High Grade Soft Tissue Tumor – Myxofibrosarcoma Presented As a Recurrent Forearm Swelling

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Abstract

Myxofibrosarcoma is one of the most common sarcomas in the extremities of elderly people and is characterized by a high frequency of local recurrence. We report a rare case of high-grade myxofibrosarcoma involving the forearm and presenting as discrete mass lesion and diffuse infiltrative nature precluded early recognition of tumor. To our knowledge, such a presentation with an extreme infiltrative growth pattern in the forearm has not been reported before.

Key Words: Myxofibrosarcoma, extremities, sarcoma.

INTRODUCTION

Myxofibrosarcoma is variant of the group of malignant fibrous histiocytomas. It is one of the most aggressive types of soft tissue neoplasms. The clinical presentation is not pathognomonic and the histological aspects are highly heterogenous, frequently delaying the diagnosis or leading to misdiagnosis. Complementary histochemical and immunohistochemical staining are mandatory to achive the diagnosis of myxofibrosarcoma. [1]

CASE REPORT:

In this study, we present a rare case with a history of a chronic hematoma after the trauma. In this case a 60 year old man came to the casualty after the trauma with the development of a small mass in the right forarm for which incision and drainage was done and the excised mass was sent for histopathological examination. The mass was Measuring 2.5x2x2 cm. It was sticky and gelatinous in consistency. The results lead us towards the possible diagnosis of spindle cell myxomatous neoplasm – intermediate to high grade (FNCLCC), for which few differentials came in front : 1) High grade myxofibrosarcoma 2)Malignant peripheral nerve sheath tumor, , 3) Malignant solitary fibrous tumor. For further diagnosis IHC was done which showed positive result for MUC-4, CD- 34 and SMA . The patient came back with the recurrence of the mass at the same site at the cytology sections of our hospital for which the material was aspirated and examined which showed presence of spindle cells in the studied smear.Wide local excision was done. The mass was gelatinous and sticky and sent for histopathological examination which measured 4.8x4.5x2 cm the HPE resulted as myxofibrosarcoma (Fedration Nationale des centers de LutteContre Le Cancer) – GRADE- III) with Immunohistochimestry confirmation: 1) strong cytoplasmic positivity for MUG -4, 2) Intra cytoplasmic positivity for SMA 3) membranous positivity for CD34 4) Negative result for S100.

DISCUSSION:

Myxofibrosarcoma represent a highly heterogeneous group of soft tissue neoplasm with a very aggressive loco- regional behaviour. Tumor growth may occur over several months, sometimes reaching 5 to 10

cminsize at the time of diagnosis. Myxofibrosarcoma is considered the most common malignant mesenchymal neoplasm in elderly patients, with slight male predominance. It usually presents as painless, slow growing, skin colored or erythematous nodules or tumors. Most lesions are located in the lower limbs and rarely on the trunk, head and neck[2]. About two-thirds of the cases develop in the dermal and subcutaneous tissues and one third in the fascia and skeletal muscleon [3]. On histopathological examination the differntial diagnosis can be : Nodular fasciitis, myxoma, neurofibroma, etc Bening conditions; other malignant tumor like myxoid liposarcoma, can also be an important differential diagnosis

CONCLUSION:

In conclusion, myxofibrosarcoma is a great clinical mimicker and may present histological nightmare. A large series of histochemical and immunohistochemical staining is recommended. Extensive surgical excision with adjuvant radiotherapy presents the optimal therapeutic option. [5]

REFERENCE:

[1] Article on high recurrence rate of myxofibrosarcoma: the effect of radiotherapy is not clear.
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HISTOPATHOLOGY IMAGES:

