



## Undifferentiated Pleomorphic Sarcoma of Chest Wall –A Rare Case Report

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### Abstract

*Undifferentiated pleomorphic sarcoma (Malignant fibrous histiocytoma) is a sarcoma that arise from bone or soft tissue. it is the most common type of sarcoma of extremity while MFH of bone is very rare. It occurs most frequently in age group of 45-55 yrs of age. The origin of tumor is not known, and one of the major predisposing factors is radiation. Other Causes include paget's disease, non-ossifying fibroma and fibrous dysplasia. Post radiation sarcoma accounts for 0.5% to 5.5%.Osteosarcoma, malignant fibrous histiocytoma and fibrosarcoma are the most common .Because adjuvant RT after breast conserving surgery plays a significant role in treatment of early – stage breast cancer, sarcoma of the breast, chest wall, sternum, axilla or supraclavicular region have been reported as a complication of treatment for breast cancer. We report here a rare case of undifferentiated pleomorphic sarcoma of chest wall in a 51yr old woman who had underwent 6 cycles of chemotherapy and radiation following modified radical mastectomy for carcinoma breast*

**Keywords:** *Undifferentiated pleomorphic sarcoma, Malignant fibrous histiocytoma (MFH), Radiation therapy (RT).*

### Introduction

Post-radiation sarcomas are rare complications of radiotherapy (RT) that occur within a previously irradiated field after several years of latency<sup>(1)</sup>. Post-radiation sarcoma accounts for 0.5% to 5.5% of all sarcomas<sup>(3)</sup>. Murray et al.<sup>(2)</sup> proposed revised criteria for post radiation sarcoma which included the following: 1)The radiation must have been given previously, and the sarcoma that subsequently developed must have arisen in the area included within the 5% isodose line; 2) No evidence that the sarcoma was likely to have been present before the

onset of irradiation; 3) All sarcomas must be proven histologically and must clearly be of a different pathology than that of the primary condition.

Osteosarcoma, malignant fibrous histiocytoma, and fibrosarcoma are the most common histological subtypes<sup>(2,3)</sup>. Because adjuvant RT after breast-conserving surgery plays a significant role in the treatment of early-stage breast cancer<sup>(4)</sup>, sarcomas of the breast, chest wall, sternum, axilla, or supraclavicular region have been reported as a rare complication of RT for breast cancer<sup>(5-7)</sup>. In total, 1,831 cases of radiation-induced sarcoma of the breast have been published in the English

literature<sup>(8)</sup>. However, only a few cases of post-radiation sarcoma after RT for breast cancer have been reported in Kerala. Here we report a case of post radiation sarcoma of chest wall in a 51 yr old woman who had underwent six cycles of chemotherapy and radiation for carcinoma breast.

### Case Report

A 51 yr old female presented with swelling left chest wall for 2 months which was rapidly increasing in size and painful. There was no history of trauma or any other illness. There was no pallor, icterus, cyanosis or oedema. Her pulse and BP were normal. Regarding the past history she was diagnosed with carcinoma breast of left breast in the year 2012 and under went 6 cycles of chemotherapy and radiation at government medical college Thrissur, Kerala.

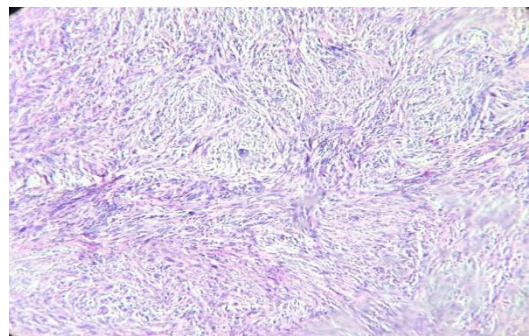
She developed these symptoms 6 yrs later and further investigations were done. Routine blood and biochemical parameters were within normal limits. FNAC done from swelling showed atypical cells suspicious of malignancy. CT thorax showed heterogeneously enhancing soft tissue density lesion posterior to sternal head of pectoralis major muscle with evidence of erosion of medial end of left clavicle, possibly chest wall recurrence. Wide local excision of chest wall tumor was done under GA and sent for HPE

Gross specimen: Received single grey white soft tissue with attached bone whole measuring 4.5\*4\*4.5 cm. Bone measured 3\*2.5\*1.5 cm, cut section showed grey white growth attached to bone measuring 4\*4\*4.5cm. Resected end of bone was free of neoplasm. Cut section of bone and marrow were free of neoplasm.

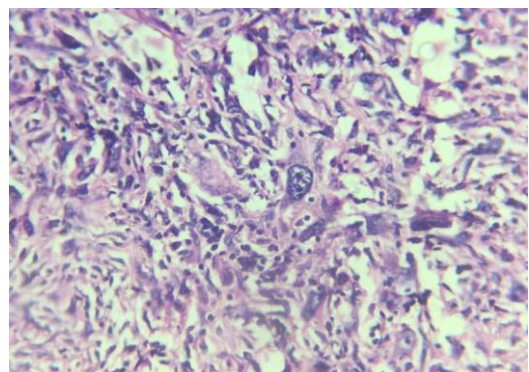
Histopathological examination revealed an infiltrating neoplasm composed of cells arranged in storiform pattern and short fascicles. Individual cells have scant to moderate amount of cytoplasm pleomorphic vesicular nuclei with prominent nucleoli. Mitosis seen 3-4/hpf. Atypical mitosis and bizarre cells seen. Stroma was desmoplastic and showed lymphoplasmacytic infiltrates. Underlying

bone, bone marrow and resected end of bone was free of neoplasm.

