

A Case of Mistaken Identity: Henoch-Schonlein Purpura Masquerading as Urticaria

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Abstract

Henoch-Schonlein purpura (HSP) is a common small vessel vasculitis encountered in children. It typically presents with purpura, arthralgia, and abdominal pain following an upper respiratory tract infection. We present the case of an 8-year-old boy with a history of atopy who presented with urticaria, purpura, and arthralgias following an upper respiratory infection. Subsequent punch biopsies revealed the diagnosis of HSP. While it is uncommon for HSP to present with urticaria, the pathogenesis behind the development of urticaria and IgA-associated vasculitis are similar. Furthermore, a history of atopy has been found to be associated with a significant number of patients with HSP. We present this case to highlight an atypical presentation of HSP and to discuss factors that may play a role in the development of this disease.

Keywords: Henoch-Schonlein purpura; Urticaria; Vasculitis

Introduction

Henoch-Schonlein purpura (HSP) is the most common small vessel vasculitis found in children [1]. Common symptoms included abdominal pain, arthritis or arthralgias, and purpura on the lower extremities and dependent areas [2]. Sequelae include intussusception, gastrointestinal hemorrhage, and nephritis, which can be delayed up to 3 months after initial presentation [2,3]. Histological examination confirms the diagnosis by showing an IgA deposit in vessel walls [3]. Here we present a case of a young boy with urticaria as well as symptoms of HSP. We highlight this case to exemplify an atypical presentation of HSP as well as discuss risk factors associated with the development of the disease.

Case

An 8-year-old boy with a history of seasonal allergies and atopic dermatitis presented to the emergency department with acute back pain and two weeks of bruising, joint swelling, and arthralgias. The patient complained of pruritus over the ecchymoses. He reported cough, congestion, and diarrhea the two weeks prior to admission. He denied fevers, night sweats, weight loss, headache, vision changes, abdominal pain, or gross hematuria. His family history included an unspecified autoimmune disorder in his father. He had no recent travel history, but he had multiple animal and environmental exposures including mice, birds, raccoons, stray cats, and mold in the home.

On examination, the patient was afebrile and vital signs were within normal limits. His bilateral ankles and dorsum of his feet had non-pitting edema and tenderness to palpation, with normal range of motion. His elbows, fingers, and knees were tender to palpation, but without restricted range of motion. Skin exam was notable for petechiae, palpable purpura, and ecchymoses at various stages of

healing on the trunk, bilateral upper and lower extremities, buttocks, and genitalia; sparing the face, palms, and soles (Figures 1 and 2). Evanescent urticarial plaques were also noted on the body throughout the hospital stay.



Figure 1: Palpable purpura (thin arrow) and urticaria (thick arrow) on the patient's arm.



Figure 2: Palpable purpura (thin arrow) and urticaria (thick arrow) on the patient's legs.

Initial labs revealed normal CBC, BMP, urinalysis, hematologic and rheumatologic work ups. Significant findings included a low C4 complement and elevated ASO titers. Punch biopsies were taken from purpuric and urticarial lesions on the abdomen and thigh. Hematoxylin and eosin (H&E) stains were significant for a peri and intravascular infiltrate of neutrophils and eosinophils involving the superficial and deep vascular plexuses (Figures 3 and 4). Direct immunofluorescence (DIF) revealed an IgA vascular deposition (not shown). These findings were consistent with Henoch-Schonlein Purpura (HSP).

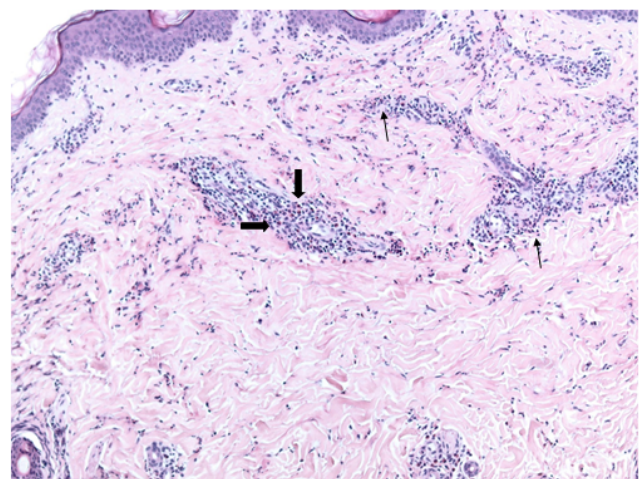


Figure 3: Punch biopsy of the abdomen showing an infiltrate of neutrophils (thin arrows) and eosinophils (thick arrows) involving the superficial and deep vascular plexuses (H&E stain, 10 × magnification).

