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CASE REPORT

Primary Leiomyosarcoma of the Inferior Vena Cava: Rare Case Report

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ABSTRACT

Primary leiomyosarcoma is rare tumor approximately 1/100000 people. It is reported that only 2% of leiomyosarcoma is vascular in origin arising from inferior venacava (IVC). Considering its incidence, it is more common in females and middle segment of IVC is most commonly involved. We report here, case of 37-year-old female presented with right flank pain for one year then after radiological imaging neoplastic mass noted arising from infrahepatic and juxtarenal IVC and ultrasound guidance biopsy was taken that proved leiomyosarcoma of IVC. Surgery was performed with clear resection margins and regular follow-up was performed in our oncology OPD with the necessary imaging and laboratory work. No recurrence seen after 6 months of postoperative surgery, but we still need follow-up for any late recurrence.

INTRODUCTION

Primary leiomyosarcoma is extremely rare malignant smooth muscle tumor of inferior vena cava. By means of literature review, less than 400 cases are reported.[1] Leiomyosarcoma is the most common primary tumor of the inferior vena cava and the second most frequent retroperitoneal neoplasm in adults. [2, 3]First case was reported and originated from the wall of the inferior vena cava by Perl in 1871.[4]Symptoms and clinical history is often nonspecific and recommended treatment is surgical resection.

Radiological imaging has important role in preoperative staging and extent of involvement of IVC. CT scan is pivotal for diagnosing and follow-up of intravascular leiomyosarcoma of IVC. Most typical appearance is lobulated intravascular heterogeneous enhancing mass resulting vein dilatation with total or near total obstruction with area of hemorrhage or necrosis. MRI is also useful for localized small size tumors for surgical approach.

Surgical resection remains treatment of choice with reconstruction of IVC. IVC reconstruction usually considered in patients requiring radical resection.[5] long term anticoagulation may be required after surgery to prevent venous thrombosis with intravenous heparin followed by warfarin.[6]

CASE REPORT

A 37-year-old hypertensive female patient presented with emergency OPD at the Sindh Institute of Urology and Transplantation (SIUT) with complaints of right flank pain and swelling since one year. No significant history of taking amlodipine 5 mg Od for hypertension. Soft non-tender palpable mass was noticed at physical examination. All baseline investigations are normal on laboratory work.

Radiological findings revealed that initially, ultrasound was done with color doppler, which showed heterogeneous mass with internal vascularity and necrosis in the right hypoch-

ondric region, raising the possibility of neoplastic mass. Triphasic CT scan was performed to determine further characterization, morphology and origin of this mass.

Then tripashic CT scan was performed showing a heterogeneous enhancement of lobulated soft tissue mass resulting from intrahepatic portion of IVC (Figure 1). Inferiorly, it is exophytic and extends to the right lumbar region within the peritoneum (Figure 2). Poster laterally is inseparable from the right kidney and its proximal ureter. It is closely related to hepatic flexure laterally. Medially, it's inseparable from descending aorta. It measures about 14.5x10.2x9.5 cm in craniocaudal, anterio-posterior and transverse dimensions.

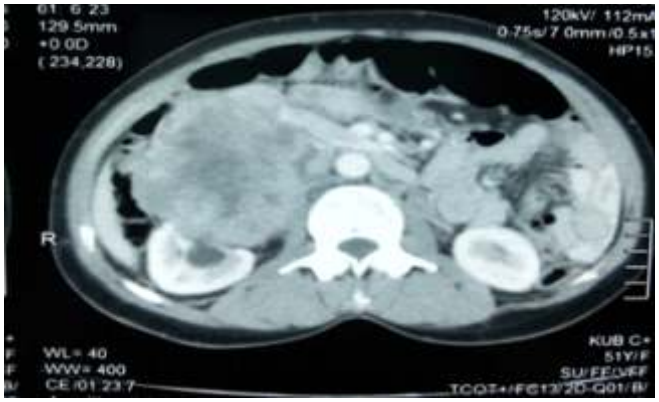

Figure 1: MDCT axial view; heterogeneously enhancing mass involving infrahepatic portion of IVC and inseparable from right k idney hilum.

