CASE REPORT

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Adult-onset Henoch-Schonlein purpura: A case report

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ABSTRACT

Background: Demographic and geographic constraints make access to specialists difficult, reinforcing the role of general practitioners (GPs) in dermatology. The purpose was to investigate dermatology practice, referral patterns to dermatologists, training needs in this field, and their opinions on teledermatology.

Case: A 21-year-old female patient presented with palpable purpuric lesions on the lower abdomen, buttocks, and lower limbs. In addition, the patient complained of abdominal pain and joint pain. The patient had suffered an upper respiratory tract infection with spontaneous resolution three days before the skin lesions. Laboratory tests showed leukocytosis and elevated anti-Streptolysin O titer. The patient was diagnosed with HSP and given oral methylprednisolone, antibiotics, and symptomatic treatment. She showed clinical improvement after taking medication.

Conclusion: Henoch-Schönlein purpura is typically a self-limiting disease and infrequent in adults, but it has the potential to manifest into life-threatening conditions such as end-stage renal failure. An integrated multidisciplinary approach is needed for early diagnosis and management.

Keywords: Henoch-Schonlein purpura, IgA vasculitis, palpable purpura.
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INTRODUCTION

Henoch-Schönlein purpura (HSP) is a specific form of cutaneous small vessel vasculitis (CSVV) with vascular immunoglobulin A (IgA) deposition.[1] It is characterized by the clinical tetrad of non-thrombocytopenic palpable purpura, abdominal pain, arthritis or arthralgia, and renal involvement.[2] Cutaneous involvement is the characteristic sign of the disease and the most common presensation, although patients may present with involvement of other organ systems. Henoch-Schönlein purpura is the most common childhood vasculitis, but it can occur at any age. The incidence of HSP among children ranges from 30 to 270 cases per million per year. The median age of onset is around 6 years, and 90% of patients are younger than 10 years, with males being more frequently affected than females at a ratio of 1.5:1.[1–3] Adult-onset HSP has been rarely reported, with an incidence of 3.4 to 14.3 per million per year.[4] In Indonesia, the incidence of HSP is not well known. Based on data obtained at Cipto Mangunkusumo Hospital in Jakarta, new cases are increasing. From July to December 2006, 10 new cases of HSP were found, compared to only 23 new cases found in the previous five years.[5]

The exact etiology of HSP is unclear. Various infectious agents, drugs, vaccines, food allergens, and insect bites may be precipitants triggering the onset of this disease in genetically predisposed individuals. Genomic studies have found an association with HLA-DQA1 and DQB1 intergenic zone, the HLA-DRB1*01:11/B1*13 loci, and DQA1*01:01/DQB1*05:01/DRB1*01:01 haplotype.[3] In more than 75% of patients, upper respiratory tract or gastrointestinal infections precede the onset of disease. Multiple bacterial and viral infections have been described as triggers of HSP, such as group A streptococcal infection (most common), infectious mononucleosis, subacute bacterial endocarditis, hepatitis, Mycoplasma pneumoniae infection, Campylobacter enteritis, Helicobacter pylori infection, Yersinia infection, Shigella infection, Salmonella infection, Brucellosis, Legionella species, parvovirus, adenovirus, varicella-zoster virus infection, rotavirus, etc.[6] More recently, HSP has also been found in association with COVID-19 infections.[6,7]

Although HSP is typically self-limited and managed with supportive care, it might lead to serious complications. Renal involvement is the most important complication and the main cause of morbidity and mortality among patients suffering from HSP, and thus the main prognostic factor.[3] In adults, HSP is often more complicated and likely to cause long-term renal disease.[8] Prompt diagnosis and multidisciplinary intervention can lead to appropriate management and mitigate potential complications of HSP. Herein, we report a case of adult-onset HSP with preceding streptococcal infection to increase the knowledge about HSP, especially in adults.

CASE REPORT

A 21-year-old female presented with a skin eruption over the lower abdomen, buttocks, and lower limbs, which appeared 14 days before consultation. The lesions were warm, mildly itchy, and painful to the touch. She also complained of diffuse abdominal pain, constipation,

and arthralgia involving both ankles. The patient had suffered a preceding upper respiratory tract infection (malaise, sore throat, and fever) with spontaneous resolution three days before the skin lesions. She had no history of taking any new medication or vaccination. She denied any history of allergies. She had no significant medical or surgical history or relevant family history.

