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CASE REPORT

Dermatofibrosarcoma Protuberance of the Breast: a Diagnostic Challenge

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Abstract Dermatofibrosarcoma protuberans (DFSP) is an uncommon slow growing neoplasm of the dermis with tendency to invade the subcutaneous tissues. It presents during the third to fourth decade of life and is commonly seen over the trunk, extremities and head and neck. DFSP presenting as a breast lump is rare but few cases have been reported in the literature. Pre-operative diagnosis with mammography, ultrasonography and FNAC is challenging. We report a case of a DFSP of the right breast in a middle aged lady with history of recurrent breast lumps excised and diagnosed in the past as benign. She presented with progressively increasing right breast lump of 2 months duration. She underwent wide local excision and histology revealed dermatofibrosarcoma protuberans. In view of its local aggressiveness with incomplete surgical margin, mastectomy was performed.

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Keywords Dermatofibrosarcoma protuberans · Breast · Immunohistochemical stains · Mastectomy

Introduction

Dermatofibrosarcoma protuberans (DFSP) of the breast is a rare neoplasm that can remain indolent and stable for years; however, it can be accompanied by rapid growth and fibrosarcomatous changes [1]. It is known to be locally aggressive, though metastases are rare and when occur affect the lungs and the lymph nodes [2] .It usually presents in the middle age between the fourth to sixth decade but can occur at all ages [2–4]. Its presentation varies from an asymptomatic skin-coloured plaque, to a nodular lesion which may sometime ulcerate [2]. Differentiating pre-operatively from other benign and malignant lesions of the breast by routine investigation may not be possible. Computerised tomography (CT), magnetic resonance imaging (MRI) and magnetic resonance spectroscopy (MRS) have also been reported as an investigation that would aid in the diagnosis of DFSP [4, 5]. Histological examination and immunochemical stains are required to confirm the diagnosis.

Case Report

A 47-year-old lady presented with history of progressively growing painless right breast lump of 2 months duration. She had past history of multiple excisions for breast lump in both her breasts almost 20 years back and were reported

