

## Primary Angiosarcoma of Retroperitoneum: A Case Report

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### Abstract :

**Background :** Angiosarcomas represent only 2% of Soft Tissue Sarcomas. Angiosarcoma of the retroperitoneum are rare and very few cases of it have been reported in the literature. Commonest soft tissue sarcomas in the retroperitoneum are liposarcoma, leiomyosarcoma and malignant fibrous histiocytoma. Here we present one of the rarely occurring, retroperitoneal angiosarcoma. Our patient was a 30-years-old woman who complained of abdominal pain. USG guided true cut biopsy from the mass was performed. On microscopic examination, it was found to be a high grade angiosarcoma. Final confirmatory diagnosis was obtained on immunohistochemical(IHC) examination.

**Keywords :** Angiosarcoma, Soft tissue sarcomas, Retroperitoneum.

### Introduction :

Soft tissue sarcomas (STS) comprise 1-2% of all cancers and can appear in any type of connective tissue throughout the body<sup>(1)</sup>. Angiosarcomas are very rare and [encompasses smallest compartment] of STS. Angiosarcomas arise in endothelium of blood vessels and represent 2% of STS.<sup>(2)</sup> Reported risk factors for development of angiosarcoma include **prior exposure to ionizing radiation or toxic chemicals, longstanding lymphedema** (Stewart-Treves syndrome), and **venous stasis**. Many patients are diagnosed as angiosarcoma with no identifiable risk factors.<sup>(2, 3)</sup> Although angiosarcoma can appear at any age but most often it occurs in second to seventh decade of life.<sup>(3)</sup>

The clinical presentation of angiosarcoma is heterogenous and multidisciplinary treatment is often necessary.<sup>(2)</sup> Angiosarcomas are most commonly found in the scalp, face, neck, extremities, or breast.<sup>(3)</sup> But angiosarcoma in retroperitoneum is exceedingly rare. Very few cases of angiosarcoma of the retroperitoneum have been reported in the literature<sup>(4)</sup> Most of the retroperitoneal angiosarcomas present with abdominal mass or pain.<sup>(5,6)</sup> Biopsy is required for definitive diagnosis. The defining histologic features of

angiosarcoma are an abnormal appearance of the endothelial lining and cellular dedifferentiation.<sup>(7)</sup> In high grade angiosarcoma only histological features are not conclusive, it should be differentiated from sarcomatoid carcinoma, amelanotic melanoma and other high grade sarcoma.<sup>(4)</sup> Generally, positive immunohistochemical staining for the normal endothelial hallmarks, [ Factor VIII related antigen, vimentin, CD31, CD34, Ulex europaeus agglutinin 1 and Vascular Endothelial Growth Factor (VEGF) ] confirm the diagnosis.<sup>(7)</sup>

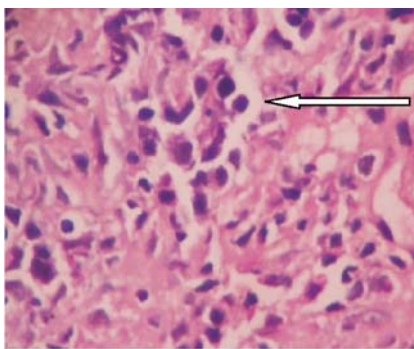
### Case report :

A 30 yrs old female presented with complain of abdominal pain. Her USG abdomen suggested malignant pelvic mass or abscess. Her CT scan showed primary malignant mass in left iliac region adhered to local soft tissue and mild left pleural effusion. MRI of both hip & sacro iliac joints with screening of lumbar spine showed a huge lobulated mass measuring 17.8x11.3x11.7cm in size, in left iliac wing, adhered to surrounding soft tissues (gluteus minimus, pyriform & part of iliac muscle with displacement of adjacent psoas muscle) and also enlargement of multiple common iliac, external iliac and internal iliac lymphnodes were seen. MRI suggested possibility of neoplastic lesion with multiple enlarged lymphnodes. USG guided biopsy from pelvic mass was carried out. On histology, multiple freely anastomosing small thin walled vascular channels, lined by atypical endothelial cells, showing hyperchromasia and mitosis were seen. It invaded

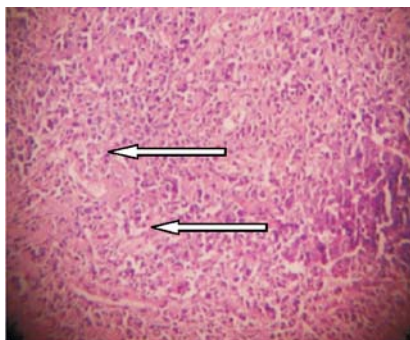
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striated muscles. These findings were consistent with histological picture of angiosarcoma. This was confirmed by IHC study which showed tumour cells' positivity for vimentin, factor VIII and CD34. On conclusion of angiosarcoma patient was advised for radiotherapy and treatment started. After two months of therapy the local advantages developed but patient developed electrolytes disturbances resulting in cardiac arrest followed by patient's death.

