

Leiomyosarcoma of the Inferior Vena Cava - An Experience from a Tertiary Care Centre in South India

Aleena Elizabeth Andrews¹, Juvaina Puthiyakam², Naufal Perumpalath³, Devarajan Ellezhuthil⁴

¹Department of Radio Diagnosis, Government Medical College, Thrissur, Kerala, India.

^{2, 3, 4} Department of Radio Diagnosis, Government Medical College, Kozhikode, Kerala, India.

ABSTRACT

BACKGROUND

Leiomyosarcomas (LMS) of inferior vena cava (IVC) are rare smooth muscle sarcomas with less than 300 cases described in the literature. Leiomyosarcoma of IVC is, often detected late in the course due to its indolent manifestation. This study intends to discuss the experiences and challenges in diagnosing this rare entity primarily by radiological imaging in a tertiary centre in India.

METHODS

This is a retrospective analysis of computed tomography (CT) and ultrasound findings in 6 cases of leiomyosarcoma of inferior vena cava who were referred for multidetector computer tomography (MDCT) scan in the Department of Radiodiagnosis of Government Medical College Calicut over the past 7 years by the treating physician. The patients were identified using a prospectively maintained database.

RESULTS

In this study, there were six patients diagnosed as leiomyosarcoma of IVC, age ranging from 35 and 64 years (mean 47.8 years, SD 10.7) with 4 (66.6 %) females and 2 (33.33 %) males. The mean size of the tumour at the time of diagnosis was 8.4 cm. The segment of IVC most commonly involved was middle segment in 5 patients (83.3 %). In this series, only a single case had tumour entirely confined within the lumen of inferior vena cava without extraluminal extension. Two out of six cases (16.66 %) had lung metastasis at the time of diagnosis. Two cases without metastasis or significant infiltration to adjacent organs were amenable to resection.

CONCLUSIONS

Leiomyosarcoma of IVC is a rare tumour, often detected late in the course due to its indolent manifestation. High index of suspicion may help in the early diagnosis of so that early treatment can begin and improve the clinical outcome.

KEYWORDS

Leiomyosarcoma, Inferior Vena Cava, Mesenchymal Neoplasms

Corresponding Author:

*Dr. Aleena Elizabeth Andrews,
Mekkattukunne House,
Chakkoth Lane, Poonkunnam,
Thrissur - 680002, Kerala, India.
E-mail: aleenaelizabeth26@gmail.com*

DOI: 10.18410/jebmh/2021/380

How to Cite This Article:

Andrews AE, Puthiyakam J, Perumpalath N, et al. Leiomyosarcoma of the inferior vena cava - an experience from a tertiary care centre in south India. J Evid Based Med Healthc 2021;8(24):2023-2028. DOI: 10.18410/jebmh/2021/380

Submission 19-02-2021,

Peer Review 26-02-2021,

Acceptance 26-04-2021,

Published 14-06-2021.

Copyright © 2021 Aleena Elizabeth Andrews et al. This is an open access article distributed under Creative Commons Attribution License [Attribution 4.0 International (CC BY 4.0)]

BACKGROUND

Leiomyosarcomas are mesenchymal neoplasms of smooth muscle cell origin.¹ Retroperitoneum is a common location for sarcomas, attributing to about 12 – 69 % of cases.² Primary vascular LMS is a rare entity of which, inferior vena cava (IVC) is the most common site.³ Leiomyosarcoma of the inferior vena cava, described in fewer than 300 patients in the literature, is a rare clinical scenario.⁴ It is a slow growing tumour, with indolent clinical manifestation. They are identified incidentally, once they are large enough to cause mass effect on adjacent structures.⁵ However, imaging done for non-specific complaints, is probably the only way to catch these lesions at its inception, offering best possible surgical cure preserving adjacent vital organs.

This study intends to discuss the experiences and challenges in diagnosing this rare entity primarily by radiological imaging.

METHODS

A retrospective chart review of patients who underwent multidetector computer tomography scan in the Department of Radiodiagnosis of Government Medical College Calicut over the past 7 years between January 2013 to December 2020, referred by treating physician was performed in this descriptive case series. The patients diagnosed with leiomyosarcoma of IVC were identified using a prospectively maintained database.

MDCT was performed (16 row, slice thickness 5 mm, rotation time 0.5 s, collimation 16 × 1.25 mm, pitch 1.75 : 1, interval 2.5 mm) before and after administration of intravenous iodinated contrast medium with enhanced scans acquired at 30 and 70 s (50 ml contrast medium bolus, flow rate of 3 ml/s) was performed.

Ultrasound images provided in some cases, were obtained prior to acquisition of CT, using 3.5 Hz curvilinear probe.

RESULTS

A total of 75,562 patients underwent contrast enhanced computed tomography of abdomen from January 2013 to December 2020. Excluding cases of CT performed for abdominal trauma, known case of gastrointestinal/hepatic/pelvic malignancy there were 59,962 cases who were evaluated for non-specific complaints. Using prospectively maintained data base, only 6 cases were diagnosed as leiomyosarcoma of IVC.

The mean age of patients diagnosed as leiomyosarcoma of IVC was 47.8 years (SD 10.7) ranging from 35 and 64 years. There were 4 (66.6 %) females and 2 (33.33 %) males. The mean size of the tumour at the time of diagnosis was 8.4 cm. The most common presentation was non-specific abdominal pain.

Physical examination revealed lower limb oedema in 4 subjects (66.6 %). Palpable mass on physical examination

was detected in 2 out of 6 cases. The segment of IVC most commonly involved was middle segment in 5 patients (83.3 %). In this series, only a single case had tumour entirely confined within the lumen of inferior vena cava without extraluminal extension. Two out of six cases (16.66 %) had lung metastasis at the time of diagnosis. Two cases without metastasis or significant infiltration to adjacent organs were amenable to resection.

A summary of the demographic data, clinical presentation, and treatment of all the participants is given in Table 1.



Figure 1. A 48-year-old female with complaints of right sided abdominal pain and a large mass palpable in the right lumbar and epigastric region: coronal contrast enhanced computed tomography of abdomen shows the intraluminal component of lesion (black arrows) distending the middle segment of IVC with the ill-defined extraluminal component (yellow arrow) of the lesion abutting the abdominal aorta



Figure 2a. A 38-year-old female complaining of abdominal pain of 2 months duration (a): ultrasound with colour doppler showing a well-defined hypoechoic lesion in the right anterior pararenal space with intraluminal extension into infrahepatic inferior vena cava



Figure 2b. A 38-year-old female complaining of abdominal pain of 2 months duration (b): axial contrast enhanced computed tomography of abdomen showing heterogeneously enhancing lesion (black arrow) at the right suprarenal location closely abutting the IVC



Figure 3. A 55-year-old male complaining of right sided abdominal pain, vague abdominal mass, and bilateral pedal oedema, for a duration of one month: coronal contrast enhanced computed tomography of abdomen shows the heterogeneously enhancing lesion (black arrow) involving the suprarenal IVC (white arrow)

Parameter	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Age	48	38	55	47	35	64
Sex	F	F	M	M	F	F
Symptoms	Abdominal pain	Abdominal pain	Right sided abdominal pain	lower abdominal pain	Back pain	Right sided abdominal pain and bilateral pedal oedema
Physical examination	Large mass palpable in the right lumbar, epigastric and umbilical region.	Nil	vague mass in the right hypochondrium and bilateral pedal oedema	Bilateral lower limb oedema	Oedema of bilateral lower limbs	Bilateral pedal oedema
Duration	One month	Two months	One month	One month	One month	Two months
Ultrasound	Mass extending to retroperitoneum and intraluminal into hepatic IVC.	well defined hetero echoic lesion in the right anterior pararenal space with luminal extension of lesion into IVC	Heteroechoic lesion in the retroperitoneum, in the right sub hepatic region filling and distending the hepatic and intrahepatic IVC	Heteroechoic lesion in the right sub hepatic location.	Heteroechoic lesion involving right suprarenal location with extension to lumen of IVC.	Hypoechoic lesion in the paraaortic location filling the lumen of IVC
MDCT	Heterogeneously enhancing soft tissue density lesion with necrosis epicentred in the anterior pararenal space of retroperitoneum measuring 17.7 x 12.5 cm x 16.5cm, with a component extending into and distending the lumen of middle segment of IVC.	Heterogeneously enhancing lesion filling the luminal aspect of IVC with extraluminal component measuring 2.5 X 3.8 x 2.7 cm.	Heterogeneously enhancing lobulated soft tissue density lesion with areas of necrosis filling the lumen of middle segment of IVC causing its distension and compressing right renal vein measuring 6.9 x 5.6 x 5.8 cm.	Lobulated heterogeneously enhancing soft tissue density lesion measuring 8.7 x 13.5 x 13.7 cm in the right side of retroperitoneum with areas of necrosis; the lesion was involving hepatic, suprarenal and infrarenal IVC, with extension to right renal vein.	Heterogeneously enhancing lesion in the right suprarenal location measuring 2.7 x 4.2 x 3.6 cm with extension of the lesion into the suprarenal segment of IVC	Heterogeneously enhancing soft tissue density lesion with necrosis in the intraluminal compartment of IVC with extraluminal extension of middle segment of IVC measuring 4.5 x 3.2 x 3.4 cm
Involved IVC segment	Middle segment of IVC	Middle segment of inferior vena	Middle segment of IVC	Upper, Middle and lower segment of IVC	Middle segment of IVC	Middle segment of IVC
Extra luminal extension/ organ involvement	Hepatic and lung metastasis	Well defined small extraluminal component without infiltration of adjacent organs.	In the sub hepatic location with ill-defined fat planes with adjacent liver, infiltrating into lumen of suprarenal IVC, extensive deposits in mesentery and nodules in lung.	Intraluminal extension up to the iliac vein and extraluminal to right lobe of liver and right kidney	Nil	Extraluminal component of lesion having well defined fat planes with adjacent structures
Management	USG guided biopsy showing atypical spindle cell neoplasm with smooth muscle differentiation consistent with leiomyosarcoma: Inoperable -palliative care with chemotherapy	IVC excision of tumour with reconstruction of vein. Histopathological analysis showed atypical spindle cell neoplasm with smooth muscle differentiation, SMA positive and MIB labelling index around 90%	USG guided biopsy showing atypical spindle cell neoplasm with smooth muscle differentiation. Inoperable-chemotherapy	USG guided biopsy showing spindle cell neoplasm. Inoperable-chemotherapy.	IVC excision of tumour with reconstruction of vein. Histopathological analysis showed atypical spindle cell neoplasm with smooth muscle differentiation, SMA positive	USG guided biopsy showing spindle cell neoplasm. Chemotherapy

