International Surgery Journal
Hassanesfahani M et al. Int Surg J. 2025 Oct;12(10):1784-1786
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20253030

Case Report

A misleading presentation: skin sarcoma presenting as a breast mass

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Received: 22 July 2025
Revised: 19 August 2025
Accepted: 02 September 2025

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ABSTRACT

Atypical fibroxanthoma (AFX) is a rare cutaneous malignant neoplasm which manifests as solitary lesions in sun-exposed areas of elderly patients, commonly on the head and neck. Given AFX's rarity and lack of specific immunohistological markers, there are no established standardized guidelines for the management of AFX, often leading to a misdiagnosis of more aggressive variants such as pleomorphic dermal sarcoma (PDS). The prognosis is generally favorable, with most cases responding well to complete surgical excision alone. We present a rare case of AFX in the breast, a 68-year-old woman with a left breast mass, which the patient described as a small pimple. A computed tomography (CT) scan revealed a superficially located lesion in the upper medial quadrant of the left breast, measuring 4.2×2.2×2.1 cm (Figure 1A, B). At the patient's request, she proceeded with upfront wide local excision without prior biopsy or any breast imaging. Histopathological examination revealed a well-circumscribed tumor measuring 3.8×3.3 cm, with no connection to the overlying epidermis (Figure 1C). Given that the surgical margin was less than 1 cm, the patient underwent a re-excision with sentinel lymph node biopsy. Final pathology was negative for positive margins and any lymph node involvement. At one-year follow-up, there was no clinical or radiological evidence of recurrence or metastasis. This case highlights the importance of including cutaneous lesions in the differential diagnosis of breast masses located near the skin surface. Accurate diagnosis requires a high index of suspicion and meticulous histopathological evaluation.

Keywords: Atypical fibroxanthoma, Cutaneous sarcoma, Pleomorphic dermal sarcoma, Breast mass

INTRODUCTION

AFX is a very rare, less aggressive, superficial variant of undifferentiated pleomorphic sarcoma. First documented in the 1960s, this skin tumor generally emerges as a solitary lesion that develops over sun exposed areas. Approximately 80% of AFX cases are localized to the head and neck, with the scalp being the most common site. AFX primarily presents among elderly patients, particularly those in their 70s and 80s. Men are notably more affected, with an incidence rate of two-thirds to three-fourths of cases.[8] Tumors typically present as firm, painless nodules, with a skin-colored or reddish-brown appearance and often ulcerated surfaces.[10] The diagnosis of AFX can be difficult due to the absence of specific immunohistological markers. It is important to differentiate it from a more aggressive variant, pleomorphic dermal sarcoma (PDS). Previous genetic research showed shared characteristics between the two, including the presence of 9p and 13q deletions in both the tumors, suggesting a possible common origin.

However, the absence of K-RAS and H-RAS mutations in atypical fibroxanthoma, as opposed to their presence in undifferentiated pleomorphic sarcoma, helps in differentiating the two conditions.[9] AFX is generally located within the dermis and superficial subcutaneous tissue and does not exhibit perineural or angiolymphatic invasion while PDS is known for its infiltrative nature, affecting deeper layers of subcutaneous fat and potentially demonstrating perineural or vascular infiltration.[3,4] Authors present a case of atypical

fibroxanthoma manifesting in an uncommon location as a breast mass. This case highlights the diagnostic complexities of AFX, emphasizing the need for meticulous histopathological evaluation to ensure accurate diagnosis and effective management.

CASE REPORT

A 68-year-old female with a history of fibromyalgia presented with a left breast mass initially described as a small pimple. Breast sonogram and mammogram were refused by the patient due to pain. A CT scan revealed a breast lesion close to the skin in the upper medial left breast, measuring 4.2×2.2×2.1 cm, with surrounding fat stranding and thickening of the overlying skin (Figure 1 A, B). Per patient's request, she underwent upfront wide local excision without biopsy. The pathological report identified a 3.8×3.3 cm well-circumscribed nodular tumor without connection to the overlying epidermis, with surrounding fat stranding and thickening of the overlying skin (Figure 1 C).

