



LOW DOSE CORTICOSTEROID FOR PEDIATRIC HENOC SCHONLEIN PURPURA TREATMENT: A CASE REPORT AND REVIEW OF THE LITERATURE

Edwin Destra¹, Charity Harlim¹, Yohanes Firmansyah², Irene Dorthy Santoso³, Amelia Setiawati Soebyanto³

¹Medical Co-Assistant, Faculty of Medicine Tarumanagara University, Jakarta, Indonesia

²Medical Doctor, Faculty of Medicine Tarumanagara University, Jakarta, Indonesia

³Dermatovenereologist, Sumber Waras Hospital, Jakarta, Indonesia

Corresponding Author: Edwin Destra, Universitas Tarumanagara

Received 28 Oktober, 2021; **Accepted** 31 Oktober, 2021; **Online Published** 28 Januari, 2022

Abstract

Henoch-Schonlein Purpura (HSP) is a common cause of pediatric microvascular vasculitis. A four-year-old boy presented with abdominal pain, swelling, and rashes on both legs, all of which were associated with multiple episodes of vomiting over the preceding ten days, as well as a history of melena and joint pain. We describe a 4-year-old boy with Henoch Schonlein purpura and severe genitourinary edema. He was successfully handled with about half of the normal steroid dose, which proved to be effective for this situation. Early detection and treatment are associated with a better prognosis in cases of Henoch Schonlein Purpura.

Keywords: Henoch Schonlein Purpura, Steroids, Methyl-Prednisolone, Low Dose Treatment, Children, Vasculitis

Introduction

Henoch Schonlein Purpura is the most frequent kind of systemic vasculitis in children; it is characterized by the deposition of immunological complexes including the antibody IgA; the precise origin of this occurrence is unclear. Purpura, rheumatoid arthritis, and stomach discomfort are

together referred to as the Henoch Schonlein Purpura "classic trio." Scrotal illness, lung disease, carditis, and CNS involvement are uncommon manifestations of HSP. Historically, children with HSP who exhibit no unusual symptoms have been treated with symptomatic medications. Steroids are used for the treatment of severe symptoms at a

dosage of 1-2 mg/kg/day. We describe an unusual case of a 4-year-old child who initially showed with the typical triad of HSP and had genitourinary edema on clinical examination. The swelling disappeared with half the prescribed dosage of the steroid. The purpose of this article is to explain HSP and its therapy, as well as to conduct a literature review.

Case Report

A 4-year-old kid, a primary school student, was referred by the Primary Public Health Service Grogol Petamburan to the Department of Dermatology's Outpatient Department Sumber Waras Hospital with a history of Rash on both feet six days earlier that progressively extended to the knees, thighs, and buttocks. The rash was followed by swelling and discomfort in the knees. A few days later, the child's mother noticed some swelling of the penis and scrotum with erythema. Multiple bouts of nausea and vomiting preceded the start of the rash, but there was no history of hematuria, genital injuries, or urinary tract infection. Later in the boy's life, he complained of minor stomach discomfort and bloody diarrhea. The abdominal pain was centered around the umbilicus, was rapid in onset, and was non-radiating. The kid was not in distress upon arrival, and his vital signs were within

normal limits. On physical examination, both legs had moderate bilateral nonpitting edema. A non-tender, non-blanching petechial/purpuric rash was seen, spreading widely across both feet and going up to the knees, buttocks, and lower abdomen (Figure 1). His abdominal examination revealed no discomfort. The white blood cell count was normal, and the erythrocyte sedimentation rate was 21 mm/hour. Platelet count increased significantly to $417 \times 10^9/L$.

The American College of Rheumatology and European League Against Rheumatism/Pediatric Rheumatology Society criteria were used to diagnose HSP. Additionally, the clinical appearance of the rash and its history fit the "classic triad" of Henoch Schonlein Purpura. The patient was given 0.5mg/kg/day of oral steroid, half the usual dosage. The patient received steroid treatment and was seen three days later. Purpura and bilateral swelling of the legs gradually resolved, although hyperpigmented macules remained. Abdominal discomfort has subsided, as has genitourinary edema and erythema. Due to the remarkable recovery, the steroid dosage was reduced to 0.25mg/kg/day (Figure 2). After a week after the last control, there were no additional issues, and the steroid was discontinued (Figure 3).



Figure 1 (A) Swelling and erythema of penis and scrotum, (B and C) Purpuric rash on both feet



Figure 2 (A) Penis and scrotum has been resolved, (B and C) Purpuric rash has gradually improved and hyperpigmented macules



Figure 3 (A) Penis and scrotum has no further problem, (B and C) Purpuric rash has been resolved with hyperpigmented macules as remains

Discussion

Henoch Schonlein purpura (HSP) is a non-blanching rash that most frequently affects children aged 3-15 years. The skin and connective tissues, scrotum, joints, gastrointestinal system, and kidneys are all affected by HSP. HSP is often preceded by an illness, such as strep throat. The distinctive non-thrombocytopenic purpura that

occurs in almost all patients across the limbs and buttocks is the disease's hallmark. Following the prodrome, a variety of symptoms emerge, including rash (95-100 percent of cases), stomach pain and vomiting (35-85 percent), joint pain (60-84 percent), subcutaneous edema (20-50 percent), scrotal edema (2-35 percent), and bloody stool.

Our patient often exhibits the Henoch Schonlein tetrad. Purpura, arthralgia, and stomach discomfort, as well as edema of the scrotum and penile region. Clinically, the non-blanching rashes present as palpable purpura on the lower legs and arms. Joint involvement is usually characterized by pain and swelling in the joints, with the knees and ankles being the most commonly affected. Abdominal discomfort is the most common gastrointestinal symptom, followed by vomiting and intestinal hemorrhage. The American College of Rheumatology 1990 criteria and the European League Against Rheumatism/Pediatric Rheumatology Society 2006 criteria were used to diagnose HSP.

