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Original Research Paper

Dermatofibrosarcoma Protuberans Masquerading Skin Tag

Medical Science

KEYWORDS : Dermatofibrosarcoma protruberans, mesenchymal tumor, skin tumor, Skin tag.

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is an uncommon malignant mesenchymal tumor, characterized by high propensity for local relapse and low metastatic potential. It is seen most commonly in individuals aged between 20 and 50 years, the usual location being trunk and it is limited to the dermis. Histopathology is characterized by fascicles of monomorphous spindle cells arranged in a storiform patten. Wide radical excision is the preferred surgical therapy. Herein, we report a 45 years old male with DFSP over thigh wide local excision was performed and the diagnosis was confirmed by histopathological examination. This case is being reported for its rarity and also its histopathological section revealed more than a variant.

CASE REPORT

A 45-year-old male, teacher by occupation, presented with asymptomatic brown raised skin lesion over left thigh for past one year, insidious in onset, which gradually progressed to develop nodules on its surface. There was no history suggestive of trauma, surgery, vaccination or burns prior to the onset of skin lesion. Cutaneous examination revealed a well-defined, hyper pigmented indurated plaque of size 3 x 2.5 cms size, with fleshy erythematous nodule on its surface, located over the medial aspect of middle one third of left thigh [Fig 1]. On palpation, it was not tender but firmly fixed to the subcutaneous tissue. We considered the following differential diagnosis – achrochordon, dermatofibroma, dermatofibrosarcoma protuberans and fibrosarcoma. With proper consent, under aseptic precautions, we performed wide local excision [Fig 2 (a) and (b)] and sent for histopathological examination, which revealed poorly circumscribed tumor in the dermis, the cells were elongated, spindle shaped, distributed as sweeping interlacing bundles, [Fig 3 (a)] Myxoid changes were also seen [Fig 3 (b)]. Tumour giant cells are noticed and occasionally touton giant cells were also seen. Mitotic figures varying from 0 to 3 per 10 high power field was noted, signifying intermediate malignant potential.

DISCUSSION:

DFSP is a rare locally aggressive cutaneous tumour with high rates of local recurrence and low risk of metastasis1,2. It was first described by Sherwell and Taylor in 1890. Darier and Ferrand in 1924, described this tumor as progressive and recurrent dermatofibroma. Hoffman coined the term dermatofibrosarcoma protuberens based on the tendency of the tumor to produce protruding nodules. DFSP is the most common sarcoma of cutaneous origin that constitutes <0.1% of all malignancies and 1% of all soft tissue sarcomas3.

DFSP most commonly occurs between 20 and 50 years of age. It has an equal sex distribution; however slight male predominance is seen in some series4. About 40% to 50% of DFSP is located on the trunk, commonly on the chest and shoulders, which is followed by proximal portions of the limb (30-40%) and head and neck (10-15%)5.

DFSP is a slow growing tumour with an early slow growth phase followed by a rapid growth phase with multiple nodules. It initially appears as an asymptomatic indurated plaque that may have a violaceous red, blue or brown appearance which over a period of time grows with development of multiple nodules within the plaque6. Murphy et al and Moureau –Zabottoa et al studied the clinical characteristic at the earliest stage and they classified three different forms of nonprotruding DFSP: 1. Morphea like, 2. Atrophoderma like 3. Angioma like7. DFSP ranges in size from 2 to 5 cms and is usually fixed to overlying skin but not to deeper structures. In histopathology, DFSP appears as poorly circumscribed tumor, with diffuse infiltration of the dermis and subcutis, yielding a honeycomb pattern. It is composed of fascicles of densely packed, monomorphous spindle cells arranged in a spongiform pattern. Mitotic activity is generally limited typically to less than 5 mitoses per 10 high power fields8.

Histologic subtypes are1

1. Pigmented DFSP (Bednar tumour)

2. Giant cell fibroblastoma

3. Atrophic DFSP

4. Sclerosing DFSP

5. Granular cell variant

6. Fibrosarcomatous DFSP

Immunohistochemistry of DFSP shows CD- 34 expression in about 80% to 100% of tumor9,10. Cytogenetic analysis shows reciprocal translocation t (17; 22) (q 22; q 13) or a supernumerary ring chromosomes 17 and 22 with result-

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ant formation of COL1A1 –PDGFB fusion transcripts which can be detected either by Fluorescent In Situ Hybridization on interface nuclei and by multiplex Reverse Transcriptase-Polymerase Chain Reaction. This serves as a useful tool for the differential diagnosis of DFSP with other CD-34+ cutaneous tumors and for treatment for treatment with imatinib11,12,13.

