Unusual purpuric eruption in a child

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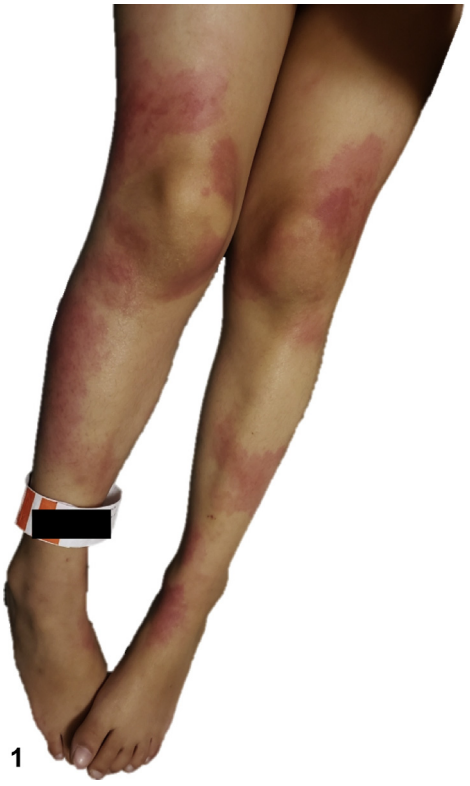

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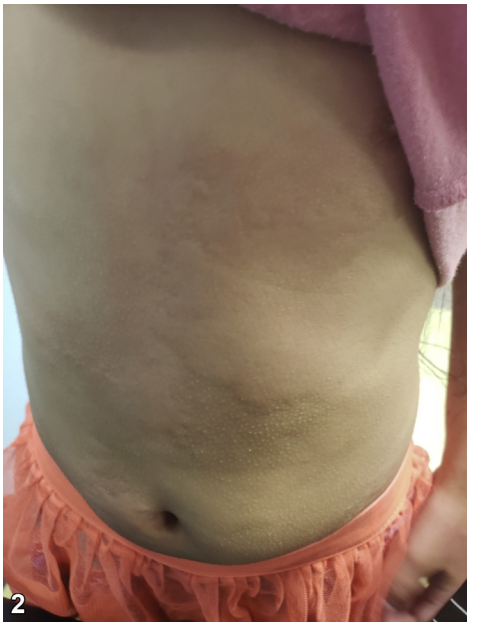

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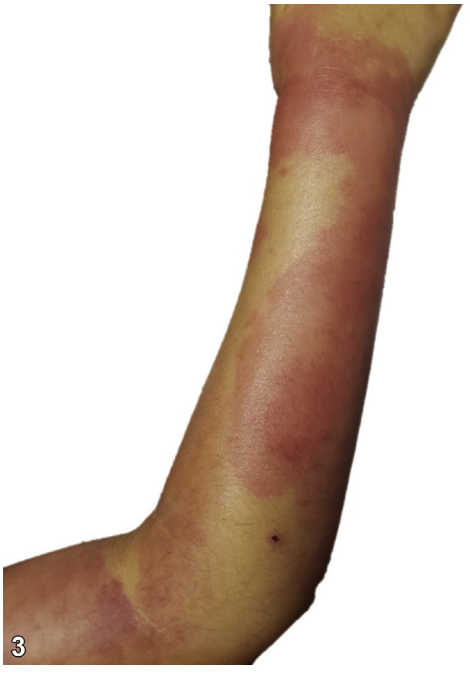

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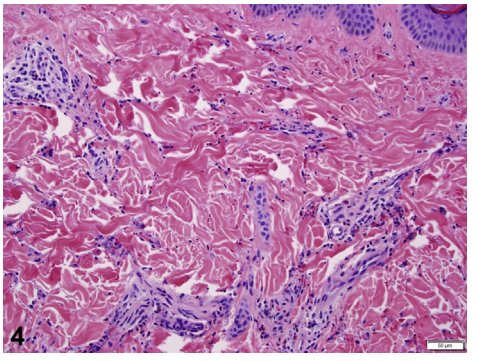

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A 5-year-old, previously healthy girl presented with new-onset, persistent, red, burning hives covering large segments of her body. Associated symptoms included angioedema, shortness of breath, and diarrhea. The lesions resolved after 24 hours, leaving behind areas of bruising. On physical examination, the patient appeared well, with scattered, variably sized, purpuric patches and urticarial plaques on the torso and extremities (Figs 1-3). Laboratory evaluation revealed leukocytosis, elevated erythrocyte sedimentation rate, normal C1q antibody levels, and decreased C4 levels. Two weeks after the initial presentation, she developed the same purpuric eruption accompanied by acute hypoxic respiratory failure.

Question 1: What is the most likely diagnosis?