The patient was fully alert, conscious, and well-hydrated during the physical examination. Her vital signs were normal. No abnormalities were found on the chest and abdominal examination. Swelling around both ankles was present. Dermatological examination of the lower abdomen, gluteal region dextra sinistra, femoral region dextra sinistra, and cruris region dextra sinistra revealed multiple palpable purpura varying in size from 0.5 to 1.5 cm (**Figure 1**). The diascopy test was negative, as there was no blanching on pressure over the lesion (**Figure 2**).

Laboratory results showed the following: anemia (hemoglobin 10.4 g/dL); neutrophilic leukocytosis (white blood cell count 14,900/uL with neutrophils accounting for 72.5%); normal platelet count (327,000/uL); normal erythrocyte sedimentation rate (ESR) (9 mm/hour); normal blood urea nitrogen (12 mg/dL); normal creatinine serum level (0.69 mg/dL); normal estimated glomerular filtration rate (eGFR) (125 mL/min/1.73m^2); normal aspartate aminotransferase (AST) (18 U/L); normal alanine transaminase (ALT) (24 U/L); elevated anti-streptolysin O titer (800 U/mL). Urinalysis was not performed during the initial visit due to the menstrual period.

The patient's clinical presentation was consistent with the classification criteria established by the European League Against Rheumatism/Paediatric Rheumatology International Trial Organisation/Pediatric Rheumatology European Society (EULAR/PRINTO/PRES). Thus the diagnosis of Henöch-Schonlein purpura was made. She was treated with methylprednisolone 32 mg per day intraorally, which was then gradually tapered. Amoxicillin clavulanate 625 mg three times a day intraorally, lansoprazole 30 mg once a day intraorally, cetirizine 10 mg once a day intraorally, and topical desoximetasone twice a day were added as treatment. The patient was advised to limit activity, elevate affected extremities, and rest. Within a week of starting medication, the patient's condition improved. The existing lesions were beginning to heal, and the patient did not report any new lesions. There were no longer any complaints of abdominal and joint pain. The patient was then encouraged to have regular follow-up examinations.

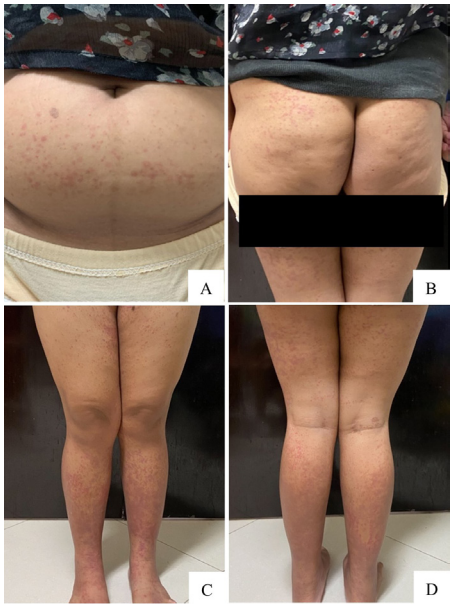

Figure 1. Dermatologic examination. Appearance of multiple palpable purpura varying in size on the lower abdomen (A), both gluteal region (B), and both lower extremities (C and D).

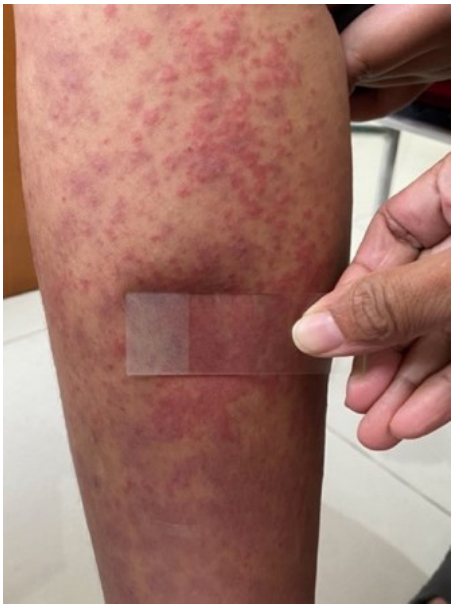

Figure 2. Negative diascopy. The lesions do not blanch when a glass slide is pressed.

