Title
Multiple eruptive dermatofibromas

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Case presentation

Multiple eruptive dermatofibromas

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Abstract

We report a 34 year-old woman with psoriasis, systemic lupus erythematosus (SLE), and recent anti-TNFa therapy, who presented with multiple, eruptive dermatofibromas (MEDF). Although the pathogenesis of MEDF remains unknown, there is substantial evidence that this phenomenon represents an aberrant immune response. Like the more common presentation of solitary dermatofibromas, these lesions are benign, and no treatment is required. However, MEDF is increasingly recognized as a sign of immune dysregulation and an appropriate work-up should be initiated to identify an underlying cause in patients without a known trigger.

Case synopsis

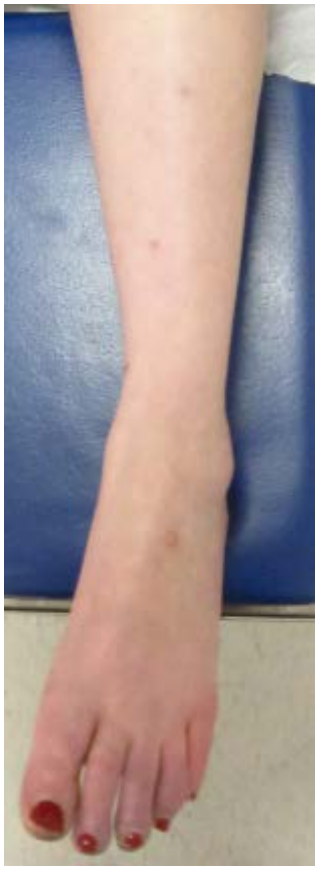
HISTORY: A 32-year-old woman was referred to the Dermatology Clinic at Bellevue Hospital Center for the management of psoriasis. She had previously been treated with several systemic medications, which included methotrexate, adalimumab, and etanercept, without success. Most recently, she had been treated with ustekinumab for four months, which was discontinued in the setting of a severe, allergic reaction. Owing to the failure of multiple medications, her dermatologist recommended that she come to our clinic for further management. Apart from mild scale and pruritus of the scalp, she denied any active, psoriatic lesions. However, she did note a six-month history of eruptive, slightly-tender, hyperpigmented papules on her left lower leg and the dorsal aspect of the left foot. The lesions had appeared abruptly within four to five months of one another.

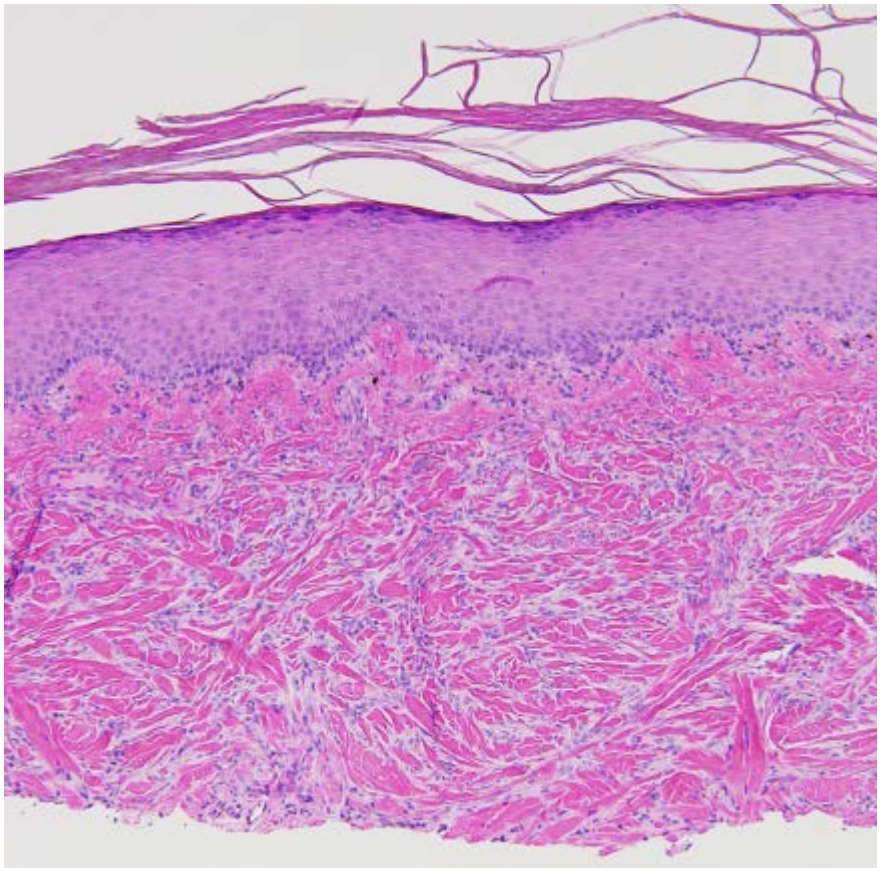
Past medical history included systemic lupus erythematosus for which she was treated with prednisone 5mg daily and was followed by the Rheumatology Service. She denied any systemic symptoms, which included weight loss, fevers, chills, fatigue, joint pain, and gastrointestinal symptoms. A shave biopsy was obtained from a lesion on the dorsal aspect of the left foot.

