

Case Report

Inflammatory myofibroblastic tumor of right leg and calcified fibroadenoma of left breast removed from a postmenopausal-women: a rare case report and review of literature

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ABSTRACT

Inflammatory myofibroblastic tumors are uncommon neoplasms; presentation of these tumors in the lower extremities is extremely rare. Treatment varies depending on type, location and patient factor. Here the case of 60-year-old female post-menopausal (P2L2) presented with swelling over anterior aspect of right leg which was exophytic in nature and left breast lump. Combined histopathology and immunohistochemistry findings favoured myofibroblastic tumor of borderline category- inflammatory myofibroblastic tumor. Mammography of left breast showed presence of fibroadenoma at 12 o' clock position. Patient underwent wide local excision of right leg swelling followed by split thickness skin grafting and excision of left breast swelling (lumpectomy). Inflammatory myofibroblastic tumors are of varied biologic potentials. They are generally classified as tumours of intermediate biological activity with uncertain malignant potential. There are many factors which affect treatment and prognosis of the patient. Five types of standard treatment for sarcoma are used: surgery, radiation therapy, chemotherapy, targeted therapy, immunotherapy. Fibroadenomas are benign breast neoplasms, typically present in women from ages of 20-35 years old (premenopausal).

Keywords: Spindle cell neoplasm, Inflammatory myofibroblastic tumor, Fibroadenoma, Mammography, Exophytic mass, Post menopausal, Immunohistochemistry

INTRODUCTION

The etiology of inflammatory myofibroblastic tumor is unknown. They may be associated with local recurrence, infiltrative growth, vascular invasion, and malignant sarcomatous transformation although they are generally benign. A soft tissue sarcoma is a presence of swelling in soft tissue of the body. Some inherited disorders can increase the risk of developing soft tissue sarcoma. Main diagnostic stay is biopsy. According to the WHO, inflammatory myofibroblastic tumors are classified as tumors of intermediate potential due to a tendency for local recurrence; however, they rarely metastasize.¹⁻⁴ They are composed of myofibroblastic and fibroblastic spindle cells accompanied by an inflammatory infiltrate

of plasma cells, lymphocytes, and/or histiocytes. They are classified as tumours of intermediate activity with uncertain malignant potential. They are typically benign. They are generally found in mucosal areas, mostly in lungs. It may be seen in extrapulmonary location out of which lower limb is very rare. Malignant transformation to soft tissue sarcoma may occur. Soft tissue sarcomas are a group of rare malignant tumors which when arise from the limbs if adequately managed have good 5-year survival rate.⁵ The mainstay of treatment for inflammatory myofibroblastic tumor is surgical resection with wide margins.¹ Radiotherapy, immunosuppression, and chemotherapy have not been proven to have any definitive benefit.

For successful treatment, early diagnosis is important.^{6,7} Diagnosis is delayed as most of them are painless, mobile, and have slow growth hence missed and biopsies are not done.⁸

Fibroadenoma of the breast is a well-circumscribed benign tumor with both epithelial and stromal growth. As it is a benign tumor, in the appropriate clinical and radiographic setting, once it is diagnosed by fine-needle aspiration or needle core biopsy, it may be left as it is. If it is causing any cosmetic problem, conservative excision can be performed.⁹

The fibroadenoma is a neoplasm of the lobular stroma with low cellularity, no atypia, and mitotic figures are absent. Histology includes myoepithelial cells and have an intracanalicular or pericanalicular pattern.¹⁰ The radiographic appearance shows an ovoid homogenous mass with sharp distinct borders.

CASE REPORT

A 60 year old postmenopausal female from Vadodara, Gujarat, India came to the hospital with complaints of right leg swelling since 5 years. There is no previous significant medical or surgical history present with no history of trauma and chronic illness. Systemic examination was normal. On examination, patient was conscious, cooperative and well oriented to time, place and person. All laboratory findings were under normal range.

