

Henoch-Schonlein Purpura in Children: The Role of Corticosteroids

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Abstract

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BACKGROUND: Henoch-schonlein purpura (HSP) is an IgA-mediated systemic small vessel vasculitis. It is the most common form of systemic vasculitis in children.

CASE REPORT: A 9 years old girl admitted to the hospital with chief complain of purplish red rash on both legs since approximately 1 week with painful knees and ankles that make the patient unable to walk. The patient was diagnosed with HSP and was treated with corticosteroid and analgesics. The patients only stayed for 2 nights at the hospital and discharged from the hospital with the ability to walk and experience no pain.

CONCLUSION: The role of corticosteroids in the treatment of HSP is still controversial. But from various research, we can conclude that the role of corticosteroid in HSP is as a symptom reliever (reduce abdominal pain and arthritis), but does not slow the progression of renal disease.

Introduction

Henoch Schonlein Purpura (HSP) is an IgA-mediated systemic small vessel vasculitis with a predilection for the skin, gastrointestinal tract, joints, and kidneys [1]. It is characterised by palpable purpura (without thrombocytopenia), abdominal pain, and arthritis. HSP is a self-limiting disease but can cause complication such as gastrointestinal haemorrhage, intussusception and end-stage renal disease (ESRD) [2]. Renal involvement in HSP affects 20-70% of patients and ranges in severity from microscopic hematuria with or without proteinuria to a nephritic/ nephrotic pattern with associated renal failure. In the majority of patients the outcome is excellent, with the incidence of severe long-term morbidity/ mortality being less than 5% [3].

Case Report

A 9 years old girl admitted to the hospital with chief complain of purplish red rash on both legs since approximately 1 week. The rash came with painful joints on the knees and ankles that make the patient unable to walk for several days. No symptom of abdominal pain and the common cold. No fever and bloody stool/ rain were reported. From physical examination: composmentis, BP 100/70, HR 80 bpm, RR 18 times per minute, and temperature 36.5°C. Physical examination from head to toe, there is a manifestation of a red purplish rash (purpura) on both of the legs below the knees, no swelling, but the reduced movement of the legs due to painful joints. No abdominal tenderness present.



Figure 1: Purpura on the leg

Routine laboratory test was performed, from the complete blood count (CBC) can be seen elevated thrombocyte count just a little (380.000 / ul), normal prothrombin time (PT) and activated partial thromboplastin time (aPTT), normal creatinine and ureum, but there is proteinuria in the urinalysis with no hematuria nor microhematuria. The patient was given medication with oral Prednisone 3 x 15 mg and ibuprofen syrup 3 x 6.25 ml. After the treatment, the painful knees disappear, but the rash still exists. And the patient was discharged from the hospital after 2 night's stay in the hospital with better clinical manifestation.

Discussion

Henoch Schonlein purpura is a self-limiting condition, usually resolving within 6 to 8 weeks. In patients with HSP, immunoglobulin A (IgA) immune complexes are deposited in the small vessels, which causes petechiae and palpable purpura. All HSP patients develop a nonpruritic rash that starts briefly as an erythematous papule or urticarial wheels and then matures into crops of petechiae and purpura. Purpura is defined as nonblanching cutaneous haemorrhages. The lesions change from red to purple to rash coloured before fading over approximately 10 days. The rash is most commonly located in dependent areas that are subject to pressure such as the lower extremities, belt line and buttocks. Non-Migratory arthritis occurs in 75% of patients. The knees and ankles are more commonly involved than small joints. The arthritis symptoms include swelling, warmth, and tenderness. Abdominal pain occurs 60-65% of patients and can mimic an acute abdomen in terms of severity [4]. According to the European League Against Rheumatism (EULAR) and Paediatric Rheumatology European Society (PRES), the diagnosis should be based on the finding of palpable purpura in the presence of at least one of the following criteria, namely, diffuse abdominal pain, arthritis or arthralgia, renal involvement (hematuria and/or

proteinuria), and a biopsy showing predominant IgA deposition [1].

In this case, the patient has a purplish red rash (purpura) on both of the legs below the knees with non-migratory arthritis on the knees and ankles. Diagnosis Criteria for HSP according to International Consensus Conference 2006 are palpable purpura in the presence of one or more of the following: Diffuse abdominal pain; any biopsy showing predominant immunoglobulin A deposition; arthritis (acute, any joint) or arthralgia; and renal involvement (any hematuria or proteinuria) [4].

In most cases, HSP is mild and self-limiting, requiring only symptomatic treatment. Bed rest and analgesics may be required for those with arthralgia or abdominal pain. The skin manifestations rarely need treatment [5]. The goals of treating HSP are to relieve acute symptoms, prevent short-term morbidity (such as abdominal complications) and prevent chronic renal insufficiency. Because HSP is characterised by leukocyte infiltration of the blood vessel walls along with immunoglobulin A disposition, and because corticosteroids inhibit inflammatory process, early treatment with corticosteroids has been postulated to be effective for all 3 therapeutic goals, but still much controversy remains [2]. There is no consensus regarding the indication of steroid use in HSP, but there were several studies that found the efficacy of corticosteroid for HSP.

According to Reamy et al, Oral prednisone at 1 to 2 mg per kg daily for two weeks has been used to treat moderate to severe abdominal and joint symptoms. A double-blind, randomised trial found that early treatment with prednisone reduced abdominal and joint pain severity in children. Although prednisone did not prevent renal disease. A meta-analysis found that corticosteroid use in children with HSP reduced the mean time to resolution of abdominal pain [4]. Bluman et al stated that current evidence does not support universal treatment of HSP with corticosteroids, as they do not appear to prevent the onset of renal disease or abdominal complications. However, corticosteroids do seem to have a role in the symptomatic management of HSP, specifically in treating abdominal pain, arthralgia and purpura [6], [7].

A study by Welch shows that steroids had no apparent effect on the development of nephritis, although there was a tendency for the renal disease to "resolve" more readily in the treated group [8]. A double-blind, randomised trial by Ronkainen J et al found that early treatment with prednisone reduced abdominal and joint pain severity in children [9]. But, from a recent large scale, RCT found that steroid treatment does not reduce the incidence and severity of nephropathy in patients with HSP [3]. It is supported by the study from Huber et al. who did not find any difference between the corticosteroid group and control group [11]. In this case, the patient was given

corticosteroid, and the painful joints disappear within 1 day; although, the purplish red rash did not diminish.

From the explanation above, the patient was having a Henoch Schonlein Purpura with the symptom of purpura on both legs, with arthritis on the knee and ankle that make her unable to walk with proteinuria in the urinalysis. She was treated with prednisone orally, and the next day the pain is gone. From many studies, it can be concluded that the role of corticosteroid is to relieve the abdominal pain and arthritis; but it is not to cure the HSP itself nor prevent the renal disease; therefore, corticosteroids were used just as a symptom reliever.

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