Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2015; 3(3):267-270 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources)
www.saspublishers.com DOI: 10.36347/sjmcr.2015.v03i03.027

Undifferentiated High Grade Pleomorphic Sarcoma of the Chest Wall: A Rare Entity

Channabasappa Kori[1*], Kiran Preet Malhotra[2], P N V Vamsidhar[3], Sameer Gupta[4], Saumya Shukla[5], Vijay Kumar[6]

[1,3]Senior Resident, Department of Surgical Oncology, King George's Medical University, Lucknow, Uttar Pradesh, India
[2,5]Assistant Professor, Department of Pathology, Dr. RML Institute of Medical Sciences, Lucknow, Uttar Pradesh, India
[4]Assistant Professor, Department of Surgical Oncology, King George's Medical University, Uttar Pradesh, India
[6]Associate Professor, Department of Surgical Oncology, King George's Medical University, Lucknow, Uttar Pradesh, India

***Corresponding Author:**
Name: Dr. Channabasappa Kori
Email: channabasappakori@gmail.com

Abstract: Undifferentiated high grade pleomorphic sarcoma is a deep seated pleomorphic soft tissue sarcoma of unknown pathogenesis. It typically occurs in the deep fascia and skeletal muscles of the extremities followed by the trunk and the head and neck. Chest wall is an uncommon site of origin. Chest wall tumors account for less than 1% of all tumors and are classified based on their tissue of origin. They may be primary or secondary and almost both occurring with equal incidence. Most common differential diagnosis of chest wall tumors is generally metastasis from carcinoma of the breast, lung, kidney, thyroid or multiple myeloma arising from ribs. Other differentiated sarcomas with pleomorphic histology which were erroneously classified categorized as malignant fibrous histiocytoma also have diverse prognosis and require exclusion. To the best of our knowledge, few cases of undifferentiated high grade pleomorphic sarcoma, primarily arising from chest wall are noted in the contemporary literature. We report a rare case, this entity arising in the right chest wall of 70 year old gentleman which was successfully treated with aggressive surgery and radiation.
Keywords: Undifferentiated high grade pleomorphic sarcoma, chest wall, malignant fibrous histiocytoma

INTRODUCTION

Malignant fibrous histiocytoma (MFH) was the term earlier used to define sarcomas with pleomorphic features on histology. As a group they compromise the most common sarcomas in adults, generally affecting extremities and retroperitoneum. Chest wall is an extremely unusual site of their occurrence. However; with development of immunohistochemical typing of sarcomas, a diagnosis of "Undifferentiated high grade pleomorphic sarcoma"(UPS) or Pleomorphic MFH is based on exclusion of other lines of differentiation. These true undifferentiated sarcomas are of rare occurrence with advancement of Immunohistochemical techniques and have a falling incidence in the contemporary literature. Few cases of UPS/ Pleomorphic MFH primarily involving chest wall are reported. Proper imaging to define the extent of the tumor and complete surgical excision is the cornerstone of management.

This article presents a rare case of malignant fibrous histiocytoma of the right anterior chest wall in a 70 year old gentleman, who was successfully treated with aggressive surgery and radiation.

CASE REPORT

A 70-year-old man, a hypertensive and a diabetic, controlled on medications, presented to us with a recurrent painless mass in the right anterior chest wall of 15 months duration. Clinical examination revealed a 20 x 20 cm, well-circumscribed mass lesion originating from the chest wall, with restricted mobility (Fig. 1a & 1b). Axillary lymph nodes were not palpable. CECT of the chest showed well defined heterogeneously enhancing lesion measuring 20 x18 x 15 cm, involving pectoralis major and having ill-defined interface with ribs and chest wall muscles. No evidence of intrathoracic extension was seen (Fig. 2).

Tru-cut biopsy from the mass was suggestive of a high-grade spindle cell sarcoma. Metastatic work up was negative. The patient was taken up for surgery and a radical *en-bloc* excision of the chest wall tumor was performed, which included removal of pectoralis major and minor muscle. The ribs appeared uninvolved during surgery, hence a clear plane of excision was obtained sparing the ribs.

