

Henoch-Schönlein Purpura with Hemorrhagic Bullous Skin Lesions

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Case Report

Patient: 53-year-old Male

Chief complaint: Abdominal pain and non-blanching palpable purpuric rashes on both legs

Physical examination: Multiple pupuric lesions were noted at the both lower extremities, arms and hands. After a few days, skin lesions had extensively spread to forearms, buttocks and thighs. Also, large, tense hemorrhagic bullae with varying size from 10 mm to 60 mm in diameter developed over lower extremities and dorsal aspects of both hands (Fig. 1).

Past medical history: His past medical history was unremarkable except for hypertension.



Figure 1. Multiple hemorrhagic bullous and purpuric lesions on the leg.

Laboratory tests: At admission, blood pressure of 140/100 mm Hg, body temperature of 36.8°C, pulse of 80/min and respiratory rate of 20/min were checked. Laboratory test revealed erythrocyte sedimentation rate (ESR) of 37 mm/h, CRP of 31.55 mg/L, white blood cell (WBC) of $17.65 \times 10^3/\text{mL}$ with neutrophilia and platelet of $465 \times 10^3/\text{mL}$. IgA level was increased by 520.8 mg/dL. BUN, creatinine, C_3 , C_4 , spot urine test, autoantibodies and tests for viral infections were all negative.

Radiologic findings: Abdomen CT showed marked thickening of gut wall with fluid collection in anterior paranephric space (Fig. 2). Upper gastrointestinal endoscopy showed erythematous and edematous mucosa with multiple shallow ulcers at bulb and second portion of duodenum (Fig. 3).

Diagnosis and treatment: Skin biopsy was compatible with leukocytoclastic vasculitis (Fig. 4). A diagnosis of Henoch-Schönlein purpura (HSP) was made with characteristic symptoms, including palpable purpura on extremities, abdominal pain, and arthralgia. Intravenous methylprednisolone of 0.5 mg/kg/day was administered to control abdominal pain and

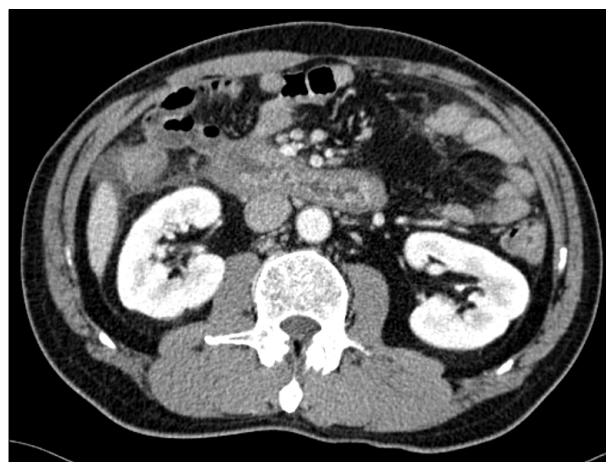


Figure 2. Edematous wall thickening of duodenal 3rd portion and terminal ileum.

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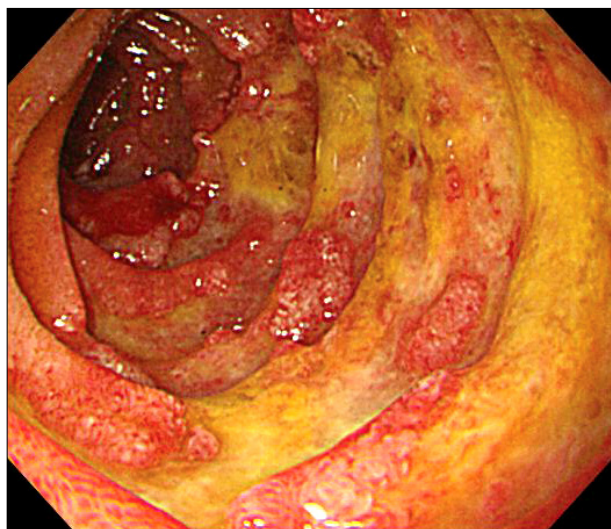


Figure 3. Several erythematous and edematous mucosa and patches were seen at the bulb and the second portion of duodenum.

cutaneous lesions. Large, tense bullae were released with needles. Owing to possibility of infection of cutaneous lesions, intravenous antibiotic treatment with ceftriaxone was introduced. Azathioprine was added, and no more spreading cutaneous lesions were observed. Also, bullae faded within the next 2 weeks and healed leaving some pigmentations and scars with tapering of corticosteroid.

Discussion

Henöch Schönlein purpura (HSP) is an immunoglobulin A (IgA)-mediated vasculitis of small-sized blood vessels and the most common acute systemic vasculitis in childhood (1,2). The major clinical features include arthritis, abdominal pain, gastrointestinal bleeding, nephritis and cutaneous lesions. Skin lesions usually present as erythematous maculopapules, petechiae, and purpura. Bulla formation in HSP is extremely rare cutaneous manifestation and more frequently noticed in adults than children (2,3). It usually develops into deep, crusted ulcers, and resolve with scarring. The treatment of bullae in HSP is not yet established definitely (1). Various treatment options are used such as dapsone, corticosteroid, and immunosuppressive agents (1-4). In this case, the patient was treated with corticosteroid and dapsone as initial therapy. Dapsone was discontinued due to gastrointestinal discomfort. After 2 weeks of corticosteroid therapy, we added azathioprine to control purpuric rashes. One more week of treatment with azathioprine, skin lesions were improved and the patient

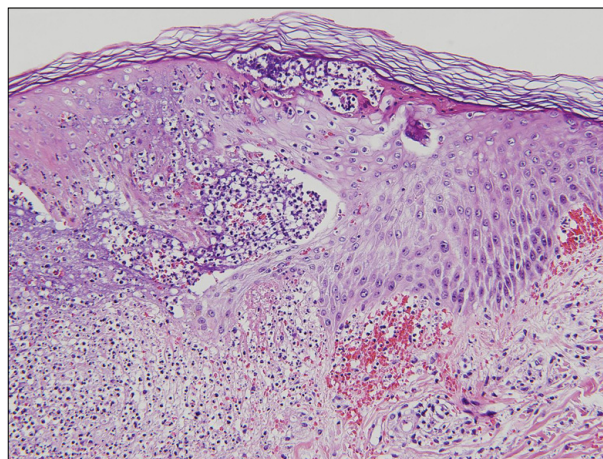


Figure 4. Biopsy from the bullae showed a necrotic epidermis, perivascular neutrophilic, lymphocytic infiltration with karyorrhexis, and endothelial swelling, which was compatible with leukocytoclastic vasculitis.

was discharged for outpatient department without any other complications.

HSP is a self-limited condition that lasts several weeks. Nephritis is the most serious long term complication of HSP and long-term prognosis of HSP is heavily dependent on the severity of nephritis. The previous reports showed that HSP patients with bullous formation underwent good prognosis without fatal systemic organ involvement such as progressive nephropathy (4). Therefore, bullous formation does not mean that the patient would suffer from a severe disease (2,4). This bullous evolution represents an unusual, but well-recognized cutaneous manifestation that may cause diagnostic challenge, even if it does not seem to have any prognostic value in the outcome of HSP.

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