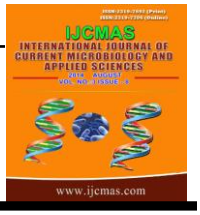
International Journal of Current Microbiology and Applied Sciences
ISSN: 2319-7706 **Volume 3 Number 8 (2014) pp. 510-517**
http://www.ijcmas.com



Original Research Article

Pleomorphic sarcoma in 60 years old male - A case report

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A B S T R A C T

Sarcomas are malignant tumors that arise from mesenchymal tissue at any of the body sites. Malignant fibrous histiocytoma (MFH) is the most common soft-tissue sarcoma of adults. It was first recognized as a distinct clinic-pathologic entity in the early 1960s as a pleomorphic sarcoma. The most common distribution site of it is extremities. In our case, patient was having pleomorphic sarcoma distal to right elbow joint which is quite rare. Here we are presenting a case where we are able to find and document the typical features of pleomorphic sarcoma (MFH). An ultrasonic examination combined with fine needle aspiration cytology should be helpful for the initial differential diagnosis of malignant fibrous histiocytoma. However, the final confirmation relies on histopathological examination.

Keywords

Soft tissue sarcoma, Pleomorphic sarcoma, Cytology, Histopathology

Introduction

Sarcomas are malignant tumors that arise from mesenchymal tissue at any of the body sites. [1] Malignant fibrous histiocytoma (MFH), the most common soft-tissue sarcoma of adults, was first described by O'Brien and Stout in the 1960s. [2] The predisposing factors are genetics, exposure to radiation or chemotherapy, chemical carcinogens, chronic irritation and lymph edema. In nearly all instances, sarcomas are thought to arise de novo and not from a preexisting benign lesion. [3, 4] It was first recognized as a distinct clinic-pathologic entity in the early 1960s as a pleomorphic sarcoma that contains both fibroblastic and histiocytic cells in varying proportions, arranged in a storiform pattern. In the 1970s, for the first time, Feldman and Norman described primary malignant tumor of bone that satisfied the histopathologic criteria of MFH. [5] The most common distribution site of MFH is extremities. [6] In Our case, patient was having soft tissue mass distal to right elbow joint which is quite rare.

Materials and Methods

60 years old male was presented to surgical OPD with rapidly growing, painless swelling, over flexor aspect of right elbow

joint for the past one month. The surgeons decided to send the patient for the FNAC. In cytology clinic we had examined that there was a swelling distal to the right elbow joint on flexor aspect measuring 8 X 7.2 X 6 cm. (Photo – 1) On palpation, it was soft to firm and not fixed to the underlying structures. Veins over the swelling were prominent and temperature was raised locally. The area to be aspirated was cleaned with spirit and a 23 gauge needle with syringe and trocar was inserted at convenient angles to the lesions and multiple hits were made within the lesion, with sufficient negative pressure, the needle was removed and pressure was applied to the area of aspiration to avoid bleeding or hematoma formation. [7] The aspirated material was placed on the glass slides. The slides were both air dried and wet fixed in alcohol. [8] Smears were stained with Hematoxylin and Eosin (H & E) stain and microscopy was performed. FNAC smears showed highly cellular smear with atypical spindle shaped cells and histiocytes with foamy cytoplasm. Spindle cells showed marked pleomorphism. Multinucleated tumour giant cells with bizarre nuclei and mitotic figures were seen in necrotic background. (Photo - 2, Photo – 3)

Overall cytological features suggested of malignant fibrous histiocytoma [undifferentiated high-grade pleomorphic sarcoma (UHPS)]. Patient underwent wide excision of tumour mass and mass sent for histopathological examination. On gross examination, there was a large globular soft tissue mass with attached elliptical skin flap which was measuring 8.4 X 7.5 X 6.4 cm with regular external surface. Cut surface showed multi-lobular grey white areas with absence of hemorrhage and necrosis. (Photo - 4, Photo – 5) Representative tissue bits were taken and examined microscopically. On microscopy, tumour consisted of spindle shaped fibroblast arranged in short fascicles and storiform pattern with plump to round shaped histiocytes showing marked pleomorphism were arranged haphazardly. There were numerous mitotic figures and many multinucleated giant cells with bizarre nuclei and areas of tumor necrosis were seen. (Photo - 6, Photo – 7)

After ruling out all possible differential diagnosis final impression was given as undifferentiated high-grade pleomorphic sarcoma (UHPS) [malignant fibrous histiocytoma]. The post operative period was uneventful, and the patient was discharged one week after the operation. He came for follow up after 15 days and started adjuvant chemotherapy.

Results and Discussion

Malignant fibrous histiocytoma is a soft tissue tumor of mesenchymal origin. It is the most common soft tissue sarcoma with the peak incidence in the 5^{th} and 6^{th} decade with male preponderance. [9] The most common anatomic sites are thigh, pelvic girdle, leg, and less commonly knee and upper arm. [10] Lower extremities are more frequently affected than upper extremities. MFH rarely arises distally to knee or elbow. In our case, the patient presented with soft tissue mass distal to the elbow joint which is rather rare.

They can be classified into superficial and profound type according to their site. The superficial form is very rare and is confined to the skin and to the subcutaneous tissue; it

may be adhered to the fascia. The profound form extends from the skin along the fascia until the muscle, or it can be located completely within the muscle. [11]

In recent years, the clinic-pathologic findings of this tumor were well defined and settled. These tumors clinically appear usually as a single, often large mass, often with areas of necrosis or hemorrhage. In our case, there was absence of hemorrhage and necrosis.

It has wide-ranging histology and morphology with total five histological variants of MFH are introduced: storiformplemorphic, myxoid, giant cell, inflammatory and angiomatoid. The storiform pleomorphic subtype of MFH is the most common subtype and most frequently seen as a deep-seated tumor of extremities in middle-aged or elderly patients. [12] It is composed of spindle-shaped and round histiocytes arranged in storiform pattern. There is high mitotic activity mimicking melanoma along with areas of haemorrhage or necrosis can occur. [13].

The diagnosis of MFH depends on precise differential diagnosis from other sarcomas. Differential diagnosis of MFH includes Pleomorphic Rhabdomyosarcoma, Fibrosarcoma, Pleomorphic Leiomyo-sarcoma. On microscopic examination, Pleomorphic Rhabdo-myosarcoma shows rhabdomyoblasts, cross striations, spindle to polygonal cells with deep eosinophilic cytoplasm and on Immunohistochemistry (IHC) it shows positive Myoglobin, MyoD1, Myosin, Myogenin. Fibrosarcoma composed of fibro blast with herringbone architecture and short fascicles, variable mitotic activity and no giant cells and IHC shows positivity for Vimentin. In Pleomorphic Leiomyosarcoma, there will be intersecting fascicles and bundles of smooth muscle cells having pale eosinophilic fibrillar cytoplasm, osteoclastic type of multinucleated giant cells and IHC shows positive Desmin, h-caldesmon and SMA. [14]

Surgical resection is the most potentially effective therapy for soft tissue sarcomas regardless of their site of origin. Ample excision is recommended because approximately 44% of the patients were presented with local recurrence and 42% with metastasis in the lungs. [15, 16, 17, 18] Presence of metastasis is usually associated with a poor prognosis. Adjuvant radiation therapy is suggested for patients with high grade sarcomas beneficial for local control. Nowadays, the role of adjuvant chemotherapy for sarcomas remains uncertain. [4]

In conclusion, this work demonstrates a case of localized MFH in the right upper extremity distal to the elbow joint which was clinically diagnosed as soft tissue sarcoma. However, anatomico-pathological and immunehistochemical studies showed that it was a malignant, poorly differentiated neoplasm based on histiocytes and fibroblasts (Malignant Fibrous Histiocytoma). The fact is that it is extremely difficult to diagnose this tumor before microscopic examination so that histopathology is the gold standard for the final diagnosis.

Photo .1 Large swelling distal to the right elbow joint on flexor aspect.

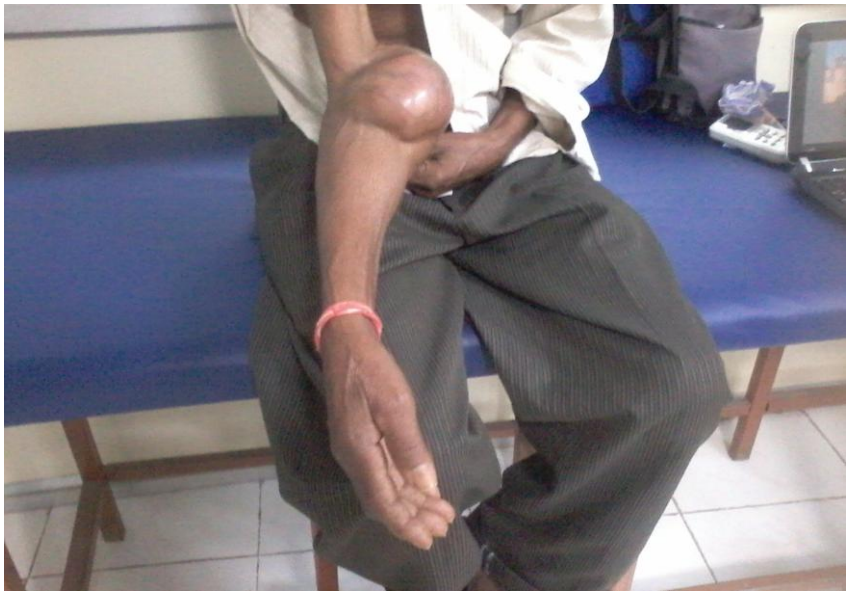


Photo.2 FNAC showed highly cellular smear with clusters of malignant tumor cells. [H & E, 4 X]

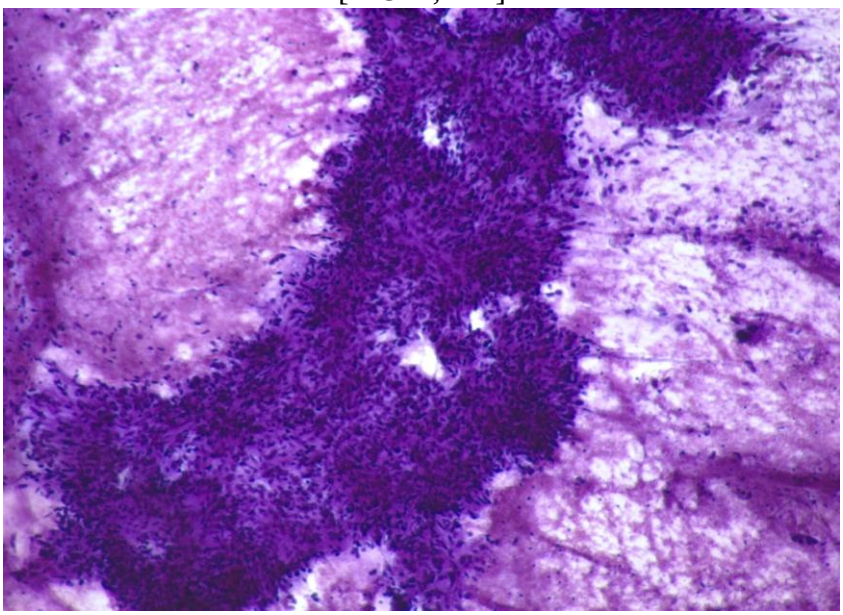


Photo.3 Smear showed histiocytes with foamy cytoplasm and atypical spindle cells with plump hyper chromatic nuclei, anisocytosis and anisonucleosis. [H & E, 4 X]

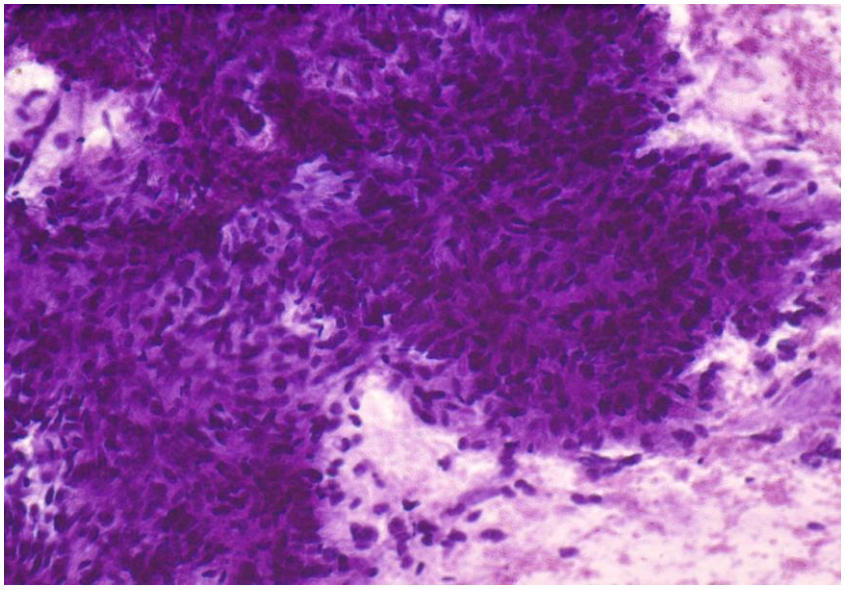


Photo.4 Gross examination showed globular skin covered soft tissue mass with regular external surface.

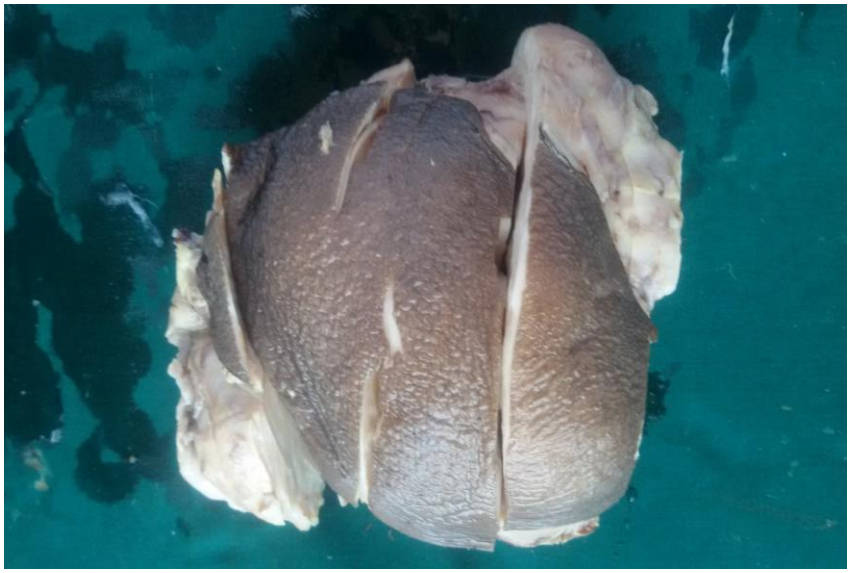


Photo.5 Whitish homogeneous fleshy cut surface of tumor.

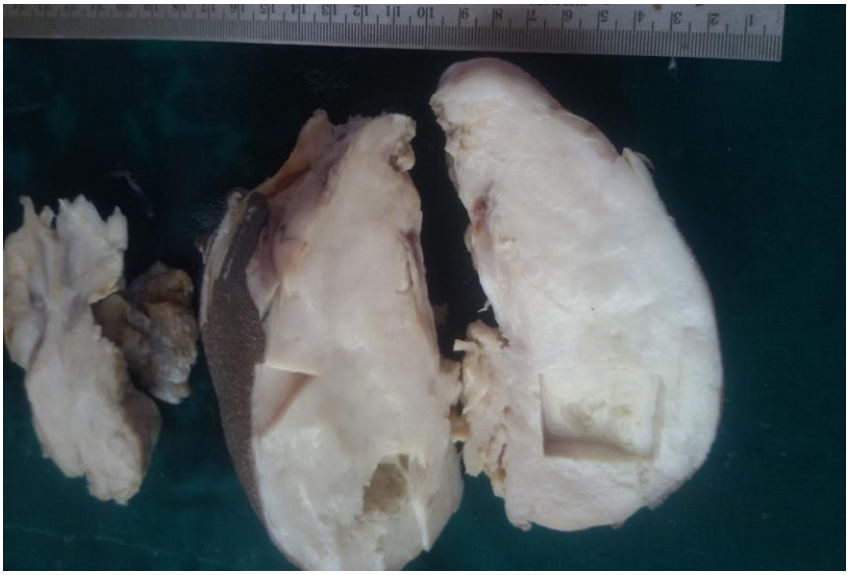


Photo.6 Tumor consists of spindle shaped fibroblast arranged in short fascicles and storiform pattern with plump to round shaped histiocytes. [H & E, 4 X]

