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Case Report

# Diagnosis and management of Henoch-Schonlein purpura in Indonesian elderly with severe complication: A rare case

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ABSTRACT

*Background:* Henoch-Schonlein purpura (HSP) in the elderly is very rare, so this case report was conducted to improve the diagnosis and management of HSP.
*Case presentation:* A 65-year-old Indonesian male with severe complications of HSP had been reported. In addition to the patient's history, physical examination, laboratory and radiological findings, he performed a left sole anterior biopsy that showed interstitial infiltrate of main neutrophils, and faint granular deposits of IgA and C3 seen within the superficial vessels. He also had severe complications of gastrointestinal bleeding, extensive leukocytoclastic skin vasculitis, renal, and neurological manifestations. He had been given a pulse steroid, cyclophosphamide. He also planned to receive rituximab but canceled due to his worsening condition.
*Discussion:* Poor prognosis in the management of HSP in the elderly needs more attention to minimize mortality.
*Conclusion:* Management of HSP in the elderly patient is challenging as it is a rare case, with a tendency for severe manifestations and rapid progression.

## 1. Introduction

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, joints, and gastrointestinal manifestations that may occur in successive episodes [1,2]. In adults, the incidence of HSP is 1.3 per 100,000 population [3]. The clinical presentation of HSP is more severe among adults and tends to be atypical [4]. The HSP in adults is associated with more frequent and severe kidney disease. One study indicated that the outcome for adults is worse than for children, requiring aggressive treatment and a longer hospital stay and resulting in persistent kidney dysfunction years later [5]. Therefore, we were interested in reporting a 65-year-old male diagnosed with HSP based on surgical case report (SCARE) 2020 guideline [6].

## 2. Case presentation

A 65-year-old male complained of colicky abdominal pain. He had been vomiting 3–5 times throughout the day and had no intake of nutrients. He had subacute intestinal obstruction secondary to ileus and a nasogastric tube was inserted. He also had a history of melena. He felt weak and could not perform daily activities independently. He noticed a purpuric rash that initially appeared on the legs and also appeared on the hands. He had complications of electrolyte imbalances and decreased urine production. He developed acute kidney injury and required dialysis. He denied a history of hypertension, diabetes mellitus, and cardiovascular complications.

Laboratory history results included ANA IF < 80, ANCA negative, C3 0.72 g/L, C4 0.224 g/L, Anti streptolysin-O 488.2 IU/mL, CRP 21.8 mg/L, procalcitonin 2.17 ng/mL. The endoscopic finding showed normal results in the stomach and duodenum (Fig. 1). A CT scan of the abdomen showed a thickening of the wall of the small bowel suggesting edema (Fig. 2). He was given emergency therapy, comprising of hydration, some medicines that stopped bleeding, gastric acid secretion inhibitor drug, and antibiotics. He was consulted by the surgery department and performed the surgery. There was thickened jejunum from the distal junction involving 30 cm of jejunum. There was ecchymosis on the surface of the jejunum and mucosal ulceration of the proximal resection area. The resected small bowel showed ulcers with vasculitis involving small and medium-sized vessels surrounded and infiltrated by predominantly neutrophils with karyorrhexis, histology negative for viral

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inclusion bodies, granuloma, and malignancy, but positive for yeast. The patient also underwent a biopsy of the left sole anterior foot that showed interstitial infiltrate of main neutrophils and faint granular deposits of IgA and C3 were seen within the superficial vessels. He was then diagnosed with HSP with severe gastrointestinal, skin, and renal complications. Because of the diagnosis of vasculitis, a decision was made to pulse with methylprednisolone 500 mg daily for 3 days and followed by cyclophosphamide 250 mg intravenously.

The physical examination showed normal blood pressure and other vital signs. There were bilateral vasculitis rashes over the fingertips, hands, toes, and feet. He also had generalized edema. The urine production measured was 150 mL in 6 hours. The laboratory results included Hb (8.1 g/dL), Hct 23.1%, MCV 82.5, MCH 28.9, leukocytes 17.720/μL, neutrophils 95.2%, platelets 108.000/μL, BUN 95 mg/dL, albumin 2.9, creatinine 3.5 mg/dL, glucose 175 mg/dL, SGOT 19 U/L, SGPT 14 U/L, Sodium 150 mmol/L, potassium 4.96 mmol/L, chloride 112 mmol/L, urinalysis: glucose negative, leucocyte negative, epithelial cells negative, casts negative, Specific gravity <1.005, blood negative, pH 7.5, protein negative, nitrate negative, and blood gas analysis (pH = 7.28, pCO2 = 19, pO2 = 94, BE = −2.4, HCO3 = 22.2, and SO2 = 99%). He was treated with tranexamic acid of 500 mg every 8 hours, esomeprazole pump of 8 mg/hour, hydrocortisone of 100 mg every 6 hours, Cavit D3 of 750 mg every 24-h, paracetamol of 1000 mg intravenously if needed, and the maintenance fluid therapies IVFD Kabiven of 400 cc/24 hour, NaCl 0.9% of 250 cc in 4 hours, and PRC Transfusion of 325 cc/day. Ceftriaxone of 1 g/12 hour and Fluconazole of 100 mg/24 hour intravenously.

