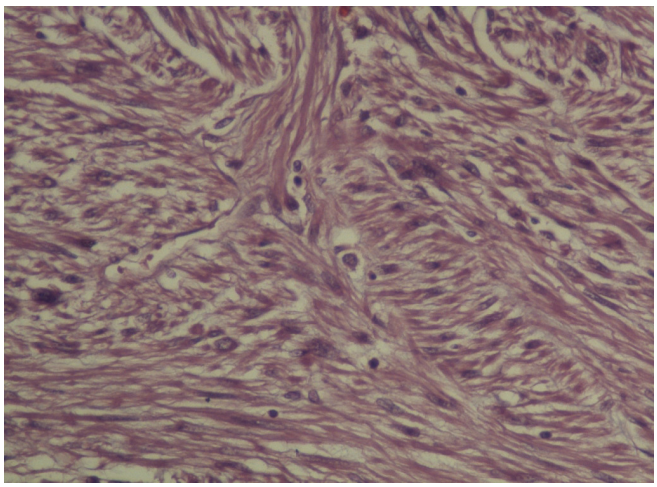

Figure 4: High power view showing spindle shaped tumor cells arranged in fascicles with nuclear atypia and significant mitotc figures. (H&E 400x)

echogenicity. CT scan of the abdomen with intravenous contrast was performed which showed a 10.5 x 6 x 6 cm lobulated, heterogeneously enhancing luminal mass with necrotic changes, in the supra-renal IVC (segment 2) distending and obliterating the lumen with extension into the distal portion of left renal vein (Figure 1). The superior margin of lesion extended into the proximal hepatic segment of IVC beyond which the veins showed normal opacification with normal right side cardiac chambers. No obvious retroperitoneal lymphadenopathy was noted. Diagnosis of primary Vena caval tumor was made with differentials included Leiomyosarcoma and angiosarcoma.

Surgical excision of the tumor was planned. On exploration a 10 x 6 cm mass was palpated in suprarenal IVC extending to left renal veins with development of collaterals. En bloc resection of the tumor with 1 cm cuff of IVC with closure of the lower end of IVC was performed with excision of a cuff of the liver which was adherent to the IVC. Reversed saphenous vein graft was anastomosed between the right renal veins to lower end of IVC in an end to side fashion. The patient tolerated the procedure well and postoperative period was uneventful and patient was on oral warfarin with frequent evaluation of PT/INR values.

Specimen was received in surgical pathology section. Grossly, it revealed a soft tissue mass adherent to vessel measuring 10 x 6.5 x 6 cm. External surface was smooth and boss elated. Cut section was nodular, grey white and firm in consistency. Small piece of liver was seen attached at one end, measuring 4 x 2 x 1.1 cm which was grossly unremarkable (Figure 2). Microscopically, sections showed a vessel wall with attached tumor mass. Tumor mass was composed of spindle shaped cells with eosinophilic cytoplasm arranged in fascicles and sheets. Tumor cells displayed nuclear atypia with hyperchromasia, increased

N/C ratio and significant mitotic activity 10-15 mitotic figures/10 hpf in cellular area) (Figure 3,4). Focal area also showed osteoclastic type of giant cells. Sections from liver showed no evidence of tumor tissue. Diagnosis of Leiomyosarcoma was made. IHC study revealed positivity for Smooth muscle actin and desmin and Ki 67 positivity in 12-15 % of tumor cells and was consistent with diagnosis of Leiomyosarcoma.

DISCUSSION

Tumors such as renal carcinomas, pheochromocytomas, or testicular tumors can secondarily invade the vena cava. Leiomyosarcoma of the IVC is a primary vascular tumor that is relatively rare.[1] It is frequently seen in the sixth decade of life with a female predominance.[2] Clinical findings are non specific and may precede the diagnosis by several years. Symptoms and resectability depend on location and extent of tumor as well as associated thrombosis. Therefore, it is better to divide the IVC into three segments i.e. a lower segment below renal veins, a middle segment from the renal vein up to hepatic veins and upper segment from the level of hepatic veins to right atrium. Leiomyosarcoma of the IVC most frequently occur in middle segment.[4] Infra renal leiomyosarcomas are often dormant for a protracted period and may cause only venous obstruction at a later stage. If the tumor is located in middle segment, the renal vein can be involved and if occlusion occurs, the patient can present with nephrotic syndrome. Tumor in the upper segment give rise to varying degree of Budd Chiari syndrome due to hepatic vein thrombosis.[2,4]

With the development of imaging modalities like USG, CT and MRI, preoperative diagnosis of Leiomyosarcoma is possible. CT scan clearly delineates the intravascular tumor, which is usually large, lobulated and sometimes heterogeneous owing to haemorrhage and necrosis. The tumor is usually hypovascular in nature, but may show peripheral enhancement following contrast injection.[2] Leiomyosarcoma with extra vascular development may be much more difficult to differentiate from retroperitoneal tumors compressing or invading IVC.[4,5]

The final diagnosis can be made by ultrasound or CT guided biopsy.[4,5] Histopathological examination reveals tumor made up of spindle cells arranged in fascicles. Typical cell of Leiomyosarcoma is elongated and has abundant cytoplasm that varies from pink to deep red in sections stained with haematoxylin-eosin. The nucleus is usually centrally located and blunt ended. By definition, Leiomyosarcoma possess some degree of nuclear atypia but mitotic activity varies considerably. However, even very low level of mitotic activity (<1/10HPF) in the face of significant atypia is sufficient for evidence of malignancy. Immunohistochemical examination reveals positivity for smooth muscle actin and desmin in one half to nearly 100% of tumors.[6] In present case, tumor cells displayed nuclear atypia with 10-15 mitotic figures/10 hpf and IHC stains were positive for smooth muscle actin and desmin.

Complete surgical resection with a tumor-free margin of 1 cm is the treatment of choice[2] which was performed in the present case. The surgical resectability is highly dependent on location of the tumor. Complete resection of the tumor is often possible in lower segment. In the middle segment, a more complicated en-block resection along with right kidney is usually needed if the renal vein is involved. If the tumor involves the upper segment, complete resection is usually not possible due to frequent extension into hepatic veins and right side of the heart.[7] In present case, right kidney was preserved and great saphenous vein graft from lower limb was put end to end with right renal vein and lower segment of IVC. Neoadjuvant therapy may be given to downsize the tumor and increase resectability rates. Nonetheless, when complete resection is not possible, debulking combined with radiation therapy still provides good palliation.[2]

CONCLUSION

Vascular leiomyosarcoma is a rare tumor, arising most frequently from IVC. Hence an accurate imaging and histopathological diagnosis is essential for improving the patient survival.

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