

A Recurrent Myxofibrosarcoma Over Chest Wall-A Rare Case.

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Abstract: Myxofibrosarcoma typically occurs on old age and the most common sites are extremities. But our case is a very rare case as in this case a 24 year young male developed a swelling in left upper antero-lateral chest wall which was diagnosed as fibrohistiocytic tumour and was operated in some other local hospital and the patient presented to us again with a recurrent swelling at the same site without evidence of metastasis after 20days. Wide local excision of the tumour was done and now the patient is all right without any evidence of recurrence. Thus giving a message how important is proper wide local excision in the treatment of myxofibrosarcoma.

Keywords: Myxofibrosarcoma, Recurrence, Wide local excision.

I. Introduction

Sarcomas are heterogeneous group of neoplasm that arise from cells of embryonic mesoderm. Soft tissue sarcomas include more than 50 histologic subtypes [1]. The most common subtypes in adults are malignant fibrous histiocytoma. Malignant fibrous histiocytoma is classified as either leiomyosarcoma, pleomorphic undifferentiated sarcoma, myxofibrosarcoma, or dedifferentiated liposarcoma based on cellular differentiation and genetics. So proper diagnosis and treatment is a big challenge to surgeons.

II. Case report

In our case a 24 year young male developed swelling in the left upper antero- lateral chest wall, which was firm, non tender, and mobile of size 3X4cm not fixed to chest wall and surrounding skin. No axillary lymphadenopathy. FNAC suggested a fibrohistiocytic tumour (pseudo-sarcomatous lesion). The patient underwent excision of the tumour. The tumour was sent for histopathology and it came out as inflammatory myofibroblastic tumour.

After 20 days the patient presented with a swelling of size 4x3 cm which was tender, rapidly progressing in size, firm, mobile not fixed to chest wall and skin in the same operative site (fig.1). On FNAC the smear showed atypical spindle cells (having ovoid to elongated nuclei with variable nuclear pleomorphism) seen in clusters and discrete manner in a myxoid background. Occasional multinucleated giant cells with few inflammatory cells found. From the above finding it was suggestive of spindle cell tumour. On high resolution sonogram there was a mixed echogenic solid intramuscular space occupying lesion of size 3.6x 2.8 cm. On CT scan, a well defined hypodense mass of size 4.6x 3.2x 2.5 cm & HU 5-20 was noted over left pectoralis major muscle less than 1cm from surface. Surrounding muscle tissue reveals normal texture & density. Contrast study did not show any enhancement (fig.2a, 2b).

So our provisional diagnosis was recurrent fibrosarcoma and we planned for wide excision of tumour. Intraoperatively it was found that tumour was intramuscular, in pectoralis major muscle. Grossly it was a white nodular mass measuring 3.4x 2.2 cm with soft gelatinous surface present within the muscle (fig.3, 4). On HPE it was a myxofibrosarcoma grade 1 as per FNCLCC system with tumour free surgical margin (fig.5, 6a & 6b). The patient was followed up to 2 months till now no evidence of recurrence or metastasis to other sites has been found.

III. Discussion

Soft tissue sarcomas include more than 50 histologic subtypes [1]. The most common subtypes in adults are malignant fibrous histiocytoma. This tumour is no longer believed to show histiocytic differentiation but rather fibroblastic differentiation. Because of the broad range of histological appearances, MFH is divided

into four subtypes; storiform–pleomorphic, myxoid, giant cell and inflammatory [2, 3]. The myxoid type, also known as a myxofibrosarcoma (MFS), is the second most common type [4, 5].

MFS is the most common soft tissue sarcoma that occurs in late adult life, peaking in the seventh decade and is mainly encountered in the lower extremities (77%), trunk (12%), retro peritoneum or mediastinum (8%) [6].

In our case the tumour was located in left upper anterolateral chest wall and the patient was 24 years old. Thus it is very rare to have the tumour in such site in such an early age. The tumour was excised but it recurred within 20 days and that too the size of tumour increased very rapidly suggesting the aggressive locally recurrent nature of the tumour. Myxofibrosarcoma is separated into two groups: superficial (dermal/subcutaneous) and deep (intramuscular/subfascial)[7]. While the superficial group of tumours infiltrate and spread extensively in a longitudinal manner, the tumours arising in the intramuscular/subfascial plane were noted to form a single discrete mass. In our case the tumour was intramuscular found in pectoralis major muscle which was nodular and formed a single discrete mass with soft gelatinous surface.