DISCUSSION

Henöch-Schonlein purpura is a complex immune-mediated vasculitis characterized by the involvement of small blood vessels in various organ systems. It involves the small vessels of the joints, kidneys, gastrointestinal tract, and skin. Henöch-Schonlein purpura can also affect the central nervous system and the lungs; however, these findings are rare. Adult-onset HSP is an uncommon disease with an incidence ranging from 3.4 to 14.3 cases per million per year, and males are more frequently affected than females at a ratio of 1.5:1.[4,9] Gender plays a role in developing immune responses. In childhood, inflammatory reactions mediated by pro-inflammatory cytokines are higher in males than females. This explains why HSP is more prevalent in boys.[10] There are few reports presenting HSP in female adults, as discussed, and in this instance, the patient's atypical demographics might delay diagnosis.

The etiology of HSP is unknown; however, it is often post-infectious, with half of all cases preceded by an upper respiratory infection. Group A Streptococcus has been one of the most commonly reported pathogens, with positive throat cultures in 10% to 30% of cases and elevated anti-streptolysin O (ASO) titers in 20% to 50% of cases.[11] In this case, the patients complained of upper respiratory tract infection before skin lesions appeared. Laboratory examination showed an increase in ASO titer, suggesting that group A streptococcal infection may have triggered HSP in this patient.

There are two criteria proposed by the American College of Rheumatology (ACR) in 1990 and the new criteria by EULAR/PRINTO/PRES in 2010. Using the EULAR/PRINTO/PRES criteria in the adult population seems feasible, given that they are more sensitive and specific than the ACR criteria.[12] Based on EULAR/PRINTO/PRES criteria, the diagnosis of HSP is made based on the presence of palpable purpura (not thrombocytopenic or petechiae) plus at least one of the

following four characteristics: (1) diffuse abdominal pain; (2) arthritis or arthralgia; (3) renal involvement (proteinuria: >0.3 g/24 h 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); (4) typical leukocytoclastic vasculitis with predominant IgA deposits or proliferative glomerulonephritis with predominant IgA deposits on histology.[2,12] The patient in this case had the clinical signs and symptoms of HSP, fulfilling the EULAR/PRINTO/PRES classification criteria. She had both the mandatory criterion (palpable purpura with lower limb predominance) and two of the supporting criteria: abdominal pain and arthralgia.

Skin involvement is present in all patients with HSP and is usually the earliest sign. The rash associated with HSP often starts as erythematous, macular, or urticarial lesions. The rash then develops into palpable purpura and petechiae that most commonly affect the buttocks and lower extremities.[6] However, purpura on the trunk and upper extremities is not uncommon, and the rash may even extend to involve a large surface area. Lesions can be asymptomatic, painful, or pruritic. Compared with children, the rash of HSP in adults can potentially become bullous or necrotic, and the bullous form has been associated with progressive, treatment-resistant renal failure.[13,14]

Gastrointestinal involvement has been reported in 37% to 65% of HSP cases in adults and may precede the onset of skin purpura.[6,13] The most common symptom is colicky abdominal pain, followed by hematochezia, diarrhea, nausea, and vomiting. The symptoms are related to vasculitis-induced bowel ischemia and edema. Potential life-threatening complications include perforations, intussusception, and bowel infarction.

Henöch-Schonlein purpura-associated joint involvement in adults has been reported in 30% to 60% of cases.[13] It is typically transient, migratory, oligoarticular, and non-destructive. Patients often present with painful, swollen joints that most commonly involve the knees and ankles. Hands and feet may also be affected. The EULAR/PRINTO/PRES classification criteria define arthritis or arthralgias as conditions with an acute onset.[6,13]

Patients with HSP usually develop renal manifestations within a few weeks after the initial presentation; however, this can be delayed several months. Microscopic or macroscopic hematuria with or without red blood cell casts is the earliest and most sensitive test suggestive of Henöch-Schonlein purpura nephritis (HSPN). Other manifestations include proteinuria, which ranges from mild to severe. Henöch-Schonlein purpura nephritis develops when the renal parenchyma is affected, and HSPN is the leading cause of morbidity from this disease. According to the 2021 Kidney Disease Improving Global Outcomes guidelines, the incidence of acute kidney injury can reach up to 32% in adults at the time of diagnosis but is rare in children. Hypertension is typically noted in one-third of cases. Chronic kidney disease is a common, long-term complication, with 10% to 30% of patients with HSPN developing end-stage renal disease at 15 years follow-up.[13]

