Original Research Paper

Volume - 14 | Issue - 04 | April - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Paediatrics

A CASE REPORT OF HENOCH-SCHÖNLEIN PURPURA

Dr. Raksha Kamat Senior Resident Doctor, Department of Pediatrics, Government Medical College and New Civil Hospital, Surat.

Dr. Panna Bulsara Associate Professor, Department of Pediatrics, Government Medical College and New Civil Hospital, Surat.

ABSTRACT **Background:** Henoch-Schönlein purpura (HSP) is the most frequent leukocytoclastic systemic small-vessel vasculitis in children. HSP can present with a wide variety of clinical symptoms, from proteinuria and microscopic hematuria without any symptoms to variable degrees of acute kidney injury (AKI) and fast progressing glomerulonephritis with a high risk of long-term renal impairment. The kind and extent of organ involvement determines therapy options in more severe situations. **Case Summary:** A 12 year old female admitted to New Civil Hospital Surat with chief complaints of rash since 8 days. Also associated with joint and abdominal pain. Patient also gave history of cough, cold, sorethroat. Investigations done, Dermatologist opinion was taken for skin biopsy, skin biopsy showed leukocytoclastic vasculitis with IgA deposits. Patient Started on prednisolone 1mg/kg/day, Patient showed improvement with prednisolone within one week and was discharged sucessfully. **Discussion:** It is crucial to define the differential diagnosis with other bullous diseases in childhood. Prognosis of HSP is good if there is no renal involvement. **Conclusion:** Henoch-Schönlein purpura is a frequent paediatric vasculitis that is multi-systemic. Although the prognosis is often quite good, some individuals may experience long-term effects, usually in the kidneys.

KEYWORDS : Vasculitis, Immunoglobulin A vasculitis, Henoch Schonlein Purpura, Pediatric, Prognosis.

INTRODUCTION

Henoch-Schönlein purpura (HSP) is the most frequent leukocy to clastic systemic small-vessel vasculitis in children. It is also referred to as anaphylactoid purpura and, more recently, IgA Vasculitis (IgAV) [1]. The condition is commonly identified by non-thrombocytopenic purpura, exhibiting characteristic purple papules that do not blanch, and are mainly found on the lower limbs and buttocks. Additionally, it may be linked to lower extremity big joint arthralgia or arthritis, lower gastrointestinal distress, and a higher chance of renal involvement. [2]

HSP can present with a wide variety of clinical symptoms, from proteinuria and microscopic hematuria without any symptoms to variable degrees of acute kidney injury (AKI) and fast progressing glomerulonephritis with a high risk of long-term renal impairment. Other possible organ involvement includes cerebral vasculitis, ureteral or bladder disease, and scrotal, penile or testicular hemorrhage. [3]

The most prevalent type of vasculitis in children is Henoch-Schönlein purpura (HSP), which affects 8 to 20 out of every 100,000 children each year.With a male-to-female ratio of 1.5:1, it is more common in men and usually affects youngsters between the ages of three and eight. Immunoglobulin A (IgA)-mediated systemic small-vessel vasculitis, Henoch-Schönlein purpura causes symptoms in the skin, joints, intestines, and kidneys due to IgA deposition in vessel walls. HSP has a seasonal pattern (nonsummer months) and is frequently preceded by an acute infectious disease, suggesting an infectious trigger even though the aetiology is unknown. [4]

The easiest way to diagnose HSP is to have one or more of the following four findings in addition to purpura or petechiae, which are typically palpable and mostly affect the lower limbs: widespread and colicky stomach discomfort, arthralgia or arthritis,renal involvement is defined as follows: >0.3 g of proteinuria in a 24-hour period; morning urine albumin or creatinine levels > 30 µmol/L; positive hematuria dipstick results; and positive histopathologic findings (predominantly IgA deposits in leukocytoclastic vasculitis on skin biopsy or proliferative glomerulonephritis on kidney biopsy). Fever, scrotal discomfort, and edoema in males are additional symptoms; pulmonary, cardiac, or neurological signs are infrequent. [5]

The majority of children's IgAV cases resolve on their own and only require supportive care rather than any special treatment. The kind and extent of organ involvement determines therapy options in more severe situations. The lack of clinical trials in this field prevents the development of strong evidence-based recommendations, despite the publication of global consensus management plans. [6]

So, here a presenting a case report of Henoch-Schönlein purpura at New Civil Hospital, Surat.

