International Journal of Science and Research (IJSR)
ISSN: 2319-7064
Impact Factor 2025: 7.089

Recurrent Henoch Schonlein Purpura in Association with Cytomegalovirus - A Case Report

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Abstract: ***Henoch-Schonlein purpura is an immunoglobulin A (IgA)-immune complex mediated leukocytoclastic vasculitis that classically manifests with palpable purpura, abdominal pain, arthritis, and hematuria or proteinuria. The condition is much more predominant in children (90% of cases) and commonly follows an upper respiratory infection. [1] It is a systemic hypersensitivity vasculitis caused by the deposition of immune complexes in small blood vessels, including the renal glomeruli and mesangium. In the skin, the presentation is with non-thrombocytopenic purpura [2] Bacterial infections, most commonly group A streptococci, viral infections (Parvovirus, Epstein Barr virus, Varicella zoster virus), allergens, drugs, vaccines have been cited as risk factors. [3][4]. The clinical presentation of IgA vasculitis in adults is similar to that in children. However, in adults, there are two main differences; abdominal symptoms are less common, but renal involvement is more significant, including end-stage kidney disease (ESKD) . Importantly, adults with no known trigger for developing IgA vasculitis should be investigated for solid-organ malignancy, a recognized risk factor [5]. We present a case of recurrent Henoch Schonlein purpura in a 24 year old male. He presented with purpuric lesions on lower limbs, bilaterally symmetrical. No prior history of sore throat or abdominal pain was there. Histopathology confirmed small vessel vasculitis. His viral markers screening showed positive for cytomegalovirus IgG antibody. There was no underlying collagen vascular disorder or malignancy. Henoch Schonlein (HS) purpura associated with cytomegalovirus (CMV) IgG positivity is a rare association with very few cases reported in literature.***

Keywords: Henoch Schonlein purpura, IgA vasculitis, cytomegalovirus

1. Case Report

A 24 year old male presented with dusky papular lesions on lower limbs. There were no accompanying complaints of fever, abdominal pain, sore throat or arthralgia. On examination the lesions were bilaterally symmetrical involving whole lower limbs. Diascopy revealed the lesion to be purpuric. [Fig 1] Lesions extended till thighs and gluteal region. [Fig 2] His hematology investigations did not show any significant abnormality. Stool for occult blood was negative. PT INR was normal ASO titre was also done. All were within normal limits. Antinuclear antibody, ds DNA, mitochondrial antibody, smooth muscle antibody, liver kidney microsomal antibody, p-ANCA and c- ANCA were all negative.

He was put on Tab Vitamin C and NSAID along with other supportive treatment. His lesions disappeared in a weeks time. In the following week, the lesions reappeared, his skin biopsy was carried out. Histopathology reported dermal capillaries infiltrated with neutrophils in the vessel wall along with RBC extravasation consistent with leukocytoclastic vasculitis. The lesions resolved and fresh crops kept appearing every 10-15 days. Patient was put on oral methyl prednisolone 16 mg od increased to 24 mg once a day. Despite oral steroids treatment, the lesions kept on appearing. His liver enzymes, SGOT rose to 155 U/L and SGPT to 740 U/L. Liver enzymes kept on rising. HbsAg, HIV, HCV were all negative. In view of repeated crops of lesions, his CMV serology was carried out IgG came out as positive, IgM negative. Also because his liver enzymes kept rising, and HbA1c became abnormal, steroids were tapered to a lesser dose and withdrawn. He was planned to start on Ganciclovir but after 2 months the lesions stopped appearing and all treatment was withdrawn, liver enzymes subsequently came back to normal. The recurrent HSP lesions were attributed to CMV positivity. He has had no recurrence since 1.5 months

