Atypical Presentation of Recurrent High-Grade Myxofibrosarcoma – A Case Report

Meenakshi Yeola[1], Kushagra Singh[2], Aditya Mehta[3], Roshani S. Manekk[4]

[1, 2, 3, 4] Department of General Surgery, Jawaharlal Nehru Medical College, Datta Meghe Institute of Medical Sciences (Deemed University), Sawangi (Meghe) Wardha, Maharashtra, India.

INTRODUCTION

Myxofibrosarcoma (MFS) is an unusual malignancy, a type of Soft Tumour Sarcoma (STS) with a proclivity for the upper and lower extremities. The World Health Organization (WHO) in the year 2002, declared a precise classification for MFS and stated it as a specific pathologic entity. A very striking clinical feature of MFS is its predilection for recurring locally. MFS is relatively uncommon and has unique sites of presentation with no standard protocols to guide the treatment decisions making approach to every case unique.

PRESENTATION OF CASE

A 65-year-old female presented with complaints of swelling over the right infraclavicular fossa since 1 month. The swelling was insidious in onset and gradually progressive in nature and was not associated with any symptoms of pain over the swelling, loss of weight, loss of appetite, breathlessness and backache. Patient is a known case of hypertension. The patient also gave history of similar swelling in the past which was excised two times, once 5 months ago and another 3 months prior to current presentation. The previous histopathology report was suggestive of "High Grade Myxofibrosarcoma". The cells were mild to moderately pleomorphic spindle cells with moderate amount of eosinophilic cytoplasm, variable mitotic activity of up to 4 per HPF (High Power Field) with less than 50 % of focal necrosis and focal myxoid areas were present. It is spindle cell tumour involving the adipose tissue and skeletal muscle. Immunohistochemistry was done, the tumour cells express SMA (Smooth Muscle Actin) and CDK 4 and are immunonegative for desmin, h-caldesmon, CD-34, S-100 protein ALK and MDM-2. [Figure 1]

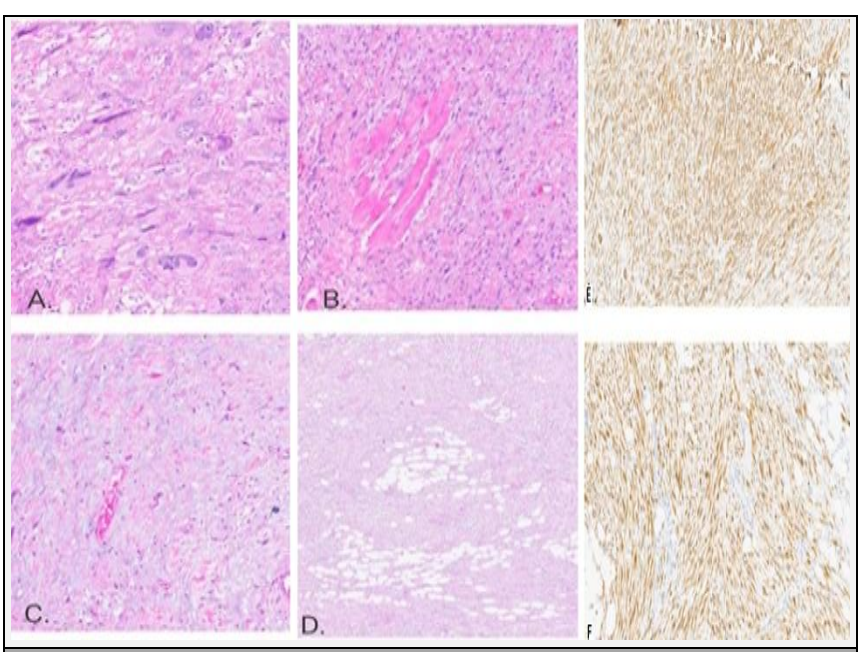

Figure 1. Histopathological Features of High-Grade Myxofibrosarcoma in the Present Case Study: A. Bizarre Cells, B. Involvement of Skeletal Muscle, C. Myxoid Areas, D. Involvement of Adipose Tissue.: Immunohistochemistry (IHC) Images E. SMA IHC, F. CDK4 IHC