The two most widely used systems for histological grading are the NCI (United States National Cancer Institute) system and the FNCLCC (French Fédération Nationale des Centres de Lutte Contre le Cancer) system. The NCI system uses a combination of histological type, cellularity, pleomorphism and mitotic rate for attributing grade 1 or 3. The FNCLCC system is based on a score obtained by evaluating three parameters selected after multivariate analysis of several histological features: tumour differentiation, mitotic rate and amount of tumour necrosis. A score is attributed independently to each parameter and the grade is obtained by adding the three attributed scores. According to the FNCLCC system in our case the tumour was of grade 1. Thus the tumour was of low grade. In myxofibrosarcoma low grade tumour has less potential for metastasis but equal chance of local recurrence as compared to high grade tumour and also if the tumour is recurrent then the chances of metastatic spread increases. Most commonly metastasis occurs by haematogenous spread. However metastasis through lymphatic spread can be seen. Most common site of metastasis is lungs and it occurs within 2 years of initial diagnosis. Other sites of metastasis are liver, brain, bones etc. In our case there was no evidence of metastasis. However the tumour was recurrent may be due to inadequate tumour free surgical margin. But after our wide local excision with adequate tumour free surgical margin till now no evidence of local recurrence is seen. The most challenging task is to identify the extent of tumour spread and the type of tumour. In our case we have used, Ultrasonography and CT scan to determine the extent of tumour spread and evidence of metastasis. Ideally MRI is the preferred imaging technique for soft tissue sarcomas but ultrasonography may be used in patients with soft tissue sarcomas for diagnostic role as well as for post operative surveillance and to guide biopsies. CT scan of chest, abdomen, pelvis should be performed to assess lung, liver, peritoneal or pelvic metastasis. MRI accurately delineates muscle groups and distinguishes among bone, vascular structures and tumour. Haemorrhagic, cystic, necrotic changes may be observed. It also distinguishes between benign lesions and their malignant counterpart. It is also valuable for determining tumour recurrence after surgery. FNAC has low diagnostic accuracy for primary tumour as compared to core needle biopsy still it is acceptable method and it is the procedure of choice to confirm and rule out the presence of a metastatic focus or local recurrence. According to TNM staging in our case it is stage IA (T1b N0 M0).

Surgery is the principal modality of treatment. Wide local excision is done for low grade myxofibrosarcoma without any metastasis. Thus in our case wide local excision was done and the specimen was sent for histopathological examination. The reports confirmed it to be a myxofibrosarcoma with tumour free margins.

If the surgical margins are not tumour free then post operatively radiotherapy is advised. However the treatment modality for high grade Myxofibrosarcoma is controversial and a multimodality approach is followed including all i.e. surgery, radiotherapy and chemotherapy. But for low grade myxofibrosarcoma without metastasis proper wide local excision is very important and can prevent recurrence.

IV. Figures



Fig.(1): Showing the mass at previous operative site.



Fig. (2a): Arrow showing the tumour mass in cross section of CT scan chest.

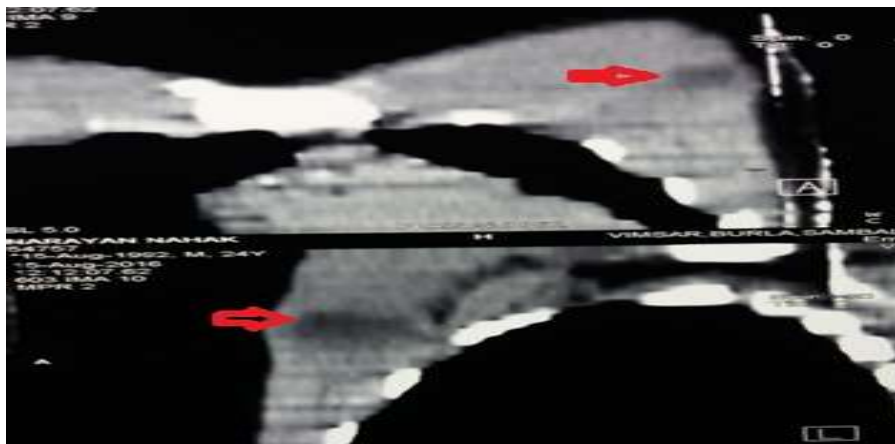


Fig. (2b): Arrow showing the tumour mass in coronal section of CT scan chest.



