

A Rare Huge Myxofibrosarcoma of Chest Wall

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Abstract: Primary soft tissue sarcoma of chest wall is an uncommon lesion. Mass of a chest wall should be considered as malignant until proven otherwise. Myxofibrosarcoma is one of the most aggressive types of soft tissue tumor. Due to its rarity over trunk wall and high possibility of malignancy, an early investigation and diagnosis is crucial. Subsequently, an early excision remains the mainstay of treatment. A multidisciplinary team approach for diagnosis and treatment is crucial.

I. Introduction

Myxofibrosarcoma (MFS) is a rare soft tissue sarcoma which accounts for less than 1% of adult malignancies [1]. MFS is one of the most aggressive types of soft tissue neoplasms commonly seen over the extremities then followed by the trunk, pelvis, head and neck region and the genital area. It is mostly seen in the elderly age group patients who presents with asymptomatic swelling of long duration. Higher possibility for delay in diagnosis or misdiagnosis due to variety of clinical presentation and histologically heterogenous features.

Prognostic factors for this disease and metastasis free survival depends on the tumour size at resection, tumour grade, positive surgical resection margins, percentage of necrosis and mitotic rate [3]. Low-grade lesions have relatively low metastatic potential while intermediate and high-grade lesions demonstrate increasing rates of distant metastases [4]. In view of MFS exhibit higher predilection for local recurrence, aggressive surgery combined with radiotherapy may contribute to more effective local control [3]. We present a case of a 70 years old male with a rare huge myxofibrosarcoma of chest wall.

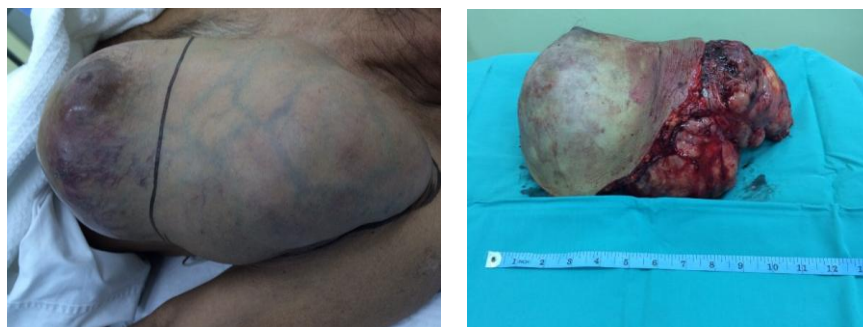
II. Case Report

A 70 years old man, with no underlying medical illness presented with painless left chest wall swelling for one and half year. The swelling was progressively increasing in size with occasional throbbing pain. Patient had no history of trauma or irradiation. There was no family history of malignancy. On examination, there was swelling over left chest wall, measuring 25 by 15cm, non tender, irregular surface, firm in consistency with dilated vessels over the surface. Systemic examinations were unremarkable. His full blood count showed haemoglobin of 8.9g/dL which was normochromic normocytic suggestive of anemia of chronic disease with leucocytosis and thrombocytosis as well. Other blood investigations were within normal range. A trucut biopsy was done which showed high grade sarcoma with fibroblastic appearances. CECT TAP done showed aggressive looking mass in left supraclavicular region. No evidences of distant metastasis to other organs.

Patient underwent excision of the tumour which measures 20 by 31cm with depth of 10cm. Pectoralis major and minor was partially excised. No breach into thoracic cavity or involvement of rib cage. Skin was unable to be approximated hence patient underwent daily povidone and liquid paraffin dressing. Patient was planned for skin grafting however he was not keen.

Macroscopic, a single large suprafascial mass (29cm x 17cm x 11.5cm), weighing 3.6kg with invasion through the fascia forming a satellite nodule. Microscopically, histological type was myxofibrosarcoma (Fédération Nationale des Centres de Lutte Contre le Cancer [FNCLCC] Grade III). Necrosis seen extent 30 – 40% of tumour. Regional lymph nodes cannot be assessed. Lymphovascular invasion not identified. The tumour was incompletely excised as it reaches the superior, antero-superior, postero-superior and medial margins. No histological confirmed distant metastasis. American Joint Committee on Cancer (AJCC) classification was pT2b pNx pMx. Final histopathological examination result was myxofibrosarcoma.

Patient was planned to be referred to oncologist but he was not keen. During the last follow up, the wound over left chest wall was clean with granulation tissues seen.



