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Case Report

Dermatofibrosarcoma Protuberans Presenting as a Breast Mass: a Case Report

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Abstract

Dermatofibrosarcoma protuberans is a rare tumor that accounts for a negligible percentage (approximately 0.1%) of all malignancies. It usually occurs on the trunk and upper and lower limbs. Its occurrence in the breast is uncommon.

A 46-year-old Nigerian woman presented with a left breast mass of 3 years duration without clinical and radiological evidence of metastasis. Initial histological diagnosis revealed a malignant phyllodes tumour. However, immunohistochemistry showed a diagnosis of dermatofibrosarcoma protuberans. She subsequently had a wide local excision with axillary clearance and has shown no clinical and/or radiological signs of recurrence of the lesion one year following surgical removal.

This case helps reiterate the need for immunohistochemistry and not only histology of all malignant breast tumours, irrespective of the nature of the presentation, mode of occurrence, and initial diagnosis. Dermatofibrosarcoma protuberans, however rare, should be considered an important diagnosis of breast malignancies. Indeed, considering the high rate of misdiagnosis of dermatofibrosarcoma protuberans, early histopathological evaluation of breast lesions is of utmost importance. Our case also demonstrates that a wide local excision that is readily available in resource-poor settings remains an extremely important option in the absence of the preferable and technical Mohs micrographic surgery (MMS).

Keywords: Dermatofibrosarcoma Protuberans; Malignant Breast Lesion; Malignant Phylloides, Breast Cancer, Nigerian; Case Report.

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Introduction

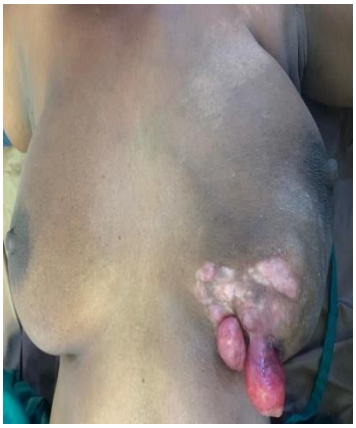
Dermatofibrosarcoma Protuberans (DFSP) is a rare, slow-growing soft-tissue tumor that accounts for 0.1% of all malignancies and below 1% of the entire soft-tissue sarcomas. [1] DFSP, which was first described by Hoffman in the year 1925 [1], is clinically a low to intermediate-grade sarcoma with an indolent course. It presents'as a small, firm, painless, raised, skin-colored plaque with the potential to invade underlying structures like fascia, muscle, periosteum, and bone, although involvement of the breast is rare. Metastasis to distant organs with a predilection for lung, brain, bone, visceral organs, lymph nodes, and soft tissues has been reported. [1, 2] Microscopically, DFSP presents as uniform spindle cell fascicles growing in a cartwheel (storiform) pattern with multiple variants and with strong and diffuse CD34 immunoreactivity. [1] DFSP is common in women than in men and people of African descent than in whites. Although it can occur from infancy to old age, it is most seen in young adults between the ages of 25 and 45 years, with a mean age of 40-43 years. [1].

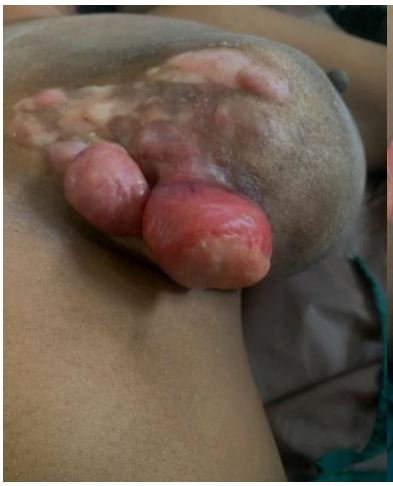
The gold standard treatment for DFSP is surgical resection irrespective of its stage. Usually, a cure is achieved whenever the tumor margins are free. In cases where surgical resection is not feasible due to advanced disease, metastasis, or functional and cosmetic reasons, radiotherapy and targeted therapy (with imatinib mesylate) are used. [1] Generally, the prognosis of DFSP is good, the five- and ten-year recurrence-free survival rates of DFSP are 86% and 76% respectively. [3] However, factors like increased age, high mitotic index, and tumor-positive margins confer a poorer prognosis. [1] Local recurrence ranges from 20-50% [3], and the mean time of recurrence from excision to recurrence is 32-38 months. [1] Because of its propensity to recur, mandatory follow-ups of 6-12 months are advised with emphasis on thorough history and examination of the primary site and draining lymph nodes; suspicious lesions would warrant imaging and biopsies to diagnose recurrence and metastasis. [1]

We hereby report the case of a 46-year-old Nigerian woman who presented with a left ulcerative and pedunculated breast mass of 3 years duration which started as a painless skin lesion.

