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# Henoch-Schonlein Purpura with Renal Involvement in a Young Woman

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**Abstract:** ***Henoch-Schonlein Purpura (HSP) is a small vessel vasculitis mediated by IgA deposition that occurs mostly in children and rarely presents in adults. This report describes a 20 years old Balinese female with HSP, who presented with cutaneous, kidney and joint manifestations of this disease. She had history of upper respiratory tract infection couple weeks before. Physical examination showed erythematous, palpable, purpuric rashes on her thighs and lower legs. Complete blood count test revealed leukocytosis and neutrophilia predominate, moreover urinalysis showed macroscopic and microscopic haematuria and also proteinuria. She was treated with intravenous methylprednisolone and showed a good outcome.***

**Keywords:** Henoch-Schonlein Purpura, Nephritis, Vasculitis, Palpable Purpura

## 1. Background

Henoch-Schonlein Purpura (HSP) is a leukocytoclastic vasculitis involving small vessels with the deposition of immune complexes containing IgA. It is characterized by the association of skin (non-thrombocytopenic palpable purpura), joint, gastrointestinal manifestations, and renal involvement.[1,2] HSP occurs mostly in children and rarely presents in adults. HSP has been associated with a history of preceding infections (especially upper respiratory tract infection) and certain drugs (e.g., penicillin, quinines, erythromycin).[10,11] A lot of studies reporting that renal involvement and joint symptoms are more common in adult cases with a lower occurrence of abdominal pain and fever. HSP with renal involvement tends to be more severe in adults with an increased risk of progression to end-stage renal disease. It may manifests as isolated haematuria and/or proteinuria, renal hypertension, nephritic syndrome and severe acute renal failure.[11]

## 2. Case Presentation

A 20 years old Balinese female presented with erythematous, nonpruritic rashes which progressed proximally from both feet to thighs and worsening within 5 days. Later the feet became swollen with moderately intense pain that aggravated by ambulation. She also complained of reddish yellow urine. She had history of upper respiratory tract infection couple weeks before.On physical examination, her vital signs were stable. The abdomen was soft and non tender. Mild bilateral non-pitting edema was present over both the legs. There was the presence of non-tender, non-blanching, palpable purpuric rashes over both legs and thighs. Laboratory tests showed mild leukocytosis with WBC count of $16x10^3$/ul; neutrophils: 84,7%; hemoglobin: 12,6 g/dl; hematocrit: 38,3%; platelets: $400x10^3$/ul; serum urea: 18 mg/dl; serum creatinine: 0,8 mg/dl; urinalysis showed macroscopic and microscopic haematuria, and also proteinuria.

The patient was diagnosed with Henoch-Schonlein purpura according to American College of Rheumatology (ACR) and European League Against Rheumatism (EULAR)/ Pediatric Rheumatology International Trials Organization (PRINTO)/ Pediatric Rheumatology Society (PRES) criteria. Patient was treated with intravenous fluids drip, methylprednisolone 62,5 mg twice a day intravenously for 5 days continuously and then tapered and switched to oral methylprednisolone, and paracetamol (as analgesia).

