International Journal of Research in Medical Sciences
Kumar R et al. Int J Res Med Sci. 2017 Apr;5(4):1702-1705
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20171292

Case Report

Primary leiomyosarcoma in the inferior vena cava- a case report

Ravindra Kumar[1]*, Suman Chaudhary[1], Hemant Kumar Mishra[1], Divesh Goyal[2]

[1]Department of Radio-diagnosis and Imaging, Mahatma Gandhi medical College and Hospital, Jaipur, Rajasthan, India
[2]Department of Oncology and Imaging, Mahatma Gandhi medical College and Hospital, Jaipur, Rajasthan, India

Received: 20 January 2017
Accepted: 03 March 2017

***Correspondence:**
Dr. Ravindra Kumar,
E-mail: ravindra2327@yahoo.com

ABSTRACT

Vascular leiomyosarcoma of IVC is a rare tumor. Only about 200 cases have been reported worldwide. 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. A 43-year-old male presented with history of vague abdominal distention, discomfort, and dyspnoea in our hospital. Ultrasound of abdomen detected a mass in retro peritoneum with mixed echogenicity inseparable from IVC. CT Imaging examination of abdomen revealed a large elongated, heterogenous mass extending from right atrium to the confluence of common iliac veins. in IVC reaching up to inferior border of right atrium. Then USG guided biopsy done and Leiomyosarcoma of IVC was diagnosed by immunohistochemistry. Due to extensive involvement of intrahepatic segment and right atrium complete resection of mass was not possible, so radiotherapy followed by chemotherapy was started. Patient started deteriorating due to intolerability with poor response to radiotherapy and chemotherapy so he was put on supportive treatment.

Keywords: Inferior vena cava, Leiomyosarcoma, Right atrium, Vascular tumor

INTRODUCTION

Leiomyosarcoma is a rare, malignant mesenchymal tumor. Only 200 cases had been re-ported as of 1996.[1] Leiomyosarcoma of vascular origin often occurs in the inferior vena cava, and one that originated from the wall of the inferior vena cava was first reported by Perl in 1871.[2]

Leiomyosarcoma are most frequently encountered in sixth decade with a female predominance.[3] The diagnosis is often challenging as patients present with non-specific complaints such as dyspnoea, abdominal discomfort, or back pain.

We herein report a case of 43-year-old male with primary leiomyosarcoma of IVC and extending up to the inferior margin of right atrium.

CASE REPORT

A 43-year-Old male presented with history of vague abdominal distension, discomfort and dyspnea since one year. He had no other significant medical and surgical history. Ultrasound of abdomen detected a mass in retro peritoneum with mixed echogenicity. Doppler study shows low resistance flow in mass.

CT scan of the abdomen with intravenous contrast was performed which shows a 19x6.7x8.5cm (L/CC X AP X TD) lobulated, heterogeneously enhancing mass lesion with intensely enhancing solid component and non-enhancing necrotic component seen in intra hepatic, supra hepatic and infra hepatic part of IVC causing widening and obstruction of lumen. The tumor is extending into right renal vein, proximal part of left renal vein, right supra renal vein and adjacent part of hepatic vein so these

are not opacified by contrast. Superiorly tumor is extending in inferior part of right atrium and inferiorly up to bifurcation.

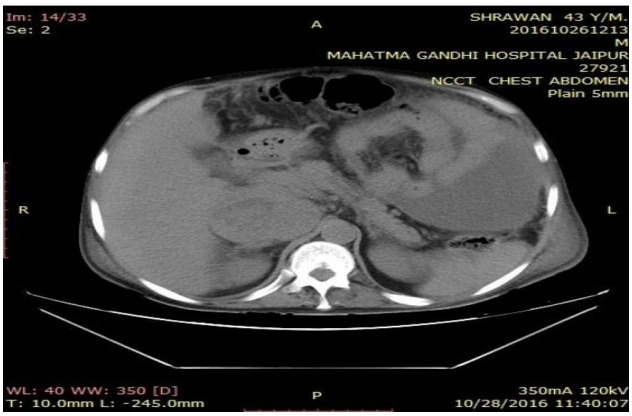

Figure 1: NCCT axial scan shows mass lesion in IVC lumen causing widening and obstruction.