**Figure1: 40x view H& E stain, showing atypical endothelial cells lining vascular space.**



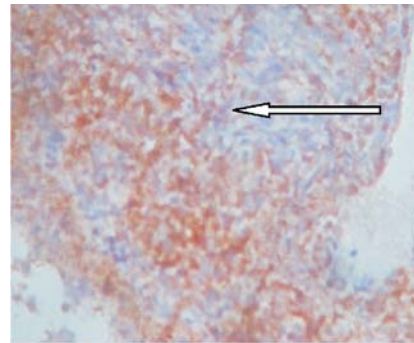
**Figure 2: 10x view H & E stain, showing multiple freely anastomosing vascular channels, lined by atypical endothelial cells.**



**Figure 3 : 10x view showing IHC- strong positive, vimentin (brown coloured staining shows positivity)**



**Figure 4: 40x view showing IHC- positive, factor VIII**



#### Discussion :

Angiosarcomas are less common malignant neoplasms and mostly found in the scalp, face, neck, extremities or breast. <sup>(3)</sup> Occurrence in the retroperitoneum is extremely rare. <sup>(4)</sup> Retroperitoneal angiosarcoma usually presents as an asymptomatic mass and grows to large sizes because a large volume of space is available so physical examination often is less remarkable. Neurological symptoms from compression of lumbar and pelvic nerves are frequently observed. Although definitive radiological findings for the diagnosis of angiosarcomas are not available but computed tomography (CT) and magnetic resonance (MR) imaging play an important role in characterization (size) and the assessment of the extent of the disease and involvement of adjacent and distant structures. <sup>(8)</sup> Differential diagnosis on retroperitoneal location includes liposarcoma, leiomyosarcoma and other high grade sarcomas. <sup>(9)</sup> So early detection by means of biopsy is very essential.

Diagnosis is based on the microscopic features and immunohistochemistry. Angiosarcoma is usually characterized by rapidly proliferating, extensively infiltrating anaplastic cells derived from blood vessels as [irregular lining of the blood filled spaces]. <sup>(9)</sup> In case of deep soft tissue angiosarcomas, have poorly differentiated areas without regular vascular channels and tumour cells may be closely packed, round, ovoid or spindle cells that resemble undifferentiated sarcoma so very difficult to diagnose on only histological features. Immunohistochemistry is often required to diagnose such cases of angiosarcoma and helps in differentiating from sarcomatoid carcinoma, amelanotic melanoma

and other high grade sarcoma. <sup>(4)</sup> In our case histological features were consist with the diagnosis of angiosarcoma and confirmed by IHC study which showed tumour cells' strong positive for vimentin, factor VIII and CD34, suggesting endothelial origin of tumour, negative for cytokeratin, epithelial membrane antigen (EMA) – markers for epithelial origin of tumour and negative for actin and desmin- markers for smooth muscle and skeletal muscle cell tumour, desmin and EMA are also, markers for malignant fibrous histiocytoma so ruling out its possibility. <sup>(4)</sup>

Angiosarcomas tend to recur locally, spread widely and high rate of lymph node metastasis compared to other soft tissue sarcomas and systemic metastasis is seen. <sup>(9)</sup> The median survival time of patients with advanced angiosarcoma is about 7-8 months. Larger tumour size (>5cm), retroperitoneal location and advanced disease, are associated with poor prognosis. <sup>(4)</sup> Our case of angiosarcoma had primary retroperitoneal location, larger size and already advanced (infiltrating surrounding soft tissue) when diagnosed, feautres favouring its poor prognosis.

### Conclusion :

Retroperitoneal angiosarcomas have asymptomatic presentation with little clinical manifestation. Moreover radiological reports that present the imagining features of this tumour are rare. Early detection by means of

biopsy is required. Histological features are not always conclusive; they should be differentiated from other high grade tumours by immunohistochemistry. This leads to late diagnosis and surgical unresectability of angiosarcoma. So only radiotherapy and chemotherapy are indicated and associated with poor prognosis.

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