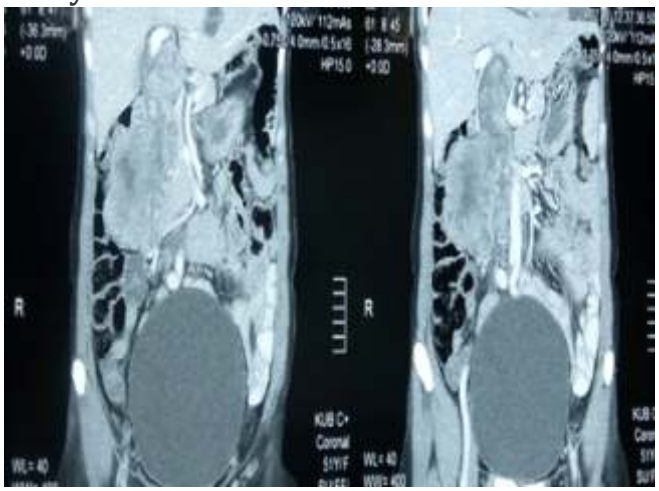

Figure 2: MDCT coronal views;Enhancing lobulated mass involving IVC extending from intrahepatic portion to infrahepatic portion with adjacent displacement of viscera and aorta

Findings were suggestive of IVC neoplastic mass possibly leiomyosarcoma. Using the above-mentioned findings, ultrasound guided mass biopsy was performed.

Histopathology findings revealed that sections examined from two cores showed hypercellular neoplastic lesion exhibiting interlacing fascicles of atypical spindle cells. These tumor cells showed marked nuclear atypica with moderate quantity eosinophilic cytoplasm and hyper chromatic cigar shaped; blunt ended vesicular nuclei with perinuclear vacuolization and inconspicuous nuclei (Figure 3). The tumor showed atypical mitosis foci with a mitotic count of 7/10 HPF. Areas of necrosis have also been appreciated. IHC markers were applied with appropriate positive controls and showed: Actin: Diffuse positive Caldesmon: Diffuse positive and Desmin: Diffuse positive (Figure 4) and CD117, CD34 and S-100: Negative (Figure 5). The morphological features and IHC profile confirmed leiomyosarcoma of IVC.

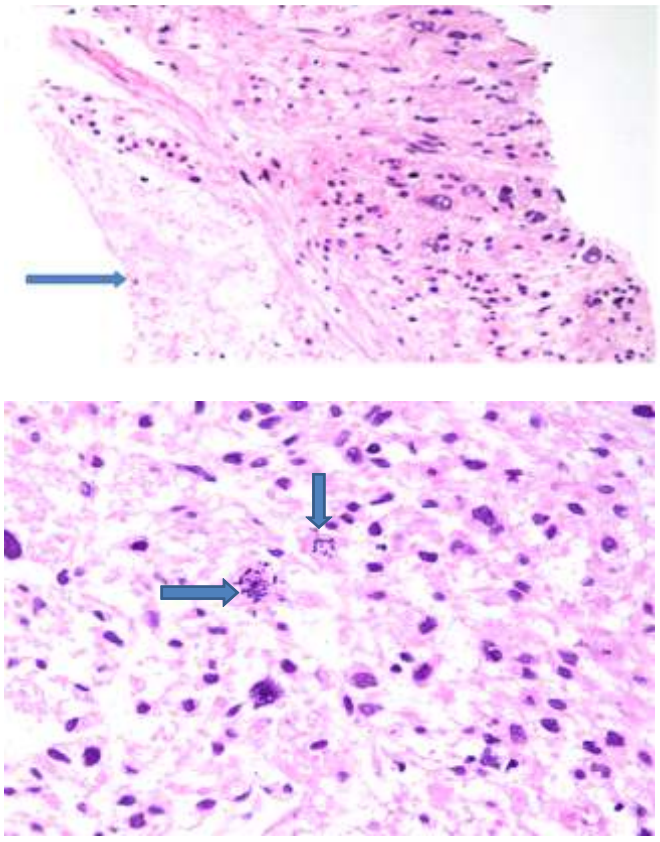

FIGURE 3: Medium and high power view pathology slides; Increased number of mitosis with nuclear atypica and eosinophilic cytoplasm and elongated hyper chromatic cigar shaped nuclei (arrows pointed).

