

中文題目：以血疱作為罕見表現的成人型 Henoch-Schönlein 紫斑

英文題目：Hemorrhagic bullae as a rare presentation of Adult-onset Henoch-Schönlein purpura

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Introduction:

Henoch-Schönlein purpura(HSP) is a systemic immunoglobulin(Ig)A-mediated small-vessel vasculitis, characterized by the clinical tetrad of palpable purpura, arthralgia or arthritis, abdominal pain and renal disease. It is the most common vasculitis in children, but is unusual in adults with a more severe course and a worse renal outcome. We reported a case with adult-onset Henoch- Schönlein purpura presenting with hemorrhagic bullae.

Case report:

A 48-year-old female with history of type II diabetes mellitus and hypertension, presented with bilateral lower leg reddish swelling for 5 days. Laboratory tests showed leukocytosis, elevated C-reactive protein and liver enzymes, but normal renal function. New non-tender, purple papules developed at both calves and then converged to hemorrhagic bullae, although the patient had taken a 7-day course of antibiotics. Under the impression of HSP, we initiated methylprednisolone 40mg twice daily. The skin biopsy reported fibrinoid necrosis of vessel walls with dense neutrophil infiltration. Direct immunofluorescence (DIF) studies showed IgA and fibrinogen deposition at superficial dermal vessels, consistent with leukocytoclastic vasculitis (LCV) and HSP. After use of steroids, the skin lesions gradually healed, and the patient was discharged with oral dexamethasone 4mg daily. Neither recurrent skin purpura nor other organ involvement were evidenced at a 3-month follow-up.

Conclusion:

The cutaneous manifestation of HSP often presents with symmetric petechiae or palpable purpura, which may converge to larger, bullous or necrotic lesions. Hemorrhagic bullae may imply serious diseases including infection, autoimmune disorders, coagulation or platelet disorders, or severe drug reactions. Compared with children, there are more renal involvement and severe courses in adult HSP. Although no evidence has indicated corticosteroids improved long-term outcome, the bullous lesions improved rapidly after use of steroids without significant renal or gastrointestinal symptoms. We presented this rare case, emphasize the diagnostic process and timely management, and share our experience with physicians in Taiwan.