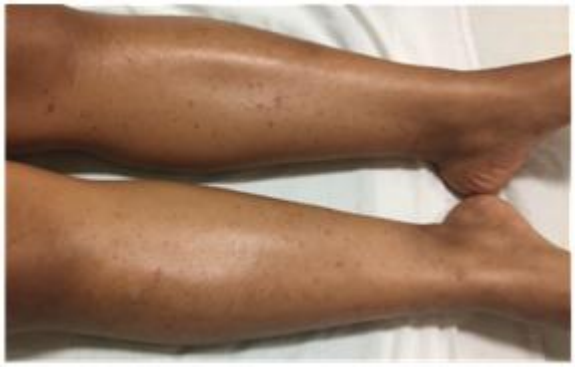

**Figure 1:** Palpable purpura of the legs

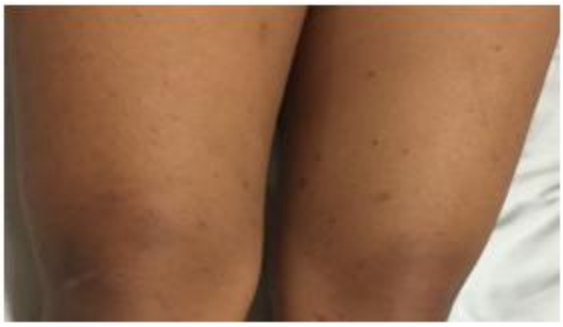

**Figure 2:** Palpable purpura of the thighs

## 3. Discussion

Henoch-Schonlein Purpura (HSP) was first described by William Heberden in 1801. Later, Schonlein recognized the association between purpura and arthritis, whereas Henoch reported a case that also included gastrointestinal symptoms along with the renal involvement.[13] HSP is the most common vasculitis of the children and rarely presents in adults. HSP is most commonly reported during late autumn to early spring seasons, though it can occur at any time. It has also been proposed that various triggers such as bacterial and viral infections (the most common is upper respiratory

tract infection), vaccinations, drugs, and autoimmune mechanisms may result in the formation of an antigen and antibody complex and the deposition of such formed immune complex in the small vessels may activate the alternate complement pathway leading to neutrophil aggregation which results in inflammation and vasculitis.[3] This process that including extravasation of erythrocytes, infiltration of tissues with neutrophils, and deposition of degenerating neutrophil fragments are well known as leukocytoclastic vasculitis (LCV). [4,5,7,8]The extravasation of blood leads to the pathognomonic purpura of the skin, while inflammation and edema leads to swelling which is usually palpable. Palpable purpura in the absence of thrombocytopenia is typically suggestive of HSP as its found in almost all patients. Purpura is usually noted in the legs and areas under pressure such as the buttocks. Other symptoms of HSP include abdominal pain and arthritis or arthralgia, though not universally present. Renal involvement occurs which manifests as isolated haematuria and/or proteinuria, renal hypertension, nephritic syndrome and severe acute renal failure. In adults, this damage may be severe leading to end stage kidney disease. It is usually detected within two months of the eruption, but sometimes may manifest as late as six months after initial onset of the disease. The most frequent pathology observed is a mesangial or endocapillary proliferative glomerulonephritis.Age of onset, the presence of renal impairment and hematuria at the onset, abdominal pain as an initial presentation, persistent eruption, renal pathology with fibrinoid necrosis and the number of sclerotic glomeruli are significant predictors of renal disease.[8,9]

The complete blood count test shows a mild leukocytosis with neutrophilia predominate. Urinalysis must be performed in order to evaluate haematuria and proteinuria. In patient with severe abdominal pain, an ultrasound examination is helpful to delineate whether an intussusception is present. Stools should be examined to evaluate visible or occult bloods. Renal biopsy should be performed in case of uncertain diagnosis or severe renal impairment such as nephrotic syndrome. Elevated serum IgA levels have been associated with HSP. Skin biopsy should be obtained from the lesion less than 24 hours and typically shows the classical leukocytoclastic vasculitis in postcapillaryvenule with IgA deposition.Inflammatory markers such as sedimentation rate (ESR) and C reactive protein (CRP) levels are often elevated. Endoscopy and/or colonoscopy play a major role in helping diagnosis of the patients with the gastrointestinal involvement as their initial presentation.[5,6,8]

In this case, 20 years old female patient had the symptoms of palpable purpuric rash over both lower legs and thighs, arthralgia which indicates joint involvement, reddish yellow urine and significant haematuria and proteinuria from urinalysis which indicates renal involvement. However, there was no sign of abdominal pain or gastrointestinal tract involvement. Leukocytosis, neutrophilia and normal platelet count were found from complete blood count test. Diagnosis of this case was made by The American College of Rheumatology (ACR) in 1990 (Table 1) and European League Against Rheumatism (EULAR)/ Pediatric Rheumatology International Trials Organization (PRINTO)/ Pediatric Rheumatology Society (PRES) in 2010 (Table 2).[1] ACR classification consists of four criterias (as seen in the table 1) and shall be diagnosed with HSP if at least 2 of these criteria are present. From EULAR/PRINTO/PRES classification, patient shall be diagnosed with HSP if there are a present of non thrombocytopenic palpable purpura (mandatory criteria) together with at least one of additional criterias as seen in the table 2.

