

• CASE REPORT •

Endoscopic findings in a patient with Henoch-Schönlein purpura

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Received: 2004-07-12 Accepted: 2005-01-21

Abstract

Henoch-Schönlein purpura (HSP) is a systemic vasculitis of the small vessels of the skin, joints, GI tract, and kidney. It preferentially affects children but may also occur in adults. We report a 60-year-old man with HSP who presented with colicky abdominal pain, bloody diarrhea, arