Case Report

Recurrent Locally Malignant Tumour of Arm : Dermatofibrosarcoma Protuberans

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ABSTRACT

Dermatofibrosarcoma protuberans (DFSP) is commonest skin sarcoma representing less than 0.1% of all tumors. Treatment includes surgical management followed by radiotherapy to decrease chances of recurrence. We present a typical case of dermatofibrosarcoma protuberans with twice local recurrence within one year after excision. 43 years old immunocompetent married male presented to us with nodular lesion over right arm since 6 years. Patient gave the history of similar lesion over the same site in past for which he was operated twice 10 and 7 years back, respectively. Patient was advised adjuvant radiotherapy but he didn't follow same. Since 6 years, patient complained of a small raised lesion over the same site which increased gradually to the present size. Hence we report this typical case of DFSP representing its inherent tendency of recurrence that could have been prevented by strict radiotherapy after surgical management.

Key-words : Dermatofibrosarcoma protuberans, radiotherapy, twice local recurrence

Introduction :

Dermatofibrosarcoma protuberans (DFSP) is a rare, slow-growing fibrohistiocyticneoplasm of low to medium malignancy affecting mainly affects young and middle-aged adults between the second and fifth decade of life. It emerges as an asymptomatic bluish or brownish erythematous multinodular lesion, involving most frequently the trunk, proximal extremities, head and neck. Recurrence after incomplete resection at the same site is common but distant metastases are rare.[1] Sites of occurrence of DFSP include surgical scars, old burns, trauma, radiation dermatitis, vaccination sites, central venous line puncture sites and even insect bites.[2]

Case History :

43-year-old married immunocompetent man came to dermatology OPD with a large nodular lesion over right arm. The patient reported an increasing size of the tumor during the preceding 6 years. The patient denied any recent weight loss, fever, night sweats or chills. Patient gave history of a peas sized lesion since 20 years for which he was first operated in 2010. Within first year of excision, there was recurrence for which he was again operated after 3 years. Again there was recurrence within first year of second excision which increased to the present size. He was advised adjuvant radiotherapy after both the surgeries but he didn't follow the same. On physical examination, a large, firm, painless, multinodular mass with no sign of localized heat or redness was found ***(Figure 1a and 1b).*** There was no lymphadenopathy. There was no familial history of malignancy. T1 & T2 weighted images were taken in sagittal & axial plains along with STIR coronal & GRE axial images. It revealed well defined heterogeneously enhancing lobulated altered signal intensity lesion of approximate size 7.1 x 2.2 cm involving skin and subcutaneous soft tissue in lateral aspect of right midarm, just abutting the underlying muscles at some places. The lesion appeared isointense to muscle on T1, mildly hyperintense on STIR and shows no diffuse restriction on DWI ***(Figure 1c and 1d).*** Microscopically, H&E stained section of skin biopsy showed unremarkable epidermis with tumor mass in the dermis. The tumor was composed of spindle cells arranged in characteristic storiform pattern. The individual spindle cells showed presence of mild nuclear

