Successful Steroid Therapy of Henoch Schonlein Purpura in Young Adult Woman with Moderate Symptoms: A Case Report

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Abstract

Henoch schonlein purpura is a rare incidence case and almost similar to the clinical symptoms of autoimmune diseases such as systemic lupus erythematus in young adult woman. A 23-year old woman patient presented with reddish spots accompanied by a burning sensation and pain in both legs, vomiting profus and abdominal pain. She also complained mild fever, frequent mouth sores, hair loss, joint pain and stiffness, frequent fatigue in the last 1 month. Laboratory tests showed mild anemia, leukocytocis, proteinuria, erythrocytes and leukocytes microscopic in urinalysis, increased of serum IgA and LED. The skin biopsy showed neutrophil cell infiltration in the capillary vessel walls with necrotic debris. Methylprednisolone oral was given since she hospitalized and decreased gradually. This case hada good prognosis because the result of serum creatinine was normal insix months latereven though had moderate symptoms.

Keywords: *Henoch schonlein purpura, moderate symptoms, adult woman*

Introduction

Henoch-schonlein purpura (HSP) is a small blood vessel vasculitis with IgA-dominant immunedeposits and involvesthe skin, gastrointestinal tract(abdominal

Until now, the etiology of HSP is still uncertain. IgA is presumed as an important role in the pathogenesis of HSP which is characterized by an increase in serum IgA concentrations, immune complexes, and IgA deposits on theblood vessels walls and renal mesangium. Several etiological factors that can be attributed to HSP are infection, drugs, food, immunization, and malignancy.t In a retrospective study of the Chinese population in Taiwan, it was found that 40.5% of HSP was preceded by infection, 8.3% was due to drug administration, and the remaining 59.5% were unknown.[4,5]

We report a young adult woman with HSP who is accompanied by mild anemia with a previous suspicion of SLE.The aim of this case report is to describe a rare incidence case, the skin biopsy and successful steroid therapy in adulthood.

Case Description

A 23-year old woman came to emergency department of Dr. Soetomo hospital complained of vomiting profusesince 2 hours earlier, more than 3 times. She also complained of reddish spots accompanied by a burning sensation and pain in both legs which appeared approximately 2 days before admission. Other complaints were mild fever, frequent mouth sores, hair loss, joint pain and stiffness especially in the morning, abdominal pain and heartburn, frequent fatigue when stressed in the last 1 month. There were no history of red

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spots on the face when exposed to the sun, cough, flu, painful urination, diarrhea, taking antibiotics or other drugs before appearing red spots, drug and food allergies, diabetes and hypertension.On physical examination, the blood pressure was 110/60 mmHg, pulse 85bpm, regular rhythm, breathing 18x/minute, axillary temperature 37.1^{0}C. Examination of the extremities revealed palpable purpura bilaterally symmetrically with varying sizes and clear borders without being surrounded by an erythematous area, painful and not blanching when pressed (figure 1).

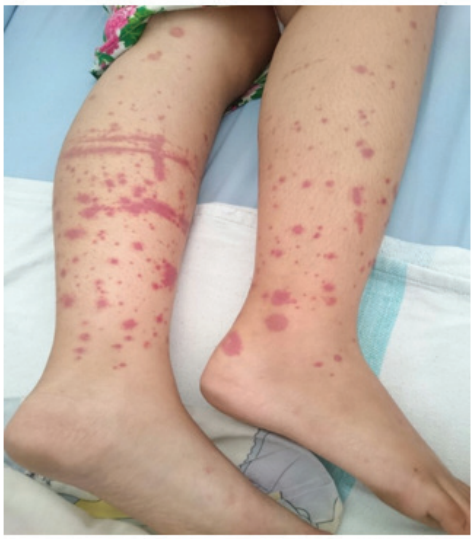

Figure 1. Patient's clinical appearance showed palpable purpura bilaterally symmetrically in both legs with varying sizes

Laboratory tests showed Hb 10.2g/dL, MCV 74.2fL, MCH 23.8pg, MCHC 32g/dL, leukocytes 13,660/mm^3, neutrophils 79.4%, platelets 335,000, AST 10 U/L, ALT 5 U/L, BUN 13 mg/dL, creatinine 0.64 mg/dL, sodium 147 mmol/L, Potassium 4.6 mmol/L. Urinalysis results obtained glucose -, protein +1, bilirubin -, ketones -, nitrite -, leukocytes-,erythrocytes microscopic 3-8, leukocytes microscopic 15-20, Crystals -, Bacteria +/ field of view.

