

Multiple eruptive dermatofibromas in an adolescent with a history of pityriasis lichenoides et varioliformis acuta



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INTRODUCTION

Dermatofibromas are dermal growths that present as hyperpigmented papules or nodules, most commonly between the third and fourth decade of life, and are often found on the legs. Multiple eruptive dermatofibromas (MEDF) are typically associated with underlying systemic disease, such as systemic lupus erythematosus, HIV, renal transplant, and hematologic malignancy, and appear more commonly in adults than they do in children.¹ We present a case of eruptive dermatofibroma in a 14-year-old boy occurring after pityriasis lichenoides et varioliformis acuta (PLEVA). To our knowledge, this is the first case of eruptive dermatofibromas reported in either an adolescent or occurring secondary to PLEVA.

CASE REPORT

A 14-year-old boy with a history of Factor VIII deficiency and alopecia areata presented with a new eruption consisting of small erythematous papules on the trunk and proximal parts of the extremities (Fig 1), some with crusting. Biopsy of these lesions showed superficial and deep perivascular and patchy band-like infiltrates in the papillary dermis predominantly of lymphocytes with extension into a hyperplastic epidermis, where spongiosis, scattered necrotic keratinocytes, pallor of the upper portion of the epidermis, and mounds of parakeratosis with neutrophils were observed (Fig 2). These histopathologic features were consistent with the diagnosis of PLEVA. He was treated with a 6-week course of doxycycline 100 mg twice daily, and the eruption resolved. Two months later, he returned with a complaint of “scarring” from the former eruption.

Abbreviations used:

MEDF: multiple eruptive dermatofibromas
PLEVA: pityriasis lichenoides et varioliformis acuta

He was noted to have 5 dark-brown, circular, barely elevated papules on his legs at the sites of prior PLEVA lesions (Figs 3 and 4). White, wavy, sclerotic centers were prominent on dermatoscopy (Fig 4). Histopathologic examination of a 4-mm punch biopsy taken from one of the lesions showed proliferation of cytologically bland, spindled cells within the superficial dermis, many of which were reactive for Factor XIIIa, consistent with a dermatofibroma (Fig 4). Due to the patient's age and grossly negative review of systems, no further work up for systemic disease was performed.

DISCUSSION

This case of MEDF in an adolescent following PLEVA suggests that this is a process that can occur in pediatric patients and be triggered by PLEVA. MEDF is defined by the presence of >5-8 dermatofibromas appearing within a period of 4 months.² It has been proposed that a dermatofibroma represents an abortive immunoreactive process triggered by dermal dendritic cells that cannot be cleared by the dampened immune system.³ It has been hypothesized that the downregulation of T-regulatory cells leads to exaggerated immune responses in other parts of the body such as the skin. This theory could be further supported by the association between MEDF and underlying

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Fig 1. Clinical photos of multiple eruptive, brown papules on the flexural aspects of the legs and arms bilaterally, some with necrotic centers.

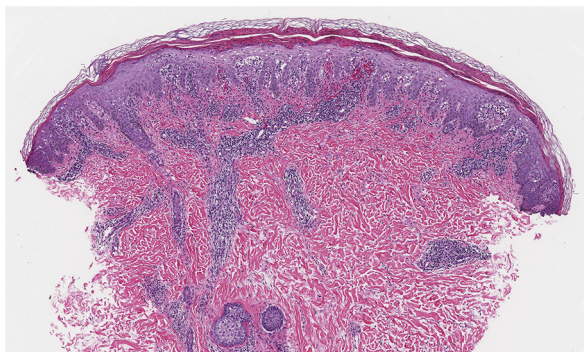


Fig 2. Histopathology of initial pityriasis lichenoides et varioliformis acuta demonstrating superficial and deep perivascular and patchy band-like infiltrates in the papillary dermis with extension into epidermis and mounds of parakeratosis with neutrophils (Hematoxylin-eosin stain; original magnification: $\times 40$).

systemic lupus erythematosus, HIV infection, renal transplant, myasthenia gravis, and pemphigus vulgaris. According to a review, more than 80% of MEDF-associated diseases were immune-mediated, suggesting that this cutaneous finding could be considered a manifestation of immune-mediated disease.⁴ While the pathogenesis of solitary dermatofibromas remains unknown, an aberrant response to minor trauma is suspected. We do not know why dermatofibromas arose in some but

not all prior PLEVA lesions. Perhaps the inflammation and skin damage was greater in those lesions than in others.

While the cause of PLEVA is unknown, it is thought that the pathogenesis involves the activation of immune response either by an infectious agent or a self-limited T-cell lymphoproliferative process. Interestingly, an association between PLEVA and autoimmune diseases has been reported, including autoimmune hepatitis,⁵ alopecia areata,⁶ periodic fever, aphthous stomatitis, pharyngitis, adenitis syndrome,⁷ common variable immunodeficiency syndrome,⁸ and idiopathic thrombocytopenia.⁹ Of note, our patient also had a history of alopecia areata. This association suggests that immune dysregulation may play a role in the pathogenesis of PLEVA. While the underlying pathogenesis of PLEVA is not completely understood, the development of MEDF post-PLEVA in our patient may point to further evidence that immune dysregulation may be present in patients with pityriasis lichenoides. It is possible that suppression of T-regulatory cells may contribute to PLEVA and MEDF.

Conflicts of interest

Dr Orlov serves on the board of 2 companies, Almirall and R2 Technologies, neither of which is involved in the development of products for PLEVA or dermatofibromas.



Fig 3. Eruptive dermatofibromas (arrows, left) corresponded to previous pityriasis lichenoides et varioliformis acuta lesions (arrows, right).

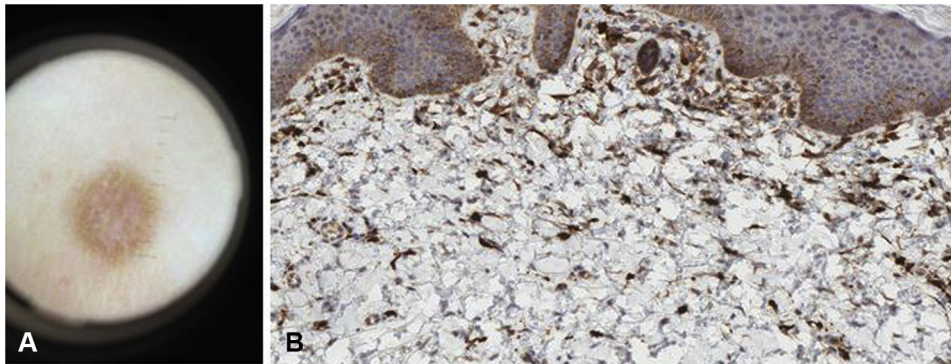


Fig 4. Dermatoscopic (A) and histopathologic (B) features of dermatofibromas. A, White, wavy, sclerotic centers were prominent on dermatoscopy. B, Factor XIIIa-positive spindle cells were present in the dermis (original magnification: $\times 200$).

Drs Haber and Meehan have no conflicts of interest to declare.

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