Case Summary

A 12 year old female admitted to New Civil Hospital Surat on 13/11/2023 with chief complaints of rash since 8 days. Rash was petechial in nature along with palpable purpura, started initially in upper limb and gradually progressed to lower limb. Also associated with joint and abdominal pain, pedal edema (non-pitting). Patient also gave history of cough, cold, sorethroat. 15 days back, took medication for same. No history of fever, blood in stools.

On examination: temperature was normal, HR=98bpm, RR=24/m, BP= 110/70mmHg, SpO2= 98%, RS: AE=BS, CVS: NAD, PA: Soft, Investigations: Hb=11gm/dl, PCV=36%, Platelet count =7lakhs, S.Cr=0.6mg/dl, urine routine- normal, no microscopic hematuria, CRP-108, RA factor-negative, ASO titre- Positive (200), Ultrasound abdomen & pelvis- kidney appeared bulky. BT, CT, PTINR, APTT-normal. Symptomatic management along with injection ceftriaxone was started.

Hematologist opinion taken. Dermatologist opinion was taken for skin biopsy, skin biopsy showed leukocytoclastic vasculitis with IgA deposit. Patient Started on prednisolone 1mg/kg/day over one week which was tapered gradually, since no renal involvement was present so nephrologist opinion was not taken.

Patient showed improvement with prednisolone and was discharged sucessfully.

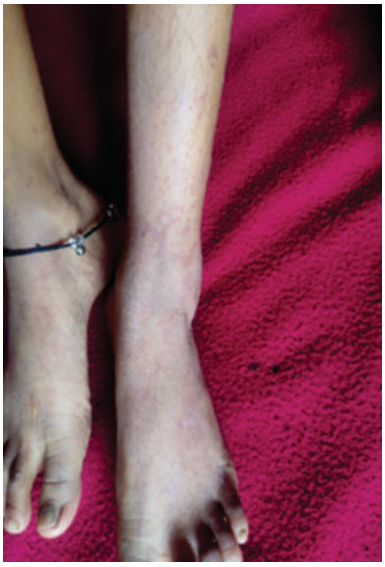

Image 1: Showing Petechial Rash On Feet And Legs.

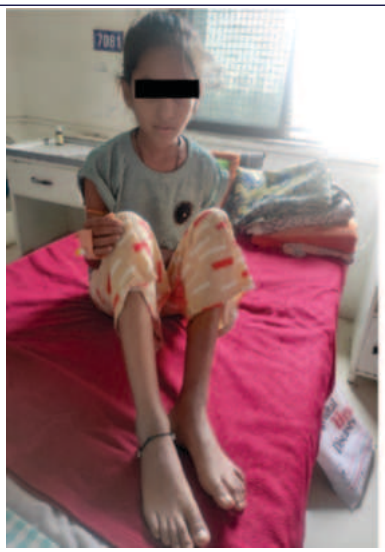

Image 2: Case of HSP

DISCUSSION

However, it is crucial to define the differential diagnosis with other bullous diseases in childhood, such as erythema multiforme, bullous impetigo, dermatitis herpetiformis, staphylococcal scalded skin syndrome, and linear IgA bullous dermatosis of children. There is no evidence linking the HSP to a worse diagnosis. A skin biopsy may occasionally be required to reach a conclusive diagnosis. [7]

There has been conjecture on whether the emergence of bullous lesions signifies a more severe course of the disease or even a different disease entity within HSP. The degree of renal involvement primarily dictates the long-term prognosis in HSP. This idea appears to be contradicted by our literature study on bullous lesions in paediatric HSP, which showed relatively less renal involvement in this patient group. Bullous lesions in the majority of patients who were reported had cured and did not recur in a matter of weeks, leading to the hypothesis that bullous HSP is a relatively benign and self-limiting condition. [8]

Uncertainty surrounds the aetiology of blistering skin lesions in HSP. Collagen may be proteolyzed by matrix metalloproteinase (MMP)-2 and MMP-9, which have been found in blister fluid. Nevertheless, nothing is known about the mechanisms that cause these enzymes to be secreted. Future studies should therefore focus on the inflammatory mediators that play a role in blister development in HSP and may be the subject of more focused treatments. It is possible to hypothesis that bullous skin lesions develop more frequently in HSP patients who have extrinsic (such as physical skin pressure) or intrinsic (such as genetic predisposition, general skin fragility) risk factors, but that the pathophysiology of HSP itself is not reflected in the appearance of bullous skin lesions. [9]

