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Clinico-Pathological Correlation in Dermatopathology

A 67-Year-Old Male with Diffuse Purpuric Vesicles and Bullae

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Vasculitis · Leukocytoclastic vasculitis · Purpuric macules

Abstract
Leukocytoclastic vasculitis (LCV) is a small-vessel vasculitis that most commonly affects the postcapillary venules in the skin. It classically presents with purpuric macules that progress to palpable purpura on the bilateral shins 7–10 days after an inciting medication or infection, or in the setting of connective tissue disease, malignancy, or inflammatory bowel disease. Up to 50% of cases have no identifiable cause. Lesions on the buttocks, abdomen, upper extremities, and face are uncommon, as are bullae and ulcers. We present a rare case of bullous LCV manifesting as grouped vesicles on the face and body mimicking varicella-zoster infection.

Case Report

A 67-year-old male with a history of cerebrovascular accident, hypertension, gout, and obesity presented with fever, a diffuse blistering rash, and altered mental status over the previous week. The patient was seen by his primary care physician for a painful lesion on his left leg and started on cephalexin for presumed cellulitis 7 days prior to presentation. On the day of cephalexin initiation, pink patches developed on the left foot, calf, and shin, and spread to the torso and other extremities. The patches subsequently blistered and became eroded.

On presentation, the patient was febrile to 101.9°F, tachycardic to the 120s, and oriented only to self. Physical examination revealed numerous vesicles and bullae approximately 1 cm in diameter over purpuric patches that coalesced over the face, torso, and extremities (Fig. 1).

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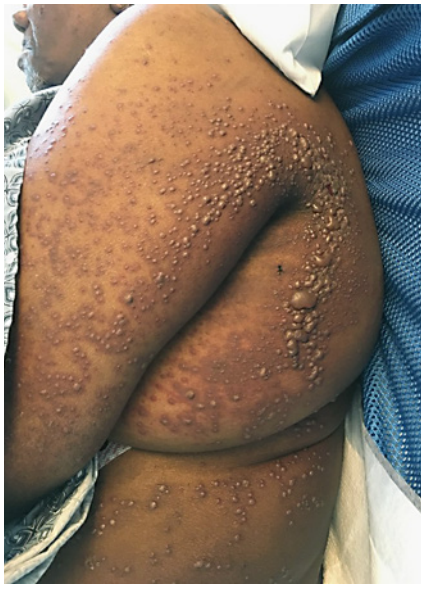

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Fig. 1. Left extensor arm and flank with tense vesicles and bullae on patches of purpura.
Fig. 2. Bilateral dorsal feet with large eroded bullae with surrounding purpura.
Fig. 3. Grouped gray vesicles with hemorrhagic crusting on an erythematous base on the bilateral cheeks.

The bilateral legs had larger bullae, and the bilateral dorsal feet had large well-circumscribed erosions without purulence (Fig. 2). There were also multiple punched-out erosions with hemorrhagic crust on the bilateral cheeks (Fig. 3).

The patient has no known drug allergies. Other than cephalexin, furosemide had been started 6 days prior to presentation. Review of systems was otherwise negative. Initial laboratory evaluation was notable for leukocytosis to 17,800 (81% neutrophils; 0.2% eosinophils). A flu swab was negative, lumbar puncture was negative for meningitis, and chest X-ray and CT of the head were unremarkable. The differential diagnosis was broad, including bacterial soft tissue infection, Stevens-Johnson syndrome, disseminated herpes simplex virus/varicella-zoster virus, IgA vasculitis, or another primary autoimmune bullous rash.

Diagnosis and Clinical Course

Punch biopsies were obtained from the back and arm, and histological examination revealed bullous leukocytoclastic vasculitis (LCV) with abundant neutrophils and no viral cytopathic changes, negative varicella-zoster virus and herpes simplex virus 1/2 stains, and no microorganisms detected on Brown Hopps, GMS or PAS/D stains (Fig. 4, 5).

The patient was started on dapsone 50 mg daily for an antineutrophil effect on hospital day 2. Intravenous methylprednisolone 150 mg daily was started on hospital day 5 due to concern for systemic (particularly renal and CNS) involvement given his rising creatinine, hematuria, and persistent altered mental status. On a combination of dapsone and steroids, the lesions on the face, trunk, and extremities crusted over, and no new lesions developed. His altered mental status and acute kidney injury also resolved. Intravenous methylprednisolone was tapered and weaned over a 10-day period. Given clinical improvement, the patient was trialed off all systemic treatment, including dapsone, starting on hospital day 22, with no recurrence of vasculitis.