Table 1. Demographics, Clinical and Imaging Features of Subjects Diagnosed with IVC Leiomyosarcoma



Figure 4. A 47-year-old male presented to our hospital with complaints of lower abdominal pain of one-month duration associated with loss of weight, bilateral lower limb oedema: coronal contrast enhanced computed tomography of abdomen showing heterogeneously enhancing lesion involving the middle and inferior segments of IVC (black arrow) with extraluminal component of the lesion (white arrow) in the sub hepatic location



Figure 5. A 35-year-old female presented with chief complaints of upper abdominal pain and bilateral pedal oedema. On physical examination she had bilateral pedal oedema. axial contrast enhanced computed tomography of abdomen showing an enhancing lesion involving the suprarenal IVC distending its lumen.



Figure 6. A 64-year-old female with comorbidities of coronary artery disease, hypertension, and diabetes, complained of right abdominal discomfort of 2 months' duration and bilateral pedal oedema. Axial contrast enhanced computed tomography of abdomen shows a lesion within lumen of the middle segment of IVC (black arrow) with a component extending outside (white arrow).

Case 1

A 48-year-old female was presented to our hospital with complaints of right sided abdominal pain. On physical examination, a large mass was palpable in the right lumbar and epigastric region. Ultrasound evaluation of abdomen showed a large sub hepatic mass extending to retroperitoneum showing both intraluminal component in the hepatic IVC and the extraluminal component.

For further characterisation of the lesion, contrast enhanced computed tomography of abdomen was performed, which showed the lesion extending to inferior vena cava distending its lumen with a large sub hepatic extraluminal component (figure 1). The lung fields bilaterally showed focal nodules of varying sizes.

She underwent USG guided biopsy of the lesion. Histopathology was consistent with atypical spindle cell neoplasm with smooth muscle differentiation, myxoid changes with SMA positive and MIB labelling index around 90 %, a diagnosis of leiomyosarcoma was made.

Case 2

A 38-year-old female complaining of abdominal pain of 2 months duration. Local physical examination revealed no significant abnormality.

On routine ultrasound evaluation, a well-defined hetero-echoic lesion was detected in the right anterior pararenal space, in the region of head of pancreas with intraluminal extension into infrahepatic IVC (figure 2a).

Further evaluation with contrast enhanced computed tomography of abdomen showed a heterogeneously enhancing lesion at the level of pancreas in the anterior pararenal space with extension into lumen of inferior vena cava (figures 2b). She underwent IVC excision of tumour with interposition of graft repair of IVC using 14 mm coated polyester graft-right renal vein reimplantation polyester graft bypass to left renal vein.

Histopathology report was consistent with leiomyosarcoma.

