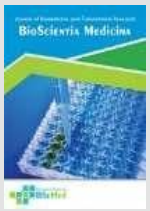
eISSN (Online): 2598-0580



Bioscientia Medicina: Journal of Biomedicine & Translational Research

Journal Homepage: www.bioscmed.com

A Henoch-Schönlein Purpura Case with Clinical Manifestation of Gastrointestinal Bleeding: A Case Report

Dian Daniella[1*], Pande Ketut Kurniari[2], Gde Somayana[3]

[1]Department of Internal Medicine, Faculty of Medicine, Universitas Udayana, Denpasar, Indonesia

[2]Division of Rheumatology, Department of Internal Medicine, Faculty of Medicine, Universitas Udayana, Denpasar, Indonesia

[3]Division of Gastroenterology and Hepatology, Department of Internal Medicine, Faculty of Medicine, Universitas Udayana, Denpasar, Indonesia

ARTICLE INFO

Keywords:
Abdominal pain
Arthralgia
Azathioprine
Henoch-schönlein purpura
Vasculitis

***Corresponding author:**
Dian Daniella

E-mail address:
dian.daniella@gmail.com

All authors have reviewed and approved the final version of the manuscript.

https://doi.org/10.37275/bsm.v7i3.786

A B S T R A C T

Background: Henoch-schönlein purpura (HSP) or also known as immunoglobulin A (IgA) vasculitis, is an autoimmune vasculitis of small blood vessels. The incidence of HSP in adults is only 5 in 100,000 adult patients. One of the causes of these low incidences is due to misdiagnosed or underdiagnosed. This is inevitable as in adults, and the disease usually presents with atypical signs and symptoms. One of the rare manifestations of this disease is gastrointestinal tract bleeding. This study aimed to describe a case of Henoch-schönlein purpura with gastrointestinal bleeding and the treatment. **Case presentation:** A rare case of a 20-year-old adult male diagnosed with HSP with the chief complaint of severe abdominal pain and gastrointestinal bleeding was presented. The patient also complained of arthralgia. Upon closer examination, the patient had palpable purpura on both the ankle and trunk. Urinalysis showed protein (+2) with erythrocyte 25-50 cells. Esophagogastroduodenoscopy showed erosive pangastritis, bile reflux, and duodenal submucosal bleeding with a normal duodenal bulb. The patient was diagnosed with HSP and was given methylprednisolone and azathioprine. On 1 month follow-up, the patient's complaints subside. **Conclusion:** Henoch-schönlein purpura should be considered in adult patients, especially in patients with skin lesions without thrombocytopenia accompanied by multiorgan involvement (gastrointestinal, renal, and joint).

1. Introduction

Henoch-schönlein purpura (HSP), or vasculitis immunoglobulin A (IgA), is a vasculitis of small blood vessels caused by autoimmune.[1] This disease is a vasculitis that generally occurs in children or begins to show clinical manifestations since childhood. This disease is so common in children that it accounts for almost half of the vasculitis that occurs in children. Although rare, HSP can also occur in adulthood. The incidence of HSP in adulthood is 5 in 100,000 patients.[2] Jithpratuck et al. stated that the low incidence of HSP in adulthood can be caused by misdiagnosis or underdiagnosis from clinicians who are not aware of the atypical clinical manifestations of HSP.[3]

Gastrointestinal bleeding is divided into upper and lower gastrointestinal bleeding. In upper gastrointestinal bleeding, bleeding occurs in the oesophagus, stomach, or duodenum (before the ligament of Treitz).[4] The most common cause of upper gastrointestinal bleeding is peptic ulcer (55-74%), either due to *Helicobacter pylori* infection or due to the

use of drugs such as non-steroidal anti-inflammatory drugs (NSAIDs) followed by oesophageal or gastric varices (5-14%). However, keep in mind that upper gastrointestinal bleeding can also be caused by vasculitis (2-3%).[5,6]

Generally, the clinical manifestations of HSP in childhood are mild and not life-threatening, but in adulthood, the clinical manifestations are generally more severe and require more aggressive therapy.[2] A typical clinical manifestation of HSP is palpable purpura (95%-100 %), abdominal pain (35%-85%), and joint pain (60%-84%).[7] In addition, there may also be atypical clinical manifestations in adult patients, such as gastrointestinal bleeding. Gastrointestinal bleeding only occurs in 18-52% of HSP patients and generally occurs together with abdominal pain.[8]

Atypical clinical manifestations with low incidence are a challenge for clinicians in diagnosing HSP. It is important for the clinician to inquire more deeply about other complaints felt by the patient. The right diagnosis is needed to be able to provide precise treatment. This case report was aimed to present a case of a patient diagnosed with HSP with clinical manifestations of gastrointestinal bleeding.

