Henoch-schönlein purpura (HSP) in an adult

C A Negara1,2* and Z Zubir1,2

1Division of Allergy and Immunology, Department of Internal Medicine, Faculty of Medicine, Universitas Sumatera Utara, Medan, Indonesia
2Haji Adam Malik General Hospital, Medan, Indonesia
*Corresponding author: abdinegara.citra85@gmail.com

Abstract. Henoch-schönlein purpura (HSP) is vasculitis of the small vessels, the most common vasculitis of the childhood and is uncommon in adults. A case of HSP is reported in a 36-year-old female with ten days history of multiple palpable purpura on region antebrachii, region femoralis and cruris dextra et sinistra. Burn sensation in both legs, pain sensation on knees joint and ankles joint and bloody stools were found. History of a cough and sore throat are often to be a presentation. Laboratory examination was mild anemia, mild leukocytes, ASTO (antistreptolysin titer O): < 200, IgA: 332 mg/dL. The patient treated by giving an injection of methylprednisolone, azathioprine, and at last this treatment apparently bears good result. The account of respiratory tract stated above presumed as the factors of the kindling of the outbreak of HSP to this patient. The prognosis in the adult is worse than children due to an increased risk of disorders of the renal.

1. Introduction
Henoch-schönlein purpura (HSP) is a leukocytoclastic vasculitis involving small vessels with the deposition of immune complexes containing IgA.[1] HSP primarily affects children, with over 50% under five years and over 75% under ten years. Its incidence is approximately between 10-24.3 every 100,000 children per years.[2] HSP is less common in adults; it incidence in adults is only 1.3 per 100,000 with a mean age at presentation of about 50 years. In childhood, the male: female ratio ranges from 1.2:1.6, but male predominance has also been reported. However, in adults, male and female are equally affected.[3] In North America, Caucasians have the highest incidence, and Africa Americans have the lowest incidence.[4]

Etiology of HSP is unknown, but it commonly follows an upper respiratory tract infection.[4] Group A β hemolytic streptococci have been the most common organism cultured in up to 36% of those tested in one series. Other organisms includehepatitis A, B, CMV (cytomegalovirus), HIV (human immunodeficiency virus), adenovirus, mycoplasma. Herpes simplex, Helicobacter pylori, Toxocara canis, Human parvovirus B19, varicella, and scarlet fever.[2] Various drugs, foods, insect bites, exposure to cold weather, trauma and genetic factors have been as triggers.[5]

The clinical features of HSP include a palpable purpuric rash, arthralgia, gastrointestinal involvement (with abdominal pain or gastrointestinal bleeding) and renal involvement).[3]

There is no specific laboratory test that has shown to be helpful in making a diagnosis.[6] It usually based on clinical features. Laboratory test includes full blood count, clotting time, electrolytes, liver profile, renal profile, anti-streptolysin titer O (ASTO), antinuclear antibody (ANA), dsDNA, IgA, urinalysis and occult bleeding in stool aimed at excluding other diagnostic possibilities and assessing the extent of organ involvement such as the kidney.[2]
The acute phase of HSP resolves spontaneously in 94% of children and 89% of adults, therefore therapy only symptomatic and supportive such as rest, adequate hydration, analgesia (nonsteroidal anti-inflammatory drugs/NSAID). But in severe cases such as renal involvement, gastrointestinal, central nervous system or severe skin rash, steroid has been used either alone or combined with immunosuppressive agents.[2,4,6]

The prognosis is excellent in the majority cases, but about 33% have a recurrence of symptoms, especially in older children and adults. The prognosis also depends on the presence or absence of renal involvement.[4]

2. Case report

36-year-old women come up to dermatology outpatients unit at H. Adam Malik hospital with a ten-day history of rash on both her legs. Burn sensation is on both her legs. Three days after getting treatment, the red rash more and more appear both of arms, pain sensation especially on knees and ankles joint, nausea and abdominal discomfort, bloody stool, are also presented. History of a cough and sore throat was presented. Past medical history treatment with oral methylprednisolone 4 mg, four tablets once a daily.

On examination, sensorium is comos mentis, blood pressure, heart rate, respiratory rate and the temperature was normal, multiple palpable purpuric with the size of lesions milliary until lenticular, discrete in the region of antibrachii dextra et sinistra, femoral dextra et sinistra and cruris dextra et sinistra.

The laboratory examination was found Hb 11.9 g/dL, leukocyte 13,080 /μL, platelets 331,000 /μL, urea 24 mg/dL, creatinine 0.57 mg/dL, HST, liver function test were normal, ANA test: 2.19, anti-dsDNA: 10.2, rheumatoid factor: negative, ASTO: < 200, IgA: 332 mg/dL, benzidine test was negative and urinalysis was normal.