There is ongoing debate on the best course of action for patients with bullous HSP. Systemic corticosteroids appear to reduce the frequency of purpura and shorten the duration of arthralgia and stomach discomfort in patients with HSP. Corticosteroids may lower the risk of end-stage renal disease, even if they do not appear to prevent renal presentation during the course of the disease. Systemic corticosteroids do not appear to be helpful in treating non-bullous skin lesions in people with Huntington's syndrome. Certain writers of documented instances involving bullous HSP proposed that prompt systemic corticosteroid therapy could be advantageous in bullous HSP by expediting the resolution of cutaneous manifestations and diminishing the creation of scars. [10]

CONCLUSION

Henoch-Schönlein purpura is a frequent paediatric vasculitis that is multi-systemic. Although the prognosis is often quite good, some individuals may experience long-term effects, usually in the kidneys. Improving the results for these kids requires a multi-institutional, multi-specialty collaborative network, with the ultimate goal being to eradicate the long-term kidney effects of : Henoch-Schönlein purpura.

REFERENCES

1. Jennette JC, Falk RJ, Bacon PA, Basu N, Cid MC, Ferrario F, Flores-Suarez LF, Gross WL, Guillevin L, Hagen EC, Hoffman GS, Jayne DR, Kallenberg CG, Lamprecht P, Langford CA, Luqmani RA, Mahr AD, Matteson EL, Merkel PA, Ozen S, Pusey CD, Rasmussen N, Rees AJ, Scott DG, Specks U, Stone JH, Takahashi K, Watts RA. 2012 revised international Chapel Hill consensus conference nomenclature of vasculitides. Arthritis Rheum. 2013;65(1):1–11. {this article available for online read only}
2. Trnka P. Henoch-Schönlein purpura in children. J Paediatr Child Health. 2013;49(12):995–1003.
3. Garzoni L, Vanoni F, Rizzi M, Simonetti GD, Goeggel Simonetti B, Ramelli GP, Bianchetti MG. Nervous system dysfunction in Henoch-Schonlein syndrome: systematic review of the literature. Rheumatology (Oxford). 2009;48(12):1524–9.
4. Ting TV. Diagnosis and management of cutaneous vasculitis in children. Pediatr Clin North Am. 2014;61(2):321–46. Epub 2014 Jan 21. {this article available for online read only}
5. Bluman J, Goldman RD. Henoch-Schönlein purpura in children: limited benefit of corticosteroids. Can Fam Physician. 2014 Nov;60(11):1007-10. PMID: 25551129; PMCID: PMC4229160.
6. Oni L and Sampath S (2019) Childhood IgA Vasculitis (Henoch Schonlein Purpura)—Advances and Knowledge Gaps. Front. Pediatr. 7:257. doi: 10.3389/fped.2019.00257.
7. López JJ, Ángel D, Lancheros D. Púrpura de Schonlein-Henoch con compromiso abdominal, descripción de un caso y revisión de la literatura. Rev CES Med. 2013; 27(2):243-254.
8. Ramelli V, Lava SA, Simonetti GD, Bianchetti MG, Ramelli GP, Milani GP. Blistering eruptions in childhood Henoch-Schönlein syndrome: systematic review of the literature. Eur J Pediatr. 2017 Apr;176(4):487-492. doi: 10.1007/s00431-017-2858-3. Epub 2017 Feb 4. PMID: 28161822.
9. Ishii Y, Takizawa T, Arakawa H, Saga R, Mochizuki H, Tokuyama K, et al.. Hemorrhagic bullous lesions in Henoch-Schonlein purpura. Pediatr Int. (2005) 47:694–7. 10.1111/j.1442-200x.2005.02133.x
10. Nothhaft M, Klepper J, Kneitz H, Meyer T, Hamm H, Morbach H. Hemorrhagic Bullous Henoch-Schönlein Purpura: Case Report and Review of the Literature. Front Pediatr. 2019 Jan 22;6:413. doi: 10.3389/fped.2018.00413. PMID: 30723709; PMCID: PMC6349767. . {this article available for online read only}