A photodistributed eruption in an immunosuppressed patient

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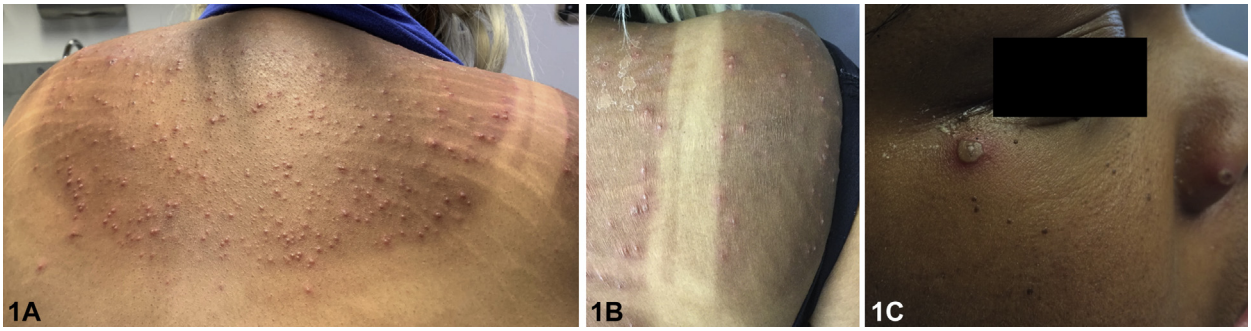
Key words: exanthem; immunosuppression; photodistributed; varicella.



A 30-year-old African American woman with a history of hidradenitis suppurativa, who was taking ustekinumab (one 90-mg subcutaneous loading dose, 1 month previously) and was 2 weeks into a prednisone taper (week 1, 40 mg daily; week 2, 30 mg daily), presented with a tender, pruritic eruption that began 5 days previously on sun-exposed areas of the shoulders then spread to her face, abdomen, and left leg. The patient discontinued prednisone 1 day after the eruption appeared. Systemic symptoms included sore throat and myalgia. She reported having had contact with her father, who had been diagnosed with zoster 2 weeks earlier. The patient had never received the varicella-zoster virus (VZV) vaccine and had not had chickenpox as a child. Pertinent physical examination findings included pustules on an erythematous base confined to the suntanned trunk (Fig 1, *A*) and left side of the shoulder (Fig 1, *B*), with scattered lesions on the face (Fig 1, *C*). Covered areas were notably spared.

Question 1: What is the most likely diagnosis?

A. Photolocalized varicella

B. Phototoxic drug reaction

C. Porphyria cutanea tarda

D. Polymorphous light eruption (PMLE)

E. Subacute cutaneous lupus erythematosus

Answers:

A. Photolocalized varicella — Correct. Varicella is a common viral infection with a characteristic exanthem. Although viral rashes aggravated by sun exposure, trauma, and inflammation have been described, photolocalized varicella is rare, and the typical distribution may not be seen.[1] As with our patient, a mild viral prodrome or

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Funding sources: None.

Accepted as a poster presentation at the American Academy of Dermatology 2019 Annual Meeting, which was then cancelled due to the COVID-19 pandemic. Subsequently, an abstract accompanying this presentation was published in JAAD (*J Am Acad Dermatol* 2020; 83: supplement AB182) on December 1, 2020.

IRB approval status: Not applicable.

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JAAD Case Reports 2021;15:19-21.
2352-5126

https://doi.org/10.1016/j.jdcr.2021.04.038

constitutional symptoms are often reported. This case highlights the importance of including varicella in the differential diagnosis of a photolocalized eruption, especially in a patient with a known history of exposure and immunosuppression.

B. Phototoxic drug reaction — Incorrect. Phototoxic drug reactions present as more similar to a sunburn than to a pruritic eruption. Commonly reported etiologic drugs include vemurafenib, voriconazole, doxycycline, hydrochlorothiazide, amiodarone, and chlorpromazine.[2] This patient was taking only ustekinumab and prednisone.