Fig.(3): Showing the mass in the pectoralis major muscle.



Fig.(4): Showing the white gelatinous mass in the excised sample.

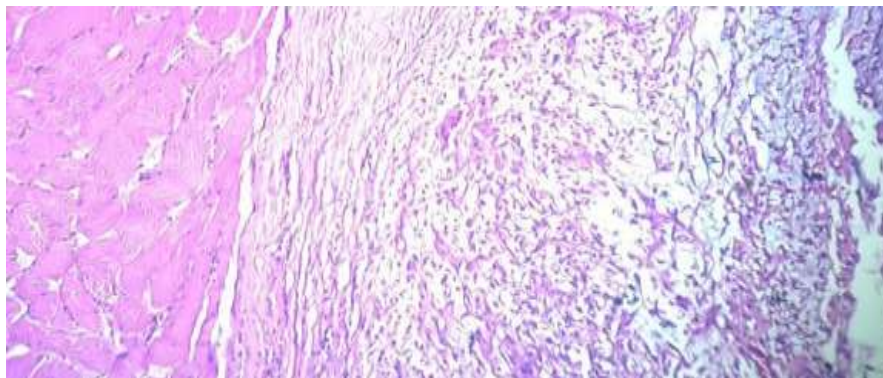


Fig.(5): Scanner view showing normal skeletal muscle on left and tumour tissue on right side.

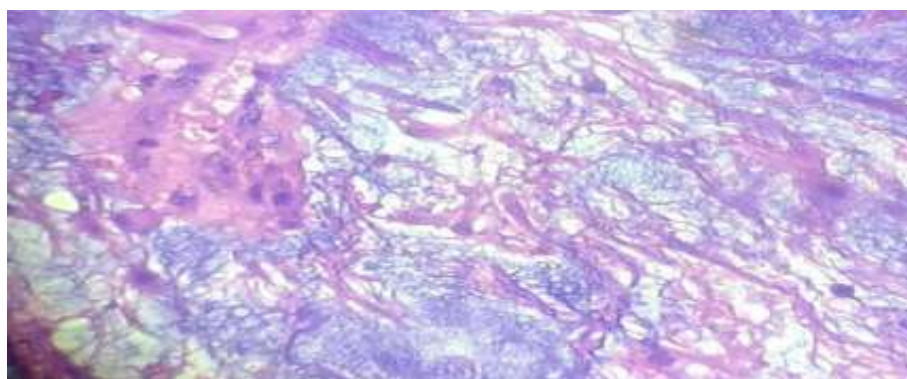


Fig.(6a): High power view(40x) showing pleomorphic cells with high nuclear cytoplasmic ratio, prominent nucleoli in a myxoid background.

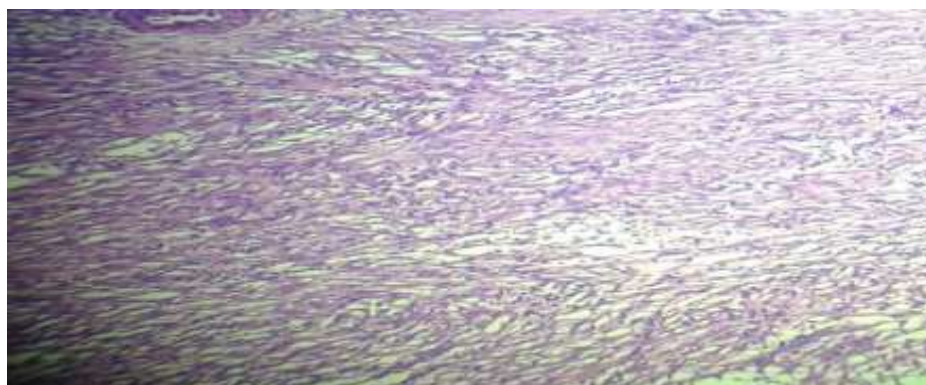


Fig.(6b): 20x view showing Tumour cells present in streaming pattern, Herring bone pattern in a myxoid background.

V. Conclusion

Myxofibrosarcoma are highly recurrent tumours and has high potential to metastasis. Hence early diagnosis with proper wide local excision is must for preventing its dreadful consequences.

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