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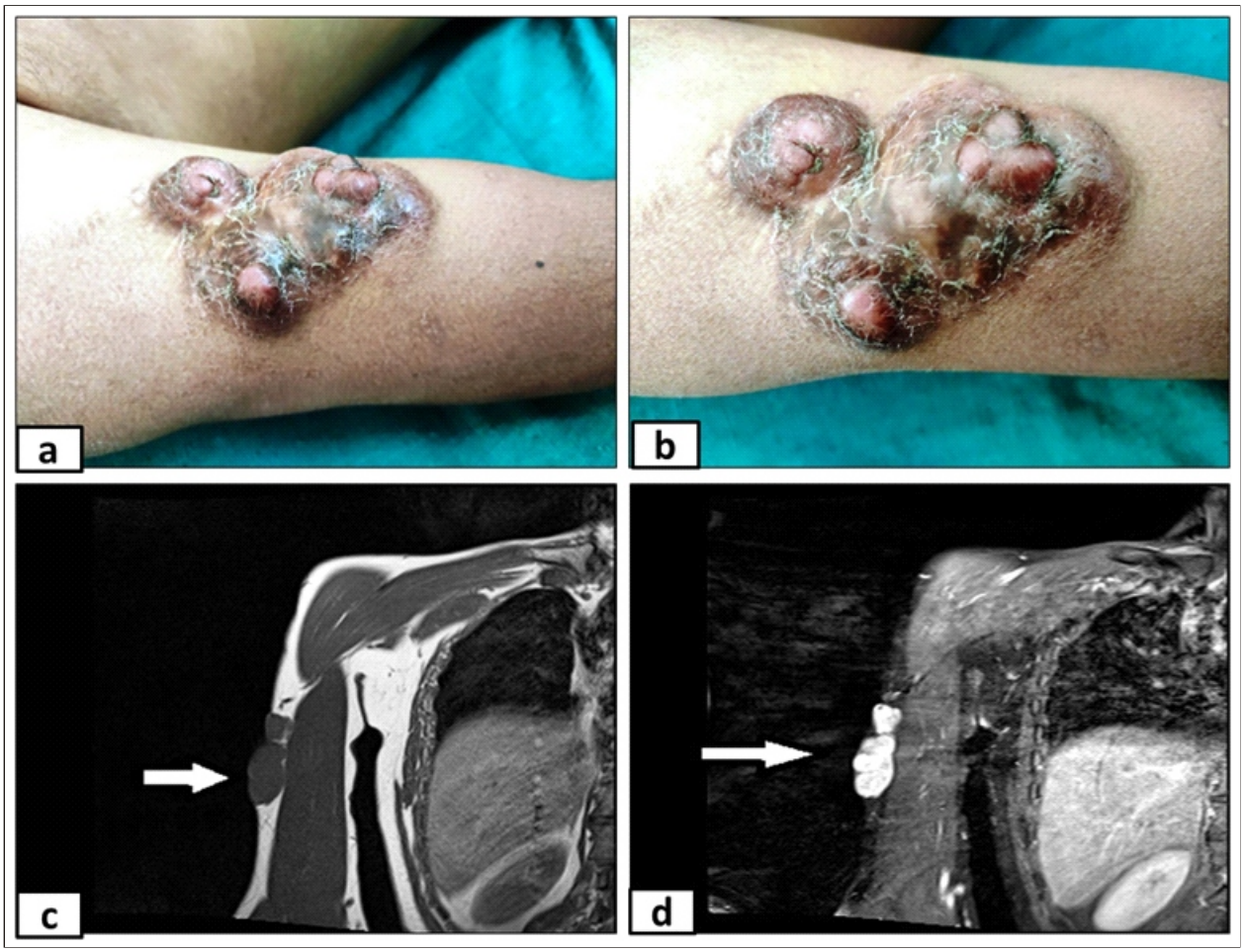

Figure 1 : **A large multinodular mass of size 7 cm x 2 cm on dorsal aspect of right arm; (a) Side view, (b) Front view, (c) The lesion appears isointense to muscles (arrow) on T1W, (d) Shows heterogenous post contrast enhancement (arrow).**

pleomorphism and low mitotic activity ***(Figure 2a and 2b).*** Immunohistochemistry showed S100 ***(Figure 2c)*** and desmin negativity ***(Figure 2d).***

Discussion :

DFSP appears as a violaceous, pink or reddish-brown plaque that develops slowly, initially restricted to the skin. With time, the tumor grows into multiple "protuberant" nodules that may infiltrate the underlying tissue, fascia, muscles and even bone.[2,3] In our case, no infiltration of the adjacent muscular or bony structures were seen.

Early stage DFSP should be differentiated from lipomas, epidermal cysts, keloids, dermatofibroma, and nodular fasciitis. In the later stages, the differential diagnosis includes pyogenic granuloma, Kaposi sarcoma and other soft tissue sarcomas.[4] Hence histological examination is the only definitive diagnostic method. Rarely DFSP might present an infiltrative subcutaneous mass. Atypia is minimal and mitoses are rarely seen.[5] Mitotic count, necrosis, and areas of fibrosarcomatous change is important in the histopathology report as they are correlated with aggressive clinical behavior and lower overall survival.[6]

Other types of DFSP are myxoid DFSP, in which myxoid characteristics predominate and Bednar tumor, which is a pigmented DFSP characterized by the presence of dendritic cells that produce melanin.[3] Immunohistochemically, tumor cells stain for vimentin, CD34, apolipoprotein D, nestin, and may

