CASE SERIES

# Leiomyosarcoma of the Inferior Vena Cava and its tributaries, a case series and review of the literature.

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## Introduction

Leiomyosarcoma of the Inferior Vena Cava (IVC) (1) and its tributaries are rare accounting for only 0.5% of all adult sarcomas. However, it is the commonest primary tumour of the IVC . Due to its rarity, large series describing such cases are lacking. The diagnosis is often delayed and incorrect diagnoses are made due to its nonspecific nature of the presentation. This report describes a series of three patients who presented with nonspecific abdominal symptoms and diagnosed to have primary leiomyosarcoma of the IVC.

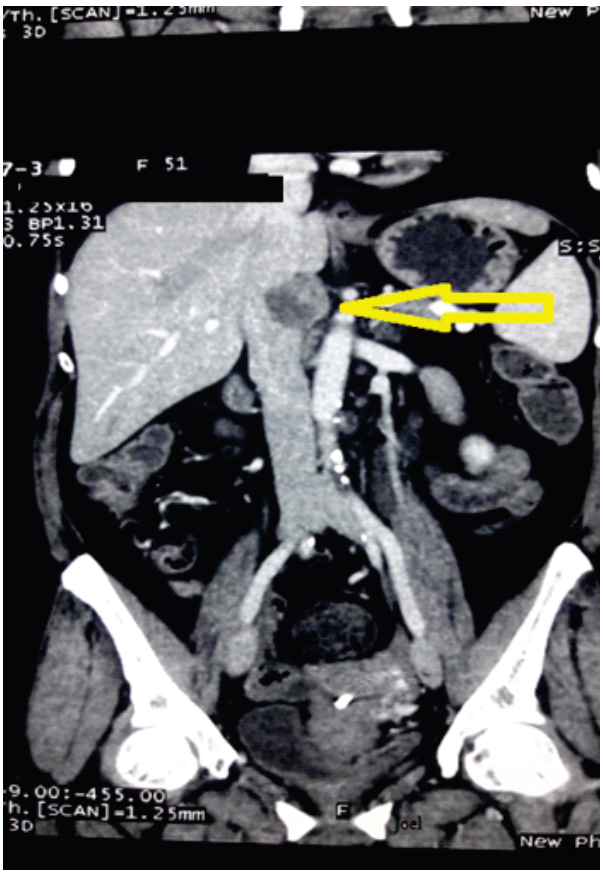


**Figure 1.** Leiomyosarcoma of the Inferior Vena Cava in the retro hepatic segment

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Received: 21-08-2023 Accepted: 01-11-2023
DOI: http://doi.org/10.4038/sljs.v41i03.9078

### Case 1

A 51-year-old female presented with nonspecific epigastric pain. An ultrasound scan of the abdomen revealed a hypo echoic, well defined, lobular mass postero lateral to the suprarenal IVC (Level II) (fig1 ). The lesion was invading the IVC. Further evaluation with contrast enhanced Computed Tomographic scan (CT scan) was done. The CT scan revealed a well-defined 3x3.5 cm mass arising in relation to the right adrenal gland. The mass was invading the postero- lateral surface of the IVC. There was no invasion into the adjacent structures.

A surgical exploration and excision of the lump was planned. During surgery the tumour was found to be in the suprarenal area posterior to the IVC, and was well defined. It was attached to the posterior wall of the IVC and was invading into it. Tumour was mobilized from the surrounding tissues. It was excised with the cuff of the IVC. The IVC was directly repaired with 4/0 polypropylene sutures.

Histology revealed a leiomyosarcoma (with malignant spindle cells, Mitotic count of 8 per10 High Power Field (HPF).The tumour was positive for smooth muscle actin (SMA) and desmin, indicating smooth muscle cell origin but it was negative for S 100 thus excluding a malignant nerve sheath tumour). The tumour was reaching the resection margin at one area. However a positron emission tomography (PET) CT scan done after 2 months did not show any residual tumour. After 308 days of follow up, the patient is well with no clinical and imaging evidence of recurrence.

### Case 2

A 52-year-old female presented with nonspecific abdominal pain. Imaging revealed a well-defined 6x4 cm dumb-bell shaped tumour invading the posterior surface of the infra renal IVC (level I). Patient underwent tumour resection with a cuff of the IVC. Histology revealed a leiomyosarcoma (tumour with atypical spindle cells with a mitotic activity of 3 to 4 per 10 HPF. The tumour was positive for SMA and desmin; it was negative for S 100). The patient was referred to the oncologist for further management. After 1230 days of follow up, the patient is well with no clinical and imaging evidence of recurrence.

**Case 3**

A 66-year-old female presented with nonspecific abdominal pain. Imaging revealed left sided retroperitoneal mass near the left kidney. A radical nephrectomy was performed. At surgery it was found that the tumour was arising from the left renal vein. Histology revealed a differentiated (pleomorphic) leiomyosarcoma extending into the renal vein. 3 months later the patient returned with nonspecific abdominal pain. A CT scan of the chest and abdomen was done. It revealed multiple metastases in the lungs and the liver. The patient was referred for palliative care.

**Summary of results**

In the above described cases, all were females (100%). The mean age at presentation was 56.3 years (51 - 66). All presented with nonspecific abdominal pain. The locations of the tumours were in the suprarenal IVC (n-1), infrarenal IVC (n-1) and the left renal vein (n-1). The mean tumour size at presentation was 4.75 cm (3.5 - 6.0). All patients underwent surgical resection in our series. Both IVC tumours were removed with a cuff of IVC. A direct repair of the IVC was done. One patient who had a primary leiomyosarcoma of the left renal vein (Case 3) underwent radical nephrectomy. At a mean follow up of 542.7 days (90 - 1230), one patient was diagnosed to have metastases in the lungs and liver two months after the surgery and died in 90 days. The other two patients are well with no clinical or imaging evidence of recurrence.

Discussion and conclusions

Leiomyosarcomas accounts for only 7% of all soft tissue sarcomas . Primary leiomyosarcoma of the IVC are rarer accounting for only 0.5% of all adult sarcomas . However, it is the commonest primary malignant tumour of the IVC .

It originates from the smooth muscle cells of the media of the vein . As it grows, it extends both into the vessel (intravascular part – i.e. tumour thrombus) and to the outside of the vein wall (extravascular). This extravascular part of the tumour could be misdiagnosed as tumours arising from the surrounding tissues e.g. Adrenal tumour. On microscopy the tumour consist of bundles of spindle cells. The mitotic activity differs depending on the tumour grade. On immuno-histochemical staining, the leiomyosarcoma is positive for desmin, vimentin and SMA .

Since the time it was first reported in 1871 , only about 400 cases are reported until now . It commonly affects females (female to male ratio of 4:1). It affects 50 to 60 year age group as the cases reported in this series .

Patients commonly present with nonspecific symptoms, while specific symptoms occur according to the location of the tumour and the completeness of the IVC occlusion i.e. Lower limb oedema, hepatic venous outflow obstruction (HVOO), etc. Due to the nonspecific nature of its presentation, the diagnosis is often missed or delayed .

Contrast enhanced CT scan of the abdomen is the imaging of choice. If supra hepatic extension (level III) is detected or suspected, a trans-esophageal echocardiography (TEE) should be performed to confirm the atrial extension, to detect the invasion into cardiac muscle and to plan the intervention. Intra operative TEE helps to monitor the tumour during surgery i.e. for migration, embolisation, etc.

For the purpose of description, the proximal extent of the leiomyosarcoma of the IVC is classified into 3 levels ( ). The level I extends from the beginning of the IVC (the confluence of the common iliac veins) to the lower border of the renal

**Table 01-** Summary of the cases

| Case | Age | Gender | Presentation | Vein | Imaging finding | Surgery | Histology | Follow up |
|---|---|---|---|---|---|---|---|---|
| 1 | 51 | F | Nonspecific abdominal pain | Supra-renal IVC | Hypo dense, well defined, 3.0 x 3.5cm, lobulated mass | Excision with IVC cuff, direct IVC repair | Leiomyosarcoma - Malignant spindle cells, mitotic count of 8 /10 HPF. Positive for SMA and desmin, negative for S 100 | Well at 1230 days |
| 2 | 52 | F | Nonspecific abdominal pain | Infra-renal IVC | 6.0 x4.0cm dumb-bell shaped tumour | Excision with IVC cuff, direct IVC repair | Leiomyosarcoma - A typical spindle cells, mitotic activity 3 to 4/10 HPF. Positive for SMA and desmin, negative for S 100 | Well at 308 days |
| 3 | 66 | F | Nonspecific abdominal pain | Left renal vein | | Radical nephrectomy | Differentiated (pleomorphic) leiomyosarcoma | Developed metastases in lungs and liver , died in 90 days |

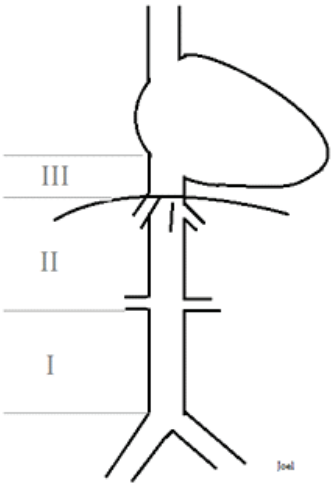


**Figure 2.** Level of the IVC involvement (according to Kieffer E, et.al.)

veins. The level II extends from the renal veins level to the lower border of the hepatic veins. The level III is above the level of the hepatic veins(1) .

Surgical excision is done with curative intent. Options depend on the extent of the tumour, the presence of IVC occlusion and the IVC wall infiltration by the tumour. For the level I tumours the proximal and distal control of the infrarenal IVC is adequate. During excision of the tumours in the suprarenal and retro hepatic area (level II), the main sources of bleeding are the renal veins and the hepatics veins. Therefore control of the infra renal IVC, renal veins, supra hepatic IVC and the hepatic hilar vessels are needed (to achieve total hepatic vascular exclusion). For the tumours extending above the hepatic venous level (Level III), sternotomy and cardiopulmonary bypass is needed.

For the intra luminal tumours which are not adherent to the wall of the IVC, the tumours can be excised with a cuff of the IVC. And for tumours which are attached to the wall of the IVC the tumour and the segment of the IVC need to be excised.

The options for IVC reconstruction include; direct repair, repair with a patch, excision of IVC and reconstruction with synthetic graft. Excision of the IVC and ligation of the ends is done if the IVC is already occluded with adequate collateral flow. However before the IVC is ligated, the distal stump pressure is measured and if the stump pressure is more than 30mmHg (1), reconstruction of the IVC is recommended to avoid venous hypertension i.e. In the lower limbs.

The consensus on adjuvant chemo and radiotherapy is lacking due to the rarity of the IVC leiomyosarcoma (8). Although in the past adjuvant chemotherapy and the radiotherapy was considered as ineffective (9), there are recent reports to suggest that there is a survival benefit with these modalities (10).

Overall the leiomyosarcoma of IVC is associated with poor prognosis with a 5 and 10 year survival of 31.4% and 7.4% (11).

Therefore leiomyosarcoma of the IVC and its tributaries should be considered as a differential diagnosis, especially if the tumour lies near the IVC or it extends into the IVC. Well planned surgical excision with vascular control provides the best chance of survival.

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