2. Case Presentation

A male, 20 years old, of Balinese ethnicity, came to the Emergency Room at Prof. Dr. I.G.N.G. Ngoerah Hospital with the chief complaint of abdominal pain. The patient had abdominal pain for eight days before entering the hospital and began to exacerbate one day before. The pain was felt throughout the abdomen and intermittently. Abdominal pain was worse after eating. The patient also complained of black and sticky stools.

The patient complained of palpable red spots that had appeared all over his body in the past two weeks before entering the hospital. The red spots were non-blanching. The patient was not aware of the area where the red spots first appeared, but the patient was aware that they were on both of the patient's ankles. Complaints were also accompanied by joint pain in the ankles without swelling, and it was exacerbated by activity, but complaints had already disappeared when the patient was admitted to the hospital. The patient also complained of gastrointestinal bleeding two weeks ago and was hospitalized.

On physical examination, the general condition was a moderate illness, and compos mentis, blood pressure at 130/70 mmHg, pulse 95 times per minute, respiratory rate 18 times per minute, and axillary temperature 36.8° Celsius, 98% oxygen saturation in room air. The patient's visual analogue scale was 8 out of 10. In the abdominal region, there was no distention with tenderness throughout the abdomen and normal bowel sounds. A digital rectal examination showed good anal sphincter tone, a smooth mucosal surface, and black stools. On the trunk, back, and extremities, multiple purpuras were found, well defined, palpable, round in shape, 0.5 – 0.8 cm in diameter, and did not disappear with pressure (Figure 1).

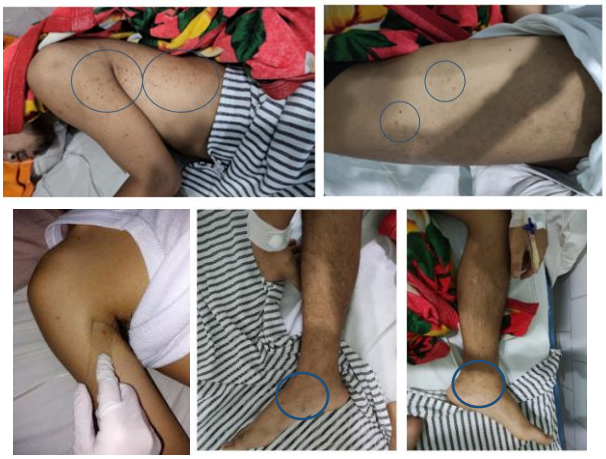


Figure 1. The clinical picture of the patient. The blue circle showed palpable purpura.

Laboratory examination showed leukocytes 24.57 x 10^3/µl, neutrophils 21.12 x 10^3/µl, lymphocytes 2.05 x 10^3/µl, haemoglobin 13.8 gr/dl, haematocrit 39.30% and platelets 430 x 10^3 /µl. Clinical chemistry examination showed serum glutamic oxaloacetic transaminase (SGOT) 70.5 U/L, serum glutamic pyruvic transaminase (SGPT) 61.50 U/L, blood urea nitrogen (BUN) 13.20 mg/dl, creatinine 0.97 mg/dl, estimated glomerular filtration rate (eGFR) 111.91 ml/minute/1.73m^2. Electrolyte examination showed sodium 136 mmol/L and potassium 4.32 mmol/L. Haemostatic physiologic examination obtained activated partial thromboplastin time (APTT) 30.5 seconds, prothrombin time (PT) 11.1 seconds, and international normalized ratio (INR) 0.97. A complete urine examination showed protein (+3), 25-50 erythrocytes, and 3-5 leukocytes with a protein creatinine ratio of 0.244. The patient underwent an esophagogastroduodenoscopy (EGD) examination. The EGD examination revealed bleeding spots of oesophageal, erosive pangastritis, and duodenal submucosal bleeding (Figure 2).