Gross examination showed tumor measuring 19 x18 x 16 cm in size (Fig. 3). Microscopy revealed a highly cellular tumor involving the dermis and subcutaneous tissue with infiltration of underlying muscle (Fig. 4). Tumor cells were highly pleomorphic and spindle shaped with high nucleo-cytoplasmic ratio and mitotic figure averaging 20-25 per 10 high power field. Immunochemistry study (IHC) showed tumor cells were positive for Vimentin (Fig. 5), but negative for markers of other sarcomas including Desmin, Pancytokeratin, SMA, S-100, CD 34, and MDM-2. Spindle cell melanoma was excluded by negativity for Melan A. Final diagnosis of UPS/ Pleomorphic MFH was made. All surgical resection margins were clear. Patient received adjuvant external beam radiotherapy and a booster dose to tumor bed (55-60 CGy over 5-6 weeks). Patient is disease free at 8 months of follow up.

Fig. 1a & 1b: Clinical photograph showing 20× 20 cm, well-circumscribed mass lesion originating from the right chest wall

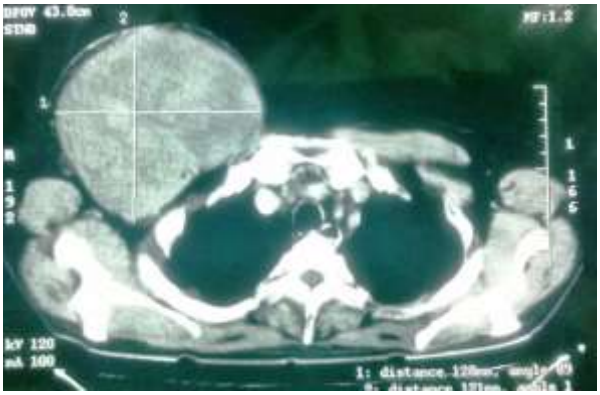

Fig. 2: CECT of the chest showing a well defined heterogeneously enhancing lesion involving pectoralis major and having ill-defined interface with ribs and chest wall muscles

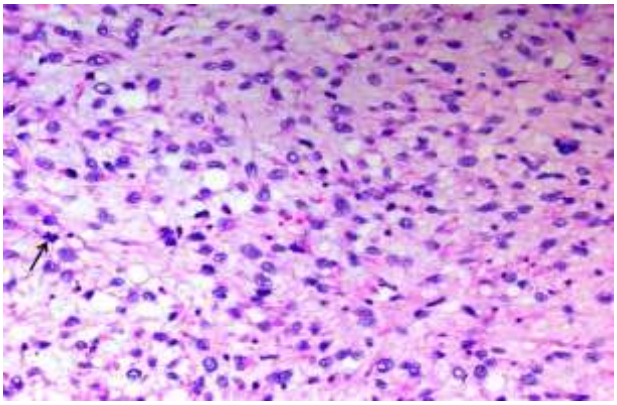

Fig. 4: Photomicrograph showing a hypercellular tumor with pleomorphic spindle cells and atypical mitosis (arrow) (H&E, x200)

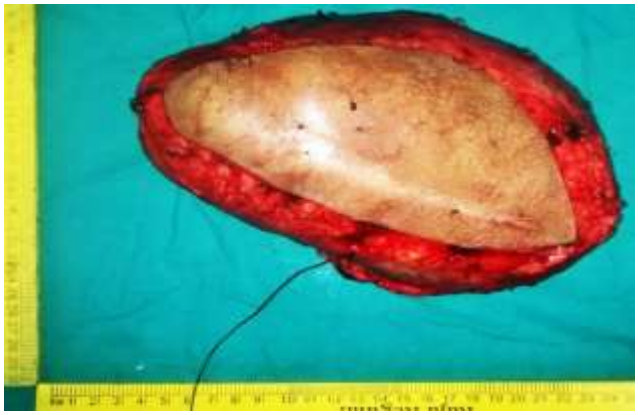

Fig. 3: Surgical resection specimen showing a tumor measuring 19 x18 x 16 cm in size

