Journal of Pakistan Association of Dermatologists. 2024;**34(3)**:790-794.

Case Report

Rapid recovery in Henoch Schonlen purpura patient with short term pulse dose corticosteroid

Ketut Kwartantaya Winaya, Sonia Elvira Salim*, Ketut Suryana**

Department Dermatology and Venereology, Faculty of Medicine Udayana University, Indonesia.
* Department of Internal Medicine, Wangaya General Hospital Denpasar, Bali, Indonesia.
** Department of Internal Medicine, Merpati Clinic, HIV and Allergy-Clinical Immunology Services Unit, Wangaya Hospital, Denpasar, Bali, Indonesia.

Abstract Henoch Schonlen Purpura (HSP) is systemic vasculitis mediated immune complexes, commonly found in children but have lower incidence in adulthood. Characterized clinically by palpable purpura, renal, joint, and gastrointestinal problem. The prognosis of HSP is excellent, typically to be self-limiting disease, therefore the treatment is usually symptomatic. Therapy glucocorticoid can be given for symptomatic improvement and used for persisting renal disease and gastrointestinal involvement. Glucocorticoid can be given 1-2mg/kgBW/day. This report presents the rapid recovery of HSP Patient in adulthood with Short Term Pulse Dose Corticosteroid. We describe a female patient with HSP, clinically presentation with palpable purpura, arthralgia, and renal involvement. Patient received symptomatic treatment and pulse dose corticosteroid that showed rapid recovery in the first three days of hospitalization. Treatment using pulse dose corticosteroid can advance recovery in adult HSP patients.

Key words
Henoch Schonlen purpura; Adulthood; Pulse dose Corticosteroid.

Introduction

Henloch-Schonlen Purpura (HSP) known as systemic vasculitis is common in childhood.[1] Incidence of HSP in children is estimated to be 15 cases per 100.000 child population with the highest incidence in 4-6 years old. The proportion of males to females is not really different, 2:1. However, HSP can be affected in adulthood. It has lower estimated incidence around 1.3 cases per 100000 population with more severe clinical presentations and worse prognosis than children.[2.3] The proportion of males to females in adulthood is 1:1.[5]

Manuscript
Received on: August 11, 2023
Accepted on: January 07, 2024
Address for correspondence
Dr. Ketut Kwartantaya Winaya SpKK (K)
Department Dermatology and Venerology,
Faculty of Medicine Udayana University,
Denpasar, Bali, Indonesia.
Email: kkwartanaya@yahoo.com

HSP is an acute immune complex systemic mediated and vasculitis leucocytoclastic of unknown etiology. HSP is generally benign because it's self-limiting, yet in some cases that delay diagnosis and getting treatment, some complications can cause death. HSP considered with post infections such as A Streptococci, Hepatitis, CMV, HSV, Human Parvovirus B19, Coxsackievirus and adenovirus.[1] Some factors can also induce deposition immune complex IgA vasculitis such as foods, insect bites, various drugs,[6] and immunizations.[4]

HSP is a vasculitis involving the small vessels, most likely small vessels of the skin, kidneys, joints and gastrointestinal tract. The classic presentations of HSP are palpable purpura, gastrointestinal problems, arthralgias and reducing renal functions. Other manifestations may be present such as fatigue, vomiting, rectal bleeding, hematemesis, headache, subcutaneous edema, scrotal edema, fever and diarrhea.[7]

HSP is generally a self-limiting disease (in 6-8 weeks). Treatment given is basically symptomatic, rest, analgesic and hydration, glucocorticoid can be administrated for symptomatic improvement severe facial or scrotal edema, or for persisting renal disease. Severe renal symptom may require monitoring.[1]

Case Report

A 19-year-old female patient, visited the emergency center of Wangaya Hospital brought by her family on 23th January 2023, with chief complaint pain in both legs since 2 days prior. Pain was felt continuously and worsened when walking, felt on both legs, particularly in area with red spots. Sometimes it was itchy on those spots. She also complained of pain on the knees and ankles. Two days before red patches appeared, she had cough and common cold. Without treatment, the fever went down the next morning, but the red patches appeared on the dorsum pedis bilaterally and then rapidly spread to upper legs. She was given some drugs by the general practitioner but there was no improvement, the skin lesions increased in number. She also complained about abdominal pain and nausea, but there was no vomiting. She had good intake of food and hydration. She did not complain about mixturation and defecation. She had never experienced any similar symptoms prior to this. She had experienced Steven-Johnson Syndrome at 5 years old, known allergic to Sulfa drugs.