Case Profile

A 46-year-old Nigerian woman presented with a left breast lump of 3 years duration to our general surgery clinic. It started as a painless, raised, hypopigmented skin lesion that gradually increased in size and involved the breast; subsequently, it became painful and ulcerated 2 months prior to presentation. There was no history of nipple discharge. She attained menarche at age 18 and still menstruating. She is married in a monogamous family setting, has 4 children, and breastfed all. The patient does not smoke or take alcohol. She had no history of previous breast disease or family history of breast or other malignancies. They were said to have procured several antibiotics and analgesics of unknown names from multiple patent drug dealers with no clinical improvement prior to her presentation. On examination, an ulcerative, pedunculated breast mass was seen in the lower inner quadrant of the left breast, with no skin or nipple changes (Figure 1-3).





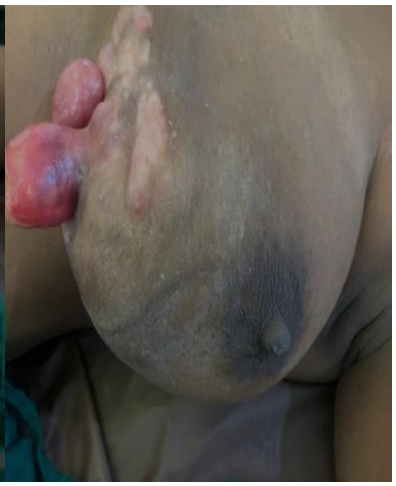


Figure 1. **Figure 2.** **Figure 3.**

Figures 1-3: Show a pedunculated breast mass in the left inner quadrant of the left breast with ulceration

There was no clinical or radiological evidence of distant metastasis. She had a tru-cut biopsy that histologically revealed a hypercellular lesion composed of moderately pleomorphic spindle-shaped cells arranged in small fascicles (Figures 4 & 5) based on which an initial diagnosis of a malignant phyllodes tumor was made.

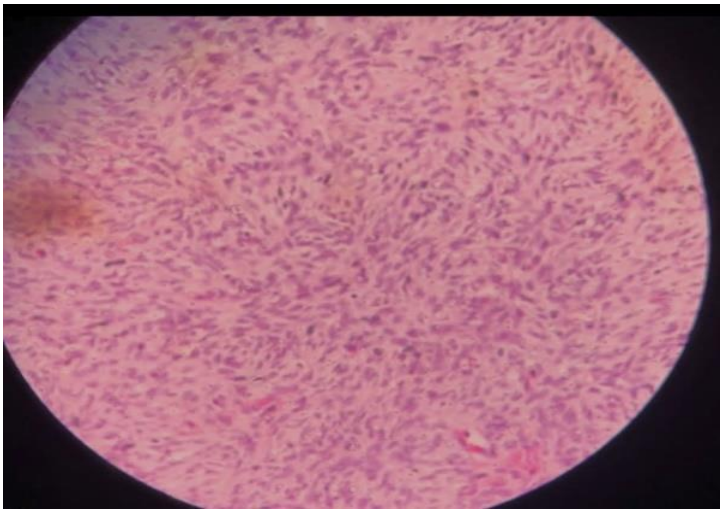


Figure 4: (H & E X 100) shows pleomorphic spindle cells with an abnormal mitotic figure (arrow).