She was treated with omeprazole20mg every 24 hours, paracetamol 500mg every 8 hours, methylprednisolone 16mg every 8 hours. Then,she was examined for peripheral blood smears, serum IgA, Albumin, ESR, CRP, urine culture, ana test, C3, C4 and skin biopsy.In peripheral blood smears, Erythrocytes were obtained: microcytichypo chromic, anisopoikilocytosis, polychromasia cells (+), normoblast (-); The leucocytes count are increase and predominantly granulocyte immature segments; The platelets count are normal and there are no giant platelet. Conclusion: Microcytic hypochromic anemia. The serum IgA was obtained 647 mg/dl, LED 85 mm/hour, Albumin 3.9 mg/dl, CRP 1.8 mg/dl, sterile urine culture, ANA test10.21 AU/mL, C3 158mg/dl, C4 36 mg/dl. The results of skin biopsy were that the epidermis showed a focus of basal cell hyperpigmentation, shortened rete ridges; and in the Dermis there is neutrophil cell infiltration in the capillary vessel walls with necrotic debris; Conclusion: according to HSP (figure 2).

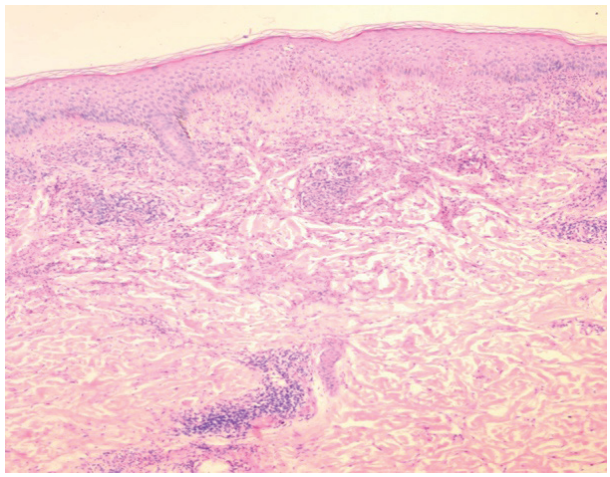

Figure 2. The skin biopsy showed neutrophil cell infiltration in the capillary vessel walls with necrotic debris

On 5th day, the complaints of pain, nausea and reddish spots on the legs were reduced so that the methylprednisolone dose was reduced to 16mg every 12 hours (figure 3). On 7th day there were no complaints and the red spots on the legs disappeared so that the methylprednisolone dose was reduced to 16mg every 24 hours (figure 4). Then,she can go home and control to outpatient unit with the dose of methylprednisolone decreased gradually. Six months later, the serum creatinine was monitored and the resultwas normal.

