



Lydia Shedlofsky DO^{1*} and Chelsea Crist²

¹Larkin Community Hospital, USA
²Southeastern Regional Medical Center, USA

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*Corresponding author: Dr. Shedlofsky, MSMedEd, Larkin Community Hospital, USA, E-mail: lshedlofsk@larkinhospital.com

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Case Report

A rare disease more common than perceived: Two case studies and brief review of IgA Vasculitis

Abstract

Immunoglobulin A (IgA) Vasculitis, more commonly known as Henoch-Schönlein Purpura (HSP), is a disorder which causes inflammation and bleeding in the small blood vessels of the skin, joints, intestines, and kidneys. We report 2 cases of IgA vasculitis found in a rural emergency department: 1) HSP in an 8-year-old male who was initially misdiagnosed with insect bites 2) HSP in an adult male patient. We present both cases and a literature review, indicating that low incidence may be secondary to under diagnosis, arguing for a need of further education on the subject.

Abbreviations

IgA: Immunoglobulin A; HSP: Henoch-Schönlein Purpura; EULAR/PreS: European League Against Rheumatism/Paediatric Rheumatology European Society

Introduction

Henoch-Schönlein Purpura (HSP) is a systemic vasculitis of unknown etiology, caused by IgA deposition in vessels. Patients of any age can be affected, but the mean age of diagnosis is 6.7 ± 2.41 years [1]. Cutaneous lesions are the most common presenting symptoms, comprised of elevated non-blanching palpable purpura typically on the lower extremities [2,3]. The classic triad for clinical diagnosis is the above rash, in addition to abdominal pain or renal involvement, and arthritis [4-6] and this is reflected in diagnostic criteria seen in table 1.

Criteria vary somewhat due to increased research into the subject, including the development that leukocytosis with IgA immune deposition in small vessels contributes to the etiology of HSP [10].

This is considered an extremely rare condition, with reported incidence estimated to be 15 cases/100,000 people per year in children, and one tenth of this incidence in adults [11]. The adult patients have a more severe condition requiring more aggressive therapy, though HSP is usually non-fatal and often self-limiting [12].

Due to this rarity, and typically benign course, HSP is often not taught in depth in medical schools, where few students are offered a dermatology course [13]. This persists into residency,

where educational focus is on most commonly seen conditions [14]. Excluding dermatitis, internal medicine physicians spend less than 0.03% of their time on dermatologic diagnoses. This indicates that each internist could go their entire careers without seeing a case of HSP [15,16]. For the past 80 years medical interns have been berated with the now infamous adage “When you hear hoofbeats look for horses, not zebras [17].” Of course, this forces physicians to develop an internal bias where IgA Vasculitis is simply not included in differentials.

Table 1: Classification Criterion for Henoch-Schönlein Purpura
Criteria Developed by Michel et al. [7].

Requiring 3 or more of 6 with specificity of 88.1% [8]		
1.	Palpable Purpura	Slightly elevated purpuric rash over 1 or more areas of the skin not related to thrombocytopenia
2.	Bowel Angina	Diffuse abdominal pain worse after meals or bowel ischemia usually including bloody diarrhea
3.	Gastrointestinal bleeding	Including melena, hematochezia, or positive test for occult blood in the stool
4.	Hematuria	Gross or microscopic hematuria
5.	Age at onset less than or equal to 20 years	Development of first symptoms at age 20 or less
6.	No medications	Absence of any medications at onset of disease which may have been a precipitating factor
European League Against Rheumatism/Paediatric Rheumatology European Society (EULAR/PreS) Criteria [9].		
Palpable purpura plus at least 1 or more of 4		
1.	Diffuse abdominal pain	
2.	Biopsy	Showing predominant IgA deposition
3.	Arthritis or arthralgia	Acute, any joint
4.	Renal Involvement	Any hematuria and/or proteinuria

We argue that it is equally possible that the true incidence is higher than published data, due to under-reporting. In fact, there are multiple published cases of misdiagnosed HSP [18–22], leading to significant morbidity for patients. This frequency of misdiagnosis contributes to the perception that HSP is so rare it can never be seen, in turn causing more misdiagnoses, lower incidence, and less teaching on the subject matter.

