

Case Report

Leiomyosarcoma of Inferior Vena Cava involving the Liver- A rare case report.

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Abstract

Sarcomas are very rare tumours and 10-30% are located in retro peritoneum. Out of all the sarcoma cases only 1% cases are of Leiomyosarcoma. We reported a case of 60 year female came with chief complaints of pain abdomen and loss of weight. On palpation there was tenderness in the right hypochondrium and was evaluated. MRI abdomen showed a well-defined heterogenous mass of size 8.8 ×7.9×8.2cm noted in the suprarenal location which was compressing inferiorvenacava (IVC) and also showed displacement of IVC with focal intraluminal extension with possibility of Leiomyosarcoma of IVC or retro peritoneum or hepato renal region. Section from trucut biopsy of Liver showed loss of normal liver architecture by pleomorphic spindle tumour cells arranged in bundles. Cells showing marked nuclear pleomorphism with enlarged irregular nucleus with many mononucleate and multi nucleate gaint cells suggestive of malignant spindle cell neoplasm. Immuno Histochemistry (IHC) staining was showed diffuse positivity for smooth muscle Actin, Desmin. Treatment was initiated with adjuvant chemotherapy. Later IVC reconstruction along with resection of supra renal mass was done.

Keywords: Leiomyosarcoma of inferiorvenacava (IVC), Malignant spindle cell neoplasm of Liver.

Introduction

Leiomyosarcoma of the inferior vena cava (IVC) is a rare neoplastic condition, accounts for less than 1% of adult soft tissue sarcomas and it affects <1/100,000 of all adult malignancies.^{1,2,3} Patients manifest with extra-luminal or intra-luminal growth usually with the invasion of surrounding structures. It has poor outcome and rate of survival for five years is between 31 to 66.7% following complete

resection.^{3,4,5}

Leiomyosarcoma of the IVC generally presents as massive tumors during the time of diagnosis. There is Female predominance and commonly seen in subjects with around 50 years of age.⁶ The potential curative modality is by only surgery. Approach to managing these cases includes preventing the obstruction to major venous flow and recognizing the efficacious adjuvant therapeutic approach to decrease the recurrence. Although the problems arise because of anatomical features of this tumor which highlights vital facts for example vascular reconstruction and multi organ resection.

In our case report, patient has undergone resection of supra renal mass with IVC resection and reconstruction along with neoadjuvant chemotherapy.

Case Report

A 60 year female came with abdominal pain for 3 months and weight loss. There were no

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complaints of fever, vomiting, loose motions or jaundice. On palpation there was tenderness in the right hypochondrium. Necessary investigations were done. USG of abdomen showed a mass arising from right adrenal encasing IVC. Magnetic Resonance Imaging (MRI) of abdomen showed a well-defined heterogenous mass of size 8.8 ×7.9×8.2cm in right suprarenal region which is compressing and displacing Inferior vena cava (IVC) with focal intraluminal extension with possibility of Leiomyosarcoma of IVC or retro peritoneum or hepato renal region. Contrast Enhanced Computed Tomography (CECT) abdomen showed a mass involving IVC, right lobe of Liver, right kidney and a complex cyst in spleen. No distant metastatic lesions noted. Complete blood picture revealed anemia (Hb-9g/dl). 2D Echo was normal.

Trucut biopsy of Liver was performed and specimen evaluated for histopathological examination. On Gross examination received multiple linear soft tissue bits measuring 1×0.5 cm in total. Entire tissue was processed. On microscopic examination loss of normal liver architecture by pleomorphic spindle tumor cells arranged in bundles. Tumor cells exhibited marked pleomorphism of the nucleus with enlarged irregular nucleus with many mononucleate and multi nucleate giant cells suggestive of malignant spindle cell neoplasm. Immuno Histochemistry (IHC) staining showed diffuse positivity for smooth muscle Actin & Desmin.

Based on histopathological features along with imaging findings, final diagnosis of leiomyosarcoma involving Inferior venacava and liver showing malignant spindle cell neoplasm of Liver in trucut biopsy of liver. Treatment was initiated with neo adjuvant chemotherapy (Gemcitabine, Doxorubicin, Docetaxel) followed by resection of IVC and reconstruction with Dacron graft along with resection of supra renal mass was performed. Patient is on regular follow-up for chemotherapy and doing well.

Figure 1: H and E stained 100x Microphotograph showing pleomorphic spindle shaped cells.