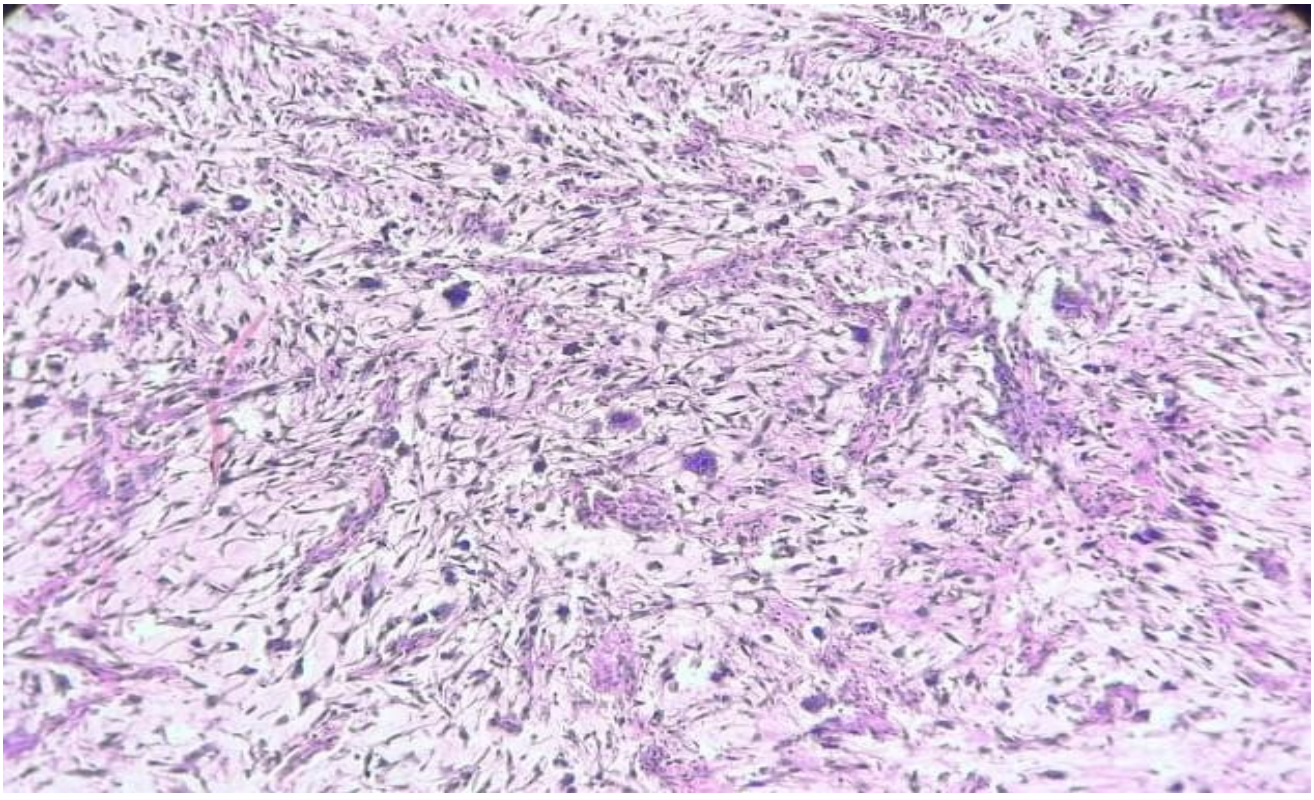

Figure 1 Spindle to stellate cells in a myexoid background.

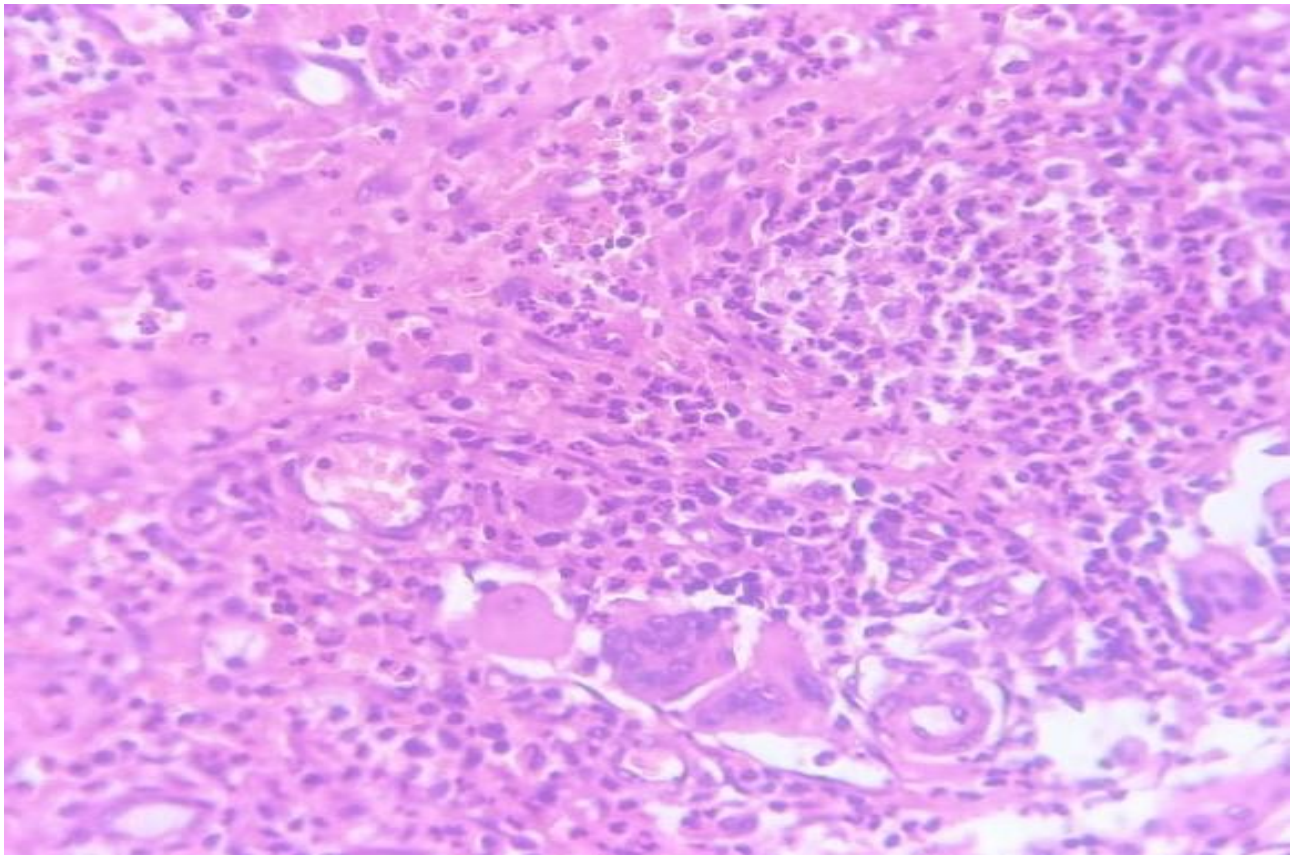

Figure 2 On 40X magnification section shows the cells are spindle to mild epitheloid in shape with hyper chromasia floated in myxoid stroma and associated with curvilinear elongated mild thick walled non hyalinised blood vessels with few bizarre and pleomorphic giant cells