C. Porphyria cutanea tarda — Incorrect. Porphyria cutanea tarda is a disorder of porphyrin accumulation and is characterized by painful vesicles that develop on sun-exposed areas of the skin, most commonly the hands and face.[3] Most cases are due to acquired or inherited uroporphyrinogen decarboxylase deficiency.[3]

D. PMLE — Incorrect. PMLE is an idiopathic photodermatosis that can present with a wide range of morphologies.[4] PMLE can affect any skin type but is more commonly reported in lighter skin types. Systemic symptoms are rare in PMLE.[4]

E. Subacute cutaneous lupus erythematosus — Incorrect. Subacute cutaneous lupus erythematosus is a subtype of cutaneous lupus erythematosus that usually presents symmetrically as an annular or papulosquamous eruption distributed on the sun-exposed sites of the skin.

Question 2: Which of the following is the most appropriate next step in the diagnostic approach for this disease?

A. Autoantibody profile

B. Biochemical porphyrin profile

C. History and physical

D. Skin biopsy

E. VZV polymerase chain reaction

Answers:

A. Autoantibody profile — Incorrect. An autoantibody profile would be more useful for the diagnosis of connective tissue disease when there is clinical suspicion. Specifically, autoantibodies are frequently associated with cutaneous lupus erythematosus. Depending on the review of systems findings and clinical systemic symptoms, further laboratory testing may be indicated.

B. Biochemical porphyrin profile — Incorrect. The diagnosis of porphyria cutanea tarda is often confirmed by obtaining a biochemical porphyrin profile. In porphyria cutanea tarda, urinary porphyrins and plasma porphyrins are elevated. Fecal porphyrins may be elevated as well. Urinary excretion of porphobilinogen is normal, differentiating porphyria cutanea tarda from other porphyrias.[3]

C. History and physical — Incorrect. Although a thorough history and physical is an important method for any diagnosis, this patient's atypical presentation and the rarity of this diagnosis required further laboratory confirmation.

D. Skin biopsy — Incorrect. Although skin biopsy can be utilized to make the correct diagnosis, a noninvasive diagnostic technique would be preferred as the next diagnostic step. On histopathologic examination, varicella will show multinucleated giant cells[5] with nuclear molding, cytopathic effect, and mild leukocytoclastic vasculitis.

E. VZV polymerase chain reaction — Correct. This patient's diagnosis was confirmed by polymerase chain reaction to detect VZV. Serologic techniques can also be used to diagnose VZV, with active infection revealing elevated IgM titers. Studies have demonstrated that immunologic techniques and molecular amplification are superior for VZV detection when compared with serologic techniques.[5]

Question 3: What is the best treatment for this patient?

A. Drug discontinuation

B. Hydroxychloroquine

C. Immediate initiation of valacyclovir

D. Phlebotomy

E. Supportive care

Answers:

A. Drug discontinuation — Incorrect. In phototoxic drug reactions, the mainstay of treatment is the identification and cessation of the causative agent. In cases where discontinuation is not possible, patients should be instructed to avoid the sun and practice strict sun protection.[2]

B. Hydroxychloroquine — Incorrect. Antimalarials such as hydroxychloroquine, chloroquine, and quinacrine are the first-line systemic therapy for cutaneous lupus erythematosus. Systemic medications are recommended in cases of cutaneous lupus erythematosus that are severe, widespread, or refractory to topical therapies.

C. Immediate initiation of valacyclovir — Correct. The treatment of choice for VZV infection in individuals at a greater risk for complications, including adults and immunocompromised patients, is antiviral therapy. The most commonly used antiviral agents are acyclovir and valacyclovir, which function by inhibiting viral DNA polymerase and subsequently VZV replication. Our patient was treated with 1 gram of valacyclovir 3 times daily for 14 days and reported significant improvement after completion of the treatment course.

D. Phlebotomy — Incorrect. The standard treatment for patients with porphyria cutanea tarda is regular phlebotomy to reduce iron and porphyrin levels in the liver.[3]

E. Supportive care — Incorrect. Varicella has a low complication rate, especially in children, and can be treated symptomatically. However, in immunosuppressed patients, antivirals should be promptly initiated to minimize the risk of complications.

Abbreviations used:

PMLE: polymorphous light eruption
VZV: varicella-zoster virus

Conflicts of interest

None disclosed.

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