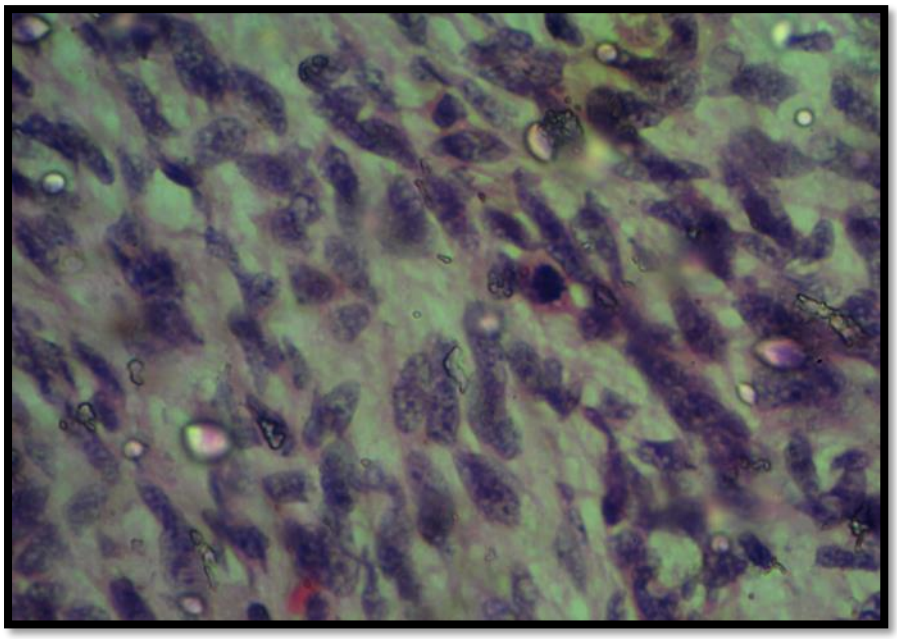


Figure 5: (H &E X 40); Shows spindle-shaped cells arranged in a storiform pattern

Histology of a subsequent excisional biopsy revealed a spindle cell neoplasm in a storiform arrangement, infiltrating the surrounding fat. The cells are mostly bland with focal atypia and a few abnormal mitotic figures with tumor-free margins and axillary nodes (Figures 6& 7).

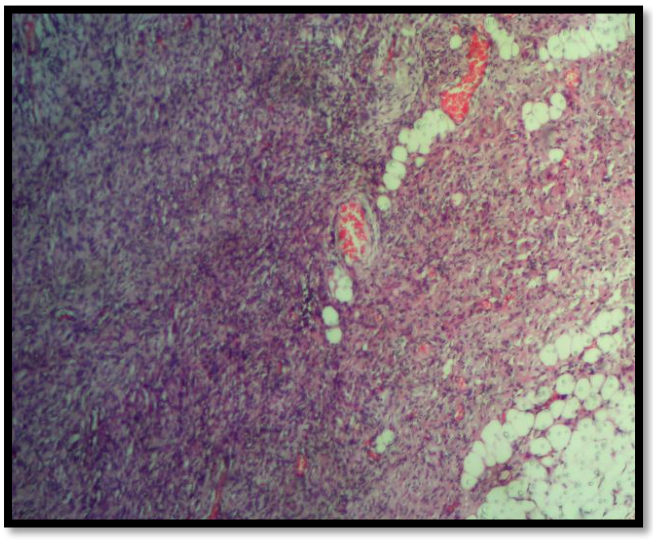


Figure 6 (H & E X 40): Shows fat cells entrapment by lesional cells

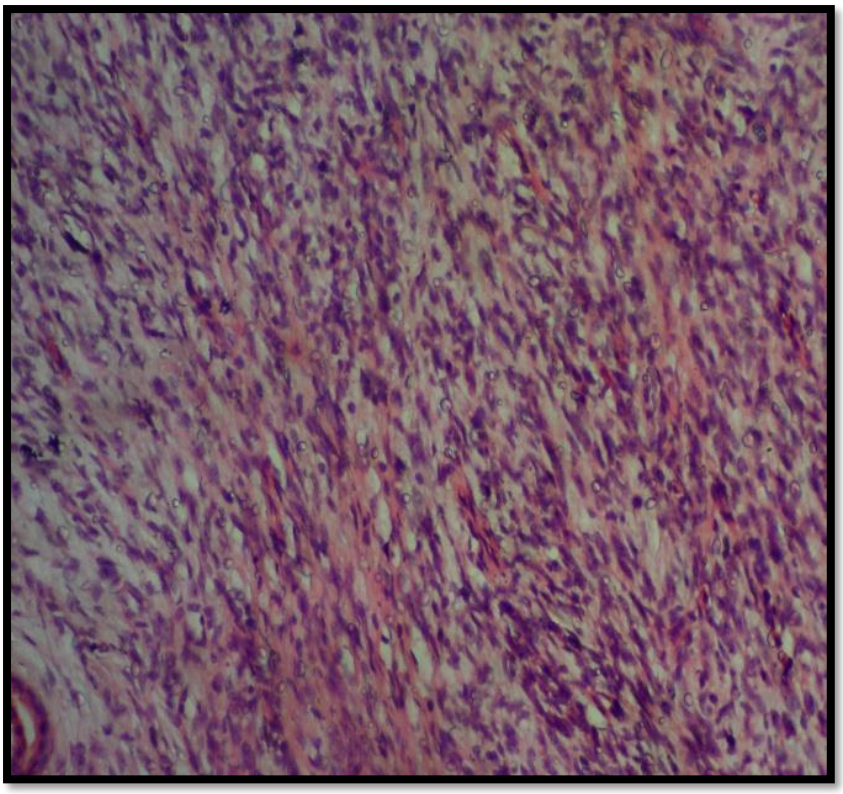


Figure 7 (H & E X 40): Shows tumour cells disposed in storiform pattern

Immunohistochemistry showed lesional cells that were diffusely and strongly positive for vimentin and CD34 but negative for pancytokeratin AE1/3, ER, and P63. Ki67 showed mitotic figures, which conformed to a final assessment of dermatofibrosarcoma protuberans presenting as a breast mass. Furthermore, we ruled out radiological evaluation as there were no signs of metastasis. Our patient subsequently had a wide local excision of the tumor with axillary clearance. Imatinib treatment and radiotherapy were not considered in our patient because there was no evidence of metastasis.

