An Asymptomatic Plaque on the Shoulder

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A 53-year-old farmer presented with a gradually progressive asymptomatic swelling over the outer aspect of the right arm for the last seven years. There was a history of trauma (bitten by grandson) at the same site approximately 12 years ago. On examination, there was a well-defined plaque measuring approximately 11 cm × 10 cm over the extensor aspect of the right arm. The plaque was skin colored to whitish yellow consisting of multiple, polysized, whitish-yellow, shiny, nodules, with intervening areas of atrophy and scarring. On palpation, the nodules were hard, non-tender, and fixed to the base of the plaque. There was no regional lymphadenopathy. There was a history of intra-lesional administration of injection triamcinolone on multiple occasions three years ago but without much improvement. His clinical and dermoscopy images are illustrated in Figure 1a and Figure 1b and histopathology and immunohistochemistry images (H and E, CD34, SMA, and S100) are given in Figure 2a, Figure 2b, Figure 2c, Figure 2d, and Figure 2e respectively.

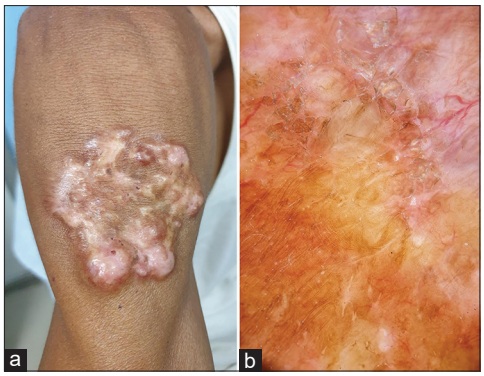

Figure 1: (a) Solitary well-defined plaque on extensor aspect of right forearm measuring 11 × 10 cm studded with red-brown to white nodules with intervening areas of atrophy and scarring. (b) Dermoscopy showing red, brown and white structure-less areas suggesting increased collagen deposition and fibrosis. Telangiectasiae are also seen

Question

What is your diagnosis?

Answer

Dermatofibrosarcoma protruberans.

Discussion

Dermatofibrosarcoma protuberans (DFSP) is a rare superficial tumor of spindle cell origin, with an annual incidence of 0.8-5 per million, often misdiagnosed clinically, and has a high rate of local recurrence but a low risk of metastasis.[1] Clinically, it often behaves as a benign, indolent tumor but microscopically, it can extend much beyond the visible clinical margins, spreading deep in the dermis and subcutaneous tissue. Chromosomal translocation t (17;22) (q22; q13), resulting in the fusion protein COL1A1-PDGFB, which causes tumor growth by increasing the levels of platelet-derived growth factor (PDGF) is implicated in genetic predisposition in the development of DFSP.[2] Acquired causes are trauma, immunization, scars, burns, chronic irritation, etc. The majority of DFSPs occur on the trunk (50%) followed by the extremities (35%).[2] DFSP usually presents as an asymptomatic slowly progressive skin-colored to red-brown, hard, indurated plaque studded with violaceous to red-brown papules and nodules. Atrophic DFSP might give the impression of morphea or scar tissue. In the early stages, the tumor may resemble a keloid. Larger tumors may be tender and can ulcerate and in advanced stages, deeper infiltration into muscle and bone can be seen. Histopathology of DFSP characteristically shows tumor cells arranged in a radial or storiform pattern, with the tumor extending into the subcutaneous fat.[3] The cells are surrounded by collagenous stroma