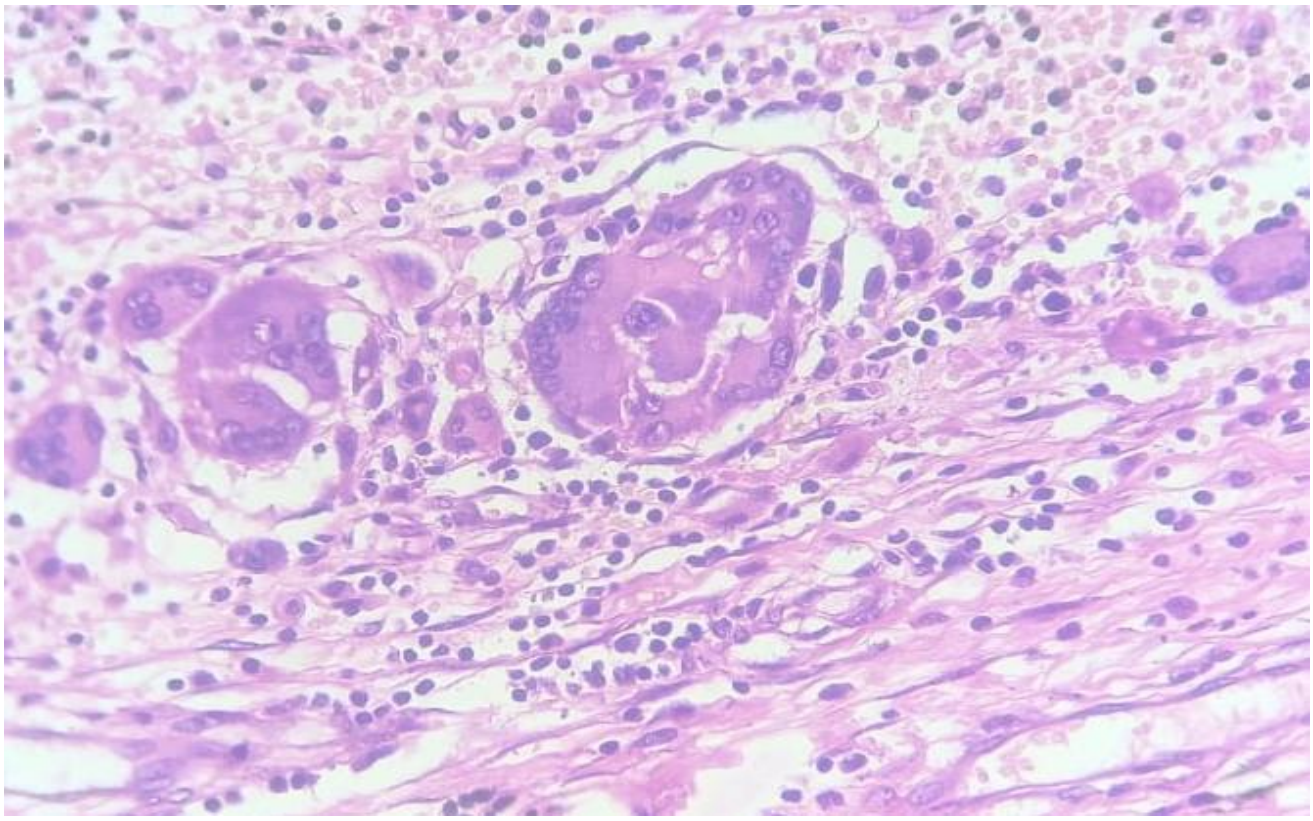

Figure 3 Bizarre cells in a myexoid inflammatory background on 40X

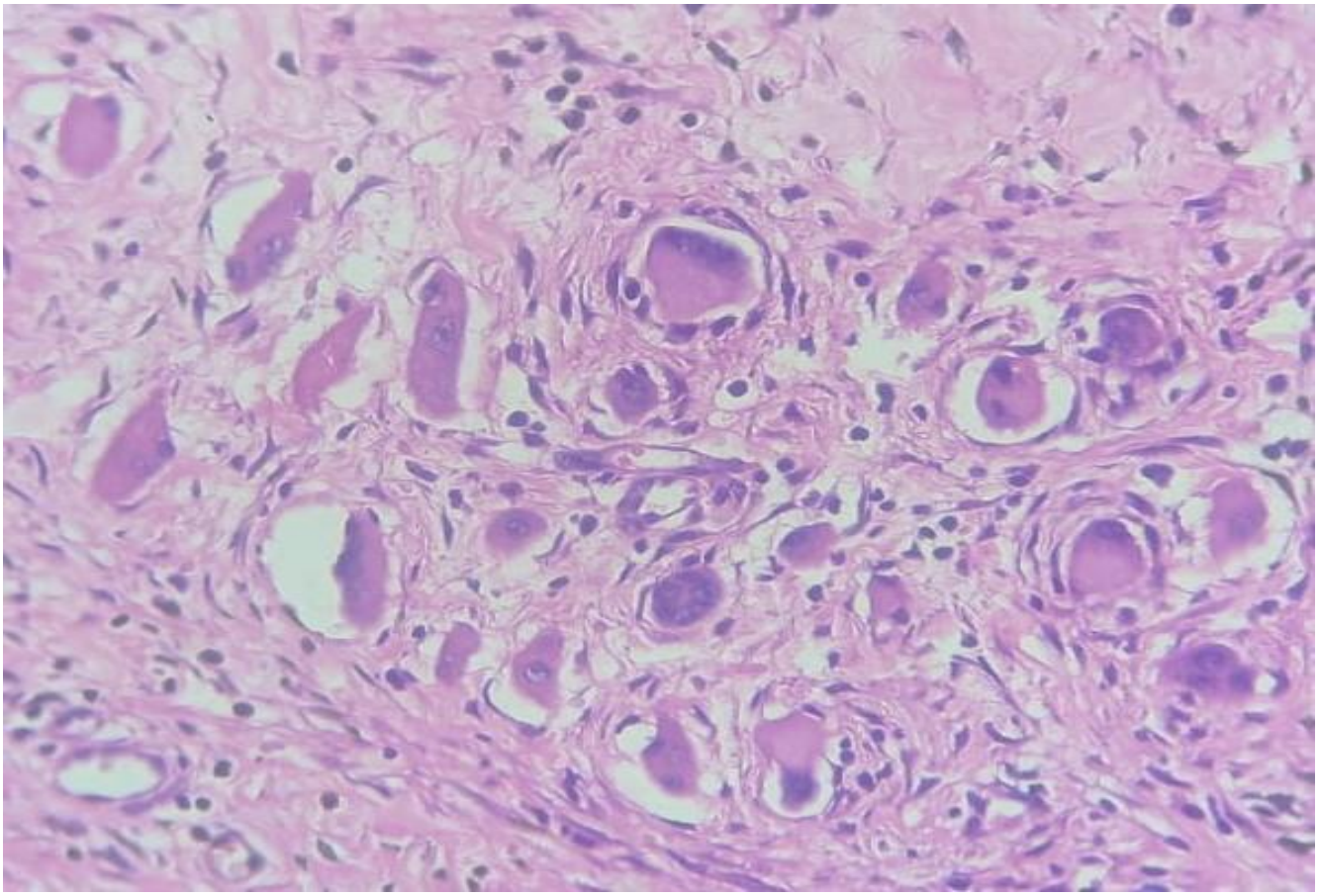

Figure 4 Giant cell in a myxoid background on 40X

