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RESEARCH ARTICLE

DERMATOFIBROSARCOMA PROTRUBERANS OF BREAST

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ABSTRACT

Dermato Fibro Sarcoma Protruberans (DFSP) is a very rare malignant tumor of subcutaneous tissue characterised by slow infiltrative growth. It presents mostly in second and fifth decade. Even though there is no well defined protocol for treatment of this tumor, wide local excision is being practiced. Present case report is of a 60 year old lady with lump in left breast for 5 years who developed ulceration over the lump progressed in 15 days underwent wide local excision. Histopathology revealed DFSP as diagnosis with CD 34 positive.

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INTRODUCTION

A slow growing, low grade tumor of dermal fibroblastic origin. DFSP very rarely metastatizes but well known to recur at local site with a rate of 26 to 60% with wide and close margins respectively. Incidence is reported to be approximately 5/million annually, with slight female predominance (Lemm *et al*., 2009; Jiang *et al*., 2014). It also occurs in other parts of the body like trunk and extremities. It has very slow growth rate for long period prior to entering rapid growth phase (Llombart *et al*., 2013). Tumor is known to have reciprocal translocation (17,22) q(22,13) or supernumerary ring chromosomes involving 17 and 22 ultimately leads to upregulation of platelet derived growth factor B(PDGFB) gene in the form of type 1 alfa 1 chain PDGFB fusion oncogene (Lemm *et al*., 2009; Jiang *et al*., 2014; Llombart *et al*., 2012).

Case report

60 year old post menopausal woman presented with history of lump in her left breast since 5 years with history of ulceration for 15 days with foul smelling discharge, without history of any sudden increase in size. Local examination revealed 10.6.4cm tumor in upper outer quadrant with multiple bosselations on its surface with well defined margin firm in consistency, non tender with 3.2cm ulcer over it, clinically negative axilla.

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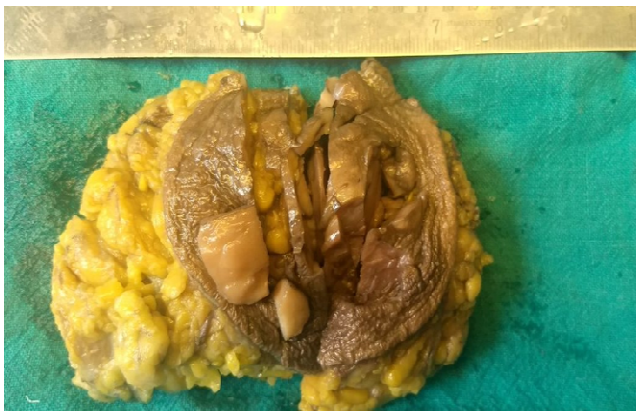
FNAC was inconclusive, trucut biopsy suggestive of benign lesion. Patient underwent wide local excision. Intra operative frozen section revealed spindle cell tumor. Wide local excision with a margin of 2-3cm was done.

Picture of the specimen



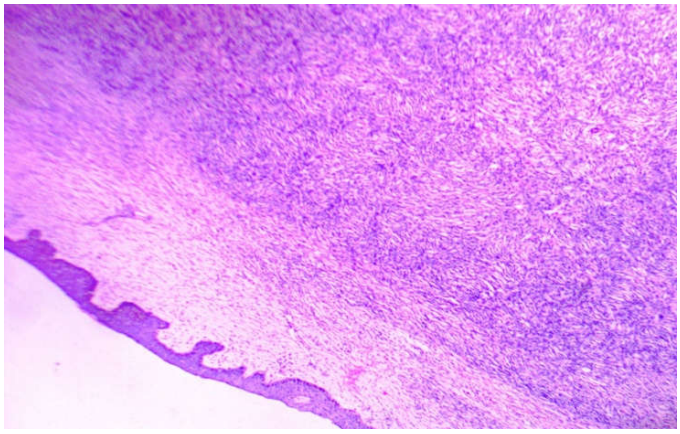
Microscopy

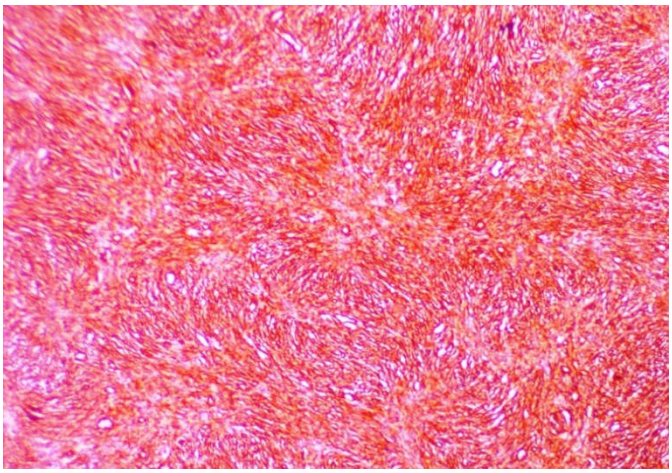
HISTOPATHOLOGY: Dermatofibrosarcoma protruberans, diffuse CD34+, Vimentin+ and focal S100+.