Corresponding Author:
Dr. Kushagra Singh,
Department of Surgery,
DMIMS (DU), Sawangi (M),
Wardha, Maharashtra,
India.
E-mail: dr.kushagrasingh@gmail.com

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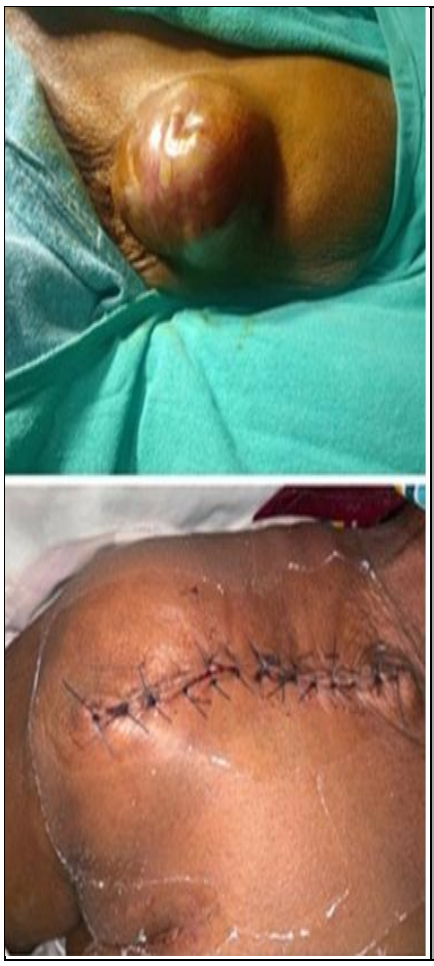

Figure 2. Preoperative and Immediate Post-Operative Clinical Picture

On examination of the right infraclavicular fossa, there was a single ovoid swelling of approximately 6 x 5 cms size, extending 4 cms lateral to sternal notch and up to 5 cms proximal to the shoulder joint in the horizontal plane. The margins of the swelling were well defined with evidence of dilated veins over the swelling. There were no visible pulsations or sinuses or scars over the swelling. The skin overlying the swelling was erythematous with multiple blebs over the skin. There was no tenderness and no local rise of temperature over the swelling. The swelling had smooth margins with irregular surface and was firm in consistency. Due to the swelling the patient had restricted abduction of the right shoulder joint. [Figure 2]

Routine haematological investigations were within normal limits. A High-Resolution Computed Tomography (HRCT) of thorax was done which was suggestive of a well-defined heterogeneously enhancing mass of size 5.7 x 4.4 x 3.9 cms with necrotic areas in the right infraclavicular region. The lesion is abutting the pectoralis major muscle posteriorly and there is no evidence of erosion to the adjacent bone. There is an evidence of a single enlarged lymph node in the precranial groove of size 1.4 x 1 cms. The Internal Jugular Vein (IJV) and the Sub-Clavian Vein (SCV) is dilated on the right side. All the features are suggestive of a possible neoplastic aetiology.

CLINICAL DIAGNOSIS

Recurrent myxofibrosarcoma over the right infraclavicular fossa

DISCUSSION OF MANAGEMENT

She was taken for wide local excision of the swelling. Curvilinear incision was given at the inferior margin of the tumor and the tumor was carefully dissected out circumferentially with 2 cms of healthy margins. Skin flap raised inferiorly and superiorly to the incision and primary closure of wound was done. The specimen was sent for histopathological examination. The histopathology report was suggestive of Malignant Fibrous Histiocytoma (MFH). A medical oncology opinion was taken, and the patient was advised radiation followed by chemotherapy with adriamycin and ifosfamide. Post operatively a full body PET (Positron Emission Tomography) scan was done which showed no evidence of any abnormal hypermetabolic or enhancing lesion in the body. The patient received radiotherapy over 2 months with dose of 60 Gy in 30 fractions over the right infraclavicular and supraclavicular fossa area. The patient is currently 5 months post-operative with a healthy scar and no signs of recurrence.

DISCUSSION

Myxofibrosarcomas has equal distribution in both sexes and were usually seen in the extremities, were deep-seated and had a mean age of occurrence at 66 years.[1] MFS exhibit a propensity for local recurrence.[2] Our patient was a 65 years old female who has presented to us with local recurrence for the 3rd time and has undergone 2 prior excisions for the tumour on same site.

