

# A young female presented with gastro-intestinal bleeding due to vasculitis of Henoch Schonlein purpura

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## Abstract

Henoch Schonlein purpura (HSP) is a small vessel vasculitis in which immune complexes of IgA and activated components of C3 complements are deposited on capillaries, venules and arterioles. We report a case of a young female who presented with acute abdominal pain, gastrointestinal bleeding, bilateral symmetrical small and large arthritis one week later while on treatment for HSP. She was treated with supportive therapy and steroids and clinically improved without any severe complications.

**Key words:** abdominal pain, vasculitis, purpura, HSP

## Introduction

Henoch Schonlein purpura is a cutaneous vasculitis of small vessels which is due to vascular entrapment of circulating immunoglobulin A. It commonly affects children. Involvement of multiple organs is reported in adults with higher frequency compared with children. In HSP, purpuric rashes appear predominantly on extensor surface of legs and buttocks, but can be seen in other regions such as arms, face, and trunk. Arthritis mostly affects the ankles, knees, and elbows. Patients who have renal involvement of the HSP can present as microscopic haematuria and proteinuria. Approximately 75% of HSP patients have gastrointestinal tract involvement, which is frequently characterized by diffuse colicky type of abdominal pain<sup>3</sup> which is a common manifestation in HSP patient.

Usually, there is no evidence on treatment of glucocorticoid for HSP in adults. The use of glucocorticoids in prevention from severe outcomes or relapse is controversial in HSP<sup>2</sup>.

## Case study

A 47-year-old woman with diabetes mellitus admitted with hematochezia (fresh blood) for two days.

Her disease course was as follows; she had history of fever 2 weeks ago which was associated with throat pain and symmetrical bilateral small and large joint pain and swelling. Involved joints were bilateral knees, elbows, and wrists. One week later she developed diffuse purpuric rashes on her lower limbs which were not faded by pressing (Figure 1, 2 & 3). She was started on symptomatic treatment without any specific treatment especially, glucocorticoids at that time. One week after the onset of rashes, she was admitted with abdominal pain, vomiting and haematochezia for 2 days. There was no haematuria or body swelling.

On admission she was afebrile. Other physical examination revealed heart rate of 120bpm and blood pressure of 130/80mmHg and examination of all the other systems were unremarkable except abdomen. There was deep tenderness in the epigastric region of abdomen without rebound tenderness and guarding. There was no splenomegaly, hepatomegaly, or free fluids. Bowel sounds were normal. Three days later, her diffuse purpuric rashes extended to her limbs and trunks. There was bilateral pedal oedema without significant joint swelling.

Her laboratory tests showed white cell count of  $20.12 \times 10^9/L$  (Neutrophils 71.5%, Lymphocytes 19.0%), haemoglobin of 14.1 g/dl, platelet of  $307000/\mu l$  and hematocrit of 42.1. Her SGOT was 10U/L, SGPT was 25U/L and total bilirubin was  $14.3\mu mol/L$ , renal function showed serum creatinine of  $96\mu mol/L$  and blood urea of  $11.5mmol/L$ , serum sodium of  $137mmol/L$  and serum potassium of  $4.5mmol/L$ . Her CRP was 213.3mg/L and ESR was  $10mm/1^{st} hr.$ , urine analysis showed albumin + and moderately field full red cells. Dysmorphic RBC showed acanthocytes of 8%. Anti-streptolysin O titre (ASOT) was more than 200IU/L, urine protein creatinine ratio was 46mg/mmol (normal range  $<20 \rightarrow 2$ years) and stool for occult blood was positive. Antinuclear antibody was negative and rheumatoid factor was negative.

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**Figure 1.** Purpuric rashes on extensor surface of the knee joint.