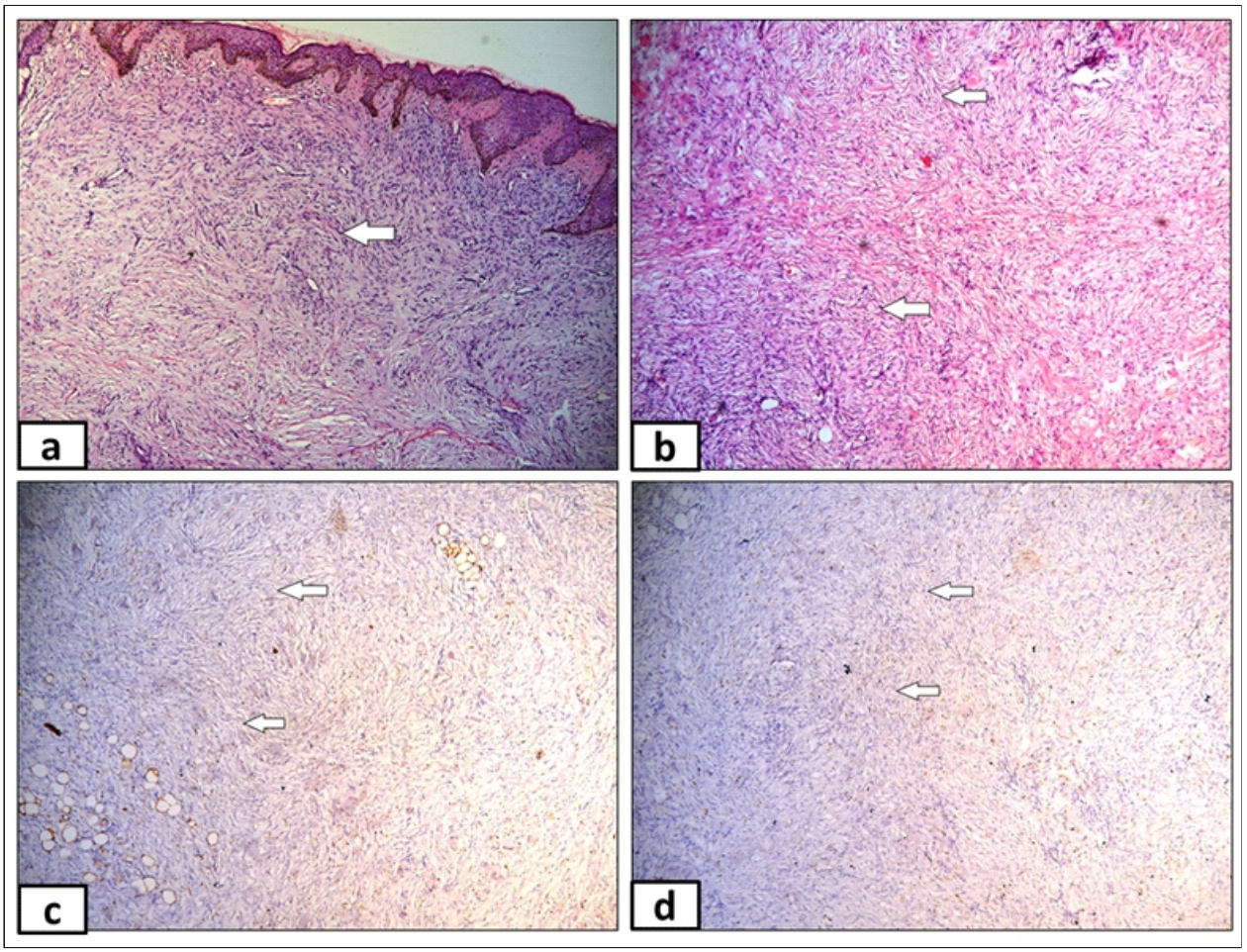

Figure 2 : **(a) Microscopy showing presence of tumor mass arranged in storiform pattern (white arrow)in dermis and unremarkable epidermis (H&E, 400X), (b) Spindle cells showing mild nuclear atypia in the dermis (H&E, 400X), (c and d) Tumor cells showing S100 negativity and Desmin negativity (IHC, 400X)**

be for EMA. Desmin, S100 protein, FXIIIa, stromelysin III, HMGA1&2, tenascin, D2-40, CD163 and keratins are negative. In our case it was desmin and S100 negative. Our Institute did not have the facility for immunohistochemistry for vimentin and CD34. The final diagnosis was made on the basis of clinical history, characteristic morphologic and available immunohistochemistry findings.

Wide local excision is treatment of choice with negative margins of 3-5 cm. If there is bone involvement, the periosteum or even a portion of the bone may be excised to achieve negative resection margins. Recurrence rate depends on the resection margins. In cases with resection margins of five cm, recurrence rates are less than 5%.[7] Reconstructive surgeries may be done to restore tissue defects. As an adjuvant treatment, imatinib mesylate, a tyrosine kinase inhibitor is used in the treatment of an unresectable, recurrent and/or metastatic disease.[2] Radiotherapy is combined with surgery in cases of positive or inadequate margins. Rate of recurrence is high appearing within the first 3 postoperative years, with 50% within the first year of surgery. However, recurrences after 5 years are also reported.[8] In our case recurrence occurred within first year after both the surgeries.

To conclude, it is important to follow-up these patients for long-term and radiotherapy must be given due to inherent tendency of recurrence.

Patient was counselled and then was referred to the surgery department for proper surgical intervention and further management.

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