She’s currently taking Methylprednisolone intravenous 125 mg twice a day for five days, intravenous Ketorolac 30 mg twice daily, Paracetamol 1000 mg three times daily, Omeprazole 20 mg twice daily, then intravenous Methylprednisolone reduced gradually become 62.5 mg twice daily for six days and added azathioprine 2x50 mg.

During hospitalization, the patient got improvement, the skin rash was reducible and turn into brownish, joint paint, abdominal discomfort was not found anymore. Then, she was discharged home and followed up as an outpatient and getting oral treatment methylprednisolone @4 mg, four tablets three times a daily, omeprazole 20mg twice a daily, Imuran 50mg twice a daily.

![Figure 1. The clinical manifestation of HSP showed.](image)
3. Discussion

HSP is a non-thrombocytopenic systemic vasculitis which primarily affects children, but it’s uncommon in adults.[1,7] Etiology of HSP unknown [1,3], but it’s commonly follow an infection such as upper respiratory tract infection, hepatitis A, B, CMV, HIV, adenovirus, mycoplasma, herpes simplex, varicella, some drugs (penicillin, erythromycin, quinines, chlorpromazine), neoplasms (leukemia, lymphoma, breast, lung, kidney, prostate, colon, cervix cancer and melanoma and several miscellaneous conditions (pregnancy, cryoglobulinemia) can also trigger HSP. A combination of genetic predisposition, environmental and immunological factors play a role in the pathogenesis of HSP.[6]

The clinical manifestations usually present with a classic tetrad of rash, polyarthralgias, abdominal pain and renal disorder.

- The rash occurs in all patients, is characterized as palpable purpura non-thrombocytopenic or coagulation disorder, often symmetrically distributed on the lower legs and arms and buttocks, it may involve face and ears but usually spares the trunk.[2,4]
- Arthritis or arthralgia are present in more than 80% of patients. They most commonly affect the large joints of the lower limbs including knees, ankles, and hips. Symptoms include pain, swelling and decreased the range of movement and left no permanent damage.[2,4]
- The reported incidence of gastrointestinal involvement is between 50-75% of cases, with the most common presentation being a colicky abdominal pain. Other symptoms include vomiting, nausea, gastrointestinal bleeding or positive stools in occult blood.[2,4]
- Renal involvement in HSP is reported to occur in 40-50%; four often presents as glomerulonephritis [4] which manifest as hematuria, proteinuria, red blood cell casts, nephrotic syndrome/nephritis, and hypertension.[2]

In this case, the 36 years old women patient during the ten days observation apparently the account of purpura at both lower and upper extremities found burning sensation, a painful sensation at the knee joints and ankles, queasy sensation, nausea and abdominal discomfort, and bloody defecation. The account of coughing and sore throat was often found also.

The laboratory test may show anemia, mild leukocytosis with a normal platelet count is found, thrombocytosis has associated with more severe disease, occasionally, eosinophilia is present, coagulation test is usually normal, a raised erythrocyte sedimentation rate, abnormal renal and liver function test, however factor XIII activity has been found to be reduced and associated with more severe disease, C3 (complement-3), C (complement-4), have been reported to be low in a few patients, in some studies throat swabs for group A β hemolytic streptococcus were positive in more than 50% patients. Elevation of IgA in the blood occurs in 50% of patients, and a definitive diagnosis is confirmed by a biopsy specimen of the skin or kidney that shows IgA deposition.[2,4,7]
At the diagnostic of the physical test was found multiple palpable purpuric with the size of lesions miliary until lenticular, discrete in region antebrachii dextra et sinistra, region femoralis sinistra et dextra and cruris sinistra et dextra.

The result of laboratory blood test was found Hb 11.9g/dl, leukocytes 13,080/µL, platelets 331,000/µL, urea 24mg/dL, creatinine 0.57mg/dL, HST, liver function test at the normal limit, ANA test: 2.19, anti-dsDNA: 10.2, rheumatoid factor: negative, ASTO: < 200, benzidine test was negative, urinalysis was normal, IgA: 332mg/dL.

In 1990, American College of Rheumatology (ACR) developed criteria for the diagnosis of HSP. According to the criteria yields a sensitivity of 87.1% and specificity of 87.7%. This classification has been modified and change by European League Against Rheumatism (EULAR) and Pediatric Rheumatology European Society (PReS).[8]

### Table 1. Diagnostic criteria of HSP (ACR and EULAR/PReS) [8].

| ACR (1990) Criteria                                                                 | EULAR/PReS (2006) Criteria                      |
|-------------------------------------------------------------------------------------|-------------------------------------------------|
| Three or more of the following criteria are needed:                                  | Mandatory criterion:                            |
| 1. Age 20 years or less at disease onset.                                            | (i) Palpable purpura                             |
| 2. Palpable purpura.                                                                 | Plus at least one of the following criteria:    |
| 3. Acute abdominal pain with gastrointestinal bleeding.                             | 1. Diffuse abdominal pain.                      |
| 4. Biopsy showing granulocytes in the walls of small arterioles or venules in superficial layers of skin. | 2. IgA deposition in any biopsy.                |