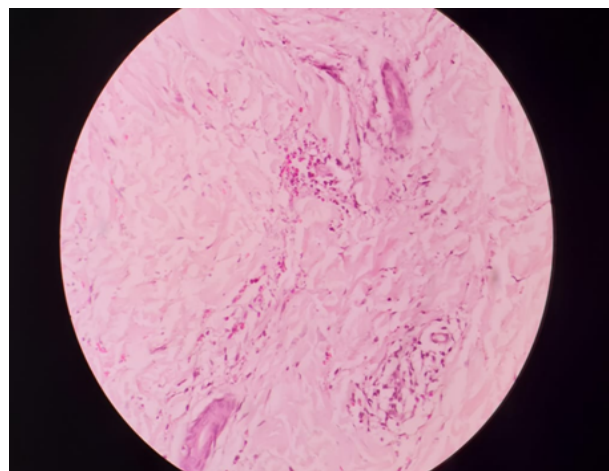


**Figure 3.** Purpuric rashes on extensor surface of right lower limb.

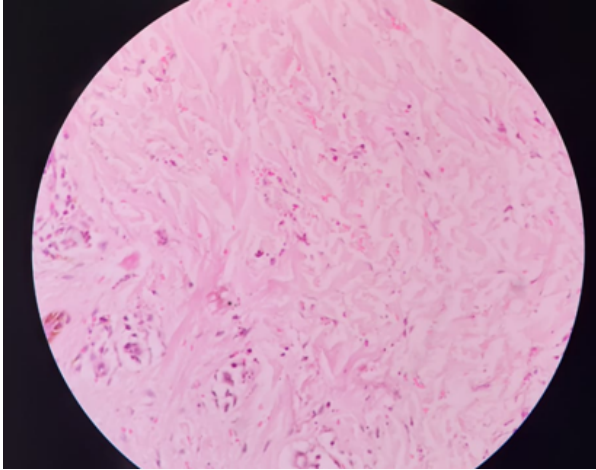


**Figure 2.** Purpuric rashes on lower limb.

Skin biopsy was taken from the purpuric rashes on the lower limb which showed perivascular lymphocytic infiltrate with infiltration of neutrophils into small vascular channels and extra vascular red blood cells. Features favour small vessel vasculitis (Figure 4 & 5). Mycoplasma antibody and Hepatitis B surface antigen were negative.



**Figure 4.** Perivascular lymphocytic infiltrate with infiltration of neutrophils into small vascular channels and extra vascular red blood cells. Features favour small vessel vasculitis.



**Figure 5.** *Perivascular lymphocytic infiltrate with infiltration of neutrophils into small vascular channels and extra vascular red blood cells. Features favour small vessel vasculitis.*

With these clinical and laboratory results, she was diagnosed with a case of adult-onset HSP involving gastro intestinal tract and complicated with gastro intestinal bleeding and was initially treated with intravenous methylprednisolone 500mg daily for three consecutive days, followed by oral prednisolone 60mg daily. She was hydrated with intravenous fluids. Her intensity of abdominal pain reduced progressively and resolved.

Purpuric rashes gradually faded away and gastro intestinal bleeding settled. She was managed with tapering dose of oral prednisolone over a period of four months and she showed complete recovery.

## Discussion

Diagnosis of HSP depends on clinical manifestations, and histopathological findings. To date, several diagnostic criteria have been used to diagnose HSP. EULAR/PRINTO/PRES criteria 2014 is the most recent gold standard<sup>4</sup>. Our patient met the standard requirements of this criteria to diagnose HSP.

Henoch Schonlein purpura manifests with pathophysiological signs including skin rash, arthritis, gastro intestinal disorders, and urinary abnormalities.

It has been considered that gastrointestinal involvement including abdominal pain, nausea, vomiting and/or gastrointestinal bleeding, occurs in more than half of patients with HSP<sup>5</sup>. Since endoscopy allows direct visualization of gastro intestinal tract which has been used widely for

diagnostic purposes<sup>6</sup>. But for our patient, we could not do endoscopic evaluation because of denial of consent by the patient.

Usually, HSP is preceded by upper respiratory tract infection. Commonly associated infective pathogens are group A streptococcus, mycoplasma, and herpes simplex virus<sup>7</sup>. Our patient's ASOT was positive.

Though the treatment is primarily supportive, hospital admission is needed for patients who present with severe abdominal pain, significant gastro intestinal bleeding or renal impairment. Supportive treatment consists of sufficient hydration and monitoring for complications of the renal and gastro intestinal system<sup>5</sup>. Because of symptomatic gastro-intestinal involvement, we kept our patient inward and managed.

Though the duration of common symptoms can be shortened by the usage of steroids but potential adverse effects must be weighed up against the benefits of steroids. Bleeding and oedema of the bowel wall and mesentery are the causes for abdominal symptoms in HSP. Therefore, the usage of steroid can contribute to reduce this oedema<sup>8</sup>. In this case, the abdominal pain began to improve with the treatment of glucocorticoids. Eventually, the rash completely disappeared and the laboratory parameters decreased to normal range.

## Conclusion

HSP is commonly seen in children and rare in adults. The pathophysiology of HSP is deposition of immune complexes in multi systems such as skin, joints, kidneys and gastro intestinal tract. Though the treatment is mostly supportive, hospital admission and usage of steroids have to be considered for the patients who are complicated with severe abdominal pain or severe renal involvement. Inward management with more potent steroids may be indicated for adults with HSP and systemic involvement.

## Overall comment

A literature search run on the topic does not seem to support the prolonged use of high dose corticosteroids over a prolonged time period as done in this case report. The disease severity here is questionable to warrant a high dose regime like the one used here. GI bleeding is not a rare manifestation.

<https://emedicine.medscape.com/article/984105-treatment#d9>

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