One-year post-surgery, our patient has no clinical sign of recurrence and/or metastasis. Also, thrice (done 1-, 4- and 6 months apart) breast CT scans and chest X-rays showed no radiological evidence of recurrence and/or metastasis. The surgical wound has healed well (Figure 7).

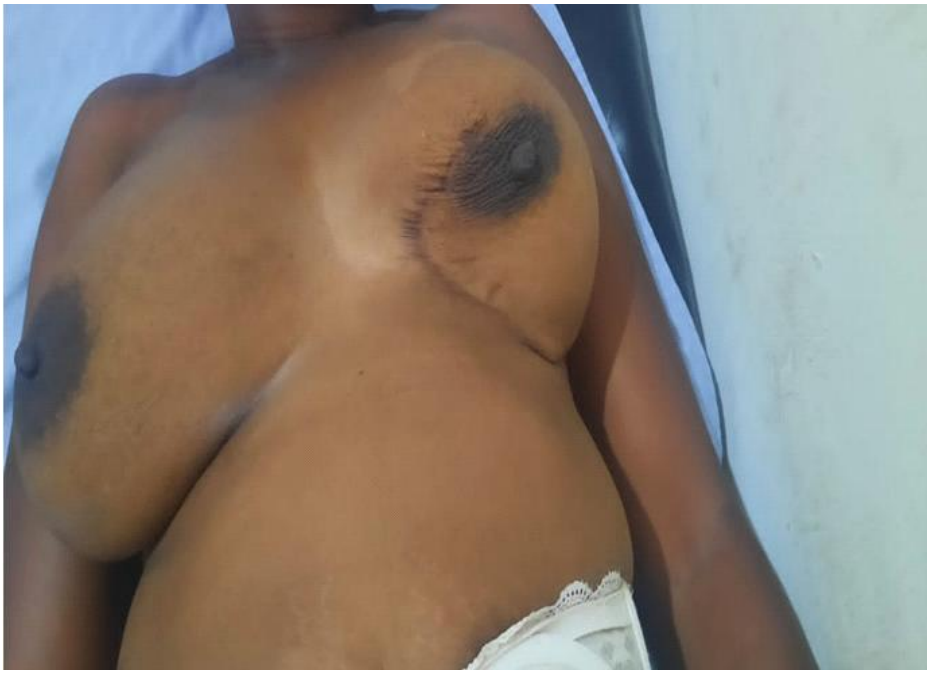


Figure 8: Shows both mammary glands with a well-healed surgical scar on the lower inner quadrants of the left breast; 6 months post-surgery.

Discussion

DFSP is regarded as a sarcoma with low-grade, malignant, aggressive biologic behavior. It has a low metastatic potential and a tendency for local recurrence post-treatment. [2, 4] While DFSP typically presents as an asymptomatic, slow-growing lesion, the clinical presentation may differ. It may initially present as a hard or firm indurated plaque, scar, or protruding mass [4, 5], looking like cysts, scars, keloids, or boils, etc. Subsequently, the lesion may develop multiple nodules, justifying the addition of the word "protuberans" to its original name of dermatofibrosarcoma. [4]

DFSP is seen more commonly in women and people of African descent, even as it mostly presents in the adult population within the 2nd to 5th decades of life. [1, 6] Our patient was a 46-year-old African woman. The majority of hospital patients present with painless multinodular bluish or brownish erythematous protruding masses, with few being ulcerated and hemorrhagic, developing over years [7], just as were seen in our patient. The tumour is frequently misdiagnosed, especially in its initial phases when it can mimic a keloid or dermatofibroma. [2]

Though DFSP most commonly occurs in the trunk, and the upper and lower extremities [8], it has been reported to seldomly occur in the head and neck region and specifically even more rarely in the facial region. [6] Localization in the breast on the other hand is very rare. [8] Our patient developed DESP in her left breast.The neoplasm demonstrates local infiltrative growth but seldom metastasizes distally [2]. Its natural history is that of low-grade malignancy, with a 26–60% local recurrence rate attributed to incomplete excision due to poor circumscription and irregular boundaries. [2] The lesion typically infiltrates well beyond its grossly visible margin into the surrounding tissue. In general, the tumor is in the dermis, but it can sometimes present as an infiltrative growth in the subcutaneous fatty tissue, forming a pastry pattern (60% of cases; neoplastic cell bands parallel to the epidermis) or a honeycomb pattern (delimitation of adipocyte islets between the tumoral tissue). [9] Its low potential for metastasis is typical,

with less than 5% probability for regional or distant metastases, with these often being restricted to the lungs and less frequently to the lymph nodes. [2] In our case, there was no evidence of metastasis.

