

CASE REPORT

Henoch-Schönlein Purpura: Management and Complication

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ABSTRACT

Henoch-Schönlein Purpura (HSP) is a disease that mainly affects children, while the incidence in adults is rarely reported. Low incidence in adults caused by undiagnosed or misdiagnosed. Course of the disease in adults is more complex, including high incidence of renal insufficiency. Renal manifestations need special attention because it can worsen the prognosis, so patients must be detected and treated as quickly as possible. Poor prognosis depends on the presence of renal clinical manifestations accompanied by an increase in the severity of renal histological grading, abdominal manifestations, and persistent purpura. We describe male patients with HSP, presenting with gastrointestinal, renal, and cutaneous manifestations. Gastroscopy showed superficial gastritis (reddish patches on almost all gastric mucosa. Skin biopsy showed lymphocytic vasculitis. Gastric biopsy shows infiltration of lymphocyte inflammation cells, histiocytes, plasma cells in the corpus and gastric antrum. Patients have received supportive therapy, steroid, and showed clinical improvement.

Introduction

HSP is a small blood vessel vasculitis mediated by deposits of IgA-immune complexes containing immune complexes and complement components. HSP characterized by clinical features of palpable non-thrombocytopenia purpura, abdominal pain, arthritis, and renal manifestations.¹⁻⁴ The etiology of HSP is still unknown, although various antigens such as infection (most are upper respiratory tract infections dental infections, and history of fever without other symptoms, where group A β -hemolytic streptococcus is the most common pathogen), vaccinations, drugs, food, and insect bites can trigger the onset of HSP.³

In children, this disorder is a self-limiting disease, while in adults is more complex, including the high incidence of renal insufficiency that occurs in nearly 50% of patients with renal involvement.²

Case report

A 45-year-old man from Tulungagung, East Java, presented with abdominal pain and heartburn since 2 weeks. Patient was taken to the doctor and given painkillers and ulcer drugs. He went to the Radiologist and diagnosed with kidney stones. No nausea, vomiting, fever, joint pain and previous cold cough. There were

red spots on the bilateral legs and arm 1 week. Patients complained of black stool 2 weeks before. No history of diabetes mellitus, but he had hypertension. On physical examination, there was red spots on upper and lower extremities (Figure 1a, 1b). Patient was awake, GCS 456, blood pressure 150/80 mmHg, pulse 100 beats per minute regular; respiratory rate 22 times per minute; axillary temperature 36.7°C

Table 1. Laboratory Findings

| Blood: | |
|-------------------|--------------------------------|
| White blood cells | 11.8x10 ³ / μ L |
| Haemoglobin | 12.9 g/dL |
| Neutrophils | 90.3% |
| Hematocrit | 36.1% |
| Platelets | 336 x10 ³ / μ L |
| SGOT | 22 U/L |
| SGPT | 33 U/L |
| BUN | 11 mg/dL |
| Serum creatinine | 1.49 mg/dL |
| HbsAg | Non-reactive |
| Urinalysis: | |



| | |
|--------------------------------|-----------------|
| hematuria | erythrocyte 3+ |
| proteinuria | protein +/- |
| Urine Sediment Flow Cytometry: | |
| Leucocyte +/-, | +/- |
| Protein 3+, | 3+ |
| Erythrocyte 3+, | 3+ |
| Albumine : Creatinine: | 150 mg/g |
| Protein : Creatinine | 500 mg/g |
| RBC, | 943.8/ μ L |
| WBC | 213.3 / μ L |
| Bacterial | 177.6 / μ L |

Gastroscopy: superficial gastritis (reddish spots on almost all gastric mucosa) (Figure 2). Skin biopsy: lymphocytic vasculitis. Gastric biopsy: infiltration of lymphocyte inflammation cells, histiocytes, plasma cells in the corpus and gastric antrum, inactive chronic gastritis, *Helicobacter pylori* negative. Echocardiography: diastolic dysfunction. patients was diagnosed with HSP based on The European League Against Rheumatism and Paediatric Rheumatology European Society, and American College of Rheumatology criteria. He was treated with intravenous metilprednisolon 1 mg/kgBW (1x62.5 mg) and started on oral metilprednisolon 16 mg every 8 hours, with resolution of his symptoms.

