Bullous Hemorrhagic Target Lesions in IgA Leukocytoclastic Vasculitis

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IgA vasculitis, formerly known as Henoch–Schönlein purpura (HSP), is the most common systemic vasculitis in children. It is defined as palpable purpura in the absence of coagulopathy or thrombocytopenia and one or more of the following criteria: abdominal pain, arthritis or arthralgia, biopsy of affected tissue demonstrating predominant IgA deposition, and renal involvement with proteinuria and hematuria or red cell casts [1].

IgA vasculitis may be preceded by an infection. It primarily occurs in children 3 to 15 years of age with a mean onset of 6–7 years. Annual incidence is 20–70 cases per 100,000. This disease occurs rarely in the summer months and shows slight male predominance with a ratio of up to 1.8:1 [2].

The most common presenting symptom is a purpuric rash on pressure-bearing areas of the skin, usually on the external aspect of the limbs and buttocks. The rash can begin as erythematous, macular, or urticarial wheals together with arthralgia. The typical dermatological presentation usually confirms the diagnosis.

Histopathology of the purpuric lesion typically shows leukocytoclastic vasculitis accompanied by deposition of IgA immune complexes with C3 and fibrin. Blood testing may reveal elevated serum IgA levels in up to 70 percent of patients [3]. Hypocomplementemia is reported in up to 15.7% of children [4].

Long-term outcomes of children with HSP are usually excellent and often resolve within one month. One-third of children experience at least one recurrence within 4 months of the initial presentation [2].

PATIENT DESCRIPTION

A previously healthy 8-year-old girl with no known hypersensitivity or other prior medical history presented to the emergency department (ED) with fever, papular lesions on all four limbs, dry cough, and abdominal pain.

Prior to this visit, she was seen at the primary care clinic where urine analysis was performed and was found to be positive for leukocyte esterase, blood, and protein. The patient was diagnosed with a urinary tract infection (UTI). First generation cephalosporins were administered pending urine cultures and were discontinued after cultures showed no growth.

On presentation to the ED, the patient presented with a fever of 38°C, vomiting, dry cough, abdominal pain, and a papular rash, 1–3 cm in diameter on the external aspects of her arms and legs [Figure 1A]. Medical history was negative for recent illness or for known exposure to coronavirus disease 2019 (COVID-19) patients.

Physical exam revealed purpura, epigastric tenderness, and sensitivity in both ankles. Ancillary testing included blood chemistry with mild hyponatremia of 130 Mmol/l. Complete blood count showed 15.45 × 10^9/L WBCs (reference value 8.5–13.5 × 10^9/L) with a differential blood count showing an elevated neutrophil count of 82% (mean reference value 52%). Hemoglobin was 12.5 GR% (reference value 12.4–16.1 GR%) and platelets 211 × 10^9/L (reference value 150–350 × 10^9/L). Sedimentation rate was elevated to 60/hour (reference value 1–20/hour) as well as elevated CRP of 15 mg/dl (reference value 0–0.5 mg/dl). Urine analysis was positive for blood and ketones with a trace of leukocytes. Chest X-ray was normal.

