INTRODUCTION
Dermatofibrosarcoma protuberans (DFSP) is a rare soft tissue tumour described in 1924 by Darier and Ferrand as progressive recurrent dermatofibroma,\(^1\) and Hoffmann in 1925 as dermatofibrosarcoma protuberans. It has tendency for local recurrence after excision.\(^2\) It can present in all parts of the body but more frequently in the region of trunk and extremities, and very rare in breast.\(^3\) Due to benign appearance and indolent behaviour, preoperative diagnosis is difficult.\(^4\) The standard treatment includes local excision with adequate margins.

CASE REPORT
A 49-year lady known case of diabetes mellitus, with anti HCV positivity presented to a surgeon, with a right breast swelling, noticed by herself five months back. That was progressive in nature over the period of five months. On physical examination, a firm well circumscribed, tender, mobile, 5x4 cm lump was noted in right upper outer quadrant of breast. On physical examination and mammography, it appeared to be benign-looking, on physical examination as well as on mammography. She underwent local surgical resection and diagnosed as DFSP on microscopic and immunohistochemical analysis with positive deep margin. She was referred for further multidisciplinary management. Case was discussed in tumour board meeting and planned for re-resection of involved deep margin. After re-resection, she has remained disease-free for last 3 years without any evidence of local or distant recurrence. In this case report, the importance of surgical resection with adequate margins, is highlighted and long term follow-up by physical examination and ultrasound for any local recurrence of DFSP in breast.

DISCUSSION
The incidence rate of progressive and recurrent dermatofibroma is 0.8 to 4.5 per 10,000 annually, comprising of 1% and <0.1% of all soft tissue sarcomas and of all malignancies, respectively. In breast, it is a very rare entity. It occurs mostly between the second and fifth decades of life. The age of this patient was 49 years, i.e fifth decade.
It infiltrates locally into subcutaneous tissue, to fascia and then to underlying muscle, and results into local recurrence. Mostly, these local recurrences are found within first three years of local resection, but this patient remained recurrence-free for the last three years. In 1-4% cases, distant metastasis are noted, more frequently in lungs and lymph nodes. Patients with non-metastatic DFSP has 5-year survival rate of upto 99%. A specific chromosomal translocation (t17-22) is detected in >90% of dermatofibrosarcoma protuberas on genetic analysis. With this translocation of chromosomes, B chain of platelet derived growth factor produced, that leads to growth stimulation of DFSP. DFSP presents usually at trunk and extremities, unusual in neck and very rare in breast. Histopathologically, DFSP includes several subtypes including Sclerosing granular cell variant, pigmented (Bednar tumor), atrophic, giant cell fibroblastoma-like, myxoid and fibrosarcomatous. Clinically, differentials of dermatofibrosarcoma protuberas include myofibroblastoma of breast, metastatic carcinoma of breast, fibromatosi, recurrent dermatofibroma and hypertrophic scars. DFSP is diagnosed initially on histopathology and confirmed by immunohistochemistry. Histopathologically, spindle cells seen that are designed in storiform manner. These spindle cells have low mitosis with a small amount of nuclear pleomorphism. This patient was also diagnosed on histopathology and immunohistochemistry. Histopathologically, differential diagnosis of DFSP includes fibromatosis, metastatic carcinoma, myoepithelioma and phyllides tumor. These lesions are differentiated on immunohistochemistry by tumour markers. In DFSP, CD34 is positive in 84-100% of cases, that was also positive in this patient, and vimentin is also positive, which indicate this tumour as the fibroblastic in nature, S-100 and desmin are negative immunohistochemical markers in DFSP that were also negative in this patient.

Due to relative resistance to chemotherapy, local resection with wide margin is the standard treatment for localised dermatofibrosarcoma protuberas. The recommended margins for complete local surgical resection are 2-3 cms, along with resection of skin, subcutaneous tissue, and fascia in three dimensions. Risk factors for increased recurrence rate included histopathologial subtype, size, increased cellularity, high mitotic rate and the head and neck location. Dragoumis et al. suggested that local recurrence rate could be decreased by adequate surgical margins, as in this patient where re-resection with wide margins was performed. Another author suggested that the Mohs surgery is a first-line management from cosmetic point of view, and to reduce the rate of local recurrence. Although, this surgery needs a particular trained team and have many stages. Dubay et al. reported that after surgical resection of DFSP either by local wide resection or by Mohs surgery, or combination of both, there was no local or distant recurrence found over the time period of 4.4 years. In this patient, after wide re-resection, no local or distant recurrence found over the period of 3 years.

Radiotherapy is an option of treatment in adjuvant setting in advanced cases where re-resection is not possible. This female patient underwent re-resection as deep resected margin was involved by tumour. Strict monitoring with follow-up every 6-12 months needed. Ultrasound and biopsy should be done as clinically indicated in cases of suspected recurrence; this patient was also followed every 6-monthly with physical examination and ultrasound.

REFERENCES