A. Urticaria multiforme

B. Urticarial vasculitis (UV)

C. Erythema multiforme

D. Henoch-Schönlein purpura (HSP) (IgA-mediated vasculitis)

E. Hereditary angioedema

Answers:

A. Urticaria multiforme — Incorrect. Urticaria multiforme typically presents in a child with large, circinate or targetoid, transient urticarial plaques, with or without angioedema. Urticaria multiforme lesions self-resolve in <24 hours with no residual pigmentation, and there is often a history of viral illness or medication usage in the days preceding development.

B. Urticarial vasculitis (UV) — Correct. UV is a subtype of cutaneous small-vessel vasculitis (Fig 4), which is uncommon in pediatric patients. Compared with lesions found in urticaria multiforme, those of UV typically last longer than 24 hours, can leave behind residual hemorrhage or hyperpigmentation, and are accompanied by burning pain and/or pruritus.

C. Erythema multiforme — Incorrect. Erythema multiforme generally arises as a hypersensitivity reaction to infection or medications. Lesions classically present as targetoid, well-demarcated, fixed red macules to papules with central necrosis and an edematous, pale surrounding ring.

D. Henoch-Schönlein purpura (HSP) (IgA-mediated vasculitis) — Incorrect. HSP is the most common multisystem vasculitis of childhood. Classic HSP presents as a triad of palpable purpura often of the lower extremities, abdominal pain, and arthritis. Although leukocytoclastic vasculitis is demonstrated on biopsy, IgA vessel deposition is found in 75% of cases of HSP, which is rare in UV.[1]

E. Hereditary angioedema — Incorrect. Hereditary angioedema is an autosomal dominant disease caused by a deficiency in C1-inhibitor protein. It presents with episodes of nonpruritic facial and peripheral edema that resolve over 48 to 72 hours. Patients may have prodromal erythema marginatum, which appears as a nonpitting, nonpruritic serpiginous rash that may manifest independently of angioedema.

Question 2: Which of the following is *not* a common systemic manifestation of hypocomplementemic urticarial vasculitis syndrome (HUVS)?

A. Chronic obstructive pulmonary disease

B. Glomerulonephritis

C. Encephalopathy

D. Uveitis

E. Arthritis

Answers:

A. Chronic obstructive pulmonary disease — Incorrect. Lung involvement is common in patients with HUVS and has been reported in between 20% and 65% of these patients. Among the most common manifestations of lung involvement are chronic obstructive pulmonary disease and pleural effusions.[2,3]

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B. Glomerulonephritis — Incorrect. Renal involvement occurs in up to 50% of patients with HUVS. Patients may present with proteinuria and hematuria and reveal various forms of glomerulonephritis on renal biopsy histologic findings.[3]

C. Encephalopathy — Correct. The nervous system is very rarely involved in HUVS. The few reports describing central or peripheral nervous system complications of HUVS include pseudotumor cerebri, aseptic meningitis, and neuropathies.[2]

D. Uveitis — Incorrect. Up to 30% of patients with HUVS have ocular involvement, and although uveitis is the most common of these manifestations, there have also been reports of associated conjunctivitis and episcleritis.[3]

E. Arthritis — Incorrect. Joint involvement is the most common systemic manifestation of HUVS, with reports describing between 50% and 70% of patients experiencing some arthralgia or arthritis.[2,3]

Question 3: What is the preferred first-line treatment for mild cases of UV?

A. Omalizumab

B. Glucocorticoids plus antihistamines

C. Azathioprine

D. Colchicine

E. Rituximab

Answers:

A. Omalizumab — Incorrect. Although successful treatment of UV with biologics, including omalizumab, has been reported in multiple case studies, this treatment has not been robustly studied in the context of UV and is less cost-effective than other treatments.

B. Glucocorticoids plus antihistamines—Correct. Corticosteroids downregulate the inflammatory response and clear deposited immune complexes linked to the pathogenesis of the disease. They have been shown to be effective for the treatment of cutaneous findings of UV in 80% of patients. Antihistamines resulted in resolution of symptoms in less than 30% of patients, but given their low side effect profile and cost, they are commonly added to the treatment regimen.[4]

C. Azathioprine — Incorrect. Azathioprine and other immunosuppressive agents are typically reserved for severe cases or cases refractory to first-line agents.[5]

D. Colchicine — Incorrect. Colchicine is usually reserved for refractory cases. The addition of colchicine to otherwise ineffective corticosteroid monotherapy has been shown to lead to clinical remission of UV.

E. Rituximab — Incorrect. Rituximab is a monoclonal antibody against the CD20 antigen on B cells. This systemic immunosuppressive has a more severe side effect profile and is not a cost-effective method of treatment in mild UV. It has been reported as an effective treatment in severe, refractory, or relapsing cases of HUVS.[5]

Conflicts of interest

None disclosed.

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