Three days later, the patient had coffee ground emesis and melena. The physical examination showed blood pressure level decreased to 100/70 mmHg. His laboratory results Hemoglobin drops to 7 g/dL and platelets count 58,000/μL. His potassium level decreased to 2.8 mmol/L. He was treated for hypokalemia with KCL 50 meq in PZ 500 ml for 24 hours, furosemide pump, and other maintenance therapies continued. He also required a transfusion of packed red cells.

On the 12th day of treatment, he had shortness of breath. He had a respiratory rate of 30 x/min. Laboratory evaluation showed hemoglobin and platelet continued to decrease by 6.8 g/dL and 38,000/μL respectively, while sodium level reached 163 mmol/L. The patient underwent echocardiography showed LVH with diastolic dysfunction, IVC 2.47 cm with collapse <50% suggesting volume overload. The patient had dialysis and transfusion of thrombopheresis and packed red cells during dialysis. Rituximab was planned to be given after dialysis.

On the 13th day of treatment, he still had melena but decreased in frequency and amount. He suddenly had a generalized seizure three times a day with a duration of 5 minutes. He was unstable at the time and could not undergo a head CT scan. His blood pressure level dropped

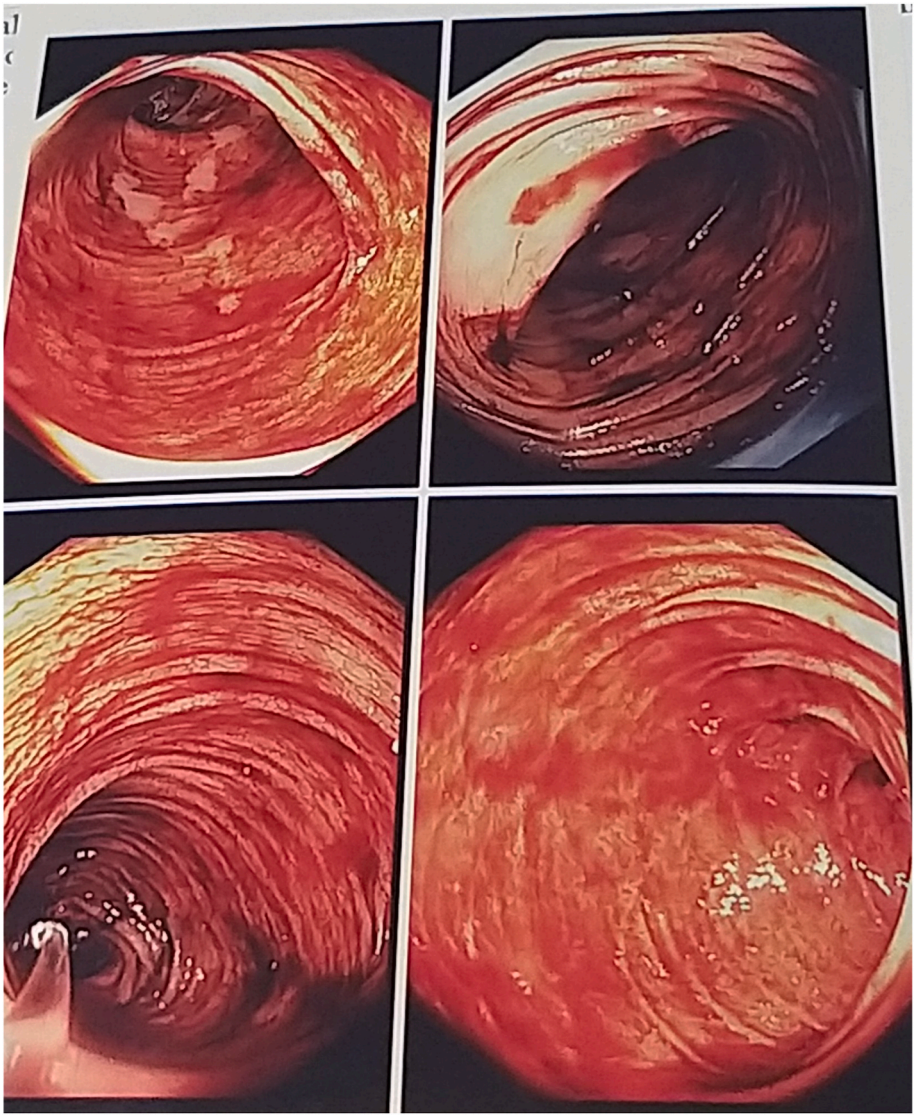

**Fig. 1.** Endoscopy showing normal condition.

to 81/49 mmHg. Laboratory evaluation showed Hb (8.8 g/dL) had increased but platelets count was low (31,000/μL). His potassium level reached (3.2 mmol/L), sodium level decreased (155 mmol/L). The patient was treated with a norepinephrine pump of 150 nano/hour with target systolic >120 mmHg. He had given emergency therapy with diazepam 10 mg intravenously and loading phenytoin 15 mg/kg at a rate of 50 mg/min, maintenance phenytoin 10 mg/kg/day, and maintenance therapies were continued. On the 15th day of treatment, the patient's condition deteriorated. The blood pressure continued to fall with the maximum dose of norepinephrine pump of 250 nano/hour until the patient passed away.

