

Henoch-Schönlein purpura: A case report of 12-year-old female child

M. Malik Basha^{1*}, S. Yasmin¹, B.P.V. Bharath Reddy¹, M. Areef¹, D. Srinath Reddy¹, N. Yamuna¹

¹ Department of pharmacy practice, santhiram college of pharmacy, Nandyal, A.P.

Submitted: 09-03-2023

Accepted: 18-03-2023

ABSTRACT: Henoch-schönlein purpura (HSP) is the most common vasculitis of childhood and is characterized by leukocytoclastic vasculitis and immunoglobulin a deposition in the small vessel in the skin, joints, gastrointestinal tract and kidney. It is a systemic disease where antigen-antibody(IgA) complexes activate the alternate complement pathway, resulting in inflammation and small vessel vasculitis. Mild disease resolves spontaneously, and symptomatic treatment alone is sufficient. We report a 12-year-old female child presented with rashes over both legs & buttocks along with joint pain. The patient was diagnosed with HSP and treated with oral prednisolone and other supportive care. The patient was finally discharged on the resolution of her symptoms. Early diagnosis and treatment favor the better outcome in cases without any renal complications.

KEY WORDS: Henoch-schönlein purpura, vasculitis, prednisolone.

INTRODUCTION:

Henoch-schönlein purpura (HSP) is an immunoglobulin-A(Ig-A) mediated systemic vasculitis affecting the vasculature of several systems including the gastrointestinal tract, renal system, skin & joints.[1] It is characterized by the presence of a vasculitic purpuric rash, abdominal pain, joint pain, renal injury, pulmonary inflammation or central nervous system involvement.[2] Around 90% of cases occur in children under age of 10 with greater preponderance for males.[1,3-5] Incidence of HSP is estimated at 14-20 per 100,000 children per year and it affects males more than females with a 1.2-1.8:1 male/ female ratio.[6]

More than 90% HSP patients are under 10 years old with the average age at onset of 6.4 years,[7] whereas HSP is relatively uncommon in adults. [8-10] The annual incidence of HSP in adults is reported to be about a tenth of that in children.[11] Since HSP is infrequent in middle

aged and the elderly, when its initial manifestation is atypical, it might be challenging to recognize. Atypical clinical presentation of HSP, including bullous lesions, anuria, and intussusception, increases the difficulty in prompt diagnosis and management. [12-14] Although gastrointestinal involvement is relatively common in adult HSP,[15] along time gap between preceding gastrointestinal presentations and delayed erupted cutaneous lesions is uncommon. [8,9,13]

It usually follows an upper respiratory tract infection caused by the group of A-beta hemolytic streptococcus, staphylococcus aureus or mycoplasma. Skin biopsies demonstrate leukocytoclastic vasculitis involving dermal capillaries and venules.

Although self-limiting in nature complications such as gastrointestinal hemorrhage and end stage renal failure may occur. Immunoglobulin-A nephritis affects 60% of patients and can vary in severity from presence of asymptomatic microscopic hematuria with proteinuria to irreversible renal failure requiring renal transplantation. Corticosteroids may be used to treat HSP with significant GI or renal involvement.

II.CASE REPORT:

A 12-year-old female child admitted to our department with complaints of joint pains since 10 days including both large & small joints with swelling, difficulty in walking an increased pain and swelling on morning times.

She had rashes over both upper and lower extremities & buttocks which are on and off in nature more in lower and upper limbs (figure 1,2,3).

She had fever since 2 days high grade relieved by taking medications. There was no history of cold, cough, vomiting and loose motions.

On examination, the general condition of the patient was fair, and vitals were stable.

Abdomen was soft and non-tender. There was presence of non-tender, non-blanching purpuric rash over both lower and upper extremities.

Lab findings were suggestive of microlytic hypochromic anemia with Hb: 9.6 gm/dl, leukocytosis with WBC: 17,900/cmm, normal RBC & platelet count. Serum-electrolytes were within normal limits. The urine routine was normal and there was no evidence of any hematuria or proteinuria.

ANA profile analysis showed negative for all antibodies. The plain X-ray abdomen & ultrasound abdomen revealed no abnormalities.

Diagnosis of HSP was made in accordance with the American college of Rheumatology and European league against rheumatism (EuLAR) and pediatric rheumatology society (PREs) criteria. [16,17]

Child was treated with oral prednisolone 20mg 2times a day for 7 days child also received other supportive care in the form of analgesics & fluids.

Our case did not have any renal involvement manifesting in the form of hematuria, hypertension or frank nephritis. Child also did not develop any neurological manifestations.