Henoch Schonlein Purpura has a worldwide prevalence of about 6–22 per 100,000 person-years in children, which is much greater than the global prevalence in adults (3.4–14.3 per 100,000 person-years). Children under the age of 17 occur at a rate of 6.2 to 70.3 per 100,000 years, with a little male preponderance (M: F = 1.2: 1.0). The peak age of onset is 4–6 years, and 90% of HSP cases occur before the age of ten, with the majority occurring before the age of five. It affects 50% of children under the age of five and 75% of children under the age of ten, and the prevalence of HSP cases declines with age. Asians have the greatest infection rate, whereas African-Caribbean's have the lowest. The rate in North America is 13.5 per 100,000 children, with the greatest rate among

Caucasians and the lowest among Afro-Americans. Winter and spring are the most frequent seasons for HSP to occur. In Indonesia, 71 individuals with HSP were included in a research; they had a little female preponderance of 1,2:1. The age ranges from two to sixteen years, with an average of seven and a half.

Henoch Schonlein Purpura treatment has been adjusted according to the degree of renal involvement. Treatment is symptomatic in the absence of renal dysfunction. If there is necrosis of the skin with ulceration, wound care may be necessary. Compression and elevation of the afflicted region may be helpful when lower leg edema develops. Individuals experiencing stomach or joint discomfort may need bed rest and pain medication. HSP therapy aims to relieve acute symptoms, decrease short-term morbidity, and avoid long-term renal damage. Oral steroids should be given to patients with a moderate rash, edema, and stomach pain (without nausea or vomiting), as well as lung, scrotum, or testicular involvement. Prednisolone is the most often prescribed steroid for the treatment of HSP. Though dexamethasone was utilized in certain instances, there is no evidence in the literature to support one over the other. Prednisone or methylprednisolone is often begun at a dose of 1 to 2 mg/kg per day for one to two weeks before being reduced to 0.5 mg/kg per day the next week and to 0.5 mg/kg every other week over the following year. HSP is often a self-

limiting illness with a favorable prognosis in individuals who do not have renal dysfunction. Patients often recover fully within four weeks, but approximately one-third of patients have a recurrence within four to six months after the original start.

Conclusion

Henoch-Schonlein Purpura is one of the most prevalent vasculitis in children, and its characteristic triad of palpable purpura, rheumatoid arthritis, and stomach involvement facilitates diagnosis. Early start of steroid therapy will aid in symptomatic alleviation and result in a favorable outcome. Renal illness may need long-term monitoring; nevertheless, the condition has a good prognosis.

Acknowledge

We would like to convey our heartfelt appreciation to the physicians and administration of Sumber Waras Hospital for their assistance. Additionally, we want to express our gratitude to Dr. Irene Dorothy Santoso and Dr. Amelia Setiawati Soebyanto, who served as our supervisors. Not to mention, we would want to express our gratitude to all of our supervisors in our dermatovenereologist rotation at Sumber Waras Hospital for sharing their expertise and experiences with us throughout our clinical rotation. Additionally, we appreciate Mrs. Muchti Widarsih's help with patient care during our

dermatovenereologist rotation at Sumber Waras Hospital.

DAFTAR PUSTAKA

1. Reamy B V, Servey JT, Williams PM. Henoch-Schönlein Purpura (IgA Vasculitis): Rapid Evidence Review. *Am Fam Physician*. 2020;102(4):229-233.
2. Sohagia AB, Gunturu SG, Tong TR, Hertan HI. Henoch-Schonlein Purpura—A Case Report and Review of the Literature. *Gastroenterol Res Pract*. 2010;2010:1-7. doi:10.1155/2010/597648
3. Chimenz R, Cannavò L, Spinuzza A, et al. Unusual presentation of Henoch-Schönlein purpura. *J Biol Regul Homeost Agents*. 33(5 Suppl. 1):69-74.
4. Ebert EC. Gastrointestinal Manifestations of Henoch-Schonlein Purpura. *Dig Dis Sci*. 2008;53(8):2011-2019. doi:10.1007/s10620-007-0147-0
5. Leung AKC, Barankin B, Leong KF. Henoch-Schönlein Purpura in Children: An Updated Review. *Curr Pediatr Rev*. 2021;16(4):265-276. doi:10.2174/1573396316666200508104708
6. Oni L, Sampath S. Childhood IgA Vasculitis (Henoch Schonlein Purpura)—Advances and Knowledge Gaps. *Front Pediatr*. 2019;7. doi:10.3389/fped.2019.00257
7. Bluman J, Goldman RD. Henoch-Schönlein

purpura in children: limited benefit of corticosteroids. *Can Fam Physician*. 2014;60(11):1007-1010.

8. Zhang F, Chen L, Shang S, Jiang K. Atypical purpura location in a pediatric patient with Henoch-Schönlein purpura. *Medicine (Baltimore)*. 2018;97(48):e13294. doi:10.1097/MD.00000000000013294
9. Çakıcı EK, Gür G, Yazılıtaş F, et al. A retrospective analysis of children with Henoch–Schonlein purpura and re-evaluation of renal pathologies using Oxford classification. *Clin Exp Nephrol*. 2019;23(7):939-947. doi:10.1007/s10157-019-01726-5
10. Roman C, Dima B, Muyshont L, Schurmans T, Gilliaux O. Indications and efficiency of dapsone in IgA vasculitis (Henoch-Schonlein purpura): case series and a review of the literature. *Eur J Pediatr*. 2019;178(8):1275-1281. doi:10.1007/s00431-019-03409-5