CASE REPORT

Dermatofibrosarcoma protuberans

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ABSTRACT

Dermatofibrosarcoma protuberans is a rare, locally aggressive skin tumor that occurs mainly over the trunk and proximal extremities, and has a tendency to recur after wide local excision and can be a therapeutic challenge. We report herein a 50-year-old female with this rare tumor.

INTRODUCTION

Dermatofibrosarcoma protuberans (DFSP) was first described as a distinct clinicopathologic entity in 1924.[1]It is a rare spindle cell tumor of the skin. It is locally aggressive, recurs after marginal or incomplete resection, and can sometimes metastasize or evolve into a tumor indistinguishable from fibrosarcoma.[2] Histologically, DFSP is a cellular dermal spindle cell neoplasm with small amounts of collagen. The tumor shows an infiltrative growth pattern into the subcutaneous fat and positive staining with CD34.[3] The treatment of choice for DFSP is surgical, including Moh's micrographic surgery, with or without postoperative radiation therapy.[4,5]Recently, the U. S. Food and Drug Administration granted approval to imatinib mesylate as a single agent for the treatment of DFSP.[6]

CASE REPORT

A 50 -year-old woman presented with a red brown painful nodule over the right shin of 6 months duration (Fig. 1). It was 2 x 1.5 cm in diameter nodule, fixed to the dermis but moved freely over the underlying tissue. She could not recall any trauma to the affected area. There was no family history of a similar lesion. There was no enlargement of the lymph nodes. The histopathology revealed on hematoxylin and eosin (H&E) stain a hyperplastic epidermis. The structures within the dermis were replaced by masses of spindle cells in storiform pattern extending to the subcutaneous fat (Fig. 2). Nuclei showed slight degree of atypicality and some mitotic figures (Fig.3). The collagen in the dermis was evident and preserved. Immunoperoxidase stains were focally positive for CD34 and negative for S-100 (Fig.4).

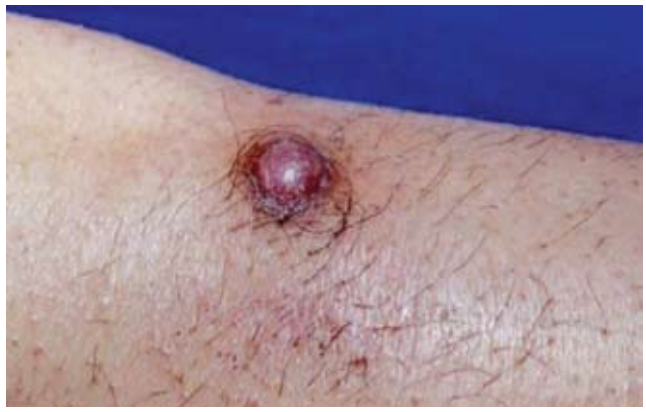

Fig. 1 Red brown dome-shaped symmetrical nodule over the shin

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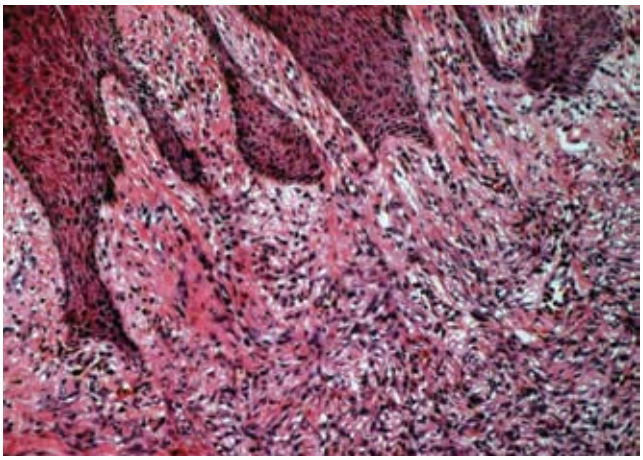

Fig. 2 Proliferation of atypical spindle-shaped cells in fascicles throughout the dermis

