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Case Report

Angiosarcoma of Deep Soft Tissue Presented as Mass Lower Leg

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ABSTRACT

We present a case of 70 year old male patient having three months history of rapidly increasing swelling in right lower leg clinically suspected of ? soft tissue tumor, ? myositis. On radiological examination large hypoechoic mass? malignancy. On histopathology diagnosed as malignant high grade soft tissue tumor - ? Malignant vascular tumor,? liposarcoma, ? Rhabdomyosarcoma. On Immunohistochemistry confirmed as angiosarcoma. Deep soft tissue angiosarcoma is extremely rare. Herewith presenting such case with clinical, radiological, histopathological and Immunohistochemistry findings. Clinically this case showed aggressive behaviour with high histological grade.

Key words: Angiosarcoma, lower extremity, soft tissue sarcomas.

INTRODUCTION

Angiosarcoma is a malignant neoplasm of endothelial origin. It is a extremely rare tumor comprise less than 1% of all sarcomas. [1] It can occur at any site, but has a strong predilection for the skin and subcutis. Its occurrence in extremities and deep soft tissue is rare. [2] Our case present with rapidly increasing deep soft tissue swelling (in calf muscle) with pain. As these tumors are aggressive and have high rate of recurrence, proper mode of diagnosis and treatment is essential.

CASE REPORT

A 70 year male patient came with history of tenderness and swelling in right lower extremity of three months duration which was rapidly increasing in size. Local examination showed firm to hard mass in calf muscle, clinically suspected of ? soft tissue tumor,? myositis, ? haematoma. There was no oedema. Patient was hypertensive and on regular treatment. There was no contributory family history. No history of trauma, radiation, infection, etc. No abnormality detected on systemic examination. Radiological examination showed large hypoechoic lesion involving medial aspect of gastrocnemius muscle with irregular margins, suspected of ? malignant soft tissue tumor. There was no evidence of regional or distant metastasis. Surgical resection of mass was done. Intraoperative

finding show firm to hard mass arising from medial aspect of gastrocnemius muscle, beneath the soleus and tendo archilis. Surrounding tissue shows fibrosis and congestion. Mass was excised and sent for histopathological study.

Gross Examination: We received a large, irregular, firm, grey white mass measuring 18x9x4.5cm with a tendon. External surface was irregular, nodular with adherent blood clot. On cut section showed grey brown tumor with variegated, fleshy appearance and areas of necrosis and haemorrhage (Figure:1).

Figure: 1- Gross photograph showing lage , grey brown soft tissue mass

Light Microscopy: Multiple section from mass showed highly vascular pleomorphic and spindle neoplastic cells arranged in sheets, clusters, epitheloid and diffuse pattern (Figure:2). The cells were highly pleomorphic having hyperchromatic or vesicular nuclei with prominent nucleoli (Figure:3). Many areas, showed atypical vascular spaces lined by endothelial cells with atypia and were filled with red blood cells (Figure:4). Areas of haemorrhage and necrosis were noted. Mitotic activity was increased. Many scattered multinucleate giant cells were seen. Epitheloid cell form with intracytoplasmic vacuoles were noted. Tumor was invading muscle tissue.

Histopathological diagnosis was given as - ? High grade angiosarcoma, ? High grade sarcoma, ? Rhabdomyosarcoma

Immunohistochemical study was done which showed positivity for Factor VIII related antigen, CD31, CD34. While it was immuno negative for pancytokeratin, myogenin, SMA, Caldesmon, Desmin, S-100 protein, HMB-45.

Final diagnosis – Angiosarcoma high grade –deep soft tissue leg.

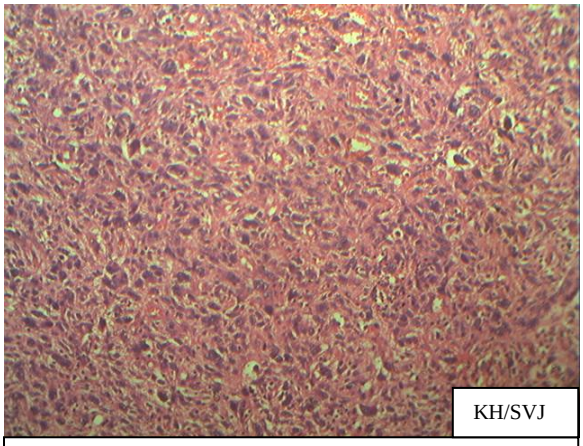

Figure 2: Photomicrograph showing neoplastic cells arranged in various pattern (Haematoxilin and Eosin, x100)

