Diffuse Red Eruptions in a Young Boy

Oliver Wisco, DO[1] ▪ Tatiana Abrantes, BS[2] ▪ Allison Robbins, MD[3]

A 10-year-old boy presented to our clinic with his parents with a 6-month history of a diffuse rash on his trunk and extremities (Figures 1-3). The rash had developed abruptly, and individual lesions were characterized as pruritic and occasionally burned.

Prior to presentation at our clinic, previous attempts to treat the rash included erythromycin, 250 mg, 3 times daily for 4 weeks, without success, followed by clobetasol, 0.05%, ointment daily for 2 weeks. Despite treatment, new lesions continued to arise. The patient was otherwise healthy.

Upon physical examination, the patient was sitting comfortably, without persistently scratching his lesions. Diffusely scattered across his entire body were reddish-brown papules and small plaques with overlying fine micaceous scale and scattered crust.

Based on the patient's history and physical examination findings, which one of the following is the most likely diagnosis?

A. Varicella
B. Pityriasis lichenoides et varioliformis acuta
C. Lymphomatoid papulosis
D. Guttate psoriasis
E. Pityriasis rosea

Correct answer: B. Pityriasis lichenoides et varioliformis acuta (PLEVA)

Discussion

PLEVA, also known as Mucha-Habermann disease, is a benign and uncommon cutaneous inflammatory disorder that is part of the disease spectrum of pityriasis lichenoides chronica.[1] PLEVA can occur in people of all ages, but it tends to predominate in boys from late childhood to early adulthood.[2] While the etiology is unknown, it has been hypothesized that PLEVA may be an inflammatory reaction triggered by foreign agents such as infections (eg, HIV, parvovirus B19) or medications (eg, estrogen-progesterone, tumor necrosis factor-α inhibitors, statins).[1] It has also been postulated that PLEVA could be an inflammatory response to a T-cell dyscrasia or an immune complex mediated hypersensitivity.[3] Febrile ulceronecrotic Mucha-Habermann disease (FUMHD) is a rare, severe, and potentially life-threatening variant of PLEVA, with a predominate T-cell infiltrate.[1,4] Although the exact etiology is unknown, FUMHD has been regarded as a low-grade lymphoproliferative disorder.

PLEVA presents as an acute eruption of bright red inflammatory papules that rapidly evolve into small blisters and pustules, which eventually ulcerate and develop necrotic crusts.[1,3,4] The lesions can appear individually or grouped and most commonly occur on the trunk, thighs, upper arms, and skin flexures. Mucosal sites are typically spared.[4] PLEVA may be asymptomatic or associated with mild pruritus, burning, and rarely, fevers or arthralgias. Because of its relapsing and remitting nature, all stages of lesions may be represented on physical examination.

While a strong clinical suspicion is imperative, a skin biopsy allows for clinicopathological correlation to confirm the diagnosis of PLEVA and eliminate the differential diagnosis (Table). Although there are no pathognomonic histologic findings associated with PLEVA, common features such as vacuolar interface dermatitis and erythrocyte extravasation aid in the diagnosis.[3,4]

AFFILIATIONS:

[1]Director of Cutaneous Oncology, Mohs Surgeon, Department of Dermatology, The Warren Alpert Medical School of Brown University, Rhode Island

[2]Medical Student, The Warren Alpert Medical School of Brown University, Providence, Rhode Island

[3]Resident Physician, Dermatology Department, The Warren Alpert Medical School of Brown University, Rhode Island

CITATION:

Wisco O, Abrantes T, Robbins A. Diffuse red eruptions in a young boy. *Consultant*. Published online November 19, 2021. doi:10.25270/con.2021.11.00009

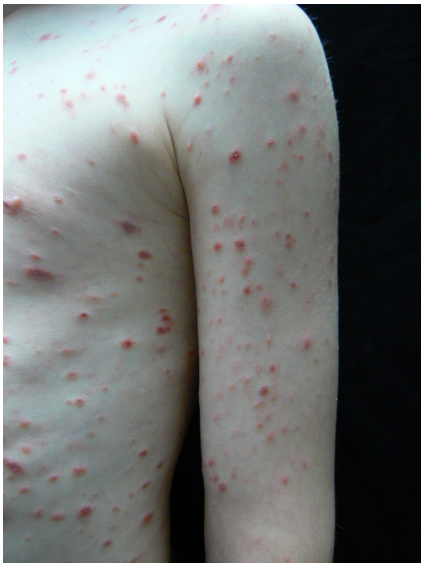
Received July 5, 2021. Accepted July 9, 2021.

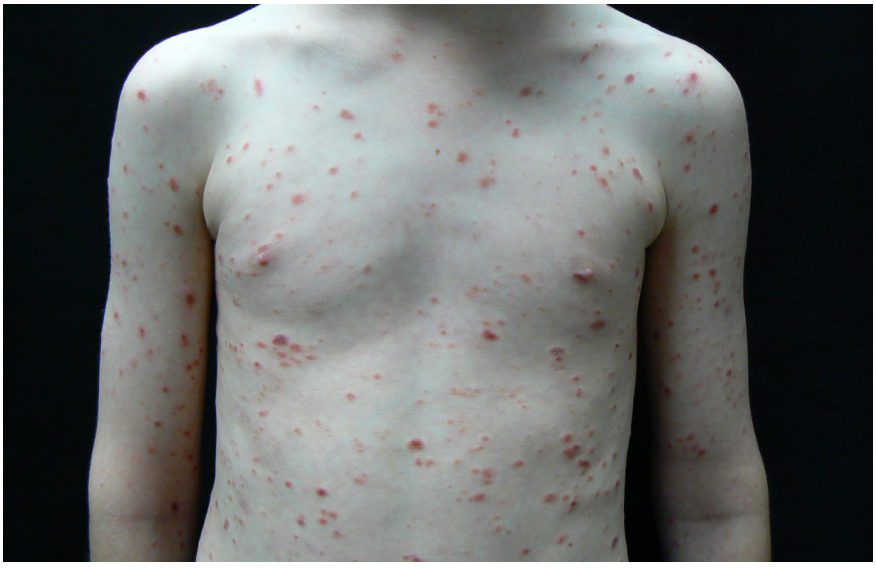
DISCLOSURES:

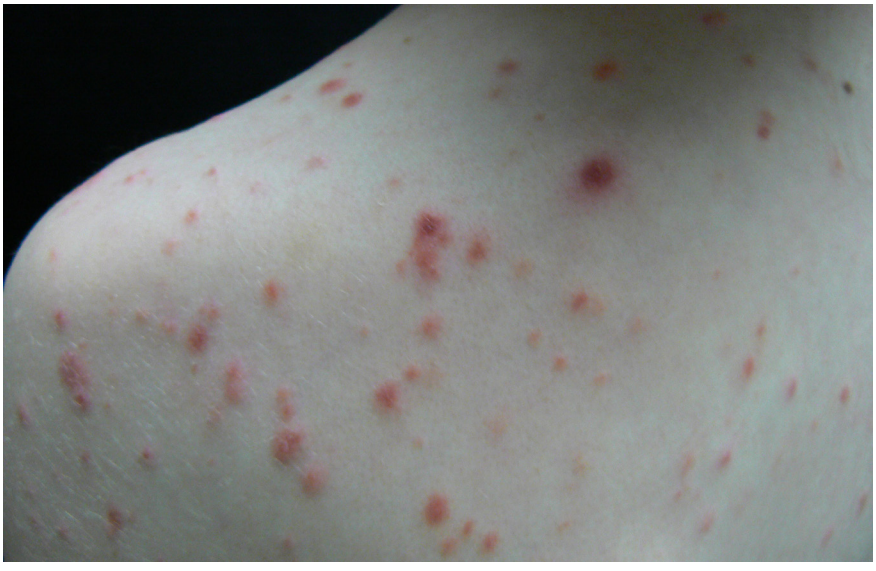
The authors report no relevant financial relationships.

CORRESPONDENCE:

Oliver J. Wisco, DO, FAAD, FACMS, The Warren Alpert Medical School of Brown University, 593 Eddy Street, APC, 10th Floor, Providence RI 02903 (Oliver_wisco@brown.edu)







Figures 1-3.

Because PLEVA is a self-limiting disorder, it will resolve without treatment but can last weeks to years. A course of antibiotics, such as erythromycin or tetracycline, and phototherapy are considered first-line treatments that can hasten the resolution of lesions. Topical corticosteroids may also be used to relieve symptoms, as well as methotrexate for patients who have failed previous treatment regimens.[1,5,6]

Varicella, though also an acute development of papulovesicles on multiple areas of skin, often presents as lesions that tend to be described as "dewdrop on a rose petal." The course of disease is shorter than PLEVA, usually lasting only 1 to 2 weeks.[7]

Lymphomatoid papulosis most closely resembles PLEVA. While the cutaneous lesions are similar to lesions seen with PLEVA, the histologic features distinguish the two. Lymphomatoid papulosis is characterized by a dermal infiltrate composed of large atypical lymphocytic blast cells that are CD30+, which are rare in PLEVA. PLEVA also occurs more often in younger patients, is relatively short-lived, and rarely, if ever, progresses to lymphoma.[8]

Like PLEVA, guttate psoriasis most often presents in children and young adults and can be asymptomatic, pruritic, or painful. Unlike PLEVA, however, guttate psoriasis lesions emerge abruptly, usually 1 to 2 weeks after a streptococcal infection. The lesions are small, round, or oval erythematous plaques with a silvery scale. Furthermore, guttate lesions are often symmetrically distributed over the trunk and proximal extremities, differing from the pattern of presentation of PLEVA.[9]

Pityriasis rosea can mimic PLEVA in its early presentation. While PLEVA is more papulonecrotic with more persistent and recurrent lesions, pityriasis rosea is composed of scaly patches or plaques that tend to follow Langer lines.[10]

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Table. Selected Differential Diagnosis of Pityriasis Lichenoides et Varioliformis Acuta[7-10]

CONDITION	CHARACTERISTICS
Varicella	Vesicular pruritic rash that begins as macules and rapidly becomes papules, followed by vesicles with "dewdrop on a rose petal" appearance
	Most lesions crust by day 6
PLEVA	Multiple erythematous macules that rapidly evolve to form inflammatory papules and vesicles, eventually developing necrotic crusts
	Most commonly occurs on the trunk, thighs, upper arms, and skin flexures
Lymphomatoid papulosis	Early grouped lesions are small, erythematous or violaceous papules that evolve to larger papules or nodules that may develop central hemorrhage, necrosis and crusting
	Histological features include CD30+ lymphoid proliferation of atypical T blast cells
Guttate psoriasis	Numerous small, inflammatory, erythematous plaques with a silvery scale
	Found predominantly on the trunk and extremities
	Most common among children and young adults with a recent streptococcal infection
Pityriasis rosea	Well-circumscribed, pink, oval, papulosquamous lesions
	Found on the trunk and proximal areas of extremities
	Scaling "herald patch" with collarette of scale is followed by several days to weeks of smaller lesions along Langer lines

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