Case 3

A 55-year-old male complaining of right sided abdominal pain, vague abdominal mass and bilateral pedal oedema for a duration of one month. Physical examination revealed pallor, a vague mass palpable in the right hypochondrium.

Ultrasound evaluation with curvilinear probe in the right hypochondrium showed a hetero-echoic lesion in the right sub hepatic location with lost fat planes within the liver.

Evaluation of the lesion by contrast enhanced computed tomography revealed heterogeneously enhancing lobulated lesion with ill-defined borders in the right suprarenal location, abutting the right kidney and infiltrating the right lobe of liver. Coronal sections confirmed the that the epicentre of the lesion was in the lumen of middle segment of inferior vena cava with a large extraluminal component showing infiltration of adjacent structures (figure 3). Further his lung fields showed few nodules and right pleural effusion.

She underwent USG guided biopsy of the lesion and the histopathology was consistent with leiomyosarcoma. Since

the tumour was extensive at the time of diagnosis with infiltration of adjacent structures and distant metastasis, she was offered with palliative chemotherapy.

Case 4

A 47-year-old male presented to our hospital with complaints of lower abdominal pain of one-month duration associated with loss of weight and physical examination revealed mild pallor and bilateral pitting pedal oedema.

On routine USG evaluation, a hetero-echoic lesion was detected in the right sub hepatic location with few hyperechoic foci within, having well-defined planes with right lobe of liver.

Contrast enhanced computed tomography of abdomen showed a well-defined hypodense lesion with few hyperdense foci, corresponding to calcification in the right suprarenal location. Coronal sections demonstrated the enhancing lesion to be in the lumen the middle and inferior segments of IVC with a large extraluminal component in the sub hepatic location (figures 4).

Ultrasound guided biopsy revealed atypical spindle cell neoplasm with smooth muscle differentiation, consistent with leiomyosarcoma. He was offered chemotherapy.

Case 5

A 35-year-old female presented with chief complaints of upper abdominal pain. On physical examination she had bilateral pedal oedema.

Routine ultrasound evaluation of abdomen revealed a lesion in the right hypochondrium.

For further characterisation of lesion, she was referred for MDCT abdomen which showed an enhancing lesion involving the suprarenal IVC distending its lumen (figure 5).

She underwent surgery with excision of the tumour with graft repair of the IVC. Histopathology was consistent with leiomyosarcoma.

Case 6

A 64-year-old female with co morbidities of coronary artery disease, hypertension and diabetes complaints of right abdominal discomfort for 2 months duration. On examination, bilateral pedal oedema was present.

Ultrasound evaluation showed a well-defined hypoechoic lesion involving the IVC in the paraaortic location.

Axial contrast CT sections of abdomen shows the lesion within lumen of the middle segment of IVC (black arrow) with a component extending outside (white arrow) (figure 6). Ultrasound guided biopsy showed features consistent with leiomyosarcoma. She was treated palliatively with chemotherapy, in view of co morbidities and lack of cooperation by her relatives for surgery.

DISCUSSION

Leiomyosarcomas are mesenchymal neoplasms of smooth muscle cell origin.¹ Retroperitoneum is a common location

for sarcomas, attributing to about 12 - 69 % of cases. It is the second most common tumour of the sarcomas affecting the retroperitoneum.² Primary vascular LMS is a rare entity of which, inferior vena cava (IVC) is the most common site.⁵ Leiomyosarcoma of the inferior vena cava, described in fewer than 300 patients in the literature, is a rare clinical scenario.⁴

It is a slow growing tumour, and being located in the retroperitoneal space, symptoms are mostly non-specific and are identified incidentally once they are large enough to cause mass effect on adjacent structures.⁶

It is usually diagnosed with imaging modalities like computed tomography, magnetic resonance imaging, alone or in combination with ultrasonography, echocardiography. Thus, it becomes challenging due to its non-specific and wide varying clinical presentation, often preceding the diagnosis by months or years.⁵ LMS of the IVC often are detected incidentally and are quite large at the time of diagnosis. In several studies, tumours were > 10 cm. In our study the average size of tumour detected was 9.3 cm.⁷

As per Teixeira et al. it most commonly affects females in the sixth decade. Occasionally, LMS of the IVC occurs in young patients, with few or no comorbidities, as localized disease.

In our study also, the majority affected were females.

The three major growth patterns described are extraluminal 62 %, intraluminal 5 % and combined 33 %. Based on the localisation along the IVC, these tumours can be further classified as involving the upper segment, spanning between hepatic veins to IVC, middle segment between the hepatic veins and renal veins and lower segment including the IVC below the renal veins.⁴

As per Yadav et al. LMS of IVC most frequently affects the middle segment, as is seen in our case.