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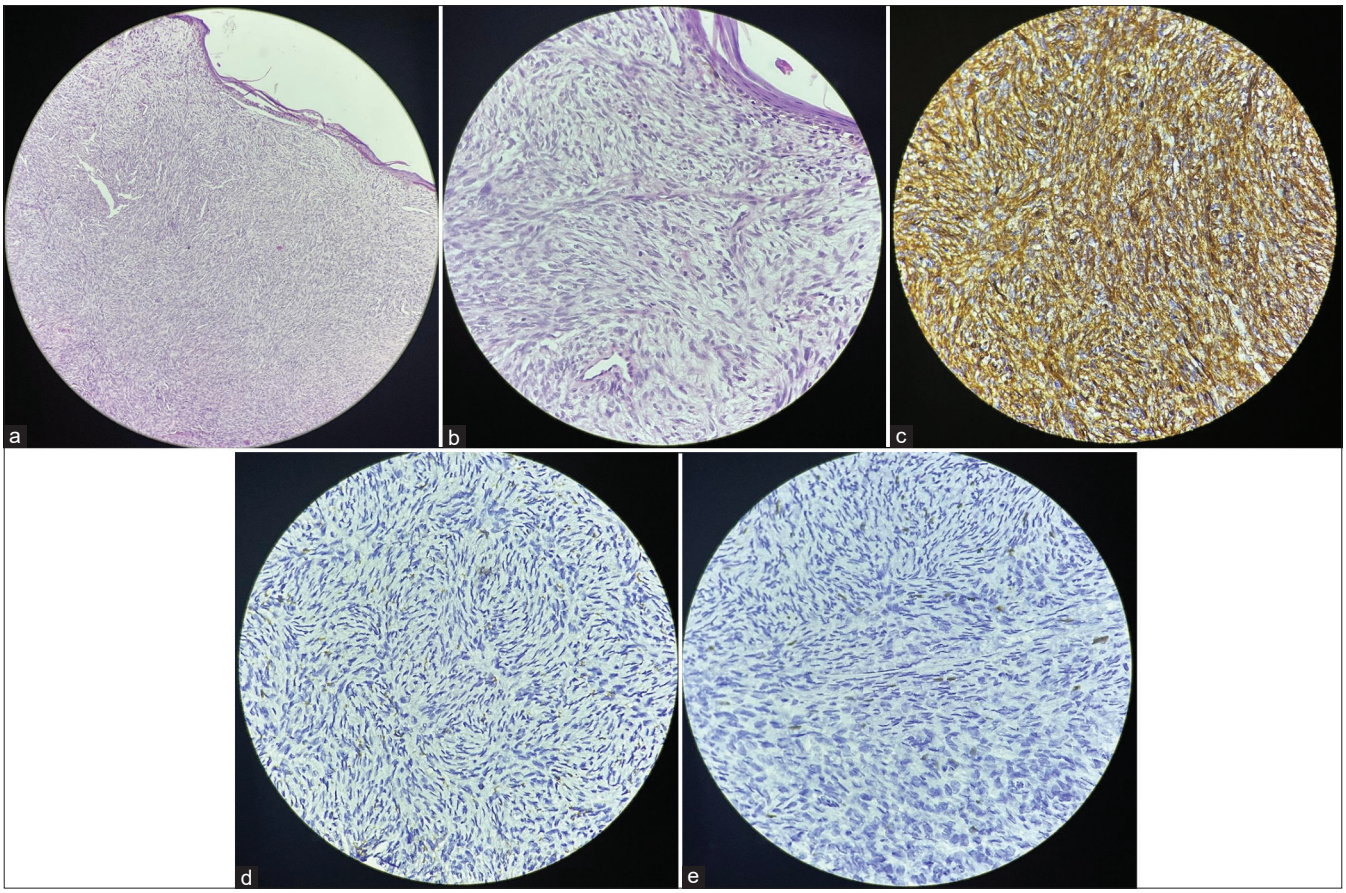

Figure 2: (a) The dermis and subcutaneous tissue are replaced by monotonous and bland tissue showing a honeycomb pattern. (H and E, 4×) (b) Skin biopsy section showing interwoven bundles of uniform, small spindle-cells. (H and E, 40×). (c) Staining with CD34 shows diffuse positivity. (IHC, 40×). (d) and (e) Smooth Muscle Antigen and S100 are negative. (IHC, 40×) (H and E, Hematoxylin and Eosin; IHC- immunohistochemistry)

which sometimes shows hyaline or myxoid degeneration. Immunohistochemistry shows CD34 positivity in 80–100% of cases, while factor XIIIa, desmin, SMA, keratins, and S100 are negative.[4] Dermoscopy features of the delicate pigmented network, arborizing vessels, structure-less light-brown areas, shiny white streaks against a pink background, and structure-less hypo- or de-pigmented areas may be a clue to early diagnosis.[5] Characteristic histopathology differentiates it from dermatofibroma, keloid scar, morphea, and fibrosarcoma. Surgery is the treatment of choice; in ideal circumstances a Mohs micrographic surgery or a wide local excision.[6] Morbidity due to local recurrence, cosmesis, and scarring post-surgery is an area of concern; however, metastasis is extremely rare.[7]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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