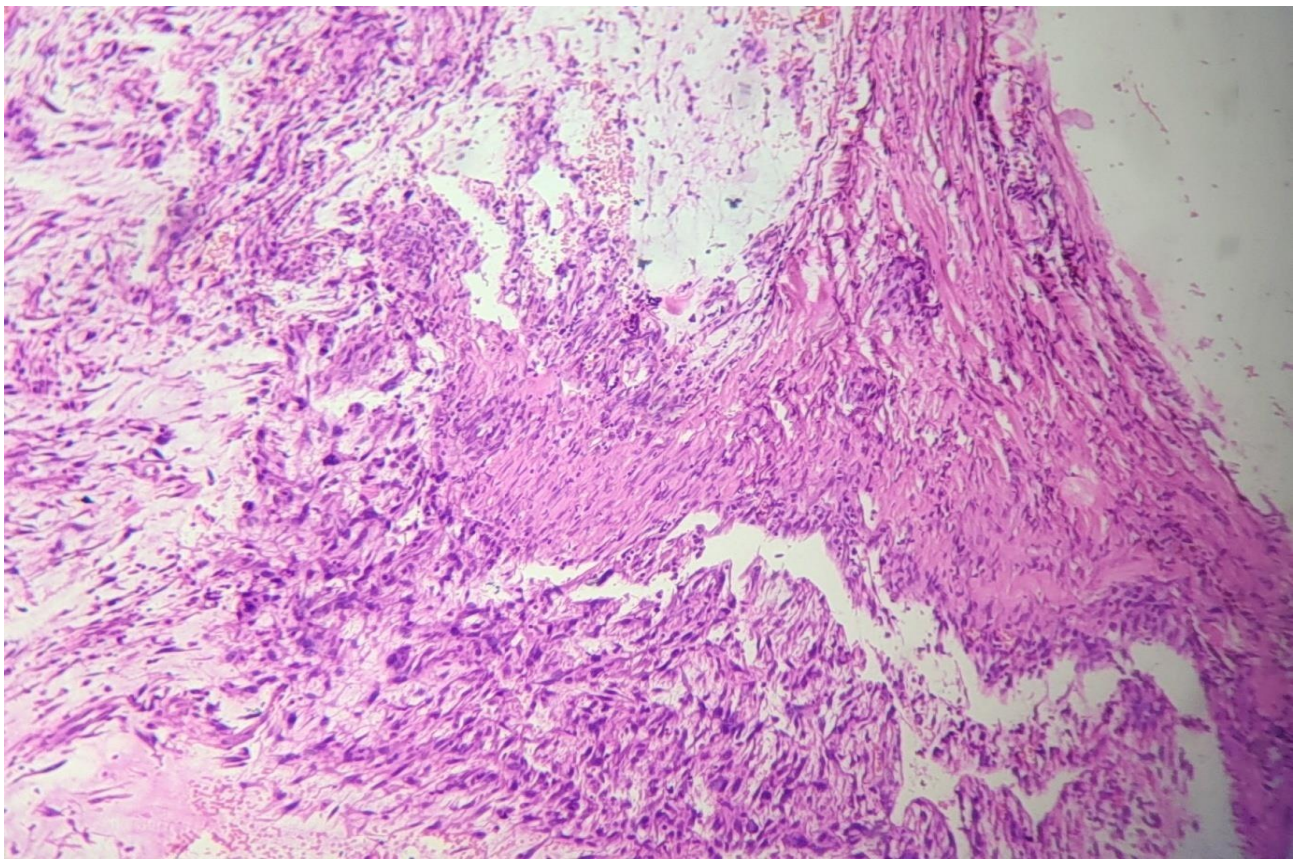

Figure 5 on 40X magnification collagen rosette is seen with steallet nuclei

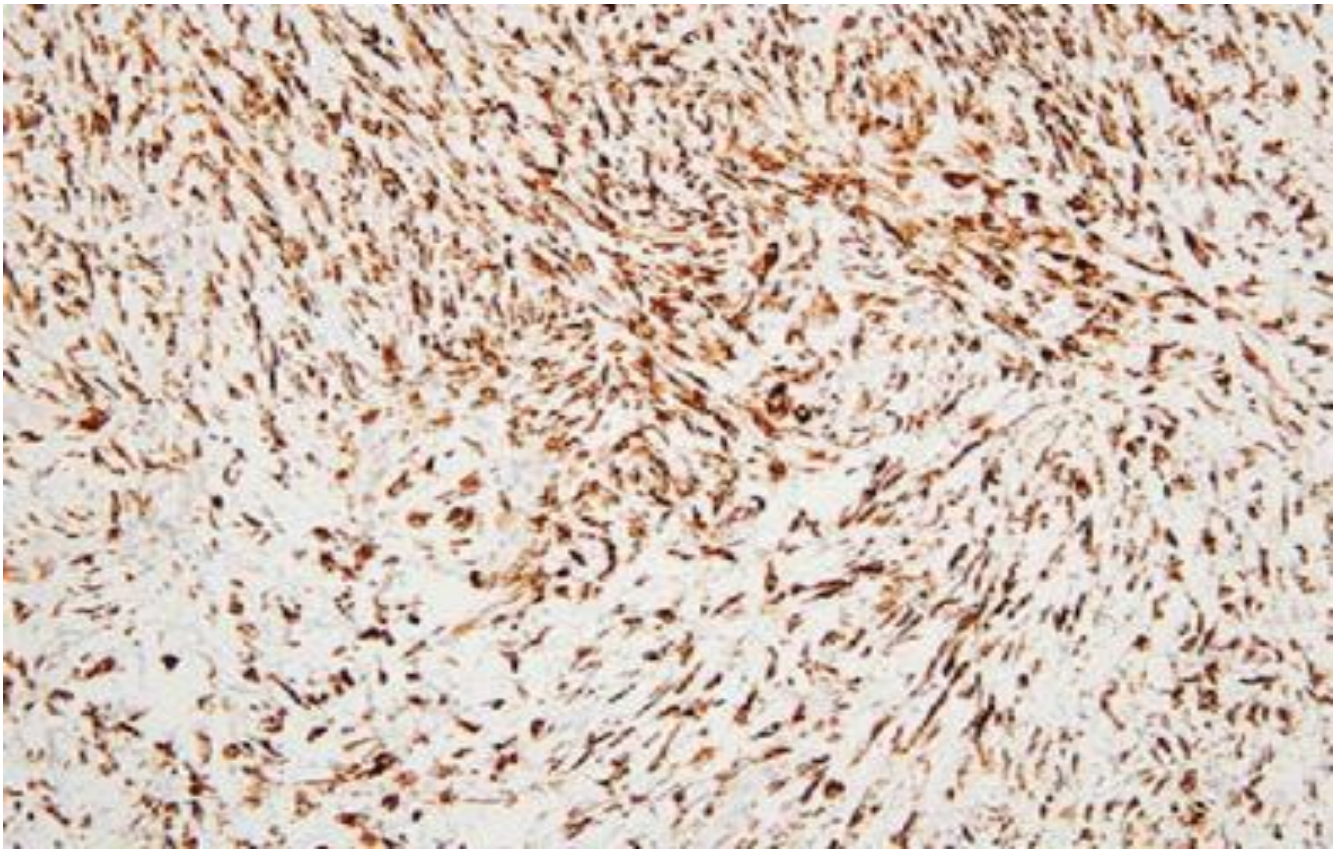