The general examination on the first day revealed an alert girl who looked weak, no sign of anemic, icteric, cyanotic and respiratory distress. There was pharynx redness, with tonsil T2-T2. There was no abnormality on thorax and abdominal examination. The dermatological examination of both lower legs unto buttocks showed purpura and redness patches. The purpura was variative, lenticular and nummular. There was edema and pain on the lesion. The blood pressure was 122/82 mmHg, pulse rate was 101 times per minute, temperature was 37.1 C, respiration 20 times/minute and saturation oxygen 98% on room air.

Laboratory examination found: hemoglobin 14 g/dL, erythrocytes: 4.93 juta/µL, hematocrit 43%, MCV: 87.2 fl, MCH: 28.4 pg, MCHC: 32.6 g/L, platelet 293.000/ uL, leucocyte: 7.980/uL, with counted basofil/ eosinofil/ neutrofil/ limfosit/ monosit, each of them: 0.3/1.9/60.1/29.3/8.4, blood sugar: 98 mg/dL, ureum: 25 mg/dL, creatinine: 1.0 mg/dL, electrolite Natrium: 136 mmol/L, Kalium 3.6 mmol/L, Clorida 107 mmol/L. On urinalysis, there is no leucocyte esteration, nitrit, protein, glucose, keton, bilirubin, blood, erythrocyt, leucocyte sediment, cylinder sediment. There was amorf crystal and bactery on urinalysis.

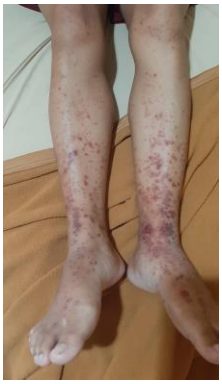

Figure 1 Purpura lesions on Day 1.

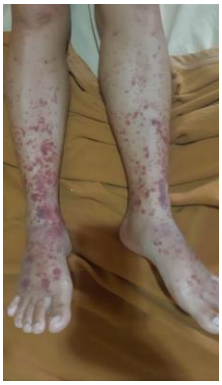

Figure 2 Purpura lesions on Day 2.

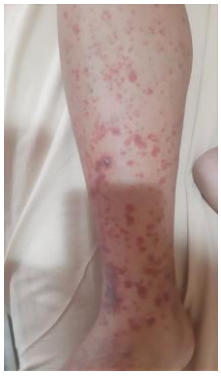

Figure 3 Multiple tenses and flaccid bullae.

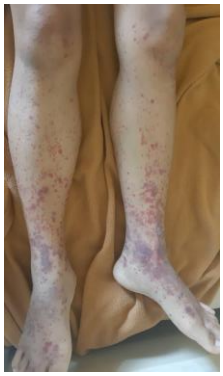

Figure 4 Purpura lesions on Day 7.

Table 1 ACR and EULAR criteria.

ACR (1990)	EULAR / PReS (2006)
Three or more criteria below:	Must have:
1. 20 years or less at onset.	(i) Palpable Purpura with lower limb predominance.
2. Palpable purpura.	At least one criteria below:
3. Acute abdominal pain with gastrointestinal bleeding.	1. Diffuse abdominal pain.
4.Biopsi shown granulocytosis in the walls of small arterioles or venules in superficial layers of skin.	2. IgA deposition (in biopsy).
	3. Arthritis or Arthralgia.
	4. Renal involvement.

The conclusion was observation purpura with differential diagnosis suspect HSP, or drug allergy. Anamnesis, physical examination and laboratory result supported the involvement of skin, gastrointestinal, joint and kidneys disorder. In this case, there was purpura redness patches on skin, arthropathy, and increase in kidney function marked by a comparison of creatinine and urea values >15 times without hematuria and proteinuria. Biopsy IgA deposition could not be performed because of a lack of facilities and funding.

The patient was treated with intravenous line NaCl 0.9% for the fluid balance, intravenous injection esomeprazol 4mg once daily, injection ceftriaxone 2gm daily, injection methylprednisolone 62,5mg twice daily, and loratadine 10mg orally twice daily. On provocation test, for injection antibiotic, it was found that patient was allergic to ceftriaxone. Therefore, the antibiotic was changed to macrolide group with low dosage, azithromycin 250mg orally twice daily.