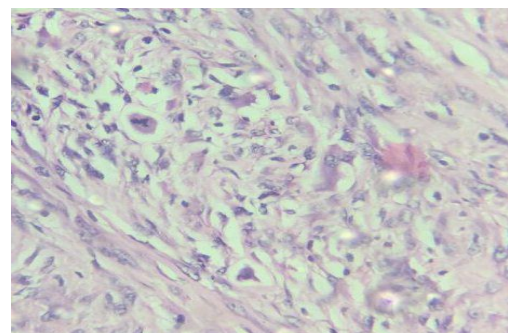
IHC was done in formalin fixed paraffin embedded tissue and 4 micrometer section were used for the procedure. Tumor cells were positive for vimentin and CD 68 with strong cytoplasmic and membrane positivity. Tumor cells were negative for SMA, CK, p63, HMB45 and S100. CK, p63 were done to rule out metaplastic carcinoma. S100 was done to rule out malignant peripheral nerve sheath tumor, SMA for leiomyosarcoma and HMB45 for melanoma.



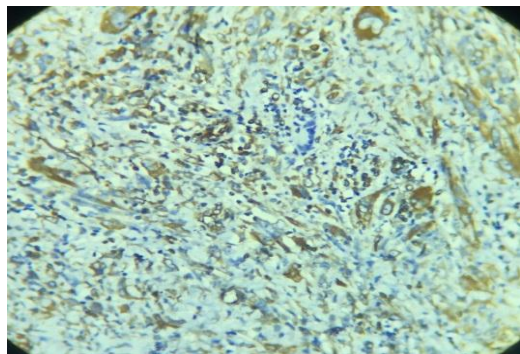
**Figure 1:** HPE image showing storiform pattern of tumor cells (10x)



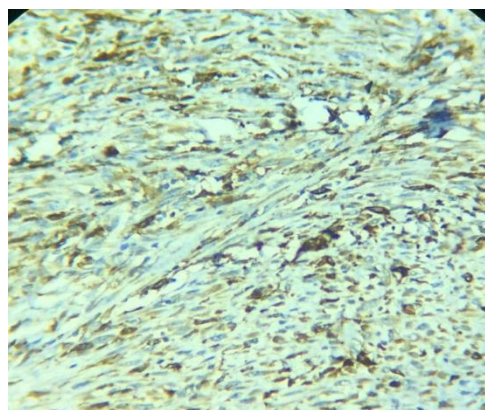
**Figure 2:** HPE image showing pleomorphic and bizarre cells (40x)



**Figure3:** HPE image showing atypical mitotic figures (40x)



**Figure 4:** IHC staining with vimentin showed strong diffuse cytoplasmic and membrane positivity



**Figure 5:** IHC staining with CD68 showed strong cytoplasmic staining

### Discussion

As survival after breast cancer treatment is typically long-term, the risk of secondary malignancies, particularly sarcomas, increases with duration of treatment. According to a large retrospective study of 16,705 patients treated for breast cancer, adjuvant RT significantly increases the rate of sarcomas and lung cancers when compared with a non-RT group<sup>(11)</sup>. Among them, 35 patients developed sarcomas similar to this case report, majority being angiosarcoma. MFH as such is very rare after radiation in case of carcinoma breast. The cumulative incidence of radiation-induced sarcoma were 0.27% and 0.48% at 10 and 15 years, respectively.

Angiosarcoma is the most prevalent histology among radiation-induced sarcomas world-wide<sup>(5,6)</sup>, though our case was not of this type. Higher radiation dose increases the risk for soft tissue and bone sarcomas after breast cancer therapy<sup>(12)</sup>. There are no distinguishable imaging features of post-

radiation sarcomas<sup>(3)</sup>. However, the presence of bony destruction with a soft tissue mass, tumor matrix mineralization at a previously irradiated area, and an appropriate latency period could be important clues for a diagnosis of post-radiation sarcoma. The prognosis of post-radiation sarcomas is generally very poor, with 5-year survival rates of 27% to 36% respectively<sup>(5-7)</sup>.

The standard treatment for radiation induced sarcoma is surgical resection, but this is often prohibited by tumour location<sup>(6,7)</sup>. Only complete surgical resection with tumour negative margins can guarantee long-term survival<sup>(14)</sup>. Chemotherapy or RT has only limited roles in treatment. Therefore, early detection and management is important to enable curative resection. Despite well-organized nationwide breast cancer databases<sup>[15]</sup>, there are no large-scale study conducted regarding post-radiation sarcomas after breast cancer treatment. Given the younger age at diagnosis and the increased use of RT as a primary treatment for breast cancer<sup>(15)</sup> longer survival after treatment and increased cumulative dose of radiation could lead to more cases of post-radiation sarcoma. Future studies should be done to investigate the incidence, distribution and clinical presentation of sarcomas after RT for breast cancer based on analysis of a large database.

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