Figure 6 Strong cytoplasmic staining for MUC 4 40X

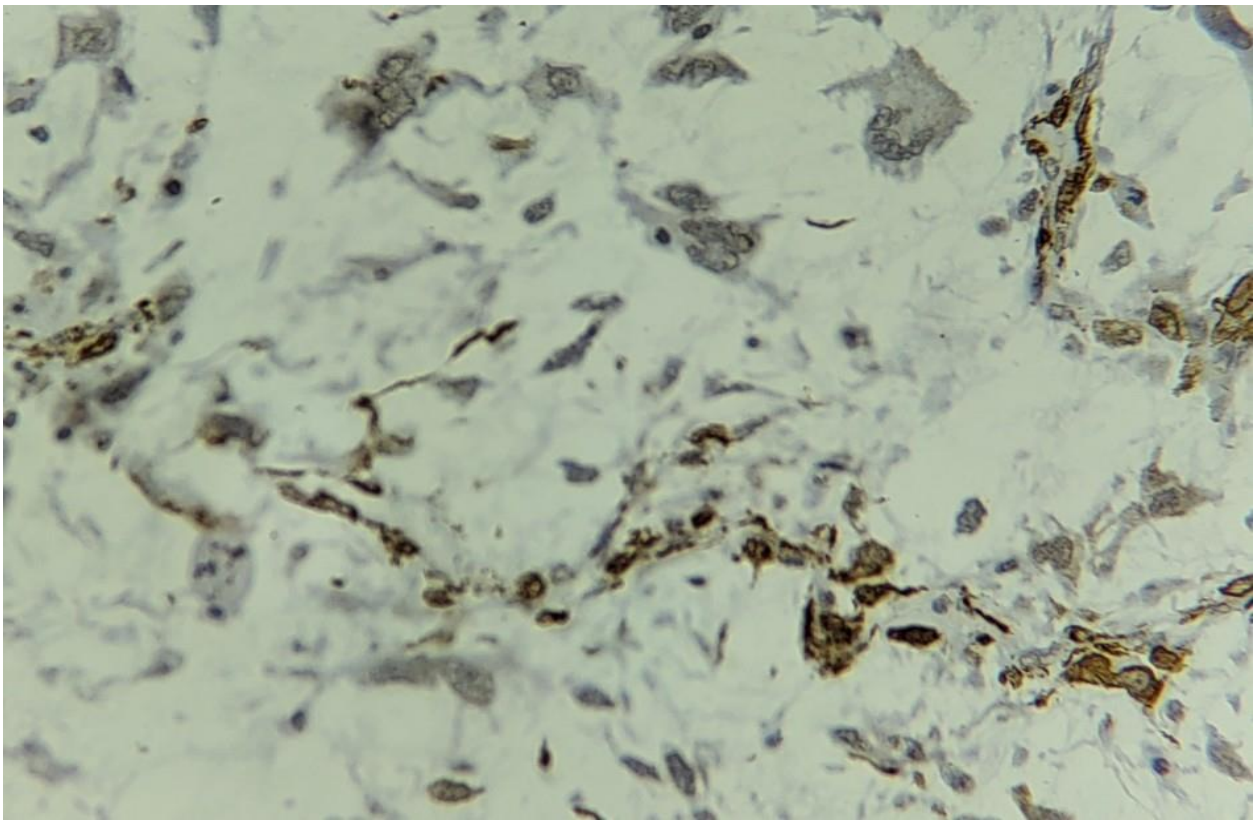

Figure 7 INTR ACYTOPLASMIC POSITIVITY FOR SMA ON IMMMUNOHISTOCHEMISTRY ON 40X

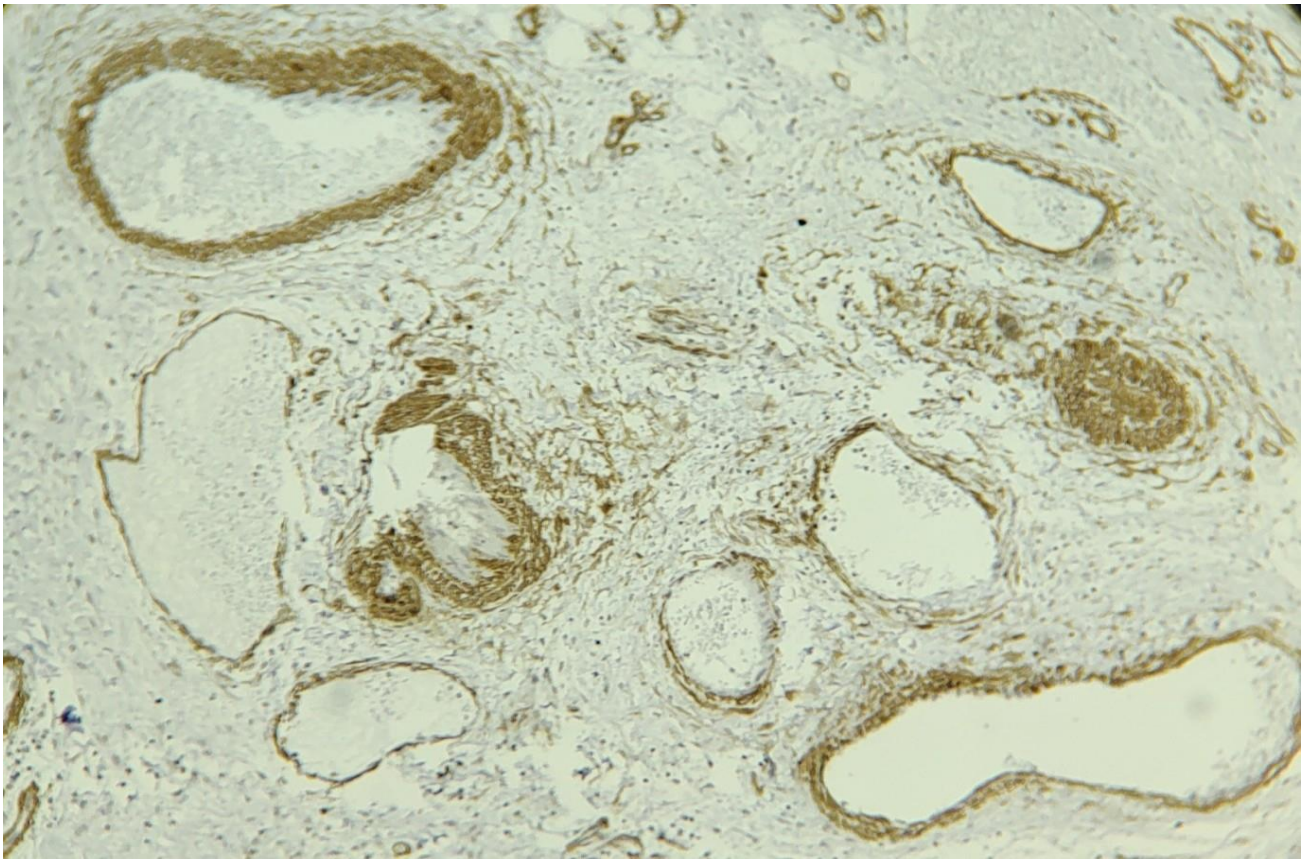