III. Discussions

MFS is a malignant mesenchymal tumor which often grouped with other malignant fibrous histiocytomas (MFH) of variable prognosis. Formerly, it was recognized as part of myxoid fibroblastic sarcoma [3]. In year 2002, World Health Organization determined that myxoid MFH without myogenic, lipoblastic or chondrogenic features diagnosed as MFS [5]. Approximately 7% of primary sarcomas arise from chest wall and account for a significant number of deaths from malignant tumours [1]. The average age of onset is 53 years old with male prevalence (66.7%) [5]. Patient usually presents with painless gradually enlarging mass. The median tumor size of 6.0 cm [3]. The clinical symptoms accompany diagnosis of soft tissue sarcoma (STS) are nonspecific [6].

There is no clearly defined aetiological factor for STS. There is a role of genetic factor in the initiation and progression of the sarcomas. Genetic mutations in pluripotent mesenchymal stem cells are believed to give rise to malignant clones, which lead to the formation of STS [6]. Mutations in tumor suppressor genes such as p53 and RB-1 and oncogenes have also showed association to STS and may also play a role in its prognosis [6]. Other risk factors include dose-dependent exposure to radiation, lymphedema, any exposure to phenoxyherbicides, chlorophenols and others [6]. Several inherited conditions such as neurofibromatosis type 1 (Von Recklinghausen disease), retinoblastoma and Gardner syndrome have STS as a component[6].

Imaging occasionally aid in definitive diagnosis by suggesting the most likely histologic STS subtype and helps in assessing the local and distant metastasis of the lesion[6]. A chest X-ray useful as screening tool although computed tomography (CT) of thorax more sensitive to detect pulmonary metastasis. Other useful tools such as magnetic resonances imaging (MRI) and positron emission tomography (PET) scan useful to identify metastasis in patients with recurrent high grade tumours [6].

Previously open surgical biopsies was considered gold standard for diagnosis however it was associated with complications such as seroma, infection, wound dehiscences and tumour implantation [1]. Alternatively, image-guided core needle biopsies have shown accurate diagnostic results [1]. Narvani et al. reported that biopsies obtained by image guidance are significantly more accurate compared with blind biopsies (95% versus 78%). Based on their report, demonstrates that image-guided core needle biopsy is a safe and highly accurate diagnostic modality to determine malignancy, histological subtype, and high grade differentiation of chest wall musculoskeletal tumors [1]. Immuno-staining is mandatory to exclude other entities that might mimic MFS [4].

The current iteration of AJCC, STS staging includes Tumor (T), Node (N) and Metastasis (M) with “a” indicating superficial and “b” indicating deep. (T) is tumour size which determined by physical examination or radiologically, subdivided to T1 for those less than or equal to 5cm and T2 for those greater than 5cm [6]. The regional lymph node involvement designated as Nx if unable to be assess, N0 for no lymph node metastasis and N1 for those with lymph node metastasis [6]. Presences of distant metastasis designated as Mx if unable to assess, M0 if no metastasis and M1 for those with metastasis [6]. Stages I to III describe localized STS, whereas Stage IV disease includes STS that has metastasized to lymph nodes and/or other distant sites [6].

Treatment of MFS consist of surgery which should be performed with a goal of attaining greater than 1-cm resection margins whenever possible [4]. Combined surgical and adjuvant radiotherapy, in order to avoid local and distant recurrences of the tumour, is absolutely recommended [5]. A possible re-excision of recurrent lesions considered mainstay therapy for disease control[5].

Five-year survival of patient with MFS was better when compared to other STS. The key determinant for survival is control of both local recurrence as well as distant dissemination [6]. For MFS, a positive or close resection margins would be associated with an increased risk of local recurrences (LR) [4]. Median time to first local recurrences was 21 months (range, 6–48 months) [4]. It appears that approximately half of patients with MFS who develop a LR will ultimately develop two or more subsequent LR [4]. An aggressive follow-up in order to search for a metastatic disease is advisable in all cases[5].

IV. Conclusion

Myxofibrosarcoma of chest wall is a rare soft tissue sarcoma with a better survival rate provided an early diagnosis with relevant investigations and subsequently generous surgical excision performed. Due to high risk of local recurrence, subsequent treatment with adjuvant radiotherapy would help. It is mandatory for a proper follow up subsequently in view of possibility of recurrences locally as well as distant metastasis.

References

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