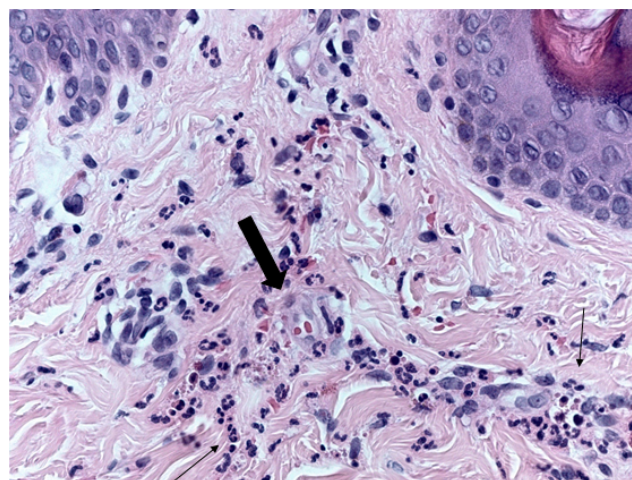


Figure 4: Punch biopsy of the abdomen showing a peri- and intravascular infiltrate (thick arrow=vessel, thin arrows=neutrophils) (H&E stain, 40 × magnification).

Discussion

EULAR/PRINTO/PRES criteria state that the patient must have petechiae or purpura with one or more of the following: diffuse abdominal pain, arthritis or arthralgia, leukocytoclastic vasculitis, renal involvement or proliferative glomerulonephritis with predominant IgA deposition [4]. Systemic manifestations such as arthritis (82%), abdominal pain (63%) or gastrointestinal hemorrhage (33%), and nephritis (40%) can occur at presentation or develop later in the disease course [2]. Renal manifestations can be delayed up to 3 months [3]. The classic cutaneous presentation includes palpable purpura on the lower extremities and dependent areas. Our patient presented with an atypical case of HSP where the cutaneous manifestations included acute urticaria.

HSP commonly follows an upper respiratory tract infection, typically *streptococcus*, but can also follow exposure to other infectious agents [5]. The pathogenesis involves the hematogenous spread of immune complexes comprised of mucosal-based IgA and microbial antigens which deposit into vessel walls inducing complement activation, mast cell degranulation, and neutrophil chemotaxis [3,6]. Proteolytic enzymes released by the neutrophils cause vessel wall damage and erythrocyte extravasation, resulting in palpable purpura [3]. Small, superficial vessel involvement can lead to urticarial lesions and purpura [7]. Deeper, dermal vessel involvement leads to hemorrhagic bullous or necrotic lesions [7].

Similar to HSP, acute urticaria develops after exposure to an infection, medication, or allergen [8]. Its pathogenesis also involves mast cell degranulation. The binding of substrates such as C5a, substance P, or allergens leads to mast cell release of histamine causing leakage of plasma into the dermis [3]. This creates the classic erythematous and evanescent wheals seen on exam [8].

Histological analysis of a skin biopsy provides a definitive diagnosis of HSP by demonstrating a neutrophilic infiltrate invading vessels of the superficial to mid-dermis on H&E with IgA deposition seen on DIF [3]. Treatment of HSP is mostly supportive, as it is a self-limited

disorder. However, up to 20% of patients will have recurrent purpuric eruptions and up to 2% will have permanent renal sequelae, thus requiring close follow up [3].

The differential diagnosis includes hematologic abnormalities (ITP, von Willebrand disease, malignancy, vitamin C deficiency), infectious agents (lyme, meningococemia, fungal), and rheumatologic diseases (JIA), however, these are less likely if the patient has normal lab work [3]. Additionally, other small-vessel vasculitides such as urticarial vasculitis, ANCA-associated vasculitides, and secondary causes of vasculitis can be excluded by skin biopsy with DIF [3].

This patient's presentation may have been due to two hypersensitivity processes occurring together. The initial mast cell degranulation induced by the IgA immune complexes may have resulted in an urticarial reaction as well. Furthermore, the patient's personal history of atopy may have played a role in his prominent mast cell degranulation and atypical cutaneous features. A retrospective study on the risk factors for IgA-associated vasculitis noted that conditions associated with immune dysregulation, such as atopy, were found in a significant portion of patients with HSP [6]. Seventy percent of the patients with infection-triggered HSP had an atopic history [6].

Conclusion

The pathogenesis behind the development of urticaria in the setting of vasculitis is currently unknown. Hence, we present this atypical case of HSP to highlight a poorly understood presentation of a common childhood disease. We hypothesize that a strong personal history of atopy may have predisposed our patient to a robust and atypical immune reaction to the streptococcal infection. While HSP is

commonly thought to involve purpura, physicians should be aware of other cutaneous presentations that can be associated with this disease and screen for vasculitides in the setting of atypical appearances. Similarly, caretakers should consider the risk factors that may predispose a patient to disease development.

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