Figure 8 CD-34 positivity around blood vessels on 40X

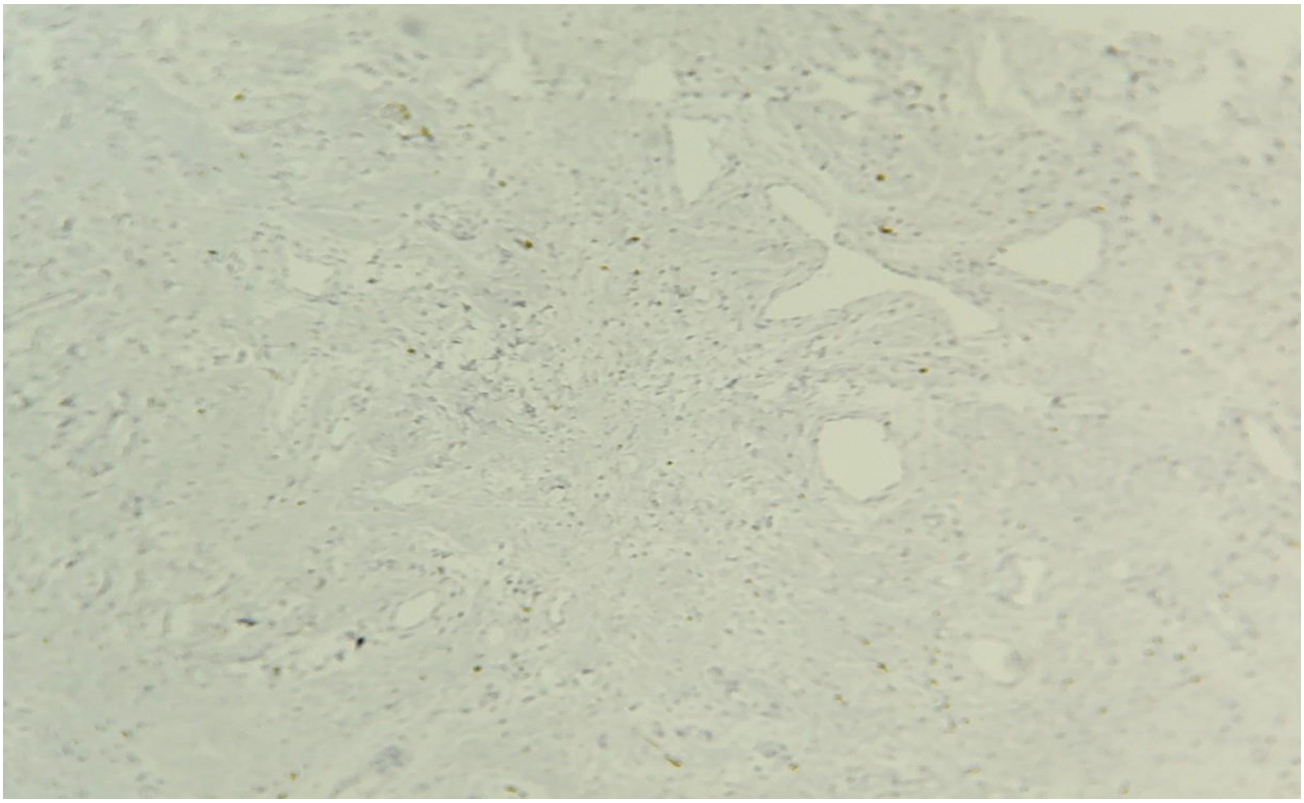

Figure 9 On 40Ximmunohisto chemistry for S-100 shows negative results

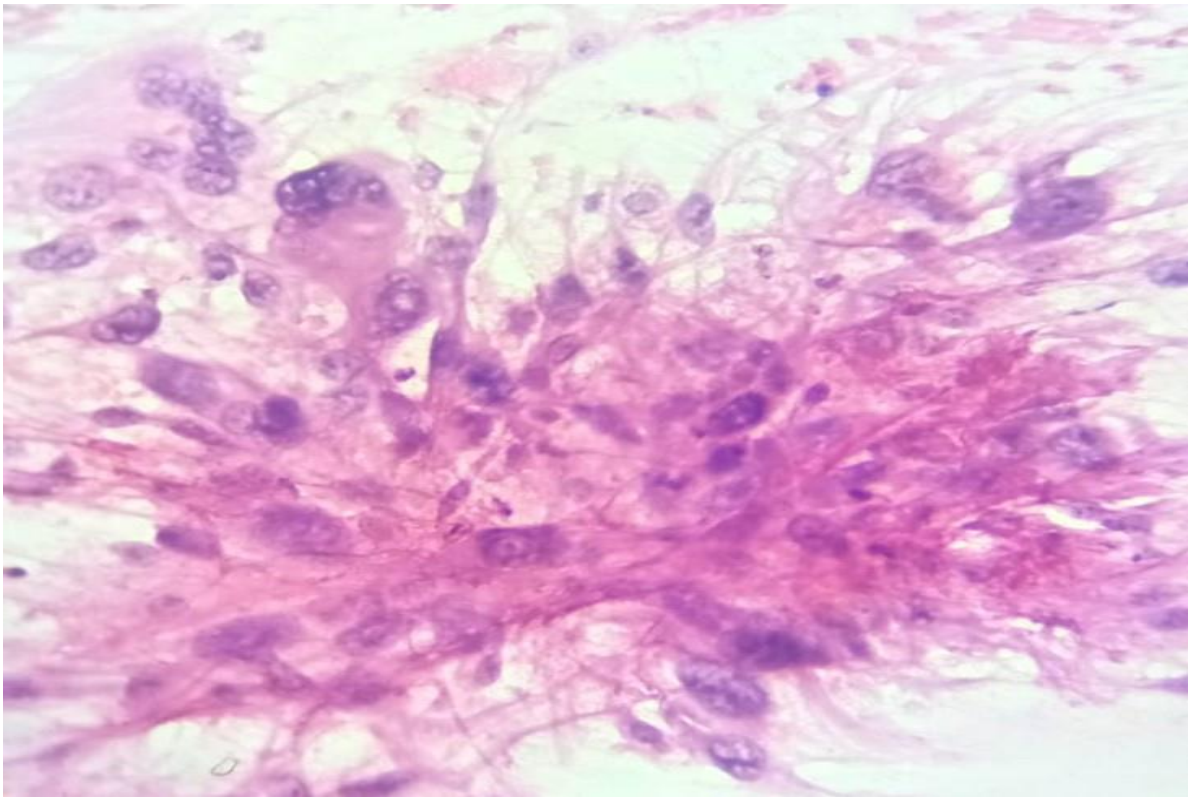