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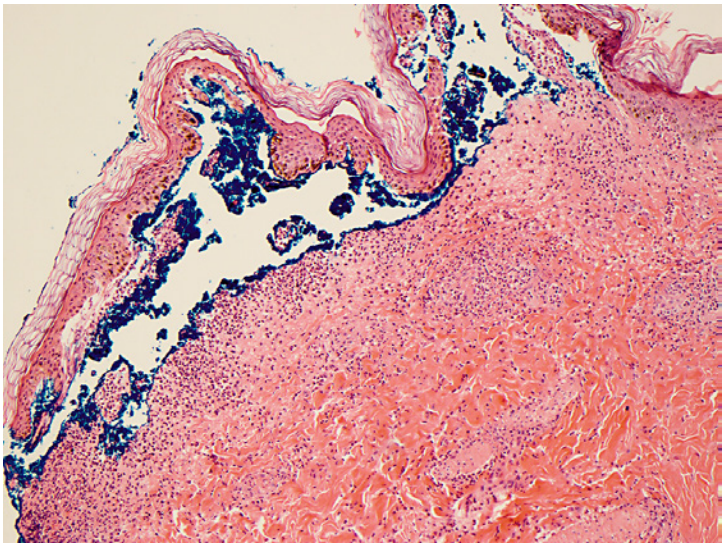

Fig. 4. Low-power histological examination showed a subepidermal blister with abundant neutrophils in the papillary dermis and destruction of small vessels in the superficial reticular dermis.

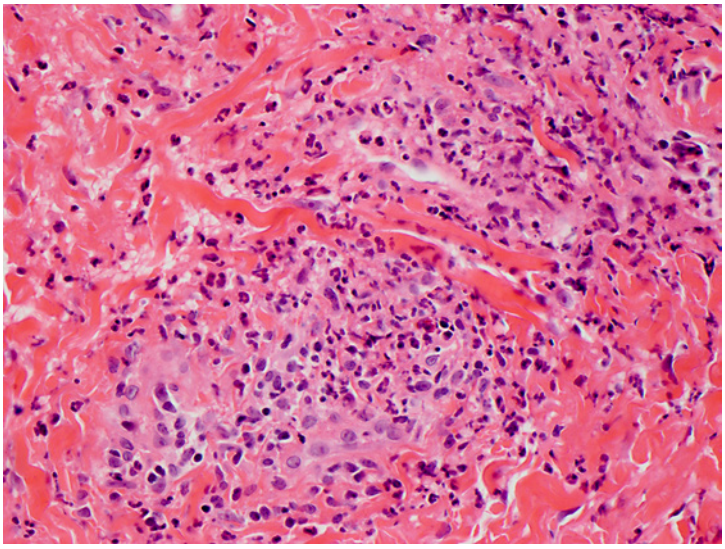

Fig. 5. High-power histological examination showed small vessel destruction with fibrin deposition, leukocytoclasia, and erythrocyte extravasation consistent with leukocytoclastic vasculitis.

Discussion

Cutaneous LCV is characterized by inflammation of the small-vessel walls by infiltrating neutrophils. It affects 30 million individuals per year with equal distribution between men and women. While the typical presentation is palpable purpura on the lower extremities, other morphologies may include ulcers, pustules, and vesicles.

Our case illustrates an unusual presentation of LCV consisting of vesicles and bullae throughout the bilateral lower extremities, upper extremities, torso, and face without clear autoimmune, rheumatological, or pharmacological etiology. To our knowledge, this is the first case of bullous LCV in the literature that notes a presentation of grouped vesicles on the face mimicking varicella-zoster virus. Previous reports of bullous LCV have linked the blistering condition to bullous pemphigoid, systemic lupus erythematosus, orlistat, ibuprofen, flu vaccine administration, and even exercise [1–7]. Our patient has no history of autoimmune or rheumatological condition, but initiation of cephalexin and furosemide 1 week prior to presentation may be inciting factors.

This case also demonstrates the importance of distinguishing skin-limited LCV from systemic vasculitis. The latter should be suspected if systemic symptoms develop, such as fever, myalgias, malaise, lymphadenopathy, abdominal pain, melena, hematochezia, diarrhea, hematuria, lower extremity swelling, or paresthesias. CNS vasculitis should be suspected given neurological symptoms such as headache, vision changes, worsening mental status, or difficulty with movement, coordination, or sensation.

Treatment of LCV depends on the level of systemic involvement. Patients with skin-limited disease may be observed or treated with supportive measures such as rest and elevation of the extremities, compression stockings, nonsteroidal anti-inflammatory drugs for pain, and antihistamines for pruritus. Patients with ulcerative lesions or end-organ involvement may require corticosteroids, anti-inflammatory agents, and immunosuppressive agents. Our case highlights the effective combination of low-dose dapsone with intravenous methylprednisolone in treating suspected systemic vasculitis with a relatively rapid taper resulting in sustained remission.

Statement of Ethics

The manuscript was prepared in compliance with all ethical and confidentiality guidelines and principles.

Disclosure Statement

Zizi Yu declares that she has no conflict of interest.
Yun Xue declares that she has no conflict of interest.
Ruth Foreman declares that she has no conflict of interest.
Daniela Kroshinsky declarses that she has no conflict of interest.

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