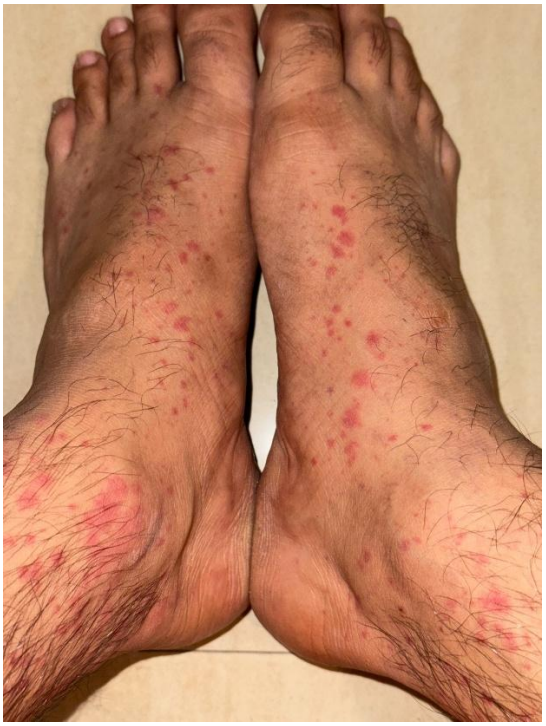

Figure 1: Palpable purpura

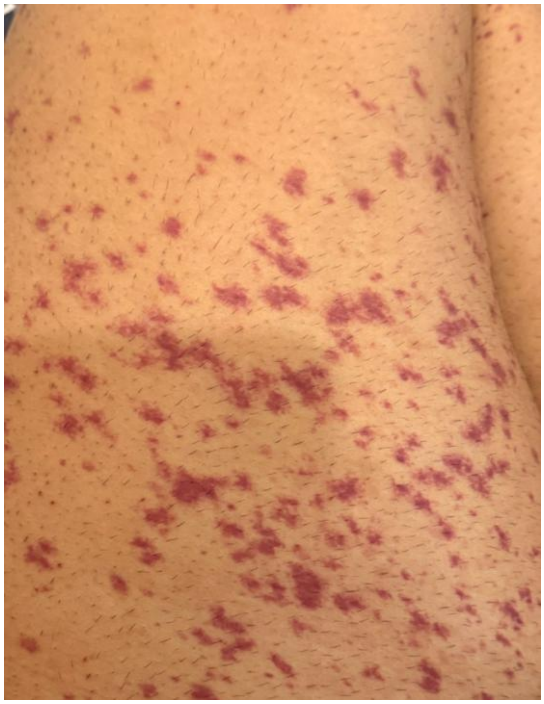

Figure 2: Purpura extending to thigh

2. Discussion

Worldwide, approximately 90% of cases of HS purpura occur in children between 3–15 years of age, with a mean age of 6 years, and also occurs less commonly in adults. [6] The pathogenesis of HSP is still unknown. Several factors such as infections, medications, malignancy, and their combinations were reported as a cause of HSP [7]. Viral infections are a common cause of HSP. COVID-19-associated IgA vasculitis has also been reported in an adult patient with significant renal involvement. [8] Very few cases of recurrent HS purpura have been reported in association with Cytomegalovirus infection. Cytomegalovirus (CMV)-induced Henoch-Schönlein purpura (HSP), is a rare form of systemic small-vessel vasculitis that occurs when a CMV infection acts as a triggering antigen, leading to the deposition of Immunoglobulin A (IgA) immune complexes in blood vessel walls. While typically triggered by upper respiratory infections in children, CMV-induced cases are more often identified in immunocompromised individuals or the elderly. [9] Active CMV infection induces an immune response that leads to an overproduction of aberrantly glycosylated IgA1 (Galactose-deficient IgA1 or Gd-IgA1). This abnormal IgA1 forms large circulating immune complexes (CICs) by binding to autoantibodies (IgG or IgA). Also CMV has the capacity to directly infect and damage vascular endothelial cells. This damage, coupled with the deposition of these IgA-containing immune complexes into the small vessels of the skin, gastrointestinal tract, and kidneys, triggers the vasculitis. The deposited complexes activate the complement system (specifically the alternative pathway) and recruit neutrophils, causing further vessel wall damage, leakage, and necrosis (leukocytoclastic vasculitis). [10]