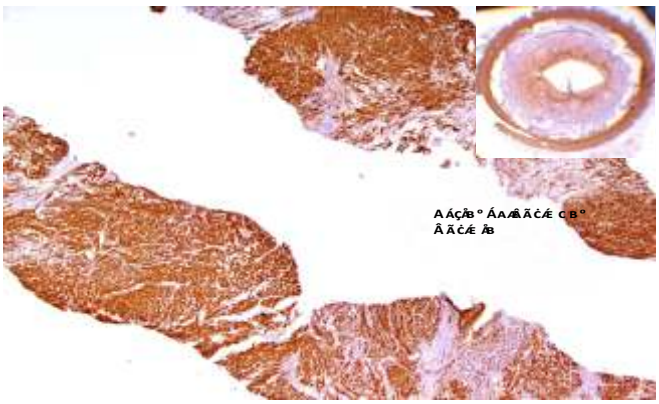

FIGURE 4; Diffuse positive desmin and caldesmon.

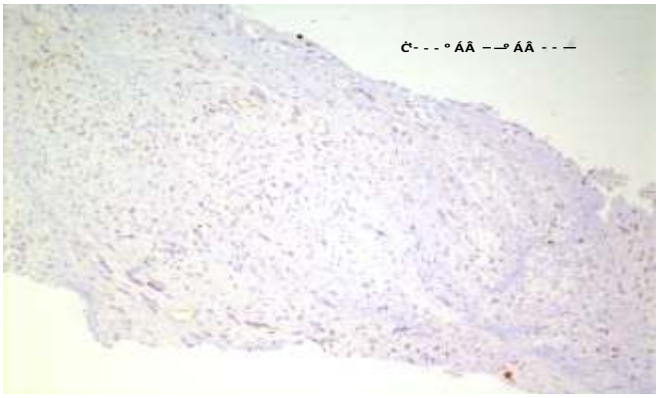

FIGURE 5: Negative CD 117,CD34 AND S-1OO.

DISCUSSION

Malignant sarcomas in retroperitoneum areunusual tumors, constituting 10% to 15% of all soft tissue sarcomaswith annual incidence of approximately 2.7 cases per million populations.[7] Primary leiomyosarcoma of IVC is categorized in three groups depending upon locations. Group one is also known as upper segment from hepatic vein to right atrium (24% cases). Group two includes middle segment from hepatic vein to renal veins (42%). Group three involves lower segment of infrarenal IVC (34%).[8]
It usually presented with nonspecific symptoms vague abdominal pain, nausea or vomiting in initial period and in late period with abdominal swelling, distention with weight loss as it commonly grows slowly and may extend and involving extra vascular viscera. Distant metastasis can be seen in liver, lung and bone but rarely.
Primary leiomyosarcoma of the IVC is rare tumors and for final diagnosis, radiological and histopathological findings must need, and as rare tumor and symptoms are vague, diagnosis is delayed and surgery would be difficult at advanced stage.[9]
As imaging has important role in identifying, diagnosing and imaging guided biopsy of these tumors for earlier management.
Histopathology is very helpful for final diagnosis and characterization of tumors. At macroscopic evaluation it usually appears as gray to white fleshy mass with whorled appearance having areas of hemorrhage or necrosis. At microscopic evaluation it appears as interlacing fascicles of atypical spindle cells and tumor cells showed marked nuclear atypical with moderate quantity eosinophilic cytoplasm with varying degrees of mitosis.[10]
Previously it was thought that it may sensitive to chemotherapy or radiotherapy and surgery alone treatment option. Preoperative chemotherapy (the combination application of doxorubicin and ifosfamide) and radiotherapy can be helpful for delbulking of tumor and ease removal of tumor.[11] Surgical resection is gold standard treatment option if tumor localized without metastasis. Prognosis is very poor due to late diagnosis with extra vascular infiltration and rarely metastasis.[12]

CONCLUSION

Primary leiomyosarcoma of IVC is rare soft tissue retroperitoneal tumor. Preoperative imaging findings and ultrasound-guided biopsy may be helpful for diagnosis, but diagnosis is based on histopathological findings. Surgery remained a major treatment option. But follow-up is necessary to rule out recurrence as the prognosis is poor.

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