The swelling was present on the anterior aspect of right leg which gradually increased in size. It was exophytic in nature. On examination, there is single well defined, nodular, non-tender, non-mobile, firm to hard approximately 5×4 cm size swelling with ulcer over the swelling, which was non-trans-illuminant and minimal serous discharge was present from the ulcer (Figure 1 and 2) All routine investigations were under normal range.



Figure 1: It shows the exophytic swelling present over right leg (5×4 cm size).



Figure 2: Lateral view of the swelling, with ulcer over the surface.

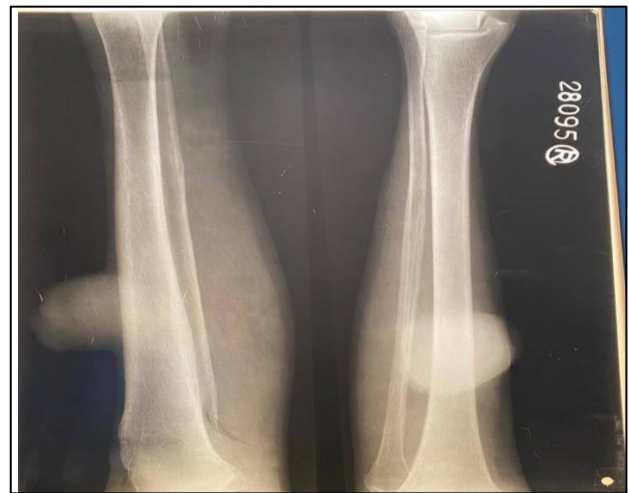


Figure 3: AP and lateral view. X-ray of right tibia and fibula showed presence of soft tissue swelling without bony involvement.

No palpable lymph nodes were present. Cytopathological report showed sarcomatous spindle cell neoplasm. Biopsy of the mass showed spindle shaped cells.

Immunohistochemistry examination revealed smooth muscle actin (1A4), muscle specific actin (Actin 88), factor 111 (EP3372) positive. BCL2 (EP36), CD34 (QBEnd/10), EMA (E29), S100 (S100B/1012), ALK (SP8), TLE-1 (ZM93) were all tested negative. Ki67 (MK167/2462) was 15-20%.

The sections show cellular spindle cell tumor with occasional areas of foamy macrophages, chronic inflammatory cell collection along with few giant cells. Cellular spindle cell areas show long fascicular arrangement of fibroelastic spindle cells. Areas of storiform type pattern was also appreciated. Occasional hypocellular myxoid area was also seen. Few areas showed haemorrhagic necrosis.

Combined histopathology and Immunohistochemistry findings favoured myofibroblastic tumor of borderline category-inflammatory myofibroblastic tumor.

Wide local excision was performed and split thickness skin grafting was done. Figure 4 shows intra-op wide split skin grafting done after wide local excision. Figure 5 shows the post-op specimen of soft tissue sarcomatous swelling.

Figure 6 shows post-op day 10.



Figure 4: Intra-op wide split skin grafting done after wide local excision.



Figure 5: Post-op specimen of the swelling.

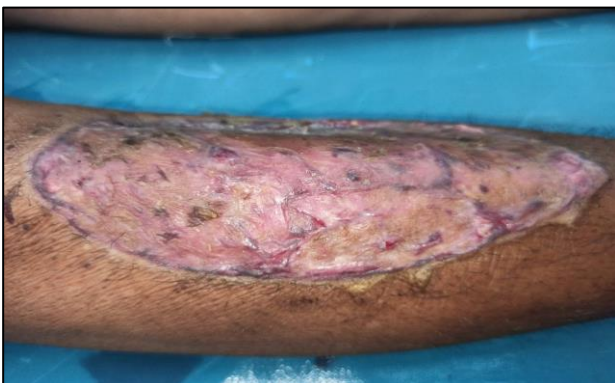


Figure 6: Post operative day 10.

Mammography of the both breasts was done. Large, well defined, irregularly margined as well as densely radio-opaque lesion was noted in upper inner quadrant of left breast. It measured approximately 75×74×71 mm (Figure 7).