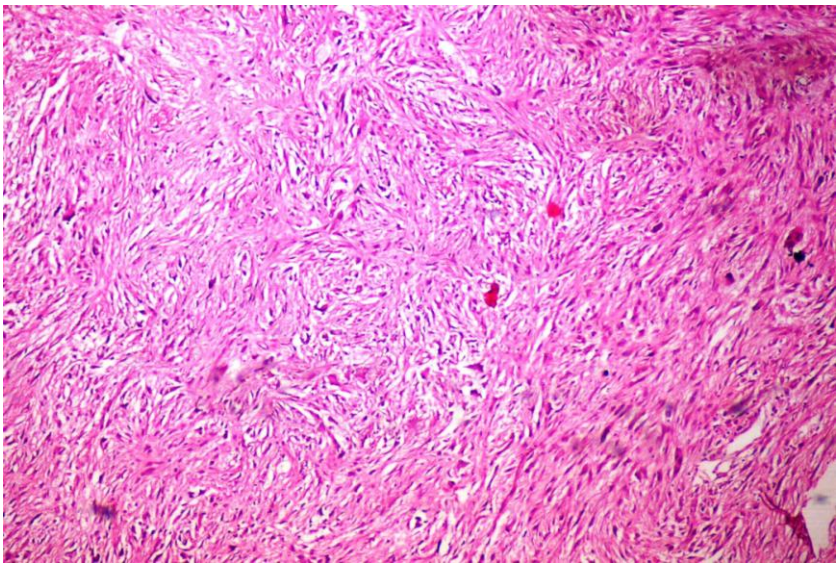
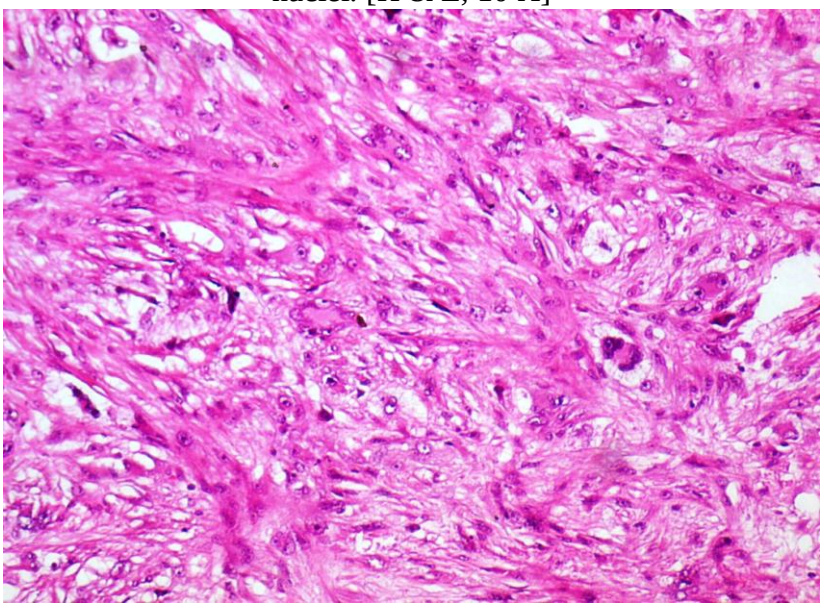


Photo.7 Section showed marked pleomorphism with haphazard arrangement of tumor cells. There were numerous mitotic figures and many multinucleated giant cells with bizarre nuclei. [H & E, 10 X]



Acknowledgement

Authors acknowledge the immense help received from the scholars whose articles are cited and included in references of this manuscript. The authors are also grateful to authors / editors /publishers of all those articles, journals and books from where the literature for this article has been reviewed and discussed.

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