On the second day of admission, patient still complained of a sore throat, itchiness on the purpura spots, pain on both legs especially on the knees and ankle. There were multiple tense and flaccid bullae and vesicles on multiple palpable erythematous purpura bases. Abdominal pain was reduced, no complaint about mixturation and defecation. At this stage, we increased injection Methylprednisolone dosage to be 125mg twice daily and added Chlorpheniramine Maleat 4mg oral twice daily to reduce the itchiness.

On the third day, patient stated that sore throat, pain and itching had already lessened. Injection pulse dosage methylprednisolone continued until third day and then was tapered off until 7th day. Other treatment continuously given until patient was discharged.

Discussion

Henoch-Shconlen purpura is a systemic vasculitis, immune complex-mediated, associated with Immunoglobulin A.[8] The clinical outcome is palpable purpura, arthralgia, gastrointestinal involvement (such as abdominal pain or gastrointestinal bleeding), and renal involvement.[9]

The current criteria modification to diagnose HSP from American College of Rheumatology (ACR) 1990 being European League Rheumatism (EULAR) and Pediatric Rheumatology European Society (PReS) 2006.

HSP commonly found in children, but it can be found in adults. Etiology of HSP has not been determined, yet it is usually associated with infection. In some cases shown there was respiratory tract infection accompanied with malaise and fever, Hepatitis A, B infection, CMV infection, HIV infection, adenovirus, mycoplasm, herpes simplex, varicella infection and some drugs (penicillin, eritromycin), neoplasm, and some spesific conditions such as pregnancy.[9,10] Some other factors that can induce HSP, are environmental factors, genetic influences, most common in Asian population. Genes also play a part associated with immunity,

certain human leucocyte antigen (HLA) alleles, influence the vascular system such as endothelial nitric oxide synthase (eNOS), angiotensin-converting enzyme, interleukin 18, chemokine monocyte protein chemo attractant protein and transforming growth factor.[12]

The final diagnosis in this case was on history and physical examination. Before purpura appeared, patient had been complaining of sore throat with fever. The next morning, purpura redness patches appeared on lower limb bilaterally and spread to upper part of the lower limb until buttock. It came with pain on the lower joints especially in ankle and knee.

Patients (21.74%) with HSP, had upper respiratory infection before redness patches appeared (Jun Cui, 2019).[11] Negara also reported there was HSP case in adults that started with upper respiratory infection.[9] According to American College of Rheumatology, the typical criteria to diagnose HSP is distributed purpura dominance in lower limb extremity, that does not go away with pressure. Bulla hemorrages can be found in some rare cases that are mostly followed by secondary infection that needed antibiotic for the treatment.[13] Joint pain was also found in 60.87% patient with HSP.[11]

Patient was given esomeprazol as a proton pump inhibitor to relieved nausea. On the first day, patient received injection methylprednisolone with dosage 2 mg/kg/BW. Early treatment with glucocorticoid has been effective to inhibit inflammation process in HSP patients.[15] Based on Oxford Rheumatology, corticosteroids have function to reduce symptomatic complaint and reduces severe oedema.[1] Some literature found giving corticosteroid effectively reduces manifestation on HSP patient which is involved in gastrointestinal and renal problem.[16] Giveing corticosteroid as early treatment can reduce risk complication in end stage renal failure and reduce average time to fully recover. Dosage of corticosteroid can start from 1mg/kg/BW over two weeks and tapering off at third and forth otherwise it can be given 2mg/kg/BW in one week and tapering off on second and third week. Both of these show good recovery within 14 days. More data is needed to exclude the harm of using corticosteroid based on the onset, duration and dosage that given.[17]

In this case, patient received pulse methylprednisolone as much as 4mg/kg/BW twice daily for 3 days, and it boosted the recovery of the skin.

Pulse corticosteroid is defined as intravenous corticosteroid administration with high doses. The aim of this concept is the use of mega doses corticosteroid, to achieve stronger and quicker efficacy while decreasing the need for the long term use of corticosteroids. It can be defined by given >250mg prednisone or it's equivalent per day, for one or more days.[18]

Conclusion

HSP is a self-limiting disease, but in some cases can cause complications such as gastrointestinal bleeding, decreased renal function until death.

In Accordance with EULAR guideline, the treatment can be supportive, bed rest, nutrition and hydration. Corticosteroid can be given to severe cases with gastrointestinal and renal related problems. In this case, giving short term pulse corticosteroid advanced recovery in adult HSP patient.

Declaration of patient consent The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship None.

Conflict of interest Authors declared no conflict of interest.

Author's contribution

KKW, KS: Identification, diagnosis and management of the case, critical review, final approval of the version to be published.