In one month at a rural emergency department two cases of HSP were initially misdiagnosed, supporting this theory.

Clinical Case Reports

Case 1

An 8-year-old male was brought to the emergency department by his mother complaining of a non-pruritic eruption to his lower extremities. Physical examination revealed scattered purplish macules and papules with diffuse petechiae covering his ankles and distal lower extremities. There were no recent illnesses, or changes in any medications. Diascopy revealed non-blanching lesions. The condition was originally misdiagnosed as secondary to insect bites. However, further history taking revealed that the patient also had bilateral knee pain and abdominal pain made worse with food. Blood work and urinalysis were all within normal limits, including ASO titer. Clinical diagnosis was made based on classic triad, in addition to meeting both the criterion developed by Michel et al., and EULAR/PReS criterion.

Case 2

A 30-year-old male truck driver with well-controlled diabetes presented to the emergency department complaining of a worsening “rash”. He reported that he had first noticed discoloration to his bilateral shins several weeks ago, but earlier in the day he saw other “spots” appear as seen in figure 1. He also admitted to fatigue.

Clinical examination revealed bilateral 1+ pitting edema up to his knees, in addition to skin findings as seen above. Anterior tibias reveal violaceous patches with superficial erosions, brown hyperpigmented patches, and scattered erythematous macules. Initial blood work revealed white blood cell count of 15, normal platelet count, and CRP of 2.3. The differential was focused on venous stasis with overlying cellulitis. However, diascopy was performed revealing that the scattered macules were non-blanching, and palpable. Additional blood work revealed an ESR of 37 and IgA of 440. Urinalysis returned showing 3+ blood, and



Figure 1: A) Image of lower extremity eruption for case 2 revealing violaceous plaques, brown patches, ulcerations, and scattered palpable purpura. B) focused image of the non-blanching purpura. Subsequently the patient was diagnosed with IgA vasculitis.

3+ protein. Patient was admitted to the hospital, and subsequent skin biopsy revealed leukocytoclastic vasculitis.

Discussion

IgA vasculitis is one of the most common type of vasculitis found in children [23], but education on the subject during training focuses on the clinical trial, and the absolute rarity of the condition. Taught that the low incidence precludes making this diagnosis, it rarely enters possible differentials. However, in a single month at a rural emergency department two cases were discovered, and nearly misdiagnosed.

Case one was a textbook example of HSP, a young male with purpura, abdominal and joint pain presenting to the emergency department. However, it was nearly misdiagnosed because of the mother’s over-riding concerns about “bed bugs”. The adult in case two has a much more difficult presentation. He is one of approximately 115,000 cases worldwide each year [11], with long-standing lower extremity discoloration and co-morbidities. Differential was focused on venous stasis with overlying cellulitis, but fortunately diascopy was performed before empiric antibiotics were begun.

Additionally, the multitude of published cases of misdiagnosed IgA vasculitis, in addition to this subjective experience, indicates that there must be additional cases worldwide that are not discovered. Misdiagnosis of IgA vasculitis leads to significant morbidity and even possible mortality for patients [24–26].

Adult patients have much more severe renal histopathological changes compared to pediatric patients [27]. One in ten adults who has biopsy proven HSP die due to the disease course [28]. This is also considered a chronic disease of the mesangium [29], so of those adults who survive, many will continue to have renal abnormalities [30]. Only corticosteroid administration during the acute stage of the illness will prevent morbidity and mortality [31], which requires accurate and rapid diagnosis.

Overall, the misdiagnosis also contributes to under-reporting, indicating that the probable incidence is higher than published data indicates. This frequency of misdiagnosis contributes to the perception that HSP is so rare it can never be seen, in turn causing more misdiagnoses, lower incidence, and less teaching on the subject matter. We strongly recommend continuing medical education on the subject, but the ideal solution is additional education while still in training, either in medical school, or in residency.

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