Right Atrial Extension of a Giant Retroperitoneal Leiomyosarcoma

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Abstract
Leiomyosarcoma of vascular origin is rarely seen occurring in the Inferior Vena Cava. We report a rare case of a young male with giant retroperitoneal leiomyosarcoma which extended into the right atrium.

Introduction
Of the many abdominal tumors which invade inferior vena cava (IVC) and extend into right atrium, renal cell carcinoma is the most frequent.¹ Leiomyosarcoma (LMS) of the IVC and adrenal gland are among the subtypes of retroperitoneal sarcomas which have a tendency to invade IVC and extend into right atrium.² ³

Case Report
A 32 years male was admitted with history of upper abdominal discomfort and pain which had progressed over a period of 2 months. A large firm mass was palpated in the right hypochondrium and lumbar region. Triple phase computed tomography of the abdomen was performed which showed a large heterogeneous infiltrating mass in the right upper retroperitoneum. The mass measured 23 cm (cranio-caudal) x 17 cm (transverse) x 17.5 cm (antero-posterior) in dimensions. The lesion extended from the undersurface of right lobe of liver (which is infiltrated by the mass) to the level of right renal hilum. Antero-posteriorly the mass extended from the right paravertebral region till the anterior abdominal wall. The hepatic flexure and ascending colon were displaced anteriorly by the mass. The right adrenal gland was not visualized separately. The upper pole of right kidney was infiltrated by the mass with posterior displacement of hilar vessels. The mass showed areas of hemorrhage, necrosis and solid enhancing nodular tissue within it (Figure 1).

The tumor thrombus in IVC extended into the hepatic part of IVC and right atrium. The azygous system was dilated as a result of the IVC obstruction. Moderate ascites was also seen (Figure 1).

The possibility of renal tumor was excluded by the fact that the epicenter of the mass was outside the kidney with secondary involvement of the renal parenchyma. The imaging differentials were primary retroperitoneal tumor, adrenal tumor and tumor arising from the IVC.

An ultrasound guided biopsy was performed and histopathological sections showed a tumor composed of spindle to oval pleomorphic cells with eosinophilic cytoplasm, indistinct cell borders and oval to elongated moderately pleomorphic nuclei. Few bizarre cells and an occasional mitosis were also noted. On immunohistochemistry, the tumor cells were positive for smooth muscle actin (SMA), epithelial membrane antigen (EMA), focally positive for Desmin and negative for cytokeratin, S-100 protein, CD117 and CD10 (Figure 2).

The histopathological impression was high grade leiomyosarcoma. Clinico-radiological correlation led to the diagnosis of LMS arising from IVC, adrenal or primary retroperitoneum. Due to the poor general health status, the patient expired without any further
intervention.

**Discussion**

LMS is a malignant tumor of smooth muscle cells which originate most commonly in the uterus and retroperitoneum.\(^1\) In retroperitoneum, most common site of origin is IVC.\(^5\) LMS of vascular origin is rarely seen occurring most commonly in the IVC. Conversely, primary malignancies of the IVC are rare, with LMS representing the vast majority.\(^6\) LMS of IVC was first described by Dzsinich et al. in 1992.\(^7\) Contrary to our case, LMS of IVC is...
four times more common in women and is mostly diagnosed in the sixth decade of life.

LMSs of the IVC are categorized by the part of IVC which is involved by the tumor (Figure 3).

Level I (lower segment): represents the portion of IVC below the renal veins.

Level II (middle segment): represents the portion of IVC between renal veins and hepatic veins.

Level III (upper segment): represents the portion of IVC between hepatic veins and right atrium.

According to the international registry of IVC LMSs, lower (44.2%) and middle (50.8%) portions of IVC are most commonly affected by LMSs, whereas the upper third region is least commonly involved (4.2%).

Due to its slow rate of growth, IVC LMSs remain asymptomatic for long time. This leads to the delayed diagnosis with dismal prognosis. LMSs involving IVC is intra-luminal in only 5% cases. In rest 95%, extraluminal growth is seen which can be mistaken for masses of adjacent organs and so the differential diagnoses should include primary tumors of these organs. Pre-operative assessment for resectibility requires modern imaging techniques such as ultrasonography, echocardiography, computed tomography (CT), and magnetic resonance imaging (MRI).

Combination of modern vascular surgery with chemotherapy and/or radiotherapy is required for proper management. Aggressive surgical removal with negative margins is essential followed by venous reconstruction by prosthetic replacement of the IVC whenever considered necessary. The material of choice for prosthetic replacement is reinforced polytetrafluoroethylene (PTFE).

Kieffer E et al evaluated 22 cases of IVC LMSs and concluded that Creation of an arterio-venous fistula eliminates the need for long-term anticoagulation therapy and ensures patency.

Conclusion

In conclusion, our case reminds us of the possibility that retroperitoneal LMSs can invade IVC and right atrium as can other renal and hepatic malignancies. The IVC should be examined by CT or MRI, especially in a case of a right retroperitoneal tumor. Image guided biopsy provides definitive pre-operative diagnosis.

References