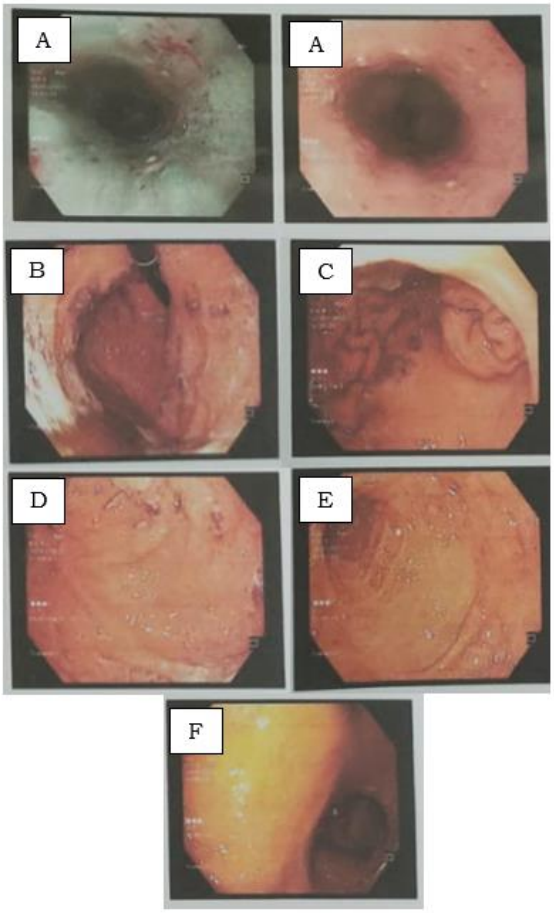


Figure 2. Esophagogastroduodenoscopy of patient. Bleeding spots in oesophagus (A); Cardia, fundus (B), antrum (C), and pylorus (D) showed oedema and submucosal bleeding; Normal duodenal bulb (E) with submucosal bleeding in duodenum pars descendens (F).

The patient was diagnosed as HSP with palpable purpura, oesophageal bleeding, gastric and duodenal submucosal bleeding, haematuria, and proteinuria. The patient received an intravenous injection of methylprednisolone 62.5 mg and continued with oral azathioprine 50 mg every 24 hours. At 1 month follow-

up after hospitalization, the patient had no complaints of abdominal pain and gastrointestinal bleeding with the therapy of methylprednisolone 8 mg every 12 hours orally and azathioprine 50 mg every 24 hours orally.

3. Discussion

Henoch-schönlein purpura is mediated by a type III hypersensitivity reaction with deposits of IgA immune complexes on the small vessel's walls. This disease generally attacks small blood vessels in the skin, gastrointestinal tract, kidneys, and joints.[9] This causes necrosis of the blood vessel walls with extravasation of erythrocytes and tissue infiltration with neutrophils.[8] The etiology of this disease is still unknown, but there are several factors that are thought to trigger it, such as an upper respiratory infection. In some cases, symptoms appear 7–14 days after an upper respiratory infection.[9] In addition to infection, drugs can also trigger HSP. Malignancies such as lung and prostate tumours and lymphoma are associated with HSP.[8]

Clinical manifestations of HSP in adults are often atypical, with more severe gastrointestinal manifestations compared to children.[10] Coppo et al. stated that in a cohort of 250 HSP patients (adults and children), 48% of patients presented with gastrointestinal complaints. Most of the gastrointestinal complaints were colicky abdominal pain, with 51% of them accompanied by gastrointestinal bleeding.[11] Generally, gastrointestinal symptoms occur within eight days of the appearance of skin lesions. It should be noted that in 10-15% of patients, abdominal pain and gastrointestinal bleeding may occur before the appearance of skin lesions. In a small proportion of patients, skin lesions do not even appear at all.[9] Gastrointestinal complaints that may appear vary from mild to severe, such as colicky abdominal pain, nausea, vomiting, diarrhoea, and constipation to complications such as perforation, ischemic vasculitis, intussusception, oesophageal ulcers, and severe gastrointestinal bleeding requiring transfusion.[12] Abdominal pain is generally colicky, aggravated by food, and localized to the epigastric and periumbilical regions. Pain occurs due to deposits of IgA in the walls of small blood vessels and polymorphonuclear cell infiltration around the blood vessels (mesenteric vasculitis), resulting in disturbances in the splanchnic circulation, causing intestinal ischemia and oedema of the gastrointestinal tract. Abdominal pain may occur with diarrhoea with profuse or occult bleeding, nausea, vomiting, and constipation. In most cases, the physical examination reveals a supple abdomen, but it should be noted that in some cases, the physical examination may resemble an acute abdomen. Gastrointestinal bleeding is generally occult, with the general complaint being melena.[9] In most cases, the bleeding that occurs is generally not severe, but there are cases with severe bleeding requiring blood transfusions.[9,13] Patients came with the main complaint of colicky abdominal pain. The pain was felt throughout the stomach. Abdominal pain was accompanied by spots on the skin, joint pain, and a history of gastrointestinal bleeding. Gastrointestinal bleeding in the patient is not massive but recurrent.