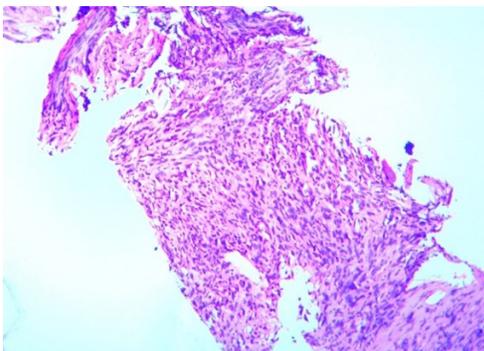
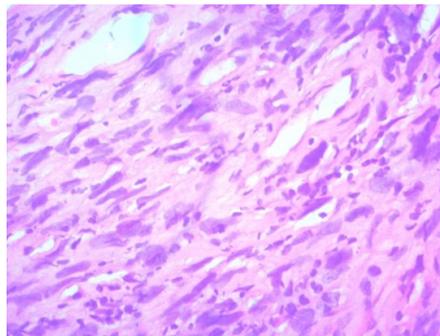


Figure 2: H and E stained 400x Microphotograph showing marked pleomorphism of nucleus with enlarged irregular nucleus.



Discussion

Leiomyosarcomas of inferior venacava are extremely infrequent malignancies and very less subjects of leiomyosarcoma of IVC are reported accounting for less than 400 cases in the literature studied till date.⁷

Leiomyosarcoma is a malignant spindle cell lesion, cell of origin being smooth muscle cell, it affects the skin, gastrointestinal tract, blood vessel and uterus.² Leiomyosarcomas arising from walls of blood vessels are vascular leiomyosarcoma accounting for only 1-2% and it affects the IVC rarely.

Leiomyosarcoma arising in veins especially the IVC occurs in tunica media and have variations in growth patterns that includes: extraluminal, intraluminal, or combined.⁷ According to surgical resection modality Leiomyosarcoma of IVC is generally classified in three segments on the basis of site of primary tumor. Segment I is infrarenal extension, segment II does not involve suprahepatic veins but involves the interrenal as well as suprarenal veins; segment III is inclusive of suprahepatic veins and can extend to cardiac region, which has the poorest prognosis.^{8,9}

In study done by Wachtel et al.¹⁰ found that overall survival in patients of leiomyosarcoma of IVC was 23 months at 1 year and 55% at five years. Disease free survival and overall survival achieved with surgical clearance of the tumor margins was being most significant. In a case series reported by Hines et al,¹¹ compared the five year (5 year) Overall survival rate among patients with positive and negative surgical margins were 0% and 68% respectively. And also in study by Hollenbeck et al¹² showed five year disease free survival in patients with positive and negative surgical margins were zero percent(0%) and thirty three percent(33%) respectively. Whereas in patients with extension of retroperitoneum, showed one year survival rate and one year primary patency was 75% and 90% respectively in patients those who had undergone

Inferior venacava reconstruction surgery in study by Ruiz et al.¹³ All these studies showed the importance of surgical clearance of tumor margins which was impacted on overall and disease free survival rate.

Frequently reported clinical symptom in the literature is abdominal pain which is nonspecific type and all other clinical symptoms such as edema of lower limbs, hepatomegaly, ascites, jaundice occurs secondary to the hepatic metastases.¹ In advanced stage of the Leiomyosarcoma of IVC, metastases occur in fewer than 50% of the patients, and frequently involves the liver, lung, lymph nodes, or bone. Usual Histopathological examination shows spindle cells arranged in fascicles having hyperchromatic nuclei. Immunohistochemical (IHC) stain helps in diagnosing the condition appropriately and utilizing imaging that helps in treatment.² Leiomyosarcoma of the IVC for the site from where it is arising needs assessment of the size, pattern of growth with the extent of narrowing of lumina of the IVC on cross sectional radiological images as in CT or MRI. On CT (radiology), Leiomyosarcoma of IVC appears as heterogeneously enhancing mass lesion having central necrosis in the retroperitoneum. MRI helps in evaluation of intraluminal tumors which might be hard to delineate on CT scan. Radiological evaluation also helps in deciding surgical and medical modality of treatment as tumor characterization, localization of tumour can be done along with evaluation of metastases.¹⁴

Treatment modality of Leiomyosarcoma IVC includes wide & complete surgical removal of the IVC, with adjuvant chemotherapy.² If metastatic disease is present surgical removal is not usually done. The management is done for prolonged survival, controlling the symptoms and inhibiting further progression of the disease. For subjects with unresectable tumours, main treatment modality is chemotherapy which includes gemcitabine, docetaxel two alternative drugs which can be utilized are ifosfamide and doxorubicin.

Conclusion

Leiomyosarcoma of Inferior Vena Cava (IVC) is a rare neoplasm, can present with intraluminal or extraluminal growth usually involving the adjacent structures such as liver. Histopathological examination combined with imaging findings aid in the diagnosis and the best management is IVC reconstruction with clear surgical margins.

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