

Multiple Eruptive Dermatofibromas Occurring in a Patient with Myelodysplastic Syndrome

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Sir,

Multiple eruptive dermatofibromas (MEDFs) are rare and the pathogenesis is unclear. It has been suggested that the presence of at least 15 dermatofibromas in a patient meets the criteria for MEDF (1). Other authors have defined MEDF as the presence of 5–8 dermatofibromas appearing within a 4-month period (2). MEDFs have been strongly associated with various systemic illnesses involving immunosuppression (2–6). We describe here a man with myelodysplastic syndrome (MDS) who developed MEDFs.

CASE REPORT

A 52-year-old man was referred for multiple pruritic, pink to red papules on the abdomen, back, arms and legs, which had been present for 3 months. Initially, he had several papules on the abdomen, but rapidly developed many similar lesions during the past several

months. He was diagnosed with MDS by a bone marrow biopsy 4 months earlier, and he was not receiving any therapy. Examination revealed approximately 40 non-tender, dome-shaped, pink to red papules ranging from 0.3 to 1.0 cm, on the abdomen, back, arms and legs (Fig. 1). Three biopsies (from the right thigh, left leg and lower abdomen) were performed.

Histopathologically, all three lesions showed similar changes and consisted of a relatively well-circumscribed collection of fibrocytes and benign-appearing histiocytes within the dermis (Fig. 2a). The overlying epidermis was hyperplastic. The histiocytes and fibrocytes were positioned between thickened bundles of collagen (Fig. 2b). Immunohistochemical stains were positive for factor XIIIa and negative for CD34, S100, keratin, leukocyte common antigen and actin. The histopathological and immunohistochemical findings were consistent with dermatofibroma. Within 4 months of follow-up, the lesions persisted, without considerable change in size or number.

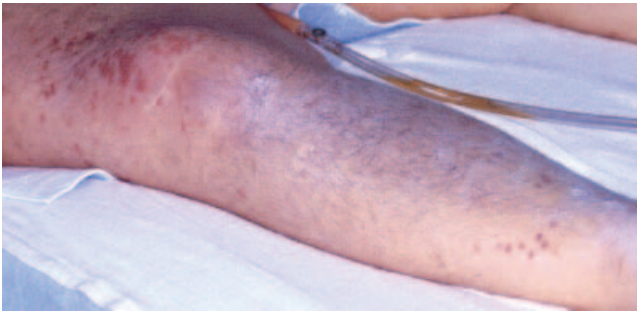


Fig. 1. Clinical appearance at presentation of multiple eruptive dermatofibromas: numerous pink to red papules distributed on the right thigh and leg.

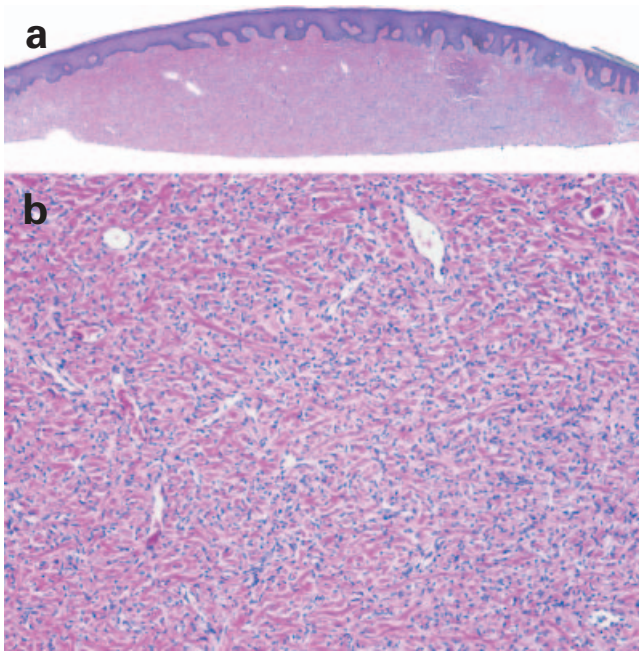


Fig. 2. (a) Epidermal hyperplasia and a dense, diffuse infiltrate of fibrocytes and histiocytes between thickened bundles of collagen within the dermis (haematoxylin and eosin stain, $\times 20$). (b) Benign-appearing fibrocytes and histiocytes positioned between thickened bundles of collagen in the dermis (haematoxylin and eosin stain, $\times 200$).

DISCUSSION

Dermatofibromas are common fibrohistiocytic lesions, usually involving the lower extremities, and present as hyperpigmented papules or nodules. However, MEDFs are very uncommon and have been associated with a variety of immunosuppressive systemic conditions. In 1970, Baraf & Shapiro (1) first described MEDFs occurring in a 39-year-old woman with no underlying systemic diseases. Subsequently, several authors have reported the presence of MEDFs in various immunosuppressive illnesses including HIV infection (2),

systemic lupus erythematosus (4), myasthenia gravis (5) and pemphigus vulgaris (6). Niiyama et al. (7) recently published a comprehensive literature review of MEDFs and noted that the incidence of these lesions was higher in patients with underlying disease compared with patients who were otherwise healthy.

It is generally accepted that haematopoietic disorders result in immunosuppression, but surprisingly, only a single case of MEDFs occurring in a patient with acute myeloid leukemia has been reported. Chang et al. (3) described a 60-year-old man who developed MEDFs on the neck, thigh and trunk after induction of chemotherapy for acute myeloid leukemia. The patient developed these lesions within a period of 2 months.

Myelodysplastic syndrome is characterized by clonal disorders of bone marrow stem cells that lead to ineffective haematopoiesis. They present as quantitative and qualitative defects of haematopoietic stem cells. Disorders of bone marrow clearly produce immunosuppression and alteration in the immunomodulation of haematopoietic cells. We describe a patient with MDS and MEDF, an association which has not been reported previously. The patient was not on immunosuppressive therapy and, with the exception of MDS, did not have other co-existing immunosuppressive diseases. Our case lends further support to the concept that MEDFs may be associated with immunosuppression. Further genetic and molecular studies may be helpful to elucidate the relationship between immunocompromised states and the development of MEDFs.

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