HSP resolver spontaneously, therefore drugs therapy is aimed for supportive and symptomatic.[4,9]Except for the renal disease or severe organ involvement.[2]

Arthralgia and arthritis usually respond with acetaminophen and non-steroidal anti-inflammatory drugs (NSAID), but in severe cases used a steroid.[2]

Skin rash rarely requires specific treatment. Dapsone has also been shown to be of benefit as steroid sparing agent, Chronic skin and joint disease used a combination of aspirin and colchicine.[2]

Abdominal pain usually settles within a few day with or without treatment. Steroids are often for the relief of abdominal pain, in severe cases associated with protein-loss enteropathy, a combination of intravenous methylprednisolone (10 mg/kg BW, max dose 500 mg) daily for three days, followed by 2 mg/kg BW oral prednisolone (max dose 80 mg) daily. Severe gastrointestinal hemorrhage has also been treated with high dose methylprednisolone 1 gr for three days followed by 40 mg prednisolone daily for a week. Also, factor XIII replacement therapy has suggested treating severe gastrointestinal tract bleeding complications. Methotrexate has been reported to be a useful steroid-sparing agent for chronic abdominal pain, Mycophenolate has also been used for unresponsive and recurrent abdominal pain.[2,10]

Treatment option for severe renal involvement include high-dose corticosteroids, either alone or combined with immunosuppressive agents such as azathioprine, cyclophosphamide or cyclosporine, high-dose intravenous immunoglobulin, plasma exchange or plasmapheresis, corticosteroids combined with urokinase and warfarin, renal transplant.[4]

In this case, the patient was treated by injecting Methylprednisolone 125 mg/12 hours for five days, injecting Ketorolac 30 mg/12 hours, Paracetamol 3x1000 mg, Omeprazole 2x20 mg, and then injecting Methylprednisolone by reducing its dose up to 62.5 mg/ 12 hours for six days while added Azathioprine (Imuran) 2x50 mg.

Prognosis is excellent because HSP is a self-limiting disease. Based on the literature, HSP resolves in 94% of children and 89% of adults.[4]

In this case, improvement of painfulness and skin rash was reducible while returning into brownish. Afterwards, the patient permitted to get outpatient while being supplied with Methylprednisolone 4 mg for 3 x 4 tab dosing, Omeprazole 2x20mg, Imuran 2x50mg.
The prognosis in this patient is *quo advitambonam, quo adfunctionambonam* and *qua ad sanationamdubiaadbonam*.

**Figure 3.** After two weeks of treatment, the skin rash (purpuric) was reducible.

4. Conclusion
It was a female patient, aged 36 years with HSP. The disorder is more common in childhood but can also affect in adults. The patient was treated with a corticosteroid (methylprednisolone) and an immunosuppressive agent (Azathioprine). The prognosis in adults is worse than children due to an increased risk of disorders of the kidneys. Therefore, HSP in adults must be addressed.

References
[1] Pillebout E, et al. 2002 Henoch-Schönlein purpura in adults: outcome and prognostic factors J. Am. Soc. Nephrol. 13 1271-8
[2] Tizard E J and Ayres M J J H 2008 Henoch-Schönlein purpura Arch. Dis. Child. Educ. Pract. 93 1-8
[3] Hung S P, et al. 2009 Clinical manifestations and outcome Henoch-Schönlein purpura: comparison between adults and children Pediatr. Neonatal. 50 162–8
[4] Roberts P F 2007 Henoch-Schönlein purpura: review article Southern Med. J. 100 821–5
[5] Trapani S, et al. 2005 Henoch-Schönlein purpura in childhood: epidemiological and clinical analysis of 150 cases over a 5 year period and review of literature Semin. Arthritis Rheum. 35 143-53
[6] Daripally V K and Shah N S 2012 Henoch-Schönlein purpura: a rare vasculitis in older adults J. R. Coll. Physicians Edinb. 42 124–7
[7] Cruz B A, et al. 2006 Henoch-Schönlein purpura in adults: a case series from a multidisciplinary study group Rev. Bras. Rheumatol. 46 380-4
[8] Lamprecht P and Gross W L 2011 Small vessel vasculitide Springer Wien Newyorkl. 389-91
[9] Sukmana N 2014 Vakulitis *Buku ajar ilmu penyakit dalam* vol 6, ed S Setiati (Jakarta: Interna Publishing) pp 519-24
[10] Niaudet P and Habib R 1998 Methylprednisolone pulse therapy in the treatment of severe forms of Henoch-Schönlein purpura nephritis Pediatr. Nephrol. 12 238–43