Arthralgia or arthritis is found in 2/3 of cases where it is commonly complained by adult patients, especially in the hips, knees, and ankles (inferior extremities), and is usually symmetrical. The symptoms are transient, migratory, and non-deforming. On physical examination, swelling and joint tenderness can be found without erythema in the joints.[9] The patient felt joint pain, but when he was admitted to the hospital, the complaints had subsided.

Skin clinical manifestations are the most common clinical manifestations, which are found in 70% of cases. The skin lesions in HSP are palpable purpura, non-thrombocytopenic and non-blanching. Commonly purpura appears in clusters and persists for three weeks. Purpura is symmetrical and occurs mainly on the lower extremities. In children, lesions may also occur on the back and buttocks.[9] In patients, multiple purpuric lesions were found on the extremities and back, which were palpable and did not disappear with pressure.

Laboratory examinations in this patient showed leucocytosis with neutrophilia and thrombocytosis accompanied by proteinuria and haematuria. In HSP patients, thrombocytopenia is not found, and leucocytosis generally occurs with a shift to the left.[1] Leucocytosis indicates inflammation and is not specific to HSP. Antinuclear antibodies, C3 and C4, generally show normal results.[14]

In 20-80% of HSP patients, renal abnormalities are found in the form of haematuria, proteinuria, nephrotic syndrome, and/or acute renal impairment.[1] The most common renal clinical manifestations are haematuria and mild proteinuria, where proteinuria that occurs without haematuria is very rare. Renal manifestations generally occur within four weeks and never precede skin lesions. Adult patients with clinical manifestations of the nephrotic or nephritic syndrome have a high rate of kidney failure which reaches 30%, and 2-5% of patients will end up with end-stage kidney disease. There are several things that increase the risk of kidney failure, such as severe proteinuria, increased serum creatinine, hypertension, and haematuria accompanied by proteinuria.[9] The patient's urine examination found proteinuria accompanied by haematuria.

In HSP patients, especially with gastrointestinal complaints, endoscopy plays an important role in diagnosis. Generally, with EGD, we will find diffuse mucosal erythema, petechiae, erosive duodenitis, bleeding, and ulcers. Purpuric lesions can be found in the duodenum pars descendens, gaster and colon.[9] The cohort by Zhang et al. in 115 adult patients showed that the most common endoscopic lesions were oedema and multiple irregular ulcers, especially in the duodenum.[15] Han et al. stated that EGD examination would show oedema (12%), erythema/petechiae (65%), and erosion/ulcer (58%).[16] It should be noted that although the lesions are generally found in the duodenum, the duodenal bulb is rarely affected. Esophagogastroduodenoscopy examination in this patient revealed a hiatal hernia, bleeding spots in the oesophageal, erosive pangastritis, bile reflux, and duodenal submucosal bleeding. Another investigation that can be done is a computed tomography (CT) scan. In HSP, you will find symmetrical and circumferential wall thickening and dilation of the mesenteric vessels. CT angiography examination shows occlusion of blood vessels.[9]

The diagnosis of HSP is established by the presence of palpable purpura, especially in the lower extremities, accompanied by any of the following symptoms: diffuse abdominal pain, IgA deposits on biopsy, arthritis/arthralgia, and renal involvement (haematuria and/or proteinuria).[17] The patient met the diagnostic criteria.