Although HSP is a clinical diagnosis, laboratory studies and imaging may help to identify complications or exclude other diseases. Baseline studies include renal function tests (urea nitrogen, serum creatinine, urinalysis), complete blood count with platelet count, coagulation profile, and IgA levels. A normal platelet count and coagulation studies, leukocytosis, eosinophilia, azotemia, elevated serum IgA levels, hematuria, proteinuria, and red blood cell casts are commonly seen. The need for blood cultures, anti-streptolysin O titers, and tests to identify other infectious agents depends on the presence of clinical indicators of specific infections. Patients with streptococcal-associated HSP may have a low C3 and elevated ASO titers.[15] Imaging can also be considered, such as renal or skin biopsy and endoscopy, which may play a role when the diagnosis is uncertain or in monitoring for possible complications and system involvement.[4,16] In contrast to other types of systemic vasculitis, HSP is self-limiting, and most patients recover from it spontaneously. The management of HSP primarily involves supportive care and includes adequate hydration, rest, and symptomatic relief of pain in most cases. Hospitalization may be required when adequate outpatient monitoring is unavailable or if dehydration, hemorrhage, or pain control requires inpatient management. A nephrology referral is recommended with significant renal involvement. In patients with severe renal disease, renal biopsy is needed to provide a definitive diagnosis and guide therapy. All patients should be followed up for at least 6-12 months, even if the initial blood pressure measurements and urinalysis are normal.[13] Treatments aim to provide acute symptom relief and prevent renal deterioration. Early aggressive therapy is recommended for children and adults with severe renal involvement. Treatment options include high-dose steroids with immunosuppressants, high-dose intravenous immunoglobulin, plasmapheresis, and renal transplant.[16]

As HSP is characterized by IgA deposition and white cell infiltration within blood vessel walls, corticosteroids can inhibit this inflammatory process.[8] It has been reported that although steroids do not prevent renal or gastrointestinal complications, they are helpful for symptomatic relief, especially abdominal and joint pains.[17] A recent algorithm is available where oral prednisolone is given for low-level vasculitis nephritis, oral/pulse steroids for moderate, and oral/pulse steroids with intravenous cyclophosphamide for severe forms.[3] For second-line therapy, rituximab, azathioprine, mycophenolate mofetil, along with steroids and cyclophosphamide, are advocated.[6] Anti-infectious agents were recommended for those who suffered from an infection.[18] Antibiotics may be effective in treating renal dysfunction if streptococci are involved.[15]

Colchicine is the treatment of choice when the skin lesions are severe. Colchicine inhibits polymorphonuclear leukocyte chemotaxis by inhibiting spindle formation, blocking lysosome formation, and stabilizing the lysosome membranes. The suppressive effect of colchicine on the inflammatory pathway may explain its efficacy on skin lesions.[4] Dapsone, an antileprotic sulfonamide used for various dermatological conditions, appears to be

of special value in diseases characterized by the accumulation of neutrophils, including leukocytoclastic vasculitis. There is evidence that it has antioxidant effects and may suppress the generation of neutrophils of toxic free radicals. It also inhibits prostaglandin D2 production and synthesis of IgG and IgA antibodies. Dapsone may also inhibit IgA–neutrophil interactions.[19]

Plasmapheresis and immunoglobulin have been used in refractory combination therapy. Intravenous immunoglobulin can also be effective in cases with rapidly progressive glomerulonephritis. Mechanisms of action include impairing autoreactive T-cells by blocking binding to antigen-presenting cells, downregulating antibody production by B-cells, and blocking the Fc-receptor-mediated immune response.[4,6] The histopathological examination did not rule this case out, thus, it is the limitation of this case report.

CONCLUSION

Henöch-Schonlein purpura is typically a self-limited illness and infrequent in adults, but it has the potential to manifest into life-threatening conditions such as end-stage renal failure. An integrated multidisciplinary approach is needed for early diagnosis and management.

CONFLICT OF INTEREST

None.

PATIENT'S CONSENT

Written informed consent was obtained from the patient for using clinical photographs and records in publication.

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None.

AUTHOR CONTRIBUTIONS

All authors contributed to this case report's case, literature, and publication.

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