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Case Report

Painless Lump over the Upper Chest - A Keloid Mimicker

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is a rare monoclonal cutaneous low grade sarcoma of fibroblast origin. The tumour has a high rate of local recurrence and late presentation due to its indolent behaviour. The estimated incidence is 0.8 per million cases in a year. Mohs, modified Mohs surgery and excision with wide margins is the treatment of choice. Hereby we are reporting a male patient with DFSP mimicking keloid.

Keywords: Dermatofibrosarcoma protuberans, low grade sarcoma, local recurrence.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) is a rare monoclonal cutaneous *low grade sarcoma of fibroblast origin which was first described by Taylor in 1890.* [1] The term Dermatofibrosarcoma protuberans was first introduced by Hoffmann in 1925. [2] The estimated incidence is 0.8 per million cases in a year. [3] The tumour has a high rate of recurrence of 24-60% after a surgical excision and very low malignant potential of <5%. [4] Hereby we are reporting a male patient with DFSP mimicking keloid.

CASE REPORT

A 28 year old male presented with painless lump over the right upper chest since 2yrs.

Clinical examination revealed an irregular hyperpigmented sclerotic plaque with hyperpigmented, dome shaped, firm, non-tender nodule in the centre of the plaque situated over the right upper lateral quadrant of the chest (Figure 1a & 1b). The lesion was fixed to the underlying skin. Axillary lymph nodes were not palpable.

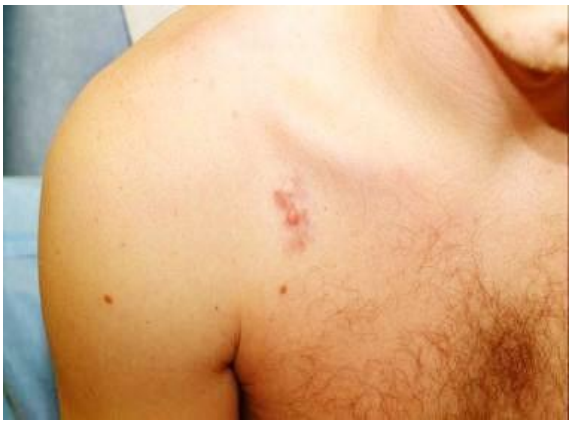

Figure 1a: solitary, hyperpigmented, dome shaped, nodule present over the right upper lateral quadrant of the chest in a 28 year old male

Histopathological examination revealed spindle cell neoplasm involving the entire dermis and extending into subcutis. Both the lobules as well as septae are infiltrated by the cells. A typical cell seen. Both lateral margins are free of

infiltrate. Lower margin shows a very thin rim of normal fibrous tissue beyond the infiltrated subcutis (Figure 2a & 2b). Routine investigations were within normal limits. Based on clinical and histopathological examination diagnosis of Dermatofibrosarcoma protuberance was confirmed.

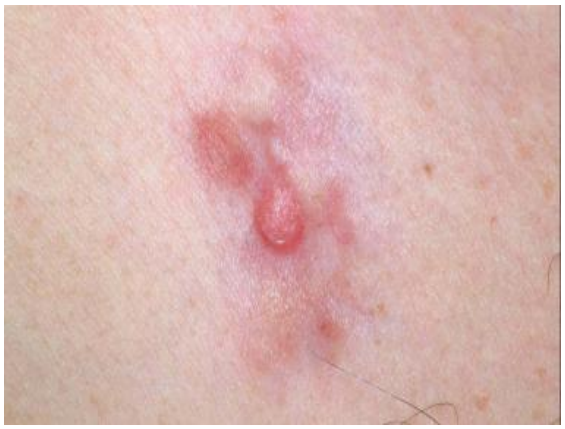

Figure 1b: close up view showing an irregular sclerotic plaque with hyperpigmented nodule in the center of the lesion

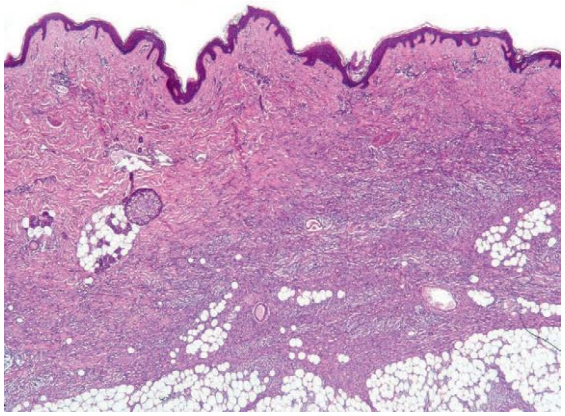

Figure 2a: H & E stain (10x) view showing spindle cell neoplasm involving the entire dermis and extending into subcutis

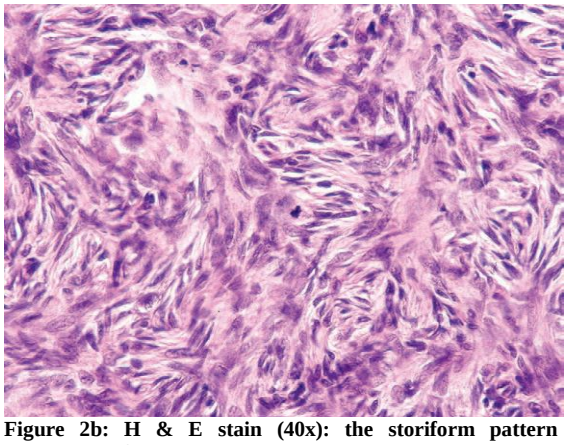

Figure 2b: H & E stain (40x): the storiform pattern comprising of cells with vesicular nuclei