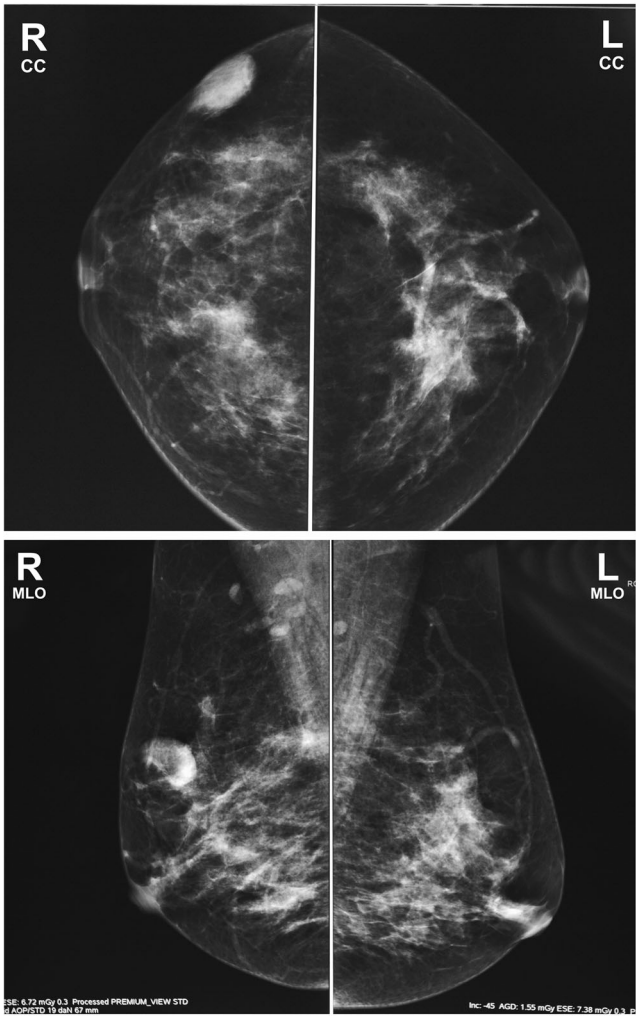

Fig. 1 Mammogram showing BIRADS 3 lesion

to be benign. She does not have any significant past medical history; however, her mother had breast cancer. On examination of the right breast, an oval-shaped, well-circumscribed, 2 × 2 cm, mobile, firm lump was noted at the 10 o'clock position very close to the surgical scar and fixed to the skin. No axillary lymph nodes were palpable. A clinical diagnosis of fibroadenoma was made. Mammogram reported a fairly well-defined lesion at the right upper outer quadrant, with no calcification (Fig 1). Breast ultrasonography revealed an ill-defined heterogeneous lesion pre-dominantly hyper echoic noted at the 10 o'clock position measured 2.1 × 2.0 × 1.3 cm with increased vascular flow on colour Doppler (Fig 2). FNAC showed hypo cellular and mixed stroma may be indicative of a phylloides tumour. She underwent wide local excision of the right breast lump. Histopathology reported as tumour cells in the skin adnexa infiltrating the subcutaneous fat (Fig 3). It was composed of bundle of uniform, mildly pleomorphic spindle cells with plump nuclei arranged in storiform, whorly and herring bone pattern (Fig 4). The mitotic figures were 10–12/10 hpf. The tumour cells showed strong immune reactivity to CD34. The final histological diagnosis was dermatofibrosarcoma protuberans (DFSP) of the breast with involvement of the resected margins. The necessity of further surgical treatment was reviewed by the tumour board and thoroughly discussed with the patient. She underwent mastectomy with axillary clearance and post-operative course was uneventful. One year follow-up of the patient has shown no local recurrence or evidence of metastasis.

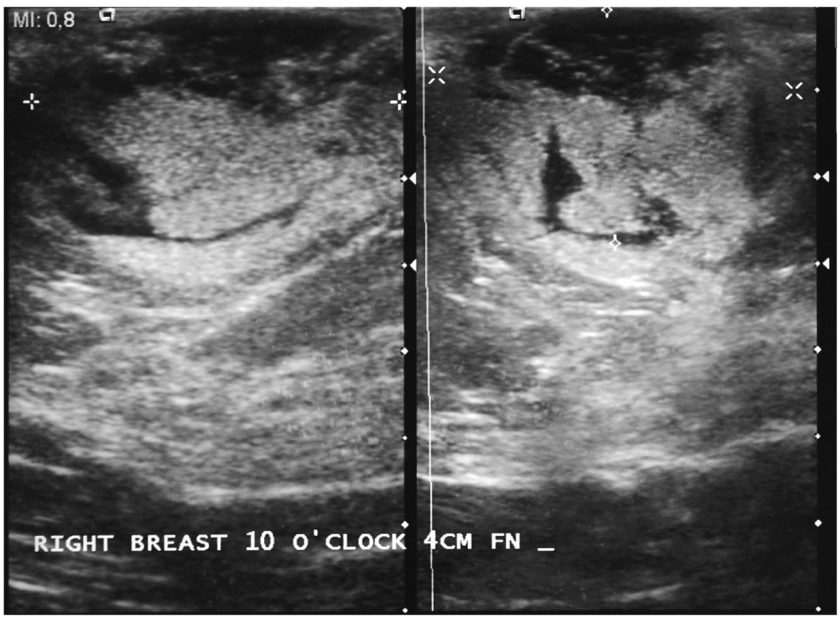

Fig. 2 Ultrasonography of the right breast showing lesion at 10 o'clock

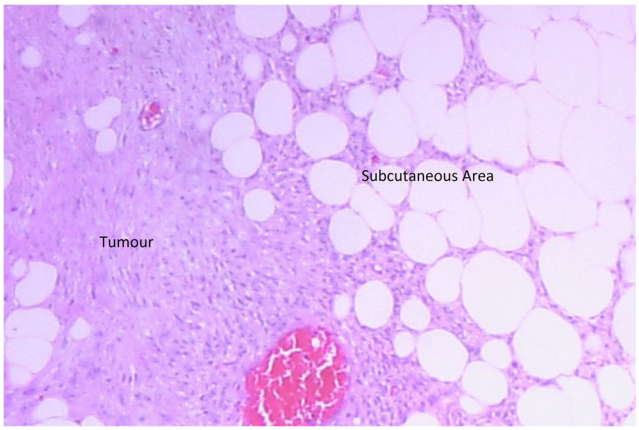

Fig. 3 H and E ×10: part of tumour with infiltrative margin into subcutaneous tissue

Discussion

Being a rare dermal tumour with varied presentation, it can cause a diagnostic dilemma and needs differentiation from other lesion-like fibrous histiocytoma, phylloides tumour and other malignant breast tumours. On sonography, if an oval mass in the subcutaneous tissue that is impinging against the skin and has a focal lobulated margin with hypo echogenicity or an irregular margin with mixed echogenicity, a diagnosis of DFSP should be considered [6]. Mammography does not show micro calcification in most of the reported case.