The standard treatment is wide surgical excision with 2-4 cms margin to investing fascia. Other varied approaches are Mohs micrographic surgery and modified Moh's microscopic technique14. Imatinib mesylate is FDA approved drug for the treatment of unresectable, recurrent and metastatic DFSP in adult patients14,15. Clinical follow up of the primary site should be done every 6-12 months and biopsy should be done in any suspicious region.

FIGURES

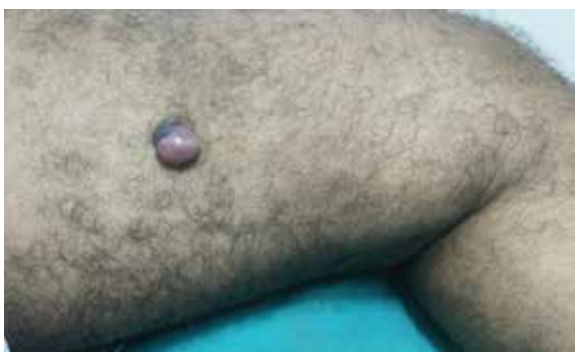

Fig 1: Indurated plaque with fleshy erythematous nodule on its surface, over the medial aspect of middle one third of left thigh

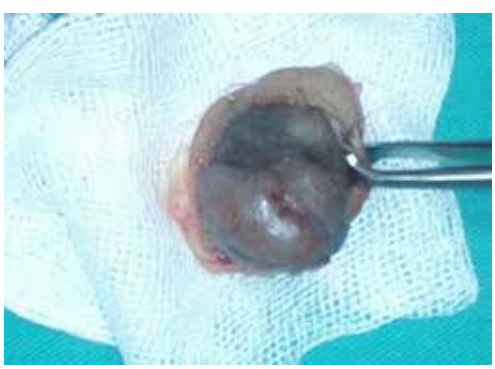

Fig 2 (a): DFSP after excision

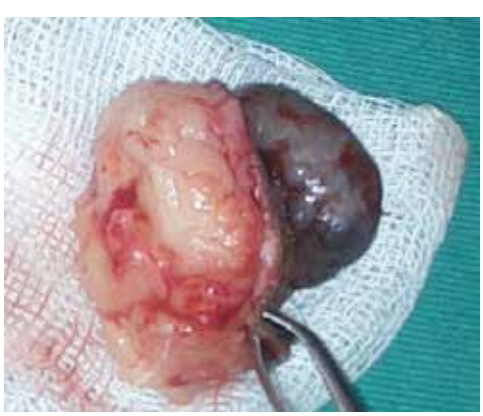

Fig 2 (b): Base of DFSP after excision

HISTOPATHOLOGICAL SECTIONS: 10x view:

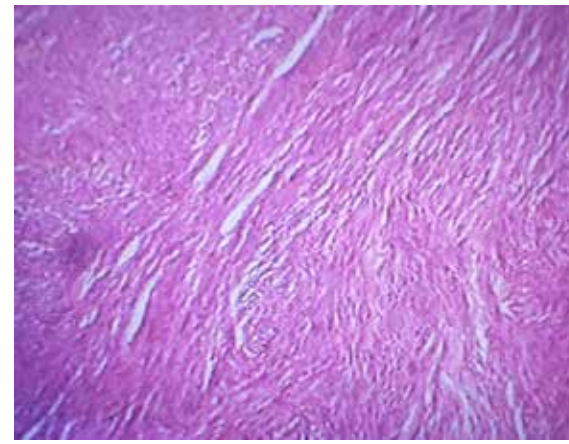

Fig 3 (a): Spindle shaped cells arranged in a storiform pattern.

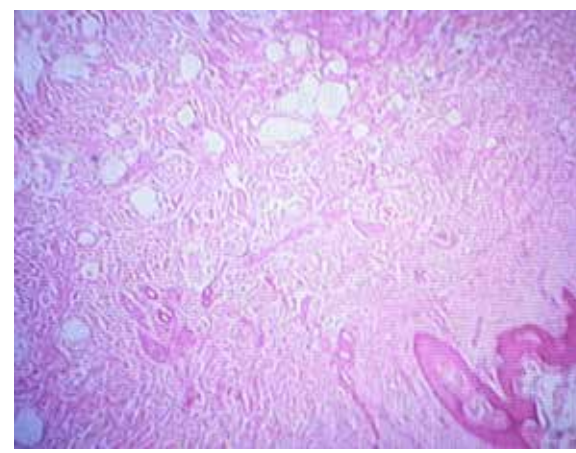

Fig 3 (b): Myxoid component in dermis

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