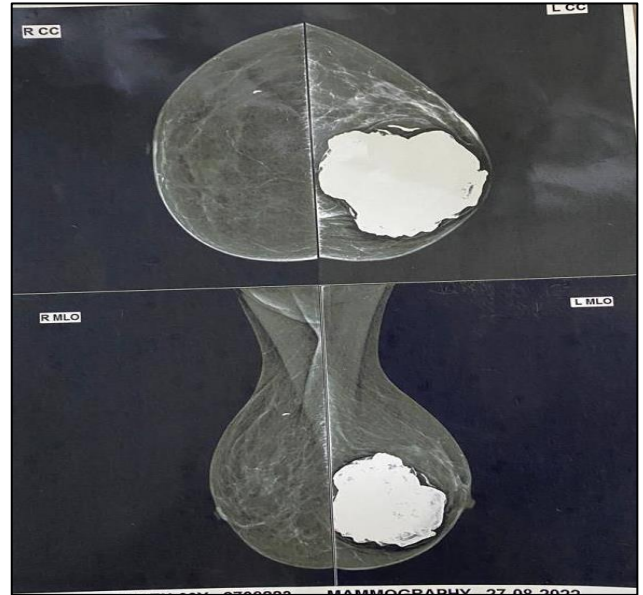


Figure 7: Mammography of left breast showed presence of calcified lump in left inner quadrant.

On USG correlation, dense calcification with strong posterior acoustic shadowing was noted. The lesion was freely mobile in upper inner quadrant of left breast causing mass effect over adjacent parenchyma suggestive of small fibroadenoma at 12 o'clock position in left breast (BIRADS-IVb). There was no evidence of abnormal ductal dilatation, nipple retraction, skin thickening. Nipple and areolar regions appeared normal. Both axillary regions were normal.



Figure 8: External appearance of the left breast swelling.

Wide local excision of breast lump was done. Figure 9 shows the large opacity left side of breast in AP view of chest X-ray.



Figure 9: Chest X-ray revealed presence of calcified left breast lump.



Figure 10: Shows excised fibroadenoma.

DISCUSSION

The pathogenesis of inflammatory myofibroblastic tumor is unclear, although there are various allergic, immunologic and infectious mechanisms postulated. The lung is the most common site of involvement but it has also been reported in other organs including the stomach, mesentery and retroperitoneum as well as the kidneys, renal pelvis, liver, spleen and lymph nodes.¹⁶ Pelvis, head and neck, trunk, and extremities are considered rare locations for the development of these neoplastic conditions. The mainstay of treatment for this tumor is surgical excision with wide margins.¹ Radiotherapy, immunotherapy and chemotherapy have not shown any definitive benefit. If transformed to sarcoma (malignancy) then they are managed with wide local excision and radiotherapy but a few that are high-grade, recurrent or involving major neurovascular structures require amputation. Amputation is a palliation procedure for patients with distressing symptoms like pain,

bleeding, fungating mass. Early diagnosis is important for successful procedure. They are best managed by surgical resection. Chemotherapy is mainly reserved for patients who present with metastasis, tissue invasion or bony involvement.^{11,12} Limb saving procedures are standard of treatment for most tissue sarcomas.

Fibroadenoma is a benign breast disease with abnormal growth of the glandular and fibrous tissue. Hormonal theories exist which implicates excess estrogen stimulation, increased estrogen receptors, or decreased estrogen antagonist activity in the breast.¹³ The typical sonographic appearance is a well-circumscribed, round or oval mobile, nontender mass with relatively uniform hypoechogenicity.¹⁴ Rarely, it may reveal heterogeneity, due to necrosis or dystrophic calcification, a finding which is common in older women. Surgical excision using a circumareolar incision is preferred in patients older than 35 years and those with large tumors. However, young women could be managed conservatively.

CONCLUSION

This case of 60-year-old postmenopausal women showed presence of exophytic swelling of right leg which was non metastatic in nature, histopathology reported only few areas of necrosis or X-ray revealed no bony involvement, so wide local excision was performed with split skin grafting. Left breast swelling depicted presence of fibroadenoma and axillary lymph nodes depicted no abnormality, excision was performed.

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