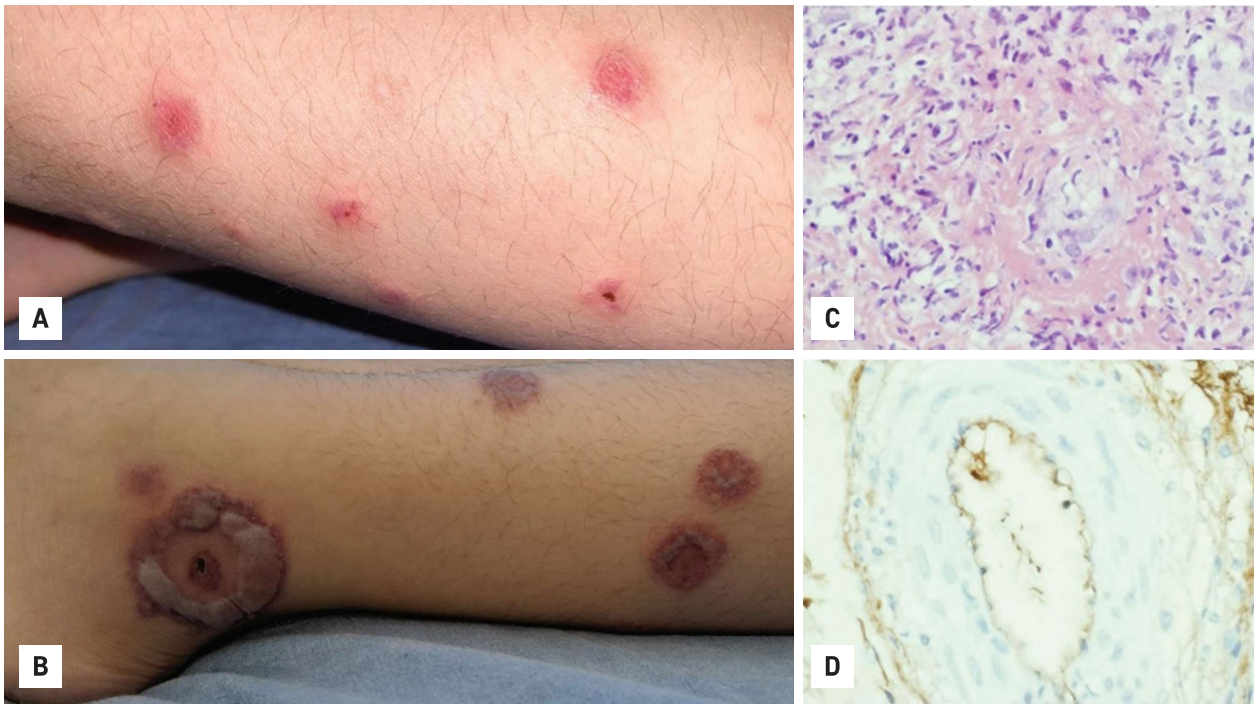
The patient was admitted with a working hypothesis of a partially treated UTI and received intravenous aminoglycosides (gentamicin).

During the patient's hospitalization, she continued to have fever and experienced a worsening of the rash, with few lesions evolving into bullous, hemorrhagic target lesions, some with central necrosis and formation of an eschar [Figure 1B] with worsening abdominal pain, cough, edema, and arthralgia of both ankles and knees. Repeated blood tests showed a rise in blood leukocytes to 23 × 10^9/L and further elevation of CRP to 35.2 mg/dl. Repeated blood and urine bacterial cultures were negative.

Further serological study for mycoplasma pneumonia, viral hepatitis, COVID-19, and rickettsia were negative. Complement levels were in the normal range, immunoglobulin levels showed high IgM values of 105 mg/dl (reference value 48–107 mg/dl). Testing for antinuclear antibody and antineutrophil cytoplasmic antibodies was negative.

Following the worsening of the patient's complaints and continued spread-

Figure 1. Clinical and histological findings. Evolution of target-like lesions with central necrosis and bullous periphery on day 1 **[A]** and day 5 **[B]** of hospitalization. Skin biopsy showing leukocytoclastic vasculitis and neutrophilic infiltration surrounding a capillary (H&E × 400) **[C]** and capillary positive for IgA (IgA deposition × 400) **[D]**



ing of the rash with no clear diagnosis, a skin punch biopsy was performed from the margins of the central lesion, revealing leukocytoclastic vasculitis with IgA deposits [Figures 1C, 1D], establishing the diagnosis of IgA vasculitis.

Following treatment with steroids the patient's fever and abdominal pain resolved. Her arthralgia began to improve. In the following days new purpureal lesions appeared, but none of them were targetoid or bullous.

After discharge from the hospital and during the follow-up period, oral steroid treatment was tapered down to a subsequent complete stop. Physical examination revealed a few healing lesions, but otherwise no findings. Blood analysis results were within normal limits.

COMMENT

We described a rare and unusual presentation of IgA vasculitis. The rash appeared relatively late in the course of the disease, which is slightly less typical in girls. The rash itself was uncharacteristic; however, bullous lesions have been described in past case reports [5]. To the best of our knowledge, no other cases with bullous hemorrhagic target lesions have been reported.

The patient's unusual presentation led us consider serum sickness or hypersensitivity vasculitis following use of cephalosporins for a suspected UTI, erythema multiforme, and sweet's syndrome.

This case presents a unique dermatological presentation of the dermatological lesions of IgA vasculitis. In contrast to typical HSP, which is usually clinically identified by the unmistakable rash, skin biopsy was crucial in this case. Without it, a definitive diagnosis would not have been made, which is important in HSP for long-term follow-up and treatment of possible sequelae.

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