Physical examination: Six, small, approximately 5-mm, hyperpigmented, papules were scattered on the left lower leg and the dorsal aspect of the foot.

Laboratory data: A complete blood count showed a hemoglobin of 11.5g/mL but was otherwise normal. A metabolic panel was normal.

Histopathology: There is a relatively well-circumscribed dermal lesion that is composed of intersecting fascicles of plump fibroblasts, with thick collagen bundles at its periphery. The overlying epidermis is hyperplastic.





Figures 1. Firm dermal papules 2. Fascicles of plump fibroblasts

Discussion

Diagnosis: Multiple eruptive dermatofibromas

Comment: Dermatofibromas (DFs), which also are known as histiocytomas, are common, benign, skin neoplasms that are characterized by small, firm, hyperpigmented papules or nodules that most commonly occur on the legs of women. DFs usually are solitary and they are not associated with internal disease.

Multiple eruptive dermatofibromas (MEDF) represent a rare variant that is characterized by the sudden development of multiple dermatofibromas. MEDF was first described in a 39-year-old woman with 61 lesions, which were predominantly on her lower legs [1]. Multiple lesions were defined to be greater than 15 [1]. However, this arbitrary criterion of 15 lesions has been the subject of debate. More recently, MEDF has been redefined as the presence of five to eight dermatofibromas that arise in a four-month period [2, 3]. Review of this disease suggests that the rapidity of lesion onset distinguishes it better than the total number of skin lesions [3].

On histopathologic examination, MEDF are indistinguishable from solitary dermatofibromas. Common features include a well-circumscribed, dermal tumor that is comprised of plump, spindle cells with collagen trapping at the outer edges of the lesion. Overlying epidermal hyperplasia also is commonly observed.

MEDF is exceedingly rare and affects only 0.3% of patients [4]. Unlike the common, solitary dermatofibroma, the majority of patients with MEDF have an underlying disease. It is estimated that 66% of patients with MEDF have an underlying immune-mediated disease [5]. The most frequently reported diseases that are associated with MEDF are systemic lupus erythematosus (SLE) and human immunodeficiency virus (HIV) infection. Other associated diseases include Sjögren syndrome, dermatomyositis, myasthenia gravis, pregnancy, mycosis fungoides, and hematologic malignant conditions [3, 5-7]. There are several reports that link immunosuppressant therapy with MEDF. Cases of MEDF in patients receiving cyclophosphamide, methotrexate, glucocorticoids, and azathioprine have been reported [5]. In 2013, the first case of MEDF that was associated with anti-tumor necrosis factor- α (anti-TNF-α) therapy was reported in a patient with rheumatoid arthritis, who was treated with etanercept [8].

To the best of our knowledge, this patient represents the first reported case of MEDF that occurred in a patient with SLE who received anti-TNF-α treatment. Autoimmune diseases and immunosuppressant therapy are well-established associations with MEDF and it is unclear if the SLE, the medication, or the combination contributes to the disease. This patient's history of psoriasis speaks further to her underlying immune dysfunction.

The diagnosis of MEDF should be considered a sign of immune dysregulation. In the absence of a known associated disease an appropriate work-up should be initiated to rule out a connective-tissue diseases, autoimmune disorders, hematologic malignant conditions, or HIV infection.

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