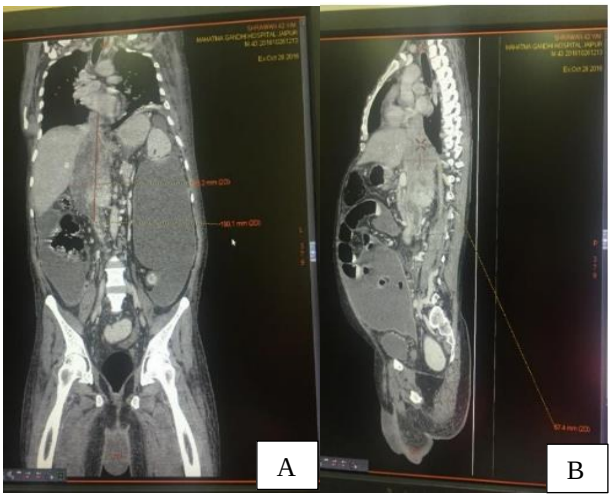

Figure 2(A, B): CECT abdomen showing heterogeneously enhancing mass lesion in inferior vena cava causing widening and obstruction reaching upto right atrium (measuring 19x6.7x8.5cm).

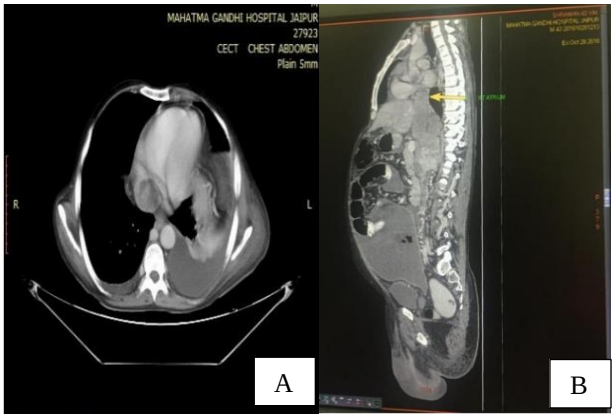

Figure 3(A, B): CECT axial scan showing the heterogenous mass lesion in IVC reaching up to inferior border of right atrium.

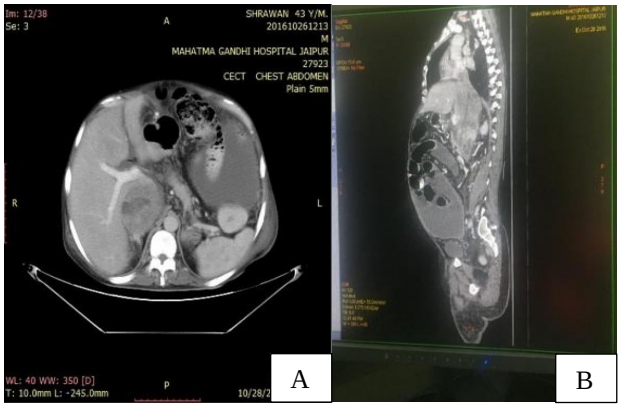

Figure 4(A, B): Axial and sagital CECT scan abdomen showing- tumor is involving the hepatic veins so are not opacified properly.

Mild right pleural effusion, moderate left pleural effusion, mild pericardial effusion and moderate to gross ascitis. Liver enlarged in size. The hepatic veins are not opacified by contrast. As the complete resection of the tumor was impossible due to hepatic IVC involvement extension into hepatic veins and right atrium.

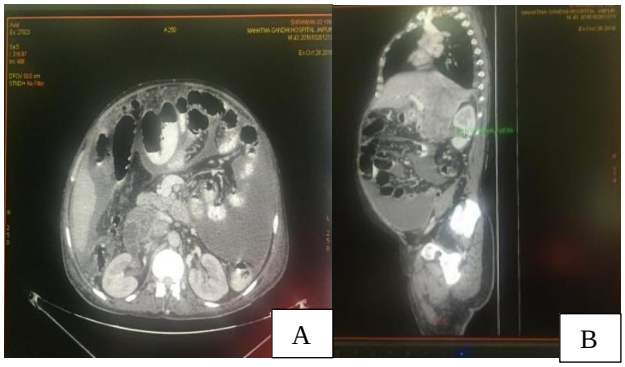

Figure 5(A, B): CECT abdomen axial and sagital scan showing the tumor is extending into right renal vein, proximal part of left renal vein, right supra renal vein.

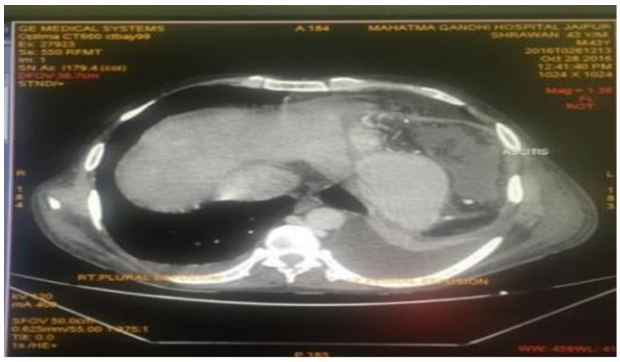

Figure 6: CECT abdomen axial scan showing mild right pleural effusion, moderate left pleural effusion, mild pericardial effusion and moderate to gross ascitis.