Figure.1:Figure.2:



Figure.3:



Figure 1,2,3 showing palpable purpura on upper and lower limbs.

III.DISCUSSION:

HSP was first described by William Herberden in 1801 later schönlein recognized the association between purpura and arthritis while a case report by Henoch also included gastrointestinal symptoms along with the renal involvement.[18] HSP is the most common vasculitis of children. 50% cases occur before the age of 5 and males are affected almost twice as common as females.[19]

The exact etiology and pathogenesis of HSP is yet to be determined. Seasonal variation depicts high prevalence rate in winter and spring and is unusual in summer months. It is also suggested that various triggers like bacterial and viral infections, drug and auto-immune mechanisms may result in the formation of antigen and antibody complex and the deposition of such formed complex in the small vessels may activate the alternate complement pathway leading to neutrophil

aggregation which results in inflammation and vasculitis.[20] The patient presents with the tetrad of rashes, poly-arthritis, abdominal pain, and renal disease. Hallmark of HSP is the non-blanching rash which clinically presents which clinically appears as a palpable purpura on the lower legs and arms.[19] Musculoskeletal involvement is generally characterized by the pain and swelling of the joints, with a predilection for large joints such as knees and ankles. GI manifestations include abdominal pain followed by vomiting and intestinal bleeding. Microscopic hematuria and albuminuria are the prominent renal finding.[21]

Our case had the symptoms of rashes over both upper and lower limbs and buttocks. Pain and swelling of joints indicating arthralgia. However, there is no signs of hematuria and albuminuria so no renal involvement. The diagnosis of HSP was made by EuLAR or PRES criteria (Table 1).

Table 1: EuLAR/PRES criteria

Mandatory criteria	Palpable purpura
Additional criteria	Diffuse abdominal pain
	Any biopsy showing predominant IgA deposition
	Arthritis or arthralgia
	Renal involvement

The patient is said to have HSP if mandatory criteria are present along with at least one of the additional criteria. Our case fulfilled the mandatory criteria along with arthralgia as the additional criteria.

There is a predominantly spontaneous resolution of all symptoms except that of the renal disease in the majority of the cases. Steroids are most often used for the relief of joint pain and skin disease.[22] The role of corticosteroids in preventing the long-term outcome of renal complications is controversial. Generally, prednisone is the commonly used steroid for the treatment of HSP. The renal involvement has a high morbidity and mortality, otherwise the disease has an excellent prognosis. A systematic review by Narchi et al stated that even if urinalysis is normal at the presentation, patient should undergo follow up urine testing for at least six months as

97% children will develop abnormal urine findings by that time.[23]

IV.CONCLUSION:

HSP is one of the most common vasculitis of the children and its classic presentation of palpable purpura, arthritis, abdominal involvement and renal features makes the diagnosis quite easier. Early initiation of treatment with steroids will help in symptomatic relief and bring a positive outcome. The renal disease may need long term follow up; otherwise, the disease has favourable prognosis.

REFERENCES:

- [1]. Saulsbury FT. Henoch-Schönlein purpura in children: report of 100 patients and review of the literature. *Medicine*. 1999 Nov 1;78(6):395-409.
- [2]. Sood R, Parekh P, Raj N, Saani I. Case Report: An Adult Presentation of Henoch-