## 3. Discussion

Diagnosis and management of HSP in adults can be challenging as it is rare, with a tendency for severe manifestations and rapid progression, and due to a lack of rigorous data regarding management. Therefore, early clinical suspicion and rapid escalation of therapy if no contraindications exist could result in favorable outcomes [7]. The pathophysiology behind HSP is not yet completely understood. HSP is generally self-limiting and harmless, but concomitant nephritis may cause severe complications [8]. The proportion of patients having renal involvement varies (20% and 80%). The estimated incidence of the nephrotic or nephritic syndrome is 7% of all HSP cases, and 1% of patients develop end-stage renal failure. HSP nephritis (HSPN) usually occurs within 1–2 months after the onset of HSP [8,9].

The use of corticosteroids is controversial and usually reserved for severe systemic manifestations [4]. Several case series have reported better outcomes in adult patients with gastrointestinal and renal involvement who were treated with corticosteroids. There have been case reports suggesting that corticosteroids pulses may be helpful in patients with massive gastrointestinal hemorrhage and widespread mesenteric vasculitis. In a randomized trial, prednisone was effective in reducing abdominal pain, as the incidence of severe abdominal pain requiring hospitalization was greater in the placebo group than in the prednisone group [10].

Analgesics, mainly acetaminophen, are useful for joint and muscle pain and fever. Nonsteroidal anti-inflammatory drugs can be used to treat arthritis but should be avoided in patients with gastrointestinal and renal manifestations. Ranitidine or H2 blocker was found to be effective in patients with moderate gastrointestinal involvement in a placebo-controlled trial [5]. A randomized controlled trial about intravenous cyclophosphamide (IVCY) demonstrated that the addition of cyclophosphamide to steroid administration provides no benefit compared with steroid administration alone [11]. On the other hand, there have been a few reports that IVCY is effective for severe HSP nephritis [12]. IVCY is effective for other types of systemic vasculitis, such as lupus nephritis and Wegener granulomatosis, with renal involvement [13].

Immunoglobulin A vasculitis (IgAV), formerly known as HSP, is a vasculitis affecting small vessels (predominantly capillaries, venules, or arterioles) with IgA1-dominant immune deposits. Additional immunosuppressive agents are reserved for moderate to severe IgAV nephritis