Imaging findings are non-specific but may be considered if metastasis is suggested clinically. Mammography is non-specific and features often mimic a benign breast lesion. DFSP appears as a well-defined oval lobulated mass with mainly hypoechoic areas and some hyperechoic areas having areas of high cellularity and tumor cells respectively with acoustic enhancement. The periphery is often highly vascular as seen in the color Doppler. CT scan appears well-defined, nodular and isodense. DFSP appears homogenous on MRI with the same intensity as skeletal muscle. [10]

Histopathology reveals relatively uniform densely grouped fusiform cells, with elongated nuclei in characteristic storiform arrangement. [1, 9] Although not very common, cellular atypia can occur in some cases. [2] The tumor is highly cellular with few mitotic figures. [2, 6] There have been reports of fibrosarcomatous transformation of DFSP associated with a more aggressive tumor. [2, 6] In line with the literature, the histology of the lesion from our patient showed hypercellularity, focal cellular atypia, moderate pleomorphism, few mitotic figures, and spindle cells in a storiform arrangement.

Microscopically, DFPS extends far beyond the assessed clinical margins, spreading in the dermis and subcutaneous tissue. [2] In our case, the margins were tumor-free. Although it has a distinctive histological appearance, DFSP can mimic other diseases. Several variants of DFSP have been described and they must be well characterized to avoid misdiagnosis with other types of tumors. These variants include pigmented, myxoid, myoid, granular cell, sclerotic, atrophic DFSP, and giant cell fibroblastoma. [4] Consequently, it is necessary to distinguish the tumor from close histological differentials. The high incidence of misdiagnosis of DFSP highlights the importance of pathological examination of early skin lesions to clarify the diagnosis. Histology with immunohistochemistry remains the gold standard for ensuring an accurate diagnosis. [2] Immunohistochemical findings are positive for CD34 and vimentin. [5] Additional immunohistochemical staining of factor XIIIa, stromelysin III, CD44, CD163 and D2–40 have been found to be positive in dermatofibroma and negative in DFSP. [4, 5] Spindle tumor cells are immunonegative for S100 protein, smooth muscle actin, desmin, cytokeratin, and epithelial membrane antigen. [5] In line with the literature, immunohistochemical evaluation of the tumor cells from our patient was CD34 and Vimentin positive but negative for pancytokeratin AE 1/3.

The mainstay of treatment for DFSP is surgical excision. Mohs micrographic surgery (MMS), a tissue-conservative procedure that guarantees total histopathologic margin control during surgery is a preferred surgical technique due to its lower rate of post-treatment recurrence. [2] However, in selected cases or in the absence of MMS, wide local excision with tumor-free margins of 2-4cm due to its infiltrative nature can be used. [2] Furthermore, in cases of positive margins, radiotherapy is a good alternative to have more positive outcomes, and in cases of unresectable, metastatic, and recurrent tumors, imatinib (a tyrosine kinase inhibitor) is preferred because it inhibits PDGF and thus decreases DFSP growth. DFSP is highly recurrent with a rate of 10-50% occurring within the first 3 years. [2, 3] The 10-year survival rate is 99%. [2] Generally, DFSP occurring in the head and neck have high rates of recurrence. Moh's surgery has been shown to have a reduced recurrence rate. Long-term monitoring is often required due to the high rate of local recurrence. [2] Our patient had local excision with a wide margin of resection. In addition, there are no signs of recurrence one-year post-surgery

Conclusion

Overall, regardless of the type of presentation, mechanism of occurrence, or initial diagnosis, all malignant breast tumours should undergo immunohistochemistry in addition to histology. Furthermore, dermatofibrosarcoma protuberans should be recognized as a significant breast tumour, despite its rarity. Also, early histological assessment of breast lesions is crucial given the high likelihood of misdiagnosis

of dermatofibrosarcoma protuberans. Indeed, our case further demonstrates that a wide local excision remains an excellent option in the absence of the preferable and technical Mohs micrographic surgery (MMS).

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