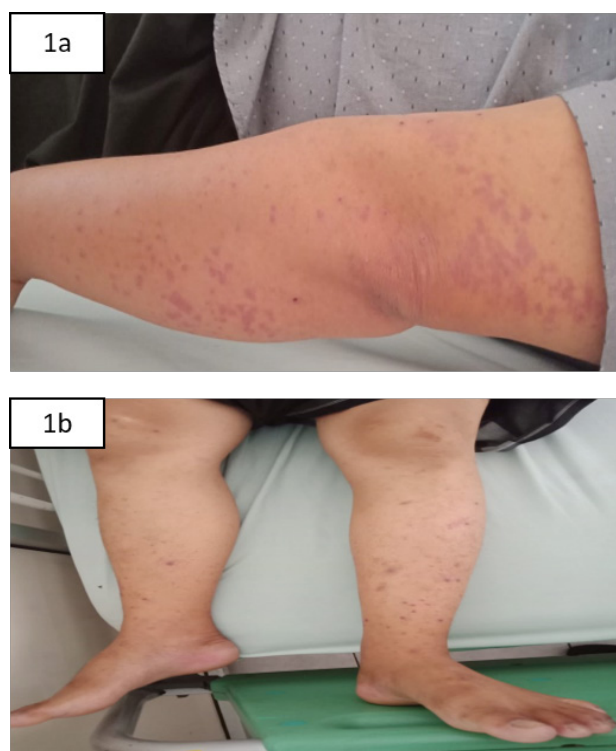


Figure 1. 1a and 1b. Red spots (purpura), over the patient's upper and lower extremities

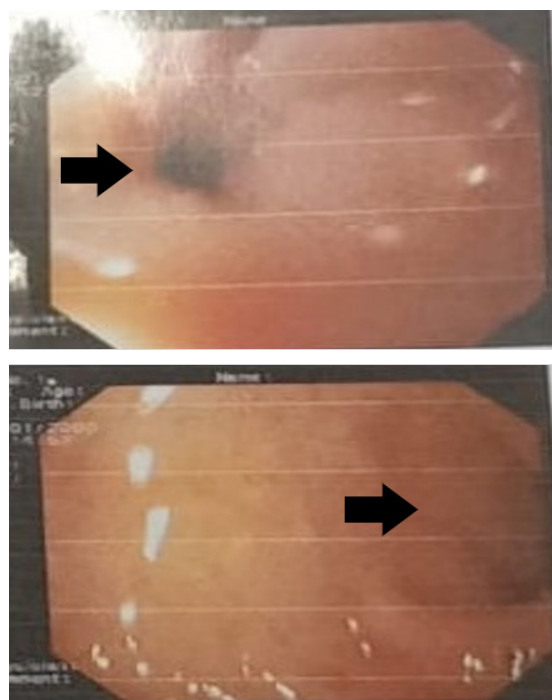


Figure 2. Gastroscopy image shows pas gastritis superficialis (red spots on almost all gastric mucosal)

Discussion

HSP is a cutaneous small vessel vasculitis with deposition of IgA and other immune factors within the vessel walls. HSP is a well-documented clinical disease in children, but is much less common in adults. The constellation of palpable purpura, arthralgia/arthritis, abdominal pain and renal complications usually aid in the diagnosis.⁵

Based on European League Against Rheumatism and Pediatric Rheumatology European Society criteria in 2006, The HSP diagnostic criteria are predominant palpable purpura in lower limb, plus one criteria: 1. Diffuse abdominal pain, 2. IgA deposits in all biopsy, 3. Renal manifestations (hematuria/proteinuria). While the diagnostic criteria based on the American College of Rheumatology in 1990, are two or more criteria of: 1. Age 20 years or less at the onset of disease, 2. Palpable Purpura, 3. Acute abdominal pain accompanied by gastrointestinal bleeding, 4. Biopsy showing granulocytes on the walls of small arterioles or venules in the superficial skin layer.² Definitive diagnosis based on clinical features and biopsy.⁵

Gastrointestinal manifestations occur in 11% of cases. *H. pylori* infection is related with the development and relapse of HSP following gastrointestinal manifestations. The classic features of gastrointestinal manifestations in Henoch-Schonlein Purpura are periumbilical colic and abdominal pain, nausea, vomiting, diarrhea, constipation, and abdominal distension.⁶ Urological manifestations mainly occur in the kidneys (about 50%).⁷