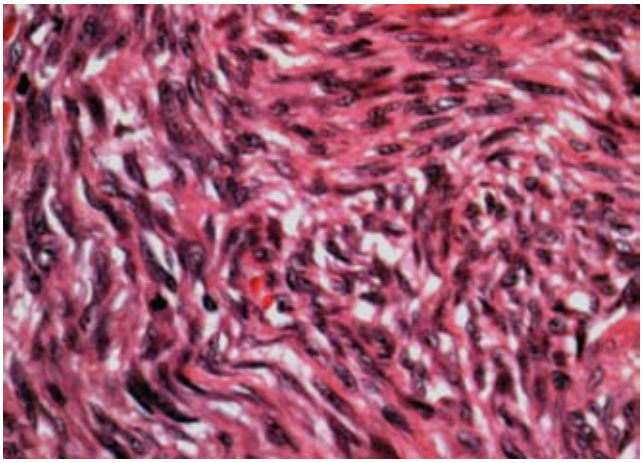

Fig. 3 Atypical spindle cells arranged in a storiform pattern

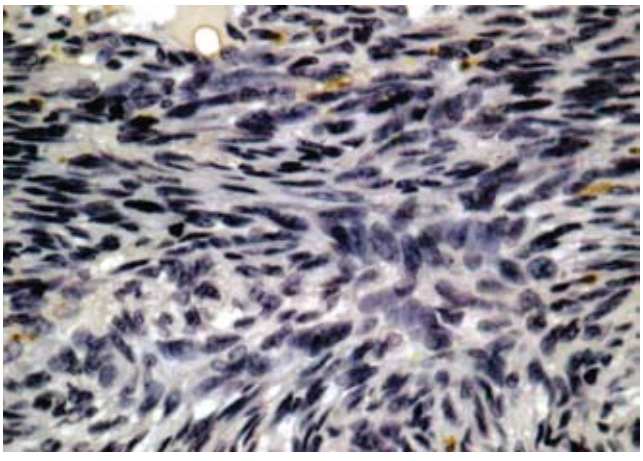

Fig. 4 Atypical spindle cells focally staining with CD34

DISCUSSION

Dermatofibrosarcoma protuberans (DFSP) is rare and often asymptomatic, slow-growing, intermediate grade cutaneous sarcoma with a distinctive fibrohistocytic appearance. Although DFSP was first reported in 1890 by Taylor, the histopathology was not defined until Darier and Ferrand did so in 1924 and it was not named until the following year by Hoffman. In 1962, Taylor and Helwig documented the distinguishing histologic feature of DFSP, spindle-shaped cells arranged in a storiform pattern.[7] It is a rare and locally infiltrative malignant cutaneous spindle cell tumor that commonly presents in younger adults as a slow-growing violaceous plaque or nodule.[7] DFSP may clinically mimic keloids, dermatofibromas, scars, morphea, hemangiomas, sarcomas, and epidermal inclusion cysts, necessitating pathologic confirmation.[8] DFSP is a spindle cell neoplasm that shows an infiltrative growth pattern. The main portion of this neoplasm shows a storiform arrangement with extension into the subcutaneous fat, creating a honeycomb pattern.[9] Expression of CD-34 antigen (human progenitor cell antigen) in DFSP is well described and has been used to support the view that these lesions are variants of nerve sheath tumors, which are distinct from benign fibrous histiocytomas that do not express CD-34.[10] The pathogenesis of DFSP is related to the rearrangement of chromosomes 17 and 22, with the fusion between the COL1A1 and the PDGF b-chain gene.[11] The treatment of choice for DFSP is surgical, including Moh's micrographic surgery, with or without postoperative radiation therapy.[4,5] Imatinib has been approved in the USA and European Union for the treatment of adult patients with unresectable, and/or metastatic DFSP, who are not eligible for surgery.[6,12] Clinical

studies of imatinib have been promising thus far showing partial to complete clinical responses in 14 of 16 treated patients.[13] In summary, physicians should be aware that DFSP can present as a small symmetrical dome shaped nodule and biopsy is critical in establishing the correct diagnosis.

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