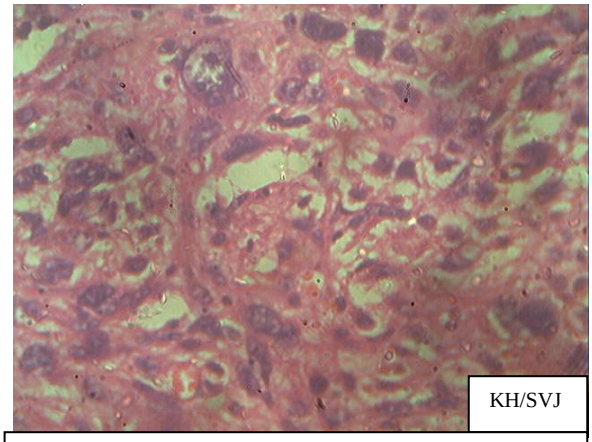

Figure 3: Photomicrograph showing highly anaplastic cells with hyperchromatic or vesicular nuclei with prominent nucleoli. (Haematoxilin and Eosin, x400)

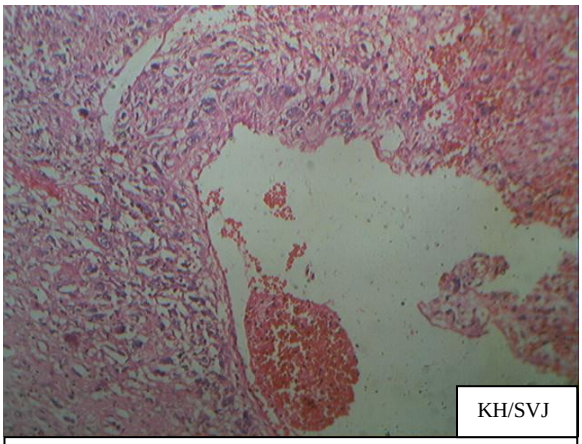

Figure 4: Photomicrograph showing atypical vascular spaces lined by endothelial cells with atypia and were filled with red blood cells. (Haematoxilin and Eosin, x100)

DISCUSSION

Angiosarcomas are one of the rarest form of soft tissue neoplasm. They show variable form from well differentiated tumors that resembles haemangiomas to those having very high grade anaplasia, making them difficult to differentiate from high grade sarcomas, carcinomas or melanomas. In contrast to deep located most soft tissue sarcomas, the angiosarcomas have predilection for skin and superficial soft tissue mostly (approximately 60%) [3] and very rarely in deep soft tissue. Angiosarcomas are more commonly seen in males, M:F ratio 3:1 and an average age of 75 years. [4]

In our case it was deeply located soft tissue mass with diffuse infiltration into surrounding muscle tissue was noted. Usually angiosarcomas are divided into cutaneous angiosarcoma(usual type), cutaneous angiosarcoma with lymphedema (lymphangiosarcoma), angiosarcoma of breast, radiation induced angiosarcoma, angiosarcoma of deep soft tissue and rare type. [5]

Variant of etiological factors like chronic lymphedema, radiation, other neoplasms, genetic or familial history, infection at site, trauma, etc. are given. However, in our case there was no such history. Also there was no oedema on leg.

Soft tissue angiosarcomas have aggressive clinical behavior. [6] Local recurrence and distant metastasis most often to the lung followed by lymph node, bone, etc. are commonly observed. The data shows angiosarcoma of soft tissue having large size, retroperitoneal location, older age, severe anaplasia are associated with poor outcome.

On the basis of histopathological morphology high grade lesions are difficult to differentiate from other, the differential diagnosis frequently included are undifferentiated carcinomas, malignant melanomas, histiocytic sarcomas and other high grade sarcomas [7] with epitheloid features. Immunohistochemistry stain such as CD31, factor VIII related antigen are useful endothelial markers to confirm angiosarcoma. [2] Our patient received treatment of surgical resection with wide surgical margin and radiotherapy. Patient was advised regular follow up.

CONCLUSION

Angiosarcoma of deep soft tissue is a rare histopathologic variant of angiosarcoma. Proper clinical, radiological, histomorphological features with immunohistochemical analysis is required to avoid misdiagnosis of these rare neoplasms.

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