Histologically, DFSP is characterised by proliferation of the dermal spindle cells arranged in a storiform or cartwheel pattern. These proliferation are made up of monotonous cells with little pleomorphism and low mitotic index [2]. Immunohistochemical stains are required to differentiate dermatofibroma (DF), DFSP and other fibrous tumours. CD34, factor XIIIa, stromelysin-3 (ST3) and apolipoprotein D (Apo D) are some of the important stains used [7]. CD34 positivity is seen in most cases of DFSP. Chromosomal abnormality mainly chromosome 17 and 22 have also been implicated to play a role in DFSP [8].

The best treatment option is wide local excision or Mohs micrographic surgery. Modified radical mastectomy is also an option in select group of patients who are likely to default follow-up. Local recurrence is high and metastasis is rare with DFSP. Other modalities of treatment used are radiotherapy and the use of imatinib in selected patients mainly in advanced disease [2, 8]. In our case, the histological and immunochemical findings favoured the diagnosis of DFSP. Keeping in mind the involvement of the resected margins, high mitotic figures, risk of recurrence, previous breast surgeries and strong family history of cancer breast, she underwent mastectomy with axillary clearance.

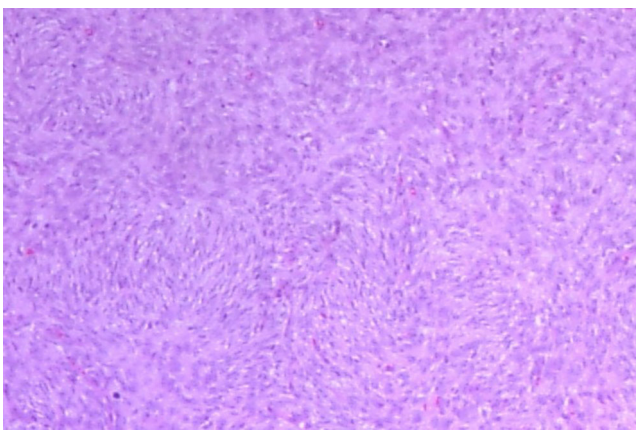

Fig. 4 H and E ×10: tumour cells arranged in storiform pattern

Conclusion

Dermatofibrosarcoma of the breast is a diagnostic challenge. In patients with recurrent tumour closely abutting the skin, it should be a differential diagnosis. Pre-operative investigations are usually non conclusive. Wide local excision with immunohistochemical stains mainly for CD34 is required to confirm the diagnosis.

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This case report has been approved and registered at the National Medical Research Register Secretariat, Malaysia with the ID as mentioned NMRR ID: NMRR-14-1054-22775.

Compliance with ethical standards

Conflict of interest There is no conflict of interest or any financial support involved in this case report. It has not been published or presented in printed or electronic form.

References

1. Yeniay L, Unlap O, Sezak M, Yilmaz R (2012) Dermatofibrosarcoma protuberans of the breast. Breast J 18(5):493–494
2. Bogucki B, Neuhaus I, Hurst E (2012) Dermatofibrosarcoma protuberance: a review of literature. Dermatol Surg 38:537–551
3. Nggada H, Bali B, Na'aya H (2006) A clinicopathological study of dermatofibrosarcoma protuberans in Maiduguri, northeastern Nigeria. Niger J Surg Res 8(1–2):78–80
4. Liu S, Ho T, Hsu S, Zhan H, Chou C (2010) Imaging of dermatofibrosarcoma protuberans of breast. Breast J 16(5):541–543
5. Ivanovic D, Prvulovic N, Stojanovic D, Sveljo O, Vicko F, Kapiel T (2009) Dermatofibrosarcoma protuberance of the breast: mammographic, ultrasound, MRI and MRS features. Arch Gynecol Obstet 280:827–830

6. Shin Y, Kim J, Sung M, Jung J (2008) Sonographic findings of dermatofibrosarcoma protuberance with pathological correlation. J Ultrasound Med 27(2):269–274
7. Kim H, Lee J, Kim S, Seo Y, Lee J, Park J, Kim H, Cinn Y, Cho K, Yoon T (2007) Stromelysin-3 expression in the differential diagnosis of dermatofibroma and dermatofibrosarcoma protuberance : comparison with factor XIIIa and CD34. Br J Dermatol 157(2):319–324
8. Mendenhall W, Zlotecki R, Scarborough M (2004) Dermatofibrosarcoma protuberans. Cancer 101:2503–2508