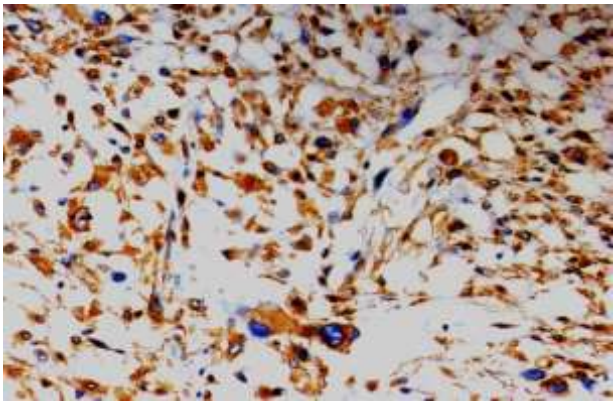

Fig. 5: The pleomorphic spindle cells are positive for Vimentin (DAB, x200)

DISCUSSION

Primary UPS/ Pleomorphic MFH of the chest wall is an uncommon entity [1, 2]. Exact aetiopathogenesis is not known and was initially thought to arise from histiocytes [3]. The earlier lesion known as MFH was first described by O' Brien and Stout in 1964 [3]. MFH was noted to affect the deep fascia, skeletal muscle or subcutaneous tissue [4, 5]. It was the most common soft tissue neoplasm in adults (20-30%) and was usually located in the extremities or the retroperitoneum. Chest wall was a rarely involved site [6].

MFH most commonly affected males in the fifth decade of life. Literature review identifies several subtypes of MFH including storiform /pleomorphic, myxoid, inflammatory and giant cell variety. Storiform/pleomorphic variety was most common histological subtype accounting for 50 to 60% of cases. Myxoid type was considered the second common variant representing 20% of cases and was least aggressive in nature. Most common presentation was slowly growing painless mass. Few cases of MFH occurring secondary to pyothorax, radiotherapy, burns or after thoracotomies have been reported in the literature [7]. Due to the diversity of morphology and prognosis in these lesions, the World Health Organization (2002), pleomorphic MFH/ UPS not otherwise specified as a distinct entity. This mandates the exclusion of other cell lines of sarcoma differentiation, representing a final pathway progression of soft tissue neoplasm's towards undifferentiation [8]. Hence the apparent incidence of correctly diagnosed pleomorphic MFH is falling. These lesions are also known to affect similar age groups and sites, with chest wall being a rare site of occurrence.

Proper imaging to define the extent of the tumor and complete surgical excision is the cornerstone of management. Plain X-ray is non specific, showing only focal soft tissue density and calcification in 5to 20% of cases. Cortical Erosion of bone is highly suggestive of diagnosis and preferentially seen in MFH and synovial sarcoma as compared to other soft tissue tumors. Contrast enhanced computed tomography (CECT) and Magnetic resonance imaging (MRI) are useful in assessment of soft tissue component. MRI better displays soft tissue extent and infiltration of bone marrow, while CECT shows accurate assessment of cortical bone involvement [9]. Role of positron emission tomography/ computed tomography (PET/CT) in assessment of primary tumor and metastases is ill defined [10]. Final diagnosis is based on histopathological examination. Immunohistochemistry (IHC) is useful to exclude other pleomorphic spindle cell lesions including synovial sarcoma, Leiomyosarcoma, Liposarcoma, Rhabdomyosarcoma, Sarcomatoid carcinoma and spindle cell melanoma.

Surgery is the definitive modality and consists of aggressive resction with wide safety margins and dissection of locoregional nodes [11, 12]. Role of chemotherapy is not well established in the literature and useful mainly in metastatic/ recurrent disease (unresectable). Radiotherapy is mainly indicated in cases of recurrences or resection with close /positive margins and in large tumors -tumor with diameter greater than 5cm, have the highest risk of disease recurrence [13].

Local recurrences are noted in 20 to 30% of all cases. Highest risk of recurrences was noted in retroperitoneal and head and neck sarcoma and lowest risk in extremity sarcoma. Prognostic factors of MFH include size, grade, depth, metastatic status and histological subtype [14]. These factors usually correlate with survival status in these patients. Oda et al. have similarly reported that tumors arising from retroperitoneum, head and neck with tumor size greater than 5cm, deep location and high grade were notable factors indicating a worse prognosis [15]. In a study by Gibbs *et al*. 10 year survival rates for low, intermediate and high grade tumors were reported as 90%, 60% and 20% respectively [15]. Inability to achieve negative margins is the most common cause of recurrence.

In the present case, we were able to achieve negative margins with wide local excision and patient was promptly treated with external beam radiotherapy. Such an approach may improve patient survival and reduce chances of recurrence as in our case.