The patient underwent radiotherapy (55 Gy/25 Fr), as well as chemotherapy comprising 3 courses of 60 mg/m^2

doxorubicin and Ifosfamide three times a weekly as first-line chemotherapy, which led to a partial response. But patient was lost to follow-up for six month. Again, came back after six month with gross ascites, bilateral pleural effusion and reinvestigation evaluation- suggestive of progressive disease.

In view of poor general condition- patient was put on only supportive treatment with due course of time death occurred. Leiomyosarcoma- Axial CT shows expansion of the IVC lumen by heterogeneously contrast- enhancing partially necrotic mass.

DISCUSSION

Leiomyosarcoma of the IVC is a primary vascular mesenchymal tumor that is relatively rare.[4] It is mainly seen in the sixth decade of life with a female predominance.[3] Clinical findings are nonspecific 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.[3,5]

With the development of imaging modalities like USG, CT and MRI, preoperative diagnosis of Leiomyosarcoma is possible. CT scan clearly delineates the intra luminal vascular tumor, which is usually large, lobulated and sometimes heterogeneous owing to haemorrhage and necrosis. The tumor is usually hypo vascular in nature, may show peripheral enhancement following contrast injection.[3] Two third of tumors will demonstrate predominantly extraluminal growth, and one third will demonstrate predominantly intraluminal growth.[10]

IVC Leiomyosarcoma with extra luminal extension may be much more difficult to differentiate from retroperitoneal tumors compressing or invading IVC. The final diagnosis can be made by ultrasound or CT guided biopsy.[5,6] Histopathological examination reveals spindle cells arranged in fascicles with variable degree of nuclear atypia and mitotic activity.

Immunohistochemical examination reveals smooth muscle actin and desmin in one half to nearly 100% of tumors.[7] In present case, tumor cells displayed nuclear atypia with 10-15 mitotic figures/10 hpf and IHC stains shows for smooth muscle actin and desmin. Complete surgical resection with a tumor-free margin of 1cm is the treatment of choice.[3]

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, like in present case complete resection is usually not possible due to frequent extension into hepatic veins and right side of the heart.[8]

Leiomyosarcoma of the IVC does not respond well to chemotherapy or radiotherapy. Surgical resection is the only choice of if possible.[9]

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.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Mingoli A, Cavallaro A, Sapienza P. International registry of inferior vena cava leiomyosarcoma: analysis of a world series on 218 patients. Anticancer Res. 1996;16:3201-5.
2. Perl L. Ein Fall von Sarkom der Vena cava inferior. Virchows Arch. 871;53:378-83.
3. Hemant D, Krantikumar R, Amita J, Chawla J, Ranjeet N. Primary leiomyosarcoma of inferior vena cava, a rare entity: Imaging features. Australas Radiol. 2001;45(4):448-51.
4. Mingoli A, Cavallaro A, Sapienza P, Di Marzo L, Feldhaus RJ, Cavallari N. International registry of inferior vena cava leiomyosarcoma: analysis of a world series on 218 patients. Anticancer Res. 1996;16(5b):3201-5.
5. Yadav R, Kataria K, Mathur SR, Seenu V. Leiomyosarcoma of Inferior vena cava: A case series of four cases. Indian J Pathol Microbiol. 2012;55(1):83-5.
6. Ahulwalia A, Saggar K, Sandhu P, Gupta K. Primary leiomyosarcoma of inferior vena cava: An unusual entity. Vascular. 2002;12(4):15-6.
7. Weiss SW, Goldblum JR. Enzinger and Weiss's soft tumor: 5th ed. Mosby Elsevier: Philadelphia, USA; 2008:545-562.
8. Monig SP, Gawenda M, Erasmi H, Zieren J, Pichlmaier H. Diagnosis treatment and prognosis of leiomyosarcoma of inferior vena cava. Eur J Surg. 1995;161(4):231-5.
9. Shindo S, Matsumoto H, Ogata K. Surgical treatment of retroperitoneal leiomyosarcoma

invading the inferior vena cava: report of three cases. Surg Today. 2002;32:929-33.
10. Smillie RP, Shetty M, Boyer AC, Madrazo B, Jafri SZ. Imaging evaluation of the inferior vena cava. RG. 2015;35(2):578-92.

Cite this article as: Kumar R, Chaudhary S, Mishra HK, Goyal D. An unusual presentation of hemoglobin SD Punjab in a Saudi Arabian adult. Int J Res Med Sci 2017;5:1702-5.