DISCUSSION

DFSP is a low grade benign mesenchymal tumor which constitutes >0.1% of all malignancies. DFSP occurs most commonly in middle aged adults, between the age groups of 20 and 50 with a slight male preponderance (3:2). [5] There are no racial differences though some reports show an increased preponderance in black population. In addition, Bednar tumour (pigmented variant of DFSP) has also been reported to occur frequently in black population. [6]

Trauma, scars, site of vaccination or pregnancy have been reported as predisposing factors for the development of DFSP. [7] The exact pathogenesis of DFSP is unknown, however chromosomal abnormalities like supernumerary ring chromosomes, abnormal clones, supernumerary ring chromosomes containing 17 sequences and translocations have been proposed. Recent studies show a translocation between chromosomes 17 and 22 (t(17:22)) resulting in over expression of platelet-derived growth factor receptor (PDGFR)- β leading to cellular proliferation, which can be found using reverse transcriptase polymerase chain reaction and florescence in-situ hybridization. [8]

The most common site of DFSP is the trunk with 25% occurring over the chest and shoulders. Less common sites involved are proximal extremities, acral, head and neck region. Enzinger and Weiss observed the following site distribution in a series of 853 patients: trunk, 47%; lower extremity, 20%; upper extremity, 18% and head and neck, 14%. [9] Initially DFSP presents as solitary, asymptomatic indurated plaques with a violaceous, red-brown or skin colour. Nodules may develop in the plaques which may ulcerate with bleeding and pain. The plaques may be 1 to 5 cms which may be fixed to the overlying skin. The tumour gradually spreads to involve deeper dermis and the subcutaneous tissue which may further

involve the fascia, muscle or bone if left untreated manifesting in the form of satellite nodules at the periphery of the lesion which may coalesce to form larger masses giving a protuberant appearance and also attributes to the incomplete excision and frequent recurrences. [4] Clinically, DFSP need to be differentiated from lipoma, sebaceous cyst, neurofibroma, lymphomas, sarcoidosis, malignant melanoma, cutaneous metastases, gumma, keloid, sclerosing hemangioma, desmoid tumor, fibrosarcoma, neurogenic sarcoma, sweat gland carcinoma, dermatofibroma, morphea, and nodular fasciitis. [10]

On histopathology, DFSP is a dermal tumour composed of monomorphic, benign appearing spindle cells arranged in irregular interwoven fascicles giving a storiform or a cartwheel appearance. [10] The other uncommon histological variants include a fibrosarcomatous variant (DFSP-FS) which show an increased risk of metastasis, a pigmented variant called Bednar tumour having melanin containing dendritic cells among spindle cells, myxoid DFSP mimicking liposarcoma or myxofibrosarcoma, atrophic variant characterized by dermal atrophy and giant cell fibrosarcoma which is seen mainly in children and histologically shows a unique combination of spindle cell patterns, myxoid areas, pleomorphic and multinucleated giant cells, distinctive sinusoid-like spaces and tentacular infiltration of adjacent subcutaneous tissue. The infiltrate may extend from the dermis to the subcutaneous tissue where in two patterns of involvement have been observed, a classical extension of spindle cells along septae or between fat cells in a honeycomb or lace-like pattern and multi-layered pattern with the bundles of slender spindle-cells arranged parallel to the skin surface. [12] Tumour may extend to underlying fascia and muscle making excision difficult. The rate of metastasis is very low which is usually <5%. [4]

Immunohistochemically the neoplastic cells react with CD34, hyaluronate, vimentin, apolipoprotein D, p75 and the absence of factor XIIIa and S100 which is very helpful in the differentiation of DFSP with fibrosarcoma, nerve sheath tumours, dermatofibromas, malignant fibrous histiocytoma and atypical fibroxanthoma. [13,14] In large recurrent lesions, magnetic resonance imaging may be useful for ascertaining deep tumour invasion. In advanced cases with pulmonary involvement a chest roentgenography may be done and computerized tomography is indicated when an underlying bone is involved.

DFSP and DFSP-FS can be staged according to American Musculoskeletal tumour society, stage IA, no extension beyond the subcutaneous tissue and stage IB, and involvement of the underlying fascia or muscle. [15] DFSP has risk for local recurrence, very rarely metastasis to regional lymph nodes and distantly to lung, brain, bone and peritracheal area via hematogenous spread.

Initial treatment of DFSP is mainly surgical and every effort should be made to remove the tumour completely. Mohs, modified Mohs surgery and excision with wide margins, typically with 2-4cm margins are the available treatment modalities. Reconstruction should be undertaken for large tumors which has undergone extensive resections after confirmation of the negative margins. DFSP with t (17:22) translocation, inoperable, recurrent and metastatic cases can be treated with a recently FDA approved drug imatinib mesylate which targets the PDGFRs and reduce the growth of the tumour. [16]

Radiation can be used as a primary modality or adjuvant to the surgery. Postoperative radiation or imatinib mesylate should be considered for positive

surgical margins and if further surgical resection is not possible.

CONCLUSION

DFSP is a rare mesenchymal tumour with a high rate of recurrence and late presentation due to its indolent behaviour. Efforts for complete removal of the tumour should be undertaken. A follow-up of the primary site will be needed every 6-12 months owing to high recurrence rates. Extensive work-up is usually not necessary because of the very low risk of metastasis. We are presenting this case for its rare occurrence and mimicking like a keloid.

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