A 45-year-old male patient with abdominal pain, heartburn, red spots on bilateral upper and lower extremities, black stool was admitted to an internal medicine emergency department. Laboratory tests revealed leukocytosis, the presence of red blood cells in urine were detected, proteinuria, increased of IgA serum

level, and vasculitis on gastroscopy and skin biopsy. This patient fulfilled criteria of The European League Against Rheumatism and the Pediatric Rheumatology European Society in 2006, the main criteria being purpura palpable, and other criteria (diffuse abdominal pain, IgA serum 434 mg/dL, hematuria and proteinuria). This patient also fulfilled 3 criteria of the American College of Rheumatology in 1990 (palpable purpura, acute abdominal pain accompanied by gastrointestinal bleeding, and skin biopsy showing lymphocytic vasculitis).

HSP have predominant polymer IgA1 immune complex deposit in capillaries of target organs (dermal, gastrointestinal, and glomerular). Seasonal predilection in children show viral etiology which rarely found in adults and there is a strong association with malignancy. IgA immune complex deposits in HSP can be caused by exposure of tumor antigens (adenocarcinomas, lymphomas (non-Hodgkin's and Hodgkin's)) and IgA myeloma. HSP can occur due to both viral infections such as hepatitis B and parvovirus B19, bacterial staphylococci, and streptococcal infections. Immunological agents, such as the hepatitis B vaccine and intravesicular Calmette-Guerin bacillus, are associated with HSP.⁸

HSP therapy mainly supportive. The specific therapies include glucocorticoids, methylprednisolone and supporting drugs (cyclosporin A, rituximab, mycophenolate mofetil), plasmapheresis, peritoneal and hemodialysis, renal transplantation. Glucocorticoids, especially prednisone is reported to reduce abdominal pain.⁷

Skin manifestations of vasculitis rarely require therapy, but bullous lesions improve with steroids. Joint manifestations (arthropathy) are often treated using non-steroidal anti-inflammatory drugs (NSAIDs). Glucocorticoid (such as prednisone) have been used to treat severe eruption, cutaneous edema, severe colicky abdominal pain, scrotal and testicular involvement. Oral prednisolone 1 mg/kgBW/day for 2 weeks reduces severity of joint complaints and shortens duration of pain.^{2,9}

Gastrointestinal manifestations are treated using low-dose oral steroids and have fast resolution in reducing abdominal pain. Oral prednisolone dose of 2 mg/kgBW/day for 1 week and tapered off for 1 week significantly shortens the duration of abdominal pain.⁹

The use of prednisolone in HSP without evidence of nephritis showed no nephropathy.⁹ Methylprednisolone pulse dose and immunosuppressive agents are used for severe or life-threatening kidney disease to prevent irreversible glomerular fibrosis with proteinuria and improvement of long-term renal outcome.^{8,10,11}

The patient has abdominal pain, black stools, red spots on bilateral upper and lower extremities. Urinalysis showed microscopic hematuria and proteinuria. He was given intravenous methylprednisolone 1 mg/kg/day (62.5 mg iv daily), continued with methylprednisolone tablets 16mg, 3 times per day, and tapered off for 1 week.

Renal prognosis for HSP nephritis is poor.³ Minimal urine abnormalities shows a good prognosis, proteinuria with/without nephrotic syndrome and/or impaired kidney function is at risk of developing chronic kidney disease.¹² The prognosis of HSP is good except in glomerulonephritis.¹¹ Although HSP is infrequent in adults, renal involvement deserves special attention because it darkens the prognosis.

Thus, these patients should be detected and treated as early as possible.¹ In our case, patients showed microscopic hematuria and proteinuria without nephrotic syndrome and/or impaired kidney function and at risk of developing chronic kidney disease.

Predictors of poor kidney development in adult patients are high creatinine levels at onset, proteinuria >1 gram/day, arterial hypertension, increased protein during follow-up, extrapapillary proliferation in renal biopsy, interstitial fibrosis, and tubular atrophy.¹

Conclusion

Pregnancy and SLE is a never ending loop both. Pregnancy in SLE should be well planned so that any complication can be prevented. Awareness of disease activities and initial management could save lives. The use of antiviral is the right step to prevent the reactivation of viral replication in hepatitis B patients with immunosuppressants. Immunoglobulin is highly recommended for newborn from mothers with HBV infection.

Conflict of Interest

The author stated there is no conflict of interest

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