Figure 10 smear shows malignant spindle cell in discohesive cluster showing marked degree of pleomorphism, irregular neuclear margin, and noticable neucleoli and having variable amount of cytoplasm 40 X magnification

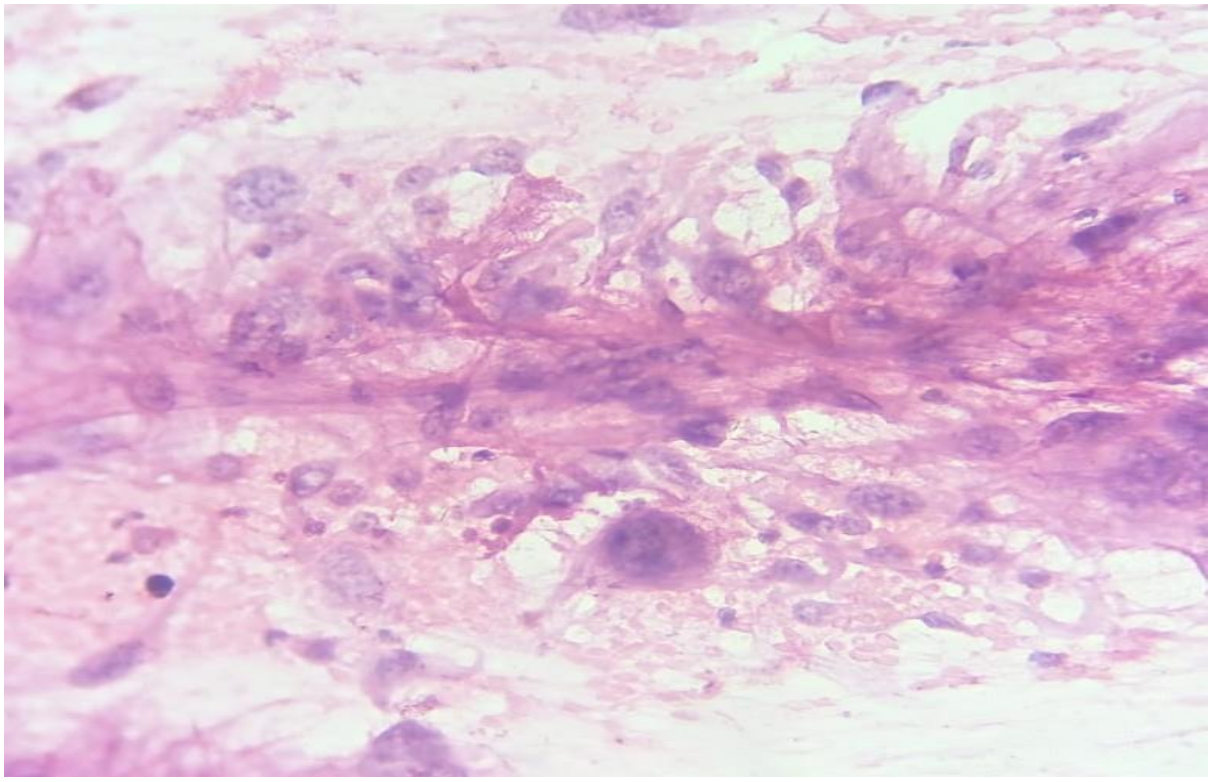

Figure 11 Smear shows spindle cell with bizarre nuclei and nuclei with multi nucleation 40X magnification