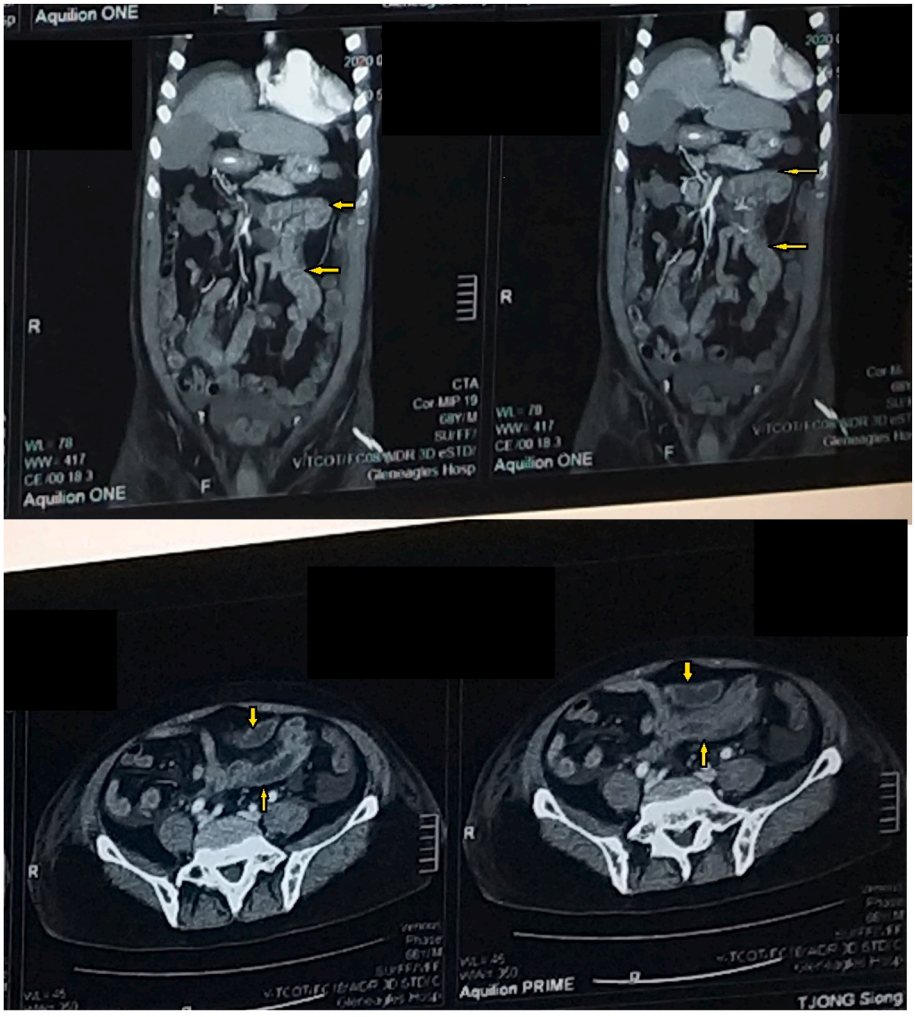


**Fig. 2.** CT Scan abdomen showing edema in the bowel.

and also for glucocorticoid-resistant situations. Other lines of therapy used in severe or refractory IgAV patients include intravenous immunoglobulins, plasma exchange, and rituximab (RTX) [14]. RTX, an anti-CD20 chimeric monoclonal antibody, has been widely used with good results in lymphoproliferative disorders, systemic lupus erythematosus, and another systemic vasculitis with an autoimmune background. In the same way that in the previous immune-mediated vasculitis, in IgAV, RTX might control disease activity by depleting B cells and, subsequently, by reducing IgA production and immune complexes containing IgA [14,15]. The main reason for RTX administration in this vasculitis was the lack of renal response to glucocorticoids or other immunosuppressive agents, while refractory gastrointestinal and neurological manifestations may lead to RTX treatment in a small proportion of IgAV patients. The presence of contraindication to glucocorticoids or cytotoxic agents was the reason for giving RTX in 9% of cases. This proportion was 27% in the previous retrospective multicenter IgAV study [16].

After RTX treatment, the majority of patients significantly have improvement in all the affected (cutaneous, articular, and gastrointestinal) territories. Concerning the nephritic involvement, hematuria and proteinuria also improve in most patients. However, the renal function remains unchanged after RTX, maybe due to established renal damage secondary to persistent or uncontrolled disease. In the present and the previous adult-onset IgAV series, the remission induction RTX regimen used for lymphoma treatment (two 1000 mg doses, two weeks apart) is more frequently prescribed than the RTX rheumatoid arthritis schedule (375 mg/m2/week, for 4 weeks). RTX efficacy is similar with both regimens since complete remission after RTX infusions is achieved by more than 70% of patients in both studies. Relapses following initial RTX doses occur in about a third of cases in ours and previously reported IgAV patients. All relapsing patients in whom RTX was given again experienced a good control of disease activity [16–18].

## 4. Conclusion

A 65-year-old male patient with severe complications of HSP has been reported. In addition to the patient's history, physical examination, laboratory and radiologic findings, the patient also performs a left sole anterior biopsy that showed interstitial infiltrate of main neutrophils and faint granular deposits of IgA and C3 are seen within the superficial vessels. He also has severe complications of gastrointestinal bleeding, extensive leukocytoclastic skin vasculitis, renal, and neurological manifestations. He has been given a pulse steroid, cyclophosphamide. He is also planning to receive rituximab but canceled due to his worsening condition. Management of HSP in the patient has been challenging as it is rare, with a tendency for severe manifestations and rapid progression.

## Ethical approval

Not applicable.

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Nothing.

## Author contribution

All authors contributed toward data analysis, drafting, and revising the paper, gave final approval of the version to be published, and agree to be accountable for all aspects of the work.

## Trial registry number

Name of the registry:-.
Unique Identifying number or registration ID:-.
Hyperlink to your specific registration (must be publicly accessible and will be checked):-.

## Guarantor

Awalia.

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Provenance and peer review

Not commissioned, externally peer-reviewed.

## Declaration of competing interest

The authors declare no competing interest.

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