The presence of IgA deposits on the vessel wall is pathognomonic for HSP. Skin biopsies should be taken within 24 hours because, in chronic lesions, damaged blood vessels may show nonspecific leakage of any Ig.[18] However, in situations where no skin biopsy was performed, as in this case, the diagnostic criteria can be used with a sensitivity of 100%. And specificity of 87%.[17]

This vasculitis is generally self-limited in 94% of children and 89% of adults. Hospitalization is performed on patients with dehydration, bleeding, or severe pain. Paracetamol and/or NSAIDs may be given for skin lesions and arthritis. If there is no improvement, the patient can be given 1 mg/day of colchicine.[7]

In mild abdominal pain, symptomatic therapy such as paracetamol can be given. However, if the pain persists, then dapsone, colchicine, or low-dose prednisone (0.5 mg/kg/day) can be given. In patients with severe gastrointestinal complaints (perforation and severe bleeding), it is necessary to evaluate the need for surgery. These patients can be given oral prednisone 1 mg/kg/day. In this case, pulsed methylprednisolone can be considered (1 gram intravenously for 3 days), cyclophosphamide (CYC), and surgery.[7] Oral prednisone can be given 1-2 mg/kg/day for one to two weeks, followed by a tapering dose of 0.5 mg/kg/day for the next week, followed by 0.5 mg/kg/day every other day for one week. In patients who cannot tolerate oral corticosteroids, intravenous methylprednisolone can

be given.[1] Various case reports state that corticosteroids have a good therapeutic response in HSP cases with gastrointestinal bleeding.[19,20] Corticosteroids in HSP reduce oedema of the intestinal wall, thereby reducing existing symptoms. However, it has not been shown to prevent the recurrence of abdominal pain.[12] Huber et al. conducted a randomized controlled trial on the use of prednisone and found that prednisone can reduce the risk of intussusception.[21] A meta-analysis of 15 studies stated that the use of corticosteroids decreases the resolution time for complaints of abdominal pain and reduces the risk of persistent kidney disorders.[22] The use of drugs that inhibit gastric acid secretion has been proven to reduce gastrointestinal symptoms.[12]

In patients with mild clinical manifestations of the kidney (haematuria, proteinuria <0.5 grams per day, normal GFR), re-evaluation can be done three months later. In patients with moderate renal clinical manifestations (haematuria, proteinuria > 0.5 grams per day, normal GFR), angiotensin-converting enzyme inhibitors (ACEi) can be given. In patients with clinical manifestations of severe kidney disease (acute and rapidly progressive renal failure) or proteinuria >1 gram per day, a combination of pulse dose methylprednisolone and low-dose prednisone and CYC is given. If treatment failure occurs, then consider cyclosporine A and rituximab.[7]

The patient was given an intravenous methylprednisolone injection and continued with oral azathioprine 50 mg every 24 hours. In various case reports, intravenous corticosteroids have been the treatment of choice in patients with gastrointestinal bleeding.[19] Azathioprine is the recommended steroid-sparing agent for steroid-resistant patients. There are no guidelines for the duration of treatment, but a case series of six recommends continuing therapy until remission for six to 15 months.[23]

4. Conclusion

This case reported a male patient, aged 20 years, with a diagnosis of HSP. The patient was diagnosed with HSP by fulfilling the diagnostic criteria of HSP, which are palpable purpura, diffuse abdominal pain, arthralgia, and kidney involvement in the form of proteinuria and haematuria. Immunosuppressant therapy showed a good clinical response. As clinicians, HSP needs to be considered in adult patients with palpable purpura without thrombocytopenia accompanied by multiorgan involvement (gastrointestinal, kidney, and joints).

5. References

1. Khader Y, Burmeister C, Patel D, Ambati A, Altorok N. Henoch-Schonlein purpura presenting as upper gastrointestinal bleed in an adult patient. Cureus. 2021; 13(3): 1–6.
2. Lei WT, Tsai PL, Chu SH, Kao YH, Lin CY, Fang LC, et al. Incidence and risk factors for recurrent Henoch-Schönlein purpura in children from a 16-year nationwide database. Pediatr Rheumatol. 2018; 16(1): 25.
3. Jithpratuck W, Elshenawy Y, Saleh H, Youngberg G, Chi DS, Krishnaswamy G. The clinical implications of adult-onset henoch-schonelin purpura. Clin Mol Allergy. 2011; 9(1): 9.
4. Stanley AJ, Laine L. Management of acute upper gastrointestinal bleeding. BMJ. 2019; l536.
5. Lanas A, Dumonceau JM, Hunt RH, Fujishiro M, Scheiman JM, Gralnek IM, et al. Non-variceal upper gastrointestinal bleeding. Nat Rev Dis Prim. 2018; 4(1): 18020.
6. Lee EW, Laberge JM. Differential diagnosis of gastrointestinal bleeding. Tech Vasc Interv Radiol. 2004; 7(3): 112–22.
7. Audemard-Verger A, Pillebout E, Guillevin L, Thervet E, Terrier B. IgA vasculitis (Henoch–Shönlein purpura) in adults: Diagnostic and therapeutic aspects. Autoimmun Rev. 2015; 14(7): 579–85.
8. Ebert EC. Gastrointestinal manifestations of Henoch-Schonlein purpura. Dig Dis Sci. 2008; 53(8): 2011–9.