CONCLUSION

It is rare case of undifferentiated high grade pleomorphic sarcoma primarily arising from the right chest wall of 70 year old man which was successfully treated with aggressive surgery and radiation. In this case, we were able to achieve negative margins with wide local excision. Patient was promptly treated with external beam radiotherapy. This approach may improve patient survival and reduce recurrence.

Abbreviations

UPS: Undifferentiated high grade pleomorphic sarcoma; MFH: Malignant fibrous histiocytoma, CD: Cluster of differentiation, SMA: Smooth muscle antigen, IHC: Immunohistochemistry.

REFERENCES

1. Shikada Y, Yoshino I, Fukuyama S, Kameyama T, Maehara Y; Complete wide resection of the malignant fibrous histocytoma of the chest wall; expect for long survival. Ann Thorac Cardiovasc Surg., 2006; 12(2): 141-144.

2. Jin-Kyung A, Ki Keun O; Malignant fibrous histiocytoma of chest wall. Yonsei Med J., 2005; 46(1):177-180.
3. O'Brien JE, Stout AP; Malignant fibrous xanthomas. Cancer, 1964; 17:1445-1455.
4. Shinjo K; Analysis of prognostic factors and chemotherapy of malignant fibrous histiocytoma of soft tissue: a preliminary report. Jpn J Clin Oncol., 1994; 24(3): 154-159.
5. Fukunaka H, Etoh T, Nakagawa H, Tamaki K; A case of subcutaneous malignant fibrous histiocytoma circumscribed by fibrous tissue. J Dermatol., 1996; 23(11): 836-839.
6. Enzinger FM, Weiss SW; Soft tissue tumors. 3rd edition, St. Louis, Mosby, 1995.
7. Nonaka M, Kadokura M, Ohkubo F, Kushihashi T, Kunimura T, Kataoka D *et al*.; Postradiation inflammatory malignant fibrous histiocytoma arising from the chest wall. Ann Thorac Cardiovasc Surg., 2001; 7(6): 371-374.
8. Fletcher CDM, van der Berg E, Molenaar WM; Pleomorphic malignant fibrous histocytoma /undifferentiated high grade pleomorphic sarcoma. In Fletcher CDM, Uni KK, Mertens F; World Health Organization Classification of Tumors: Pathology and Genetics of Tumors of Soft Tissue. IARC Press, Geneva, 2002: 120-122.
9. Kuhlman JE, Bouchardy L, Fishman EK, Zerhouni EA; CT and MR imaging evaluation of chest wall disorders. RadioGraphics, 1994; 14(3): 571-595.
10. Sen F, Akpinar AT, Alper E, Kanat O, Gürel S, Emıroğlu R *et al*.; Metastatic malignant fibrous histiocytoma to sigmoid colon presenting as a huge abdominopelvic mass on combined positron emission tomography/computed tomography. Turk J Gastroenterol., 2011; 22(1): 109-111.
11. Bielack SS, Schroeders A, Fuchs N, Bacci G, Bauer HC, Mapeli S *et al*.; Malignant fibrous histiocytoma of bone: a retrospective EMSOS study of 125 cases. European Musculo-Skeletal Oncology Society. Acta Orthop Scand., 1999; 70(4): 353-360.
12. Matsumoto S1, Ahmed AR, Kawaguchi N, Manabe J, Matsushita Y; Results of surgery for malignant fibrous histiocytomas of soft tissue. Int J Clin Oncol., 2003; 8(2): 104-109.
13. Belal A, Kandil A, Allam A, Khafaga Y, El-Husseiny G, El-Enbaby A *et al*.; Malignant fibrous histiocytoma: a retrospective study of 109 cases. Am J Clin Oncol., 2002; 25: 16-22.
14. Oda Y, Tamiya S, Oshiro Y, Hachitanda Y, Kinukawa N, Iwamoto Y *et al*.; Reassessment and clinicopathological prognostic factors of malignant fibrous histiocytoma of soft parts. Pathol Int., 2002; 52(9): 595-606.
15. Gibbs JF, Huang PP, Lee RJ, McGrath B, Brooks J, McKinley B *et al*.; Malignant fibrous histiocytoma: an institutional review. Cancer Invest., 2001; 19(1): 23-27.