The lesion is composed of a poorly differentiated pleomorphic neoplasm characterized by large, highly bizarre, and atypical spindled to epithelioid cells, with marked cellular pleomorphism, abundant eosinophilic cytoplasm, and mitotic Figures (Figure 1 D, E). Immune stain was positive for CD68 (Figure 1 F). Given one of the margins was less than 1 cm, she underwent re-excision and sentinel lymph node biopsy. The final pathology report showed negative margins and no nodal disease. On her one-year follow-up, she has no evidence of recurrence or metastasis on examination and imaging.

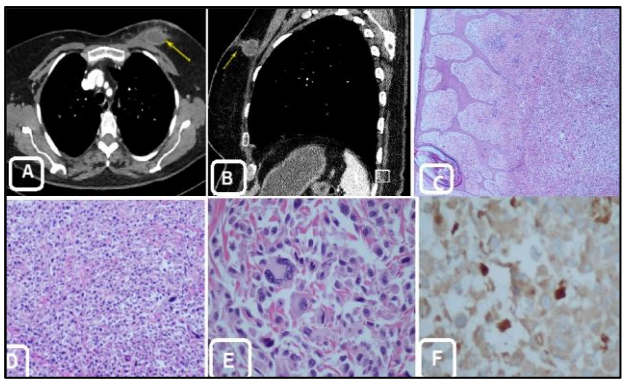

Figure 1: (A-F) Imaging and histopathological findings of atypical fibroxanthoma presenting as a breast mass.

DISCUSSION

AFX is an exceptionally rare cutaneous malignancy that poses significant diagnostic challenges, particularly when it arises in atypical locations such as the breast. As illustrated in this case, AFX, while classically associated with sun-exposed areas like the head and neck, can present as a solitary lesion in less common sites, such as the breast. The rarity of AFX necessitates a heightened level of vigilance when evaluating any skin mass, especially in elderly patients with a history of significant sun exposure. The diagnostic difficulty of AFX is compounded by its nonspecific clinical presentation and the absence of distinctive immunohistological markers. The challenge is further accentuated by the need to differentiate AFX from more aggressive sarcomas, such as PDS. PDS, with its infiltrative nature and potential for perineural or vascular invasion, can present similarly to AFX, but with notable differences in histological features and depth of invasion.[4,5]

The prognosis for AFX is generally favorable, with the majority of cases responding well to treatment. However, there are rare instances where the disease can metastasize. Mohs micrographic surgery is associated with a higher cure rate compared to wide local excision, due to its precision in removing cancerous tissue while preserving healthy skin.[6] Given the association of AFX with significant ultraviolet radiation exposure, individuals diagnosed with this condition should undergo regular follow-up examinations, ideally every six months. These evaluations are crucial for monitoring potential recurrence, detecting metastasis, and identifying any new skin cancers that may develop.[7]

The case highlights the importance of considering AFX in the differential diagnosis of skin lesions, particularly when they exhibit atypical features or occur in unusual locations. This case underscores the need to maintain a broad differential when evaluating skin masses, especially those presenting with atypical characteristics. The rarity of AFX requires a high index of suspicion and thorough histopathological examination to avoid misdiagnosis and ensure appropriate management. This vigilance is vital for achieving accurate diagnoses and optimizing patient outcomes, particularly in cases where the clinical presentation diverges from the more common patterns of malignancy.

CONCLUSION

In conclusion, AFX is a rare cutaneous malignancy that presents distinct diagnostic challenges, particularly when it occurs in atypical locations such as the breast. Its association with significant sun exposure and its potential for misidentification as more aggressive neoplasms underscore the importance of careful histopathological evaluation. While the overall prognosis for AFX is generally favorable, with a high cure rate achievable through Mohs micrographic surgery, ongoing vigilance is essential. Regular follow-up every six months is recommended to monitor for recurrence, metastasis, and the emergence of additional skin cancers. This case highlights the need for a comprehensive approach in the diagnosis and management of AFX to ensure optimal patient outcomes and effective long-term care.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Hassanesfahani M, Alla M, Bucci L, Khan N. A misleading presentation: skin sarcoma presenting as a breast mass. Int Surg J 2025;12:1784-6.