The common clinical findings seen in a study was a slow growing, painless, smooth and firm mass which was suggesting differential diagnosis such as lipoma, neurofibroma, or follicular cyst or cutaneous leiomyosarcoma. Some cases also showed papulonodular lesion indicating inflammatory changes. Also, there were cases in which there was no evidence of systemic metastatic disease at the time of presentation.[3] In another study also, done by Whelan Sharon et al[4] a painless, gradually enlarging mass was found to be the most common symptom. In our case also, the patient presented with a painless swelling which gradually progressed over a period of 1 month. The skin overlying the swelling was erythematous with multiple blebs over the skin. The swelling had smooth margins with irregular surface and was firm in consistency.

Preoperatively, evaluation of tumour with MRI or CT scan is difficult as well, leading to underestimation of tumour extension due to the specific infiltrative pattern of MFS.[5] Imaging is essential for pre-surgical planning and for a good complete resection.[6] In our case also HRCT thorax was done, but it could not provide a definitive diagnosis of myxofibrosarcoma, however it proved to be useful in providing information that, the lesion is abutting the pectoralis muscle posteriorly and there is no evidence of erosion to the adjacent bone, thus helping us plan the wide local excision.

A study shows that, some of the tumours having myxoid areas consisted well differentiated uniform spindled cells, showing a range of differentiation, from fibroblastic cells which were well differentiated to cells that had marked pleomorphic features, hyperchromasia, and multinucleate

cells. Many of the cells also had abundant eosinophilic cytoplasm. The cells showed typical and atypical mitotic figures and they ranged from ≤ 1 up to 8 cells per 10 HPF.[4] In our case also, the histopathology report of the tumour showed cells which were mild to moderately pleomorphic spindle cells with moderate amount of eosinophilic cytoplasm. There was variable mitotic activity of up to 4 per HPF with less than 50 % of focal necrosis and focal myxoid areas were present. It was a spindle cell tumour involving the adipose tissue and skeletal muscle.

A study shows that, within the various histological subtypes of "MFH", pleomorphic type shows frequent positivity for SMA and desmin.[7] Another study showed that myxofibrosarcomas that were stained for MDM2 and CDK4 were located in the deep soft tissues of extremities (limb and arm).[8] The study also states that, MDM2 immuno staining might be more sensitive than CDK4 staining but CDK4 is the more specific.[9] In our study the, tumour cells express SMA and CDK 4 and are immune negative for desmin, h-caldesmon, CD-34, S-100 protein ALK and MDM-2.

Surgical resection of the tumour with wide margins continues to be the foundation for treating MFS. Radiotherapy and chemotherapy can be given in the neoadjuvant and adjuvant settings.[10] Radiotherapy has been shown to prevent from local recurrence during the first 5 years, but not in the longer period.[5,11] In our case, the patient has undergone wide local excision with 2 cm of healthy margins and has received first dose of radiotherapy over 2 months with dose of 60 Gy in 30 fractions over the right infraclavicular and supraclavicular area. Currently the patient is 5 months post-operative and does not show any signs of recurrence of the tumour.

A study done by Odei B et al shows that, 15 – 38 % of LRs (Local Recurrence) in MFS advance to higher histologic grade with an significant increase in metastatic potential.[10] However in the present case, though the tumour was high grade, the PET scan did not show, evidence of any abnormal hyper metabolic or enhancing lesion noted elsewhere in the body.

The study done by Odei B et al, also states that, the most commonly used chemotherapy agents for MFS were ifosfamide and adriamycin.[10] In our case also, oncologist opinion was taken, and the patient was advised radiation followed by chemotherapy with adriamycin and ifosfamide.

FINAL DIAGNOSIS

Recurrent high-grade myxofibrosarcoma

CONCLUSIONS

Even though myxofibrosarcomas have predilection for extremities, it can also present proximal to the limbs as in this case. Also, despite the high local recurrence, the patient did not show any evidence of distant metastasis. Thus, further follow-up of patients and more studies are required to estimate the outcome of radiotherapy and chemotherapy in such cases.

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Disclosure forms provided by the authors are available with the full text of this article at jemds.com.

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