- Schönlein Purpura. *Cureus*. 2022 Jun 28;14(6).
- [3]. Calvino MC, Llorca J, Garcia-Porrúa C, Fernandez-Iglesias JL, Rodriguez-Ledo P, Gonzalez-Gay MA. Henoch-Schönlein purpura in children from northwestern Spain: a 20-year epidemiologic and clinical study. *Medicine*. 2001 Sep 1;80(5):279-90
- [4]. Trapani S, Micheli A, Grisolia F, Resti M, Chiappini E, Falcini F, De Martino M. Henoch Schonlein purpura in childhood: epidemiological and clinical analysis of 150 cases over a 5-year period and review of literature. In *Seminars in arthritis and rheumatism* 2005 Dec 1 (Vol. 35, No. 3, pp. 143-153). WB Saunders.
- [5]. Anil M, Aksu N, Kara OD, Bal A, Anil AB, Ün B. Henoch-Schönlein purpura in children from western Turkey: a retrospective analysis of 430 cases. *The Turkish journal of pediatrics*. 2009 Sep 1;51(5):429.
- [6]. Sivaraman V, Fels EC, Stacy P. Ardion, NELSON Textbook of Pediatrics, 21st Edition. Chapter 192.1.2019;1.
- [7]. Gardner-Medwin JM, Dolezalova P, Cummins C, Southwood TR. Incidence of Henoch-Schonlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. *The Lancet*. 2002 Oct 19;360(9341):1197-202.
- [8]. Gupta V, Aggarwal A, Gupta R, Chandra Chowdhury A, Agarwal V, Lawrence A, Misra R. Differences between adult and pediatric onset Henoch-Schonlein purpura from North India. *International Journal of Rheumatic Diseases*. 2018 Jan;21(1):292-8.
- [9]. Cao R, Lau S, Tan V, Tey HL. Adult Henoch-Schönlein purpura: Clinical and histopathological predictors of systemic disease and profound renal disease. *Indian Journal of Dermatology, Venereology and Leprology*. 2017 Sep 1;83:577
- [10]. Audemard-Vergier A, Terrier B, Dechartres A, Chanal J, Amoura Z, Le Gouellec N, Cacoub P, Jourde-Chiche N, Urbanski G, Augusto JF, Moulis G. Characteristics and management of IgA vasculitis (Henoch-Schönlein) in adults: data From 260 patients included in a french multicenter retrospective survey. *Arthritis & rheumatology*. 2017 Sep;69(9):1862-70.
- [11]. Tang C, Scaramangas-Plumley D, Nast CC, Mosenifar Z, Edelstein MA, Weisman M. A case of Henoch-Schonlein purpura associated with rotavirus infection in an elderly Asian male and review of the literature. *The American Journal of Case Reports*. 2017;18:136.
- [12]. Gulati G, Siv J, Ware AE. Bullous skin lesions in an adult male: a diagnostic dilemma. *The American Journal of Case Reports*. 2015;16:215.
- [13]. Krishnan M, Nahas J. Adult onset Henoch-Schonlein purpura and intussusception: A rare presentation. *Case Reports in Rheumatology*. 2016 Sep 26;2016.
- [14]. Weiser JA, Rogers HD, Stokes MB, Grossman ME. Henoch-Schönlein purpura presenting with anuria in an adult. *Cutis*. 2010 Oct 1;86(4):181-4.
- [15]. Audemard-Vergier A, Pillebout E, Amoura Z, Cacoub P, Jourde-Chiche N, Lioger B, Martis N, Moulis G, Rivière E, Baldolli A, Girard C. Gastrointestinal involvement in adult IgA vasculitis (Henoch-Schönlein purpura): updated picture from a French multicentre and retrospective series of 260 cases. *Rheumatology*. 2020 Oct;59(10):3050-7.
- [16]. 7 Mills JA, Michel BA, Bloch DA, Calabrese LH, Hunder GG, Arend WP, Edworthy SM, Fauci AS, Leavitt RY, Lie JT, Lightfoot Jr RW. The American College of Rheumatology 1990 criteria for the classification of Henoch-Schönlein purpura. *Arthritis & Rheumatism*. 1990 Aug;33(8):1114-21.
- [17]. Ozen S, Ruperto N, Dillon MJ, Bagga A, Barron K, Davin JC, Kawasaki T, Lindsley C, Petty RE, Prieur AM, Ravelli A. EULAR/PReS endorsed consensus criteria for the classification of childhood vasculitides. *Annals of the rheumatic diseases*. 2006 Jul 1;65(7):936-41.
- [18]. Tarvin SE, Ballinger S. Henoch-Schonlein purpura. *Current Paediatrics*. 2006 Aug 1;16(4):259-63.
- [19]. Roberts PF, Waller TA, Brinker TM, Riffe IZ, Sayre JW, Bratton RL. Henoch-Schonlein purpura: a review article. *SOUTHERN MEDICAL JOURNAL-BIRMINGHAM ALABAMA*-. 2007 Aug 1;100(8):821.



- [20]. Sohagia AB, Gunturu SG, Tong TR, Hertan HI. Henoch-Schonlein purpura—a case report and review of the literature. *Gastroenterology research and practice*. 2010 Oct;2010.
- [21]. Chen P, Zhu XB, Ren P, Wang YB, Sun RP, Wei DE. Henoch Schonlein Purpura in children: clinical analysis of 120 cases. *African health sciences*. 2013 Apr 12;13(1):94-9.
- [22]. Tizard EJ, Hamilton-Ayres MJ. Henoch–Schönlein purpura. *Archives of Disease in Childhood-Education and Practice*. 2008 Feb 1;93(1):1-8.
- [23]. Narchi H. Risk of long term renal impairment and duration of follow up recommended for Henoch-Schönlein purpura with normal or minimal urinary findings: a systematic review. *Archives of disease in childhood*. 2005 Sep 1;90(9):916-20.