DISCUSSION

First being described by Darier and Ferrand in 1924, the term DFSP was coined by Hoffmann in 1925. Because of its rarity the diagnosis is often missed by pathologist.

Mammography reveals a subcutaneous oval mass with smooth well defined margins (doi/abs/10.2214/AJR.08.2141). MRI is done when primary DFSP is located other than head, neck and upper part of thorax (Chen *et al*., 2009). USG reveals hypoechoic mass with irregular border with no peripheral or internal blood flow (doi/abs/10.2214/AJR.08.2141). DFSP metastatizes only in 2-5% case, so CT and extensive laboratory tests are not recommended (Llombart *et al*., 2013; Zhang *et al*., 2015).





DFSP is classified histologically as (Llombart *et al*., 2013)

- Pigmented (Bednar tumor),
- Giant cell fibroblastoma-like, atrophic, sclerosing, granular cell variant,
- Fibrosarcomatous and
- Myxoid DFSP.

A definitive diagnosis of DFSP is usually established on the basis of routine histopathological and immunohistochemical features. Immunohistochemical expression of CD34 has been considered as a diagnostic marker for DFSP. 80–100% of DFSP tumors express CD 34 (Llombart *et al*., 2013; Chang, 2004; Li *et al*., 2004). Factor XIIIa is useful in the differential diagnosis between DFSP and cellular fibrous histiocytoma (Li, 2004). Novel immunohistochemical markers have been identified for use in differential diagnosis, including stromelysin III, apolipoprotein D, nestin and CD163 (Llombart, 2012; Thway *et al*., 2016). Despite the presence of a fibrosarcomatous component in DFSP, DFSP differs from breast sarcoma in its cutaneous derivation (Li *et al*., 2004; Thway *et al*., 2016). Fine-needle aspiration cytology has low diagnostic accuracy for mesenchymal breast tumor. Core biopsy is considered the standard procedure for diagnosing such tumors, though adjunctive immunohistochemical analysis is often required (Lim *et al*., 2016; Al Barwani *et al*., 2016; Llombart *et al*., 2011). Diagnosis from a core biopsy is also difficult many a times. The differential diagnosis of solitary fibrous tumors (SFTs) is expansive, and a diagnosis based on core biopsy specimens can be challenging, since certain distinctive features of SFT, including alternating cellular and hypocellular architecture, and vascular pattern, may not be appreciable (Li *et al*., 2004; Lim *et al*., 2016; Llombart *et al*., 2011). Complete surgical resection with a wide margin is accepted as the treatment for DFSP. However, there is no guideline for the margin of resection. When DFSP has been excised with close margins, the local recurrence rates range between 26 and 60%. Following wide local excision (2–3 cm), reported local recurrence rates are lower (0–30%) (Snow *et al*., 2004; Farma *et al*., 2010; Fields *et al*., 2011). With the use of a standardized surgical approach, including a meticulous pathological evaluation of margins, a low recurrence rate (1%) was achievable with relatively narrow margins (median size, 2 cm), allowing primary closure in 69% of patients. Though DFSP is considered to be radiosensitive, the role of adjuvant radiotherapy in treating this neoplasm remains uncertain [(19)]. Imatinib mesylate, a tyrosine kinase inhibitor produced substantial regression of locally advanced tumors prior to surgical excision (McArthur, 2005). Multimodality treatment, particularly the use of tyrosine kinase inhibitors, could be effective, but should not be considered as curative. DFSP follow-ups subsequent to surgery are recommended for a minimum of 3 years, in 6-month intervals (Kuzel *et al*., 2015).

Conclusion

As patients ignore DFSP because of its slow growth and is often misdiagnosed as benign tumor, DFSP poses great challenge in diagnosis. So diagnosis of DFSP should be considered strongly in a breast lump with history of slow progression even with fungation. Confirmation should be considered through IHC to assist surgeon in achieving good resection margin and thus decreasing recurrences.

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