SES: Diagnosis and management of the case, manuscript writing, final approval of the version to be published.

References

1. Clunie G, Wilkinson N, Nikiphorou E, Daepak J. Henoch-Schonlein Purpura (HSP). In: Oxford Handbook of Rheumatology. United Kingdom: Oxford University Press. 2018; 5 pp. 502-4.
2. Yi F, Bai Z, Li Y, Xu X, Guo X, Qi X. A good response to glucocorticoid for Henoch–Schönlein purpura with abdominal pain and gastrointestinal bleeding in an adult: A care case report. *Medicine*. 2020; **99**:1(e18602).
3. Oni L and Sampath S. Childhood IgA Vasculitis (Henoch Schonlein Purpura)-Advances and Knowledge Gaps. *Front Pediatr 2019*;**7**:257. doi: 10.3389/fped.2019.00257
4. Langford A, Fauci A. IgA Vasculitis (Henoch Schonlein). Harrison's Principles of Internal Medicine. New York: McGraw- Hill. Ed 2017. pp 172-3.
5. Duvuru G, Stone JH. Henoch-Schonlein Purpura. In: Imboden JB, Helmann DB, Stone JH. Current Diagnosis and Treatment Rheumatology. 3rd ed. New York: McGraw-Hill Education; 2013. p. 305– 8.
6. Ball GV, Fessler BJ, Bridges SL, Jr. Oxford Textbook of Vaskulitis. In: Gonzales-Gay MA, Blanco R, eds. IgA vasculitis (Henoch Schonlein purpura). 3rd ed. United Kingdom: Oxford University Press; 2014. p. 527-42.
7. Robinson, P, Hotwagner D. Henoch Schonlen Purpura [Internet]. *StatPearls*. 2022. Available from: https://www.ncbi.nlm.nih.gov/books/NBK537252/.
8. Hocevar A, Rotar Z, Jurcic V, Joze P, Cucnik S, Vizjak A, *et al*. Ig A vasculitis in adults: the performance of the EULAR/PRINTO/PRES classification criteria in adults. *Arthritis Res Ther*. 2016;**18**:58. doi: 10.1186/s13075-016-0959-4.
9. Negara A, Zubir Z. Henoch-Schonlein purpura (HSP) in an adult. IOP Conf Series: Earth and Environmental Science. 2018;**208**:125. doi : 10.1088/1755-1315/125/1/012155
10. Brogan P, Eleftheriou S, Dillon M. Small vessel vasculitis. *Pediatr Nephrol*. 2018; **25**:1025-35. doi: 10.1007/s00467-009-1317-4.
11. Cui J, Huang L, Guo J, Rong C, Zhang B. Diagnosis and treatment of adult mixed-type Henoch-Schonlein purpura. *Cent Eur J Immunol*. 2019;**44(2)**:138–43. Doi: https://doi.org/10.5114/ceji.2019.87064.
12. Oni L, Sampath S. Childhood IgA Vasculitis (Henoch Schonlein Purpura)-Advances and Knowledge Gaps. *Front Pediat.* 2019; 7:257. doi: 10.3389/fped.2019.00257.
13. Henoch Schonlen Purpura. American College of Rheumatology. https://www.rheumatology.org/I-Am-A/Patient-Caregiver/Diseases-Conditions/Henoch-Sch%C3%B6nlein-Purpura-HSP. Cited 3 February 2023.
14. Alfarra Y. Management of Henoch-Schonlen Purpura case in children. *Jurnal Wiyata*. 2021;**8**:2.
15. Prameswari R, Indramaya DM, Sawitri S. Haemorrhagic Bullous Lesion in Henoch-Schonlein Purpura. *BIKK*. 2016:**28(2)**:161-7.
16. McPartland K, Wright G. Acute abdominal pain: Henoch-Schönlein purpura case in a young adult, a rare but important diagnosis. *Clin Med (Lond)*. 2019;**19(1)**:77-9.
17. Pamela F, James A, Xianqun L, Jon M, Chris F. Effects of Corticosteroid on Henoch-Schonlein Purpura: A Systematic Review. *National Institutes*. 2007. **120(5)**:1079-87. doi: 10.1542/peds.2007-0667.
18. Yonatan Edel, Tomer Avni, Daniel Shepshelovich, Shelley Reich, Benaya Rozen-Zvi, Michal Elbaz *et al*. The safety of pulse corticosteroid therapy-Systematic review and meta-analysis. *Semin Arthritis Rheum*. 2020;**50(3)**:534-45.