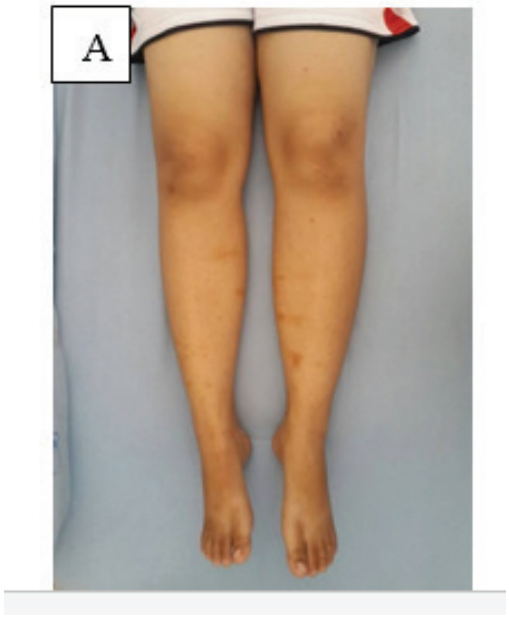

Figure 3. Patient's clinical appearance on 5th day

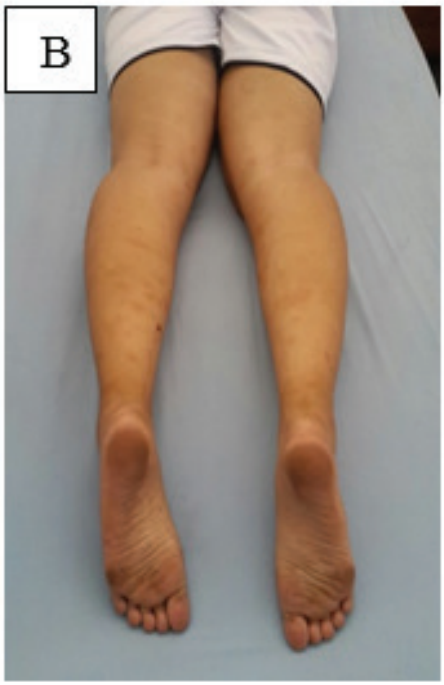

Figure 4. Patient's clinical appearance on 7th day

Discussion

Based on the 2010 EULAR, diagnostic criteria of HSP is palpable purpura, not thrombocytopenic + ≥ one of the following: diffuse adominal pain, typical leukocytoclastic vasculitis with predominant IgA deposits or proliferative glomerulonephritis with predominant IgA deposits, arthritis/arthralgias, renal involvement (proteinuria: >0.3 g/24h or >30 mmol/mg of urine albumin to creatinine ratio on a spot morning sample; and/or hematuria, red blood cell casts: >5 red cells per high power field or ≥2+ on dipstick or red blood cell casts in the urinary sediment).[6]

The differential diagnosis of HSP is very diverse. To differentiate is, first, whether the purpura on the skin is palpable or not. If it is not palpable and there is thrombocytopenia, the diagnosis is immune thrombocytopeniapurpura.Ifitispalpable,anautoimmune panel will be examined such as ANA tests, C3 and C4, if the panel is positive,the vasculitis is caused by SLE. If the panel shows a negative result, the next step is a skin biopsy examination. From the result of a skin biopsy, if a typical picture of granuloma formation is found and there is necrosis, it can be diagnosed as Wagener's granulomatosus andif the resultshows a typical picture of eosinophil infiltration (in asthma/allergic rhinitis), it is diagnosed as Churg Strauss syndrome. If the result shows the form of neutrophil infiltration with necrotic debris (leukocytoclastic), so there are two possibilities, namely hypersensitivity vasculitis and HSP (serum IgA concentration increases).[6,7,8] The diagnosis path in that theory is appropriate in this case.

HSP is actually a self-limiting disease. Currently, the management of HSP is still a matter of debate. Patients need to be hospitalized to control ongoing acute symptoms and renal involvement. Symptomatic treatment is sufficient to reduce mild symptoms such as arthritis, heartburn, and nausea by administering non-steroidal anti-inflammatory drugs and proton pump inhibitors. Skin symptoms due to vasculitis rarely require treatment, but several studies have reported successful therapy with steroids. Oral steroids are indicated for severe rashes, edema, severe abdominal pain and accompanied by nausea and vomiting. Usually, oral steroids can be started at a dose of 1 mg/kg/day and then gradually tapered off. Patients who are unable to take the oral route due to severe nausea and vomiting may be given intravenous steroids. Steroid administration can reduce abdominal symptoms within 2-3 days as well as reduce the chance of recurrence of HSP and the possibility of developing persistent renal impairment. If

accompanied by severe renal disorders such as rapidly progressive nephritis and pulmonary hemorrhage, high doses of corticosteroids are given which can be combined with immunosuppressive drugs. Immunosuppressive drugs such as cyclophosphamide in combination with steroids are usually indicated for patients with severe nephritis.[9,10,11] This case got better with steroids threrapy because she had moderate symptoms and previously suspected SLE related symptoms .

The patient's prognosis depends on age at exposure, recurrence rate, renal involvement (high creatinine on exposure, proteinuria >1g/day, hematuria and anemia when first diagnosed, membranoproliferative gromerulonephritis) and skin area, presence of hypertension, fever at exposure, neurological symptoms and immunological disorders (increased IgA accompanied by decreased IgM at diagnosis, low factor 13), purpura above the waist, persistent purpura, and increased sedimentation.[9] About 85% of adult HSP patients develop nephritis and kidney failure, which has a poor prognosis. Renal disorder usually occur in the first 6 months to 12 months so further monitoring is needed.[12] This case has a good prognosis because there is still no renal involvement even though it occurs at>20 years of age and the presence of gastrointestinal symptoms.

Conclusion

Purpura has many causes and skin biopsy is the last step in determining the cause of vasculitis. This case report demonstrates the successful steroid management of HSP with moderate symptoms in young adult woman. It is a rare case and has a good prognosis in adulthood.

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Ethical Clearance: Not required for a case report.

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