9. Cojocariu C, Maria Singeap A, Chiriac S, Sfarti C, Girleanu I, Petrea O, et al. Gastrointestinal manifestations of IgA Vasculitis-Henoch-Schönlein purpura. In: Digestive System - Recent Advances. IntechOpen; 2020.
10. Roberts PF, Waller TA, Brinker TM, Riffe IZ, Sayre JW, Bratton RL. Henoch-Schönlein purpura: A review article. South Med J. 2007; 100(8): 821–4.
11. Coppo R. Long-term prognosis of Henoch-Schonlein nephritis in adults and children. Nephrol Dial Transplant. 1997; 12(11): 2277–83.
12. Hamzaoui A, Melki W, Harzallah O, Njim L, Klii R, Mahjoub S. Gastrointestinal involvement revealing Henoch Schonlein purpura in adults: Report of three cases and review of the literature. Int Arch Med. 2011; 4(1): 31.
13. Wang S, Tang H, Du W, Ding Y. Massive gastrointestinal hemorrhage caused by Henoch-Schoenlein purpura. Medicine (Baltimore). 2021; 100(50): e28240.
14. Yang YH, Yu HH, Chiang BL. The diagnosis and classification of Henoch-Schönlein purpura: An updated review. Autoimmun Rev. 2014; 13(4–5): 355–8.
15. Zhang Y, Huang X. Gastrointestinal involvement in Henoch-Schönlein purpura. Scand J Gastroenterol. 2008; 43(9): 1038–43.
16. Han Y, Jin SY, Kim DW, Jeen YM, Kim YH, Choi IH. Endoscopic and microscopic findings of gastrointestinal tract in Henoch-Schönlein purpura: Single institute experience with review of literature. Medicine (Baltimore). 2019; 98(20): e15643.
17. Ozen S, Pistorio A, Iusan SM, Bakkaloglu A, Herlin T, Brik R, et al. EULAR/PRINTO/PRES criteria for Henoch-Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: Final classification criteria. Ann Rheum Dis. 2010; 69(5): 798–806.
18. Akbar N, Akbar S, Kubbara A. Henoch-Schonlein purpura with a negative skin biopsy. J Investig Med. 2016; 64(4): 978.2-979.
19. Yi F, Bai Z, Li Y, Xu X, Guo X, Qi X. A good response to glucocorticoid for Henoch-Schönlein purpura with abdominal pain and gastrointestinal bleeding in an adult: A CARE case report. Med (United States). 2020; 99(1): 1–5.
20. Kang HS, Chung HS, Kang KS, Han KH. High-dose methylprednisolone pulse therapy for treatment of refractory intestinal involvement caused by Henoch-Schönlein purpura: A case report. J Med Case Rep. 2015; 9(1): 1–5.
21. Huber AM, King J, McLaine P, Klassen T, Pothos M. A randomized, placebo-controlled trial of prednisone in early Henoch Schönlein Purpura [ISRCTN85109383]. BMC Med. 2004; 2(1): 7.
22. Weiss PF, Feinstein JA, Luan X, Burnham JM, Feudtner C. Effects of corticosteroid on Henoch-Schönlein purpura: A systematic review. Pediatrics. 2007; 120(5): 1079–87.
23. Fotis L, Tuttle PV, Baszis KW, Pepmueller PH, Moore TL, White AJ. Azathioprine therapy for steroid-resistant Henoch-Schönlein purpura: a report of 6 cases. Pediatr Rheumatol. 2016; 14(1): 37.