Clinical presentation mainly depends on location, and is seen to be non-specific, which includes lower limb oedema, abdomen pain, mass, back pain, renal hypertension, and dyspnoea.

The most common route of metastasis is haematogenous, and is frequently to liver, lung bone, and brain. In advanced stages, the tumour may spread through lymphatics. In our case series we have found that two cases had metastasis at the time of diagnosis, and the preferentially involved site was lungs.

Leiomyosarcoma is a slow growing hypovascular tumour, sometimes hypervascular, corresponding to the involved segment and supplying arteries. Sonography may detect the tumour as a mixed hypoechogenic mass in the retroperitoneum. Detection of luminal extension of lesion to IVC, may help in clinching the diagnosis, but with many limitations.

Radiological techniques play an important role in the accurate diagnosis of LMS, particularly with cross sectional imaging modalities like CT or MRI. It can delineate an irregular-shaped heterogeneous lobulated soft-tissue mass in the IVC, expanding its lumen accompanied with peripheral post contrast enhancement and non-enhanced necrotic or cystic areas.⁸

Leiomyosarcoma with predominant extraluminal component can be challenging to differentiate from other retroperitoneal tumours compressing the inferior vena cava.

As per Quian et al. among the differentials of intraluminal mass of IVC, includes leiomyosarcoma, angiosarcoma, tumour thrombus and bland thrombus.

The definitive diagnosis can be made only by USG or CT guided biopsy. The primary modality of treatment is surgery, and its resectability is determined by the location of the tumour, with those in the lower segment being completely resectable.

Surgery is currently the only potentially curative therapy.⁹ Combination of radical resection of tumour with adjuvant chemotherapy is the treatment of choice for disease without metastasis at the time of diagnosis. The aim when approaching IVC LMS include achieving local control, maintaining the patency of major venous flow.¹⁰ Though resistant to chemotherapy or radiotherapy, neoadjuvant therapy may be provided to downsize the tumour, prior undertaking surgical procedure.

CONCLUSIONS

Leiomyosarcoma of IVC is a rare tumour, often detected late in the course due to its indolent manifestation. However, imaging done for non-specific complaints, is probably the only way to catch this lesion at its inception, offering best possible surgical cure preserving adjacent vital organs.

Data sharing statement provided by the authors is available with the full text of this article at jebmh.com.

Financial or other competing interests: None.

Disclosure forms provided by the authors are available with the full text of this article at jebmh.com.

REFERENCES

- [1] Sephien A, Mousa MS, Bui MM, et al. Leiomyosarcoma of the inferior vena cava with hepatic and pulmonary metastases: Case report. *J Radiol Case Rep* 2019;13(5):30-40.
- [2] Marko J, Wolfman DJ. Retroperitoneal leiomyosarcoma. *Radiographics* 2018;38(5):1403-1420.
- [3] Kapoor R, Bansal A, Sharma SC. Leiomyosarcoma of inferior vena cava: case series of four patients. *J Cancer Res Ther* 2015;11(3):650.
- [4] Cortecero JM, Rubio MDG, Romá AP. Leiomyosarcoma of the inferior vena cava: AIRP best cases in radiologic-pathologic correlation. *Radiographics* 2015;35(2):616-620. <http://dx.doi.org/10.1148/rg.352130105>
- [5] Sessa B, Iannicelli E, Caterino S, et al. Imaging of leiomyosarcoma of the inferior vena cava: comparison of 2 cases and review of the literature. *Cancer Imaging* 2010;10(1):80-84.
- [6] Yadav R, Kataria K, Mathur S, et al. Leiomyosarcoma of inferior vena cava: a case series of four cases. *Indian J Pathol Microbiol* 2012;55(1):83.
- [7] Nascif RL, Antón AGS, Fernandes GL, et al. Leiomyosarcoma of the inferior vena cava: a case report. *Radiol Bras* 2014;47(6):384-386.
- [8] Teixeira FJR Jr, do Couto NSD, de Freitas PAL, et al. Leiomyosarcoma of the inferior vena cava: survival rate following radical resection. *Oncol Lett* 2017;14(4):3909-3916.
- [9] Tameo MN, Calligaro KD, Antin L, et al. Primary leiomyosarcoma of the inferior vena cava: reports of infrarenal and suprahepatic caval involvement. *J Vasc Surg* 2010;51(1):221-224.
- [10] Singla B, Bansal A. An insight into the inferior vena cava leiomyosarcoma. *Niger J Surg Sci* 2014;24(1):18-22.