Case Report

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When Urticaria Is Not Just an Allergy: A Case of IgA Vasculitis Masquerading as a Drug Reaction

Rajvi Chaudhary and Shaily Saraf *

Dr KC Patel Medical College and Research Center

* Correspondence: shaily.saraf95@gmail.com

Abstract: Immunoglobulin A (IgA) vasculitis previously called Henoch-Schönlein Purpura represents the most common vasculitis of childhood, characterized by IgA1 deposition in small vessels of the skin, joints, gastrointestinal (GI) tract, and kidney [1]. This case report presents an atypical presentation of IgA vasculitis in an eight-year-old girl, where the skin manifestations were predominated by urticarial rashes prior to the onset of the classical palpable purpura and was mistaken as a case of drug induced urticaria. She presented with an acute history of severe pain abdomen, urticarial rashes over buttocks, bilateral knee joint pain and difficulty in walking with burning sensation of both feet. She was being treated as a case of acute gastroenteritis with drug induced urticaria prior to presentation at our center. On detailed physical examination bilateral lower limb palpable purpura were noticed with prolonged capillary refill time in bilateral toes and feeble dorsalis pedis pulses. The diagnosis of IgA vasculitis was made clinically based on EULAR criteria, and she was treated with steroids and supportive management on which patient recovered well and remained asymptomatic during follow up. This case highlights the importance of considering a diagnosis of IgA vasculitis in children with atypical presentation like in our case where urticarial rashes preceded the classical palpable lower limb purpura. The multisystem presentation of disease with nonspecific symptoms and lack of definitive diagnostic tests, mandates a high index of suspicion in these children. Although the illness can be self-limiting, but the potential for late renal involvement and end-stage renal disease underscores the importance of early diagnosis and treatment.

Keywords: IgA vasculitis; atypical presentation; urticarial rash; pediatric vasculitis

Introduction

Immunoglobulin A (IgA) vasculitis previously called Henoch-Schönlein Purpura (HSP) is a multisystem small vessel vasculitis involving the skin, gastrointestinal system, joints and kidney that predominantly affects children with an average annual Incidence rate of 30 per 100,000 people, with a male predominance and higher incidence in Caucasians or Asians. [1,2] It is characterized by non-thrombocytopenic purpura predominantly involving gravity-dependent and pressure bearing surfaces of skin like buttocks and lower extremities. Gastrointestinal (GI) manifestations include pain abdomen, vomiting, and rarely GI bleeding. Joint involvement presents as arthralgia/arthritis with a predilection for large joint involvement-like knees and ankles. [3] Renal involvement usually occurs later in the disease course, and should be investigated proactively by blood pressure measurement, urine analysis (proteinuria, hematuria) and eGFR measurement. [3] A history of upper respiratory tract infection (URI) or food/drug antigen exposure may be present. [1] Renal and skin biopsies, although not routinely indicated, demonstrate leukocytoclastic vasculitis with IgA deposition. [3] Although IgA vasculitis is usually self-limited and requires only supportive management ensuring adequate hydration and nutrition, the potential for late renal involvement and end stage renal disease mandates early diagnosis, treatment and adequate follow up. Steroids are indicated for severe abdominal pain unresponsive to NSAIDs or in the presence of complications such as nephritis. [4]

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The illness usually lasts for 4 weeks, however around 3.0% - 33.0% of pediatric patients experience recurrence, most commonly involving the skin, and in a milder form than previous episode. [5]

Case Report:

An 8-year-old girl, presented to the out-patient-department with a history of severe pain abdomen and vomiting for two days which was followed by urticarial rashes over back and buttocks a day later. For these complaints she was treated symptomatically with antispasmodics and antiemetics by her physician. Upon presentation to our center at day 3 of illness, the parents also reported bilateral knee joint pain, and a burning sensation in both feet, severe enough to make her non ambulatory for last one day A history of upper respiratory tract infection was documented 10 days prior to this episode.

On Physical examination, she was conscious and oriented. Heart rate was 130/min. Pulses were well felt on bilateral upper limbs, however she had bilaterally feeble dorsalis pedis pulses and a prolonged capillary refill time in bilateral toes. Her blood pressure was 104/68 mmHg in right upper limb. She was noted to have bilaterally symmetrical, non-blanchable petechial rashes on the extensor aspect of lower limbs, bipedal edema and acrocyanosis of both great toes. Urticarial lesions were noted over the back, buttocks and thighs. Bilateral knee joints were not swollen, with normal range of motion, however pain was present on movement. The abdomen was non distended, however was diffusely painful on palpation. No guarding/rigidity/organomegaly was noted. The rest of the cardiovascular/respiratory and CNS examination were normal.

On laboratory investigations, hemoglobin was 11.6 g/dl, total leucocyte count was 16,940 cells/mm3 comprising 84% neutrophils and 14% lymphocytes, and platelet count was 3.95 L/mm3. C-reactive protein was elevated (74 mg/dl), however ESR was within normal limits (10 mm/hr.). Serum electrolytes and renal function tests were normal. Urine analysis was normal, with no hematuria or proteinuria. Coagulation parameters were normal. Ultrasound whole abdomen revealed minimal free fluid. USG doppler of bilateral lower limb was also done in view of feeble dorsalis pedis and prolonged CFT. However, no evidence of thrombosis was noted. The patient was also advised for an APLA workup, but couldn't be done due to cost concerns

As per EULAR /PRINTO/PRES CRITERIA [10] child was clinically diagnosed as a case of IgA vasculitis She was started on supportive management with lower limb elevation, antispasmodics and anti-emetics and maintenance fluids. Urticarias were treated with oral anti-histaminic and took 3-4 days for complete resolution. In view of severe pain abdomen, the child was treated with oral prednisolone for 2 weeks @ 1mg/kg/day and was then tapered and stopped over the next 4 weeks. On 2nd day of steroid therapy, dorsalis pedis pulses were well palpable and foot edema became passive. The child became ambulatory on day 3 of hospital stay. Patient was advised follow up with serial urine analysis and regular blood pressure monitoring for next 1 year in view of possible renal involvement in future. and was also advised for a pediatric rheumatology opinion.

Discussion

IgA vasculitis is a disease primarily affecting the small vessels of skin, GIT, joints and kidney. The association between arthritis, purpura and arthralgia was first described in a publication by Schönlein in 1837. Later Henoch reported cases of children with pain abdomen, purpura, joint pain and dysentery. [6] Almost 90 percent of the cases have been documented in children less than 10 years of age. [7]. Our case had an initial atypical presentation predominated by urticarial rashes and gastrointestinal symptoms which preceded the classical palpable purpura in dependent areas. Since the diagnosis is made by clinical criteria without any specific diagnostic test, our case highlights the importance of high index of suspicion for the diagnosis of IgA vasculitis.

In our case, the 8-year-old girl presented with severe pain abdomen and urticaria as the initial symptoms. The initial presence of urticaria without the classical palpable purpura was the reason for the diagnostic dilemma. (Image 3) As the disease progressed, she also developed joint pains and later

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palpable purpura when the diagnosis of IgAV was considered. Purpuric rash is seen in almost 100% of IgAV cases, arthritis in 75% cases, abdominal symptoms in 60-65% cases and renal involvement in 40-50% cases. [15](Image 1,2)

In up to 30% of the cases gastrointestinal symptoms can precede the characteristic skin rash. [14] This is important to consider, especially in children as IgA vasculitis accounts to be the most common vasculitis in pediatric age group. [13]. Previous case reports by Rahees V.K.et al., Katerina Yale, Olabola Awosika et al. have also reported the presence of urticaria as an initial symptom in HSP. By our case, we highlight the fact that skin involvement other than the classical palpable purpura should also raise the suspicion for IgAV in the presence of other relevant clinical features.[16]

The exact etiology and pathogenesis of IgA vasculitis is unclear. Genetic, immunological and environmental triggers have been implicated. [1]

Dysregulated IgA synthesis and impaired clearance of IgA- containing immune complexes have been shown to exacerbate the inflammatory response, perpetuating vascular injury and multi-organ involvement. [8] While genetic predisposition and dysregulated immune system sets the stage, various environmental factors can precipitate IgA vasculitis onset. Infectious agents, particularly respiratory pathogens like Streptococcus and viruses, have been implicated in triggering immune complex formation and vasculitis flares. Additionally, medications, insect bites, and food allergens have been reported as potential triggers, highlighting the multifactorial nature of disease initiation in susceptible individuals. [9] Clinical presentation varies widely, ranging from mild cutaneous manifestations to severe systemic involvement, including nephritis and gastrointestinal complications. Early recognition and timely intervention are critical to prevent long-term sequelae, especially renal impairment. Management strategies primarily focus on symptomatic relief, including nonsteroidal anti-inflammatory drugs (NSAIDs) for joint pain and glucocorticoids for severe systemic manifestations. In cases of renal involvement, immunosuppressive therapies may be warranted to attenuate ongoing inflammation and preserve renal function. [10]

[12] 6-12 months after the first presentation. This delayed renal involvement highlights the need for prolonged follow up with regular BP monitoring and urine microscopy. Timely treatment prevents end stage renal disease. [4]

ACR classification criteria (1990)	EULAR/PRINTO/PRES classification criteria (2010) [3
Two of the following criteria: age ≤ 20 years palpable purpura acute abdominal pain biopsy showing granulocytes in the walls of the small arterioles or venules	Purpura or petechiae AND One of the following four criteria Abdominal pain Arthritis or arthralgia Renal involvement Leucocytoclastic vasculitis with predominant IgA deposits or proliferative glomerulonephritis with predominant IgA deposits
sensitivity 87.1 %; specificity 87.7 %	sensitivity 100 %; specificity 87 %

Conclusion

This case highlights the importance of considering IgA vasculitis as a differential in children who present with the typical constellation of features involving skin, joints, GIT and renal system.

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We also highlight that skin involvement is not just limited to the classical palpable purpura but also other atypical features like urticarial predominant rashes. Early diagnosis and management help to reduce morbidity as well as possibility of renal complications including End stage renal disease

References

1. Song Y, Huang X, Yu G, Qiao J, Cheng J, Wu J, Chen J. Pathogenesis of IgA vasculitis: an up-to-date review. Frontiers in immunology. 2021 Nov 9;12:771619.
2. Xu L, Li Y, Wu X. IgA vasculitis update: Epidemiology, pathogenesis, and biomarkers. Front Immunol. 2022;13:921864. Published 2022 Oct 3. doi:10.3389/fimmu.2022.921864.
3. Petty R, Salehzadeh F. Knee Swelling and Rash. Pediatric Autoimmunity and Transplantation: A Case-Based Collection with MCQs, Volume 3. 2020:47-51.
4. Pitcher D, Braddon F, Hendry B, Mercer A, Osmaston K, Saleem MA, Steenkamp R, Wong K, Turner AN, Wang K, Gale DP. Long-term outcomes in IgA nephropathy. Clinical Journal of the American Society of Nephrology. 2023 Jun 1;18(6):727-38.
5. Hu YC, Yang YH, Chiang BL. Immunoglobulin A vasculitis: The clinical features and pathophysiology. The Kaohsiung Journal of Medical Sciences. 2024 Jul;40(7):612-20.
6. Parums DV. A review of IgA vasculitis (Henoch-Schönlein purpura) past, present, and future. Medical Science Monitor: International Medical Journal of Experimental and Clinical Research. 2024 Jan 28;30:e943912-1.
7. Basyouny A, Imam A, Abouzaid KA. An Atypical Presentation of Henoch-Schönlein Purpura With Features of Acute Hemorrhagic Edema of Infancy and Koebnerization: A Case Report. Cureus. 2025 Feb 17;17(2).
8. Jennette JC. Overview of the 2012 revised International Chapel Hill Consensus Conference nomenclature of vasculitides. Clinical and experimental nephrology. 2013 Oct;17:603-6.
9. McCarthy HJ, Tizard EJ. Clinical practice: Diagnosis and management of Henoch–Schönlein purpura. European journal of pediatrics. 2010 Jun;169:643-50.
10. Ronkainen J, Koskimies O, Ala-Houhala M, Antikainen M, Merenmies J, Rajantie J, Örmälä T, Turtinen J, Nuutinen M. Early prednisone therapy in Henoch-Schönlein purpura: a randomized, double-blind, placebo-controlled trial. The Journal of pediatrics. 2006 Aug 1;149(2):241-7.
11. Van de Perre E, Jones RB, Jayne DR. IgA vasculitis (Henoch–Schönlein purpura): refractory and relapsing disease course in the adult population. Clinical Kidney Journal. 2021 Aug 1;14(8):1953-60.
12. Sugino H, Sawada Y, Nakamura M. IgA vasculitis: etiology, treatment, biomarkers and epigenetic changes. International journal of molecular sciences. 2021 Jul 14;22(14):7538.
13. Ozen S, Sag E. Childhood vasculitis. Rheumatology (Oxford). 2020;59(Suppl 3):iii95-iii100. doi:10.1093/rheumatology/kez599
14. V. K., R., Jimo, L.N., Golmei, N., Sananganba, R.K. and Chettri, P. 2024. A unique and rare presentation of Henoch-Schonlein purpura: a case report. International Journal of Contemporary Pediatrics. 11, 10 (Sep. 2024), 1471–1473.
15. Reamy BV, Bunt CW, Fletcher S. A diagnostic approach to pruritus. Am Fam Physician. 2011;84(2):195-202.
16. Yale K, Awosika O, Rengifo-Pardo M, Ehrlich A. Genital Allergic Contact Dermatitis. Dermatitis. 2018;29(3):112-119. doi:10.1097/DER.0000000000000371