**Table 1:** The American College of Rheumatology (ACR) 1990 Criteria for HSP diagnosis [1]

| Criteria | Definition |
|---|---|
| Palpable purpura | Slightly raised “palpable” hemorrhagic skin lesions, not related to thrombocytopenia |
| Age ≤ 20 years at disease onset | Patient 20 years or younger at onset of first symptoms |
| Bowel angina | Diffuse abdominal pain, worse after meals, or the diagnosis of bowel ischemia, usually including bloody diarrhea |
| Wall granulocytes on biopsy | Histologic changes showing granulocytes in the walls of arterioles or venules |

**Table 2:** EULAR/PRINTO/PRES 2010 Criteria for HSP Diagnosis [1]

| Criteria | Definition |
|---|---|
| Mandatory criteria | Palpable purpura, not related to thrombocytopenia |
| Additional criteria | Diffuse abdominal pain |
| | Histopathology: typical LVC with predominant IgA deposits or proliferative glomerulonephritis with predominant IgA deposits |
| | Arthritis or arthralgia |
| | Renal involvement (proteinuria: >0,3 g/24h or >30 mmol/mg of urine albumin to creratinine ratio on a spot morning sample; and/or haematuria, red blood cell casts: >5 red cells per high power field or ≥2+ on dipstick or red blood cell casts in the urinary sediment) |

Management of HSP is primarily supportive therapy with rest, analgesia and adequate renal hydration. Most cases of HSP are self-limited and may require little or no intervention. Symptomatic treatment will be sufficient for symptoms such as rash and arthritis. Acetaminophen and non steroidal anti inflammatory drugs can be used.Prednisone or methylprednisolone at a dosage of 1-2 mg/ kg daily for one to two weeks, tapering down to 0,5mg/kg/day over the next week and then 0,5mg/kg every other day for one more week can be used to treat patient who doesn’t tolerate oral steroids or who has abdominal symptoms with severe GI involvement and may enhance the rate of resolution of the arthritis, however this treatment does not prevent recurrence of the disease. Intravenous corticosteroids alone are effective for gastrointestinal and joint involvements but nephritis usually requires treatment with both steroids and immunosuppressive agents such as intravenous cyclophosphamide or azathioprine. Its also recommended for rapidly progressive glomerulonephritis (RPGN) and hemorrhagic involvement of the lungs and brain.However, not all studies can confirm the benefits of treatment with steroids and immunosuppressive drugs in preventing kidney involvement compared to placebo groups. In patients with advanced disease however, a regimen consisting of high dose pulsed intravenous

methylprednisolone (250 to 1000 mg per day for three days) followed by oral prednisone for three months may be beneficial. Treatment of the renal involvement should only be considered in patients with marked proteinuria and/or impaired renal function during the acute episode. A renal biopsy is recommended in this circumstance. Routine follow-up of all patients with HSP is recommended, with blood pressure checks and urinalysis monitoring weekly or biweekly for the first one to two months after presentation.[8,11,12]

In this case, patient was treated with fluids drip, paracetamol (as analgesia) and intravenous methylprednisolone at 62,5 mg dosage, twice a day in the first three days and showed a good improvement of purpuric rash, absent of arthralgia and significantly reduced of haematuria and proteinuria. Intravenous methylprednisolone at 62,5 mg dosage was still given continuously until five days and monitoring of urinalysis was performed. Absent of haematuria and proteinuria were shown from the urinalysis after total five days of intravenous methylprednisolone treatment. Furthermore, methylprednisolone was tapered and switched to oral drugs. Patient discharged from the hospital with oral methylprednisolone and paracetamol. Patient was scheduled to follow up her urinalysis a week after.

## 4. Conclusion

In summary, this case report presents a 20 years old Balinese female diagnosed with HSP, who presented with palpable purpuric rashes, renal involvement and joint manifestations of this disease. Diagnosis was made according to ACR criteria and EULAR/PRINTO/PRES criteria. The patient was treated with intravenous methylprednisolone and then tapered and switched to oral methylprednisolone which showed a positive outcome. After discharged from the hospital, patient was scheduled to get her urinalysis monitoring a week later.

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