Consensus management guidelines suggest using oral corticosteroids for milder disease, oral, or intravenous corticosteroids plus azathioprine or mycophenolate mofetil or intravenous cyclophosphamide for moderate disease and intravenous corticosteroids with cyclophosphamide for severe disease. Angiotensin system inhibitors act as adjunctive treatment for persisting proteinuria and frequently relapsing disease may necessitate the use of immunosuppressant agents [11]

This case is reported because most cases of HS purpura associated with CMV are reported in elderly and associated with CMV duodenitis. Another case report in 27 year old male was following upper respiratory tract infection and patient had abdominal pain, vomiting and arthralgia. [12] Our patient had no other significant complaints and did not give history of prior sore throat also. Cytomegalovirus (CMV) has not been reported as a virus associated with HSP onset. However, cytomegalovirus infections may reactivate during HSP. Latent viral infection reactivation has been considered in the setting of immunosuppression. CMV complicating HSP was observed in both children and adults after high-dose steroid therapy. [13],[14]

References

[1] Gaskill N, Guido B, Magro CM. Recurrent adult onset Henoch-Schonlein Purpura: a case report. Dermatol Online J. 2016 Aug 15;22(8):13030/qt1r12k2z1.
[2] Parums DV. A Review of IgA Vasculitis (Henoch-Schönlein Purpura) Past, Present, and Future. Med Sci Monit. 2024 Jan 28; 30: e943912.
[3] Piram M, Gonzalez Chiappe S, Madhi F, et al. Vaccination and risk of childhood IgA vasculitis. Pediatrics. 2018; 142(5): e20180841.
[4] Yoneoka R, Kasai H, Hino A, et al. IgA vasculitis as a potential complication of fourth-line chemotherapy with Tegafur/Gimeracil/Oteracil (S-1) in advanced non-small cell lung cancer: A case report. Am J Case Rep. 2023; 24: e941826
[5] Pillebout E, Thervet E, Hill G, et al. Henoch-Schönlein Purpura in adults: Outcome and prognostic factors. J Am Soc Nephrol. 2002; 13: 1271.
[6] Oni L, Sampath S. Childhood IgA vasculitis (Henoch Schonlein Purpura) – advances and knowledge gaps. Front Pediatr. 2019; 7: 257.
[7] Zhang XD, Yang SY, Li W, Ming ZJ, Hou YL, Niu ZQ, Zhang YP. Adult Henoch-Schönlein purpura associated with small cell lung cancer: a case report and review of the literature. Oncol Lett. 2013; 5: 1927–1930.
[8] Jedlowski PM, Jedlowski MF. Coronavirus disease 2019-associated immunoglobulin A vasculitis/Henoch-Schönlein purpura: A case report and review. J Dermatol. 2022 Jan;49(1):190-196.
[9] Shin SB, Choi YJ, Lee J, Kwak BG, Kim YH, Ha KS, Kang JH. Henoch-Schönlein Purpura with Concurrent Cytomegalovirus Duodenitis. Infect Chemother. 2017 Jun;49(2):146-150.
[10] 10.Song Y, Huang X, Yu G, Qiao J, Cheng J, Wu J, Chen J. Pathogenesis of IgA Vasculitis: An Up-To-Date Review. Front Immunol. 2021 Nov 9; 12: 771619.
[11] Oni L, Sampath S. Childhood IgA Vasculitis (Henoch Schonlein Purpura)-Advances and Knowledge Gaps. Front Pediatr. 2019 Jun 27; 7: 257.
[12] Bhanushali JD. A unique finding of Henoch-Schönlein purpura in adults. Pan Afr Med J. 2024 Jun 13; 48: 59.

[13] Matsumura M., Komeda Y., Watanabe T., Kudo M. Purpura-free small intestinal IgA vasculitis complicated by cytomegalovirus reactivation. BMJ Case Rep. 2020; 13: e235042. doi: 10.1136/bcr-2020-235042.
[14] Mizerska-Wasiak M., Winiarska M., Nogal K., Cichoń-Kawa K., Pańczyk-Tomaszewska M., Małdyk J. IgA Vasculitis Complicated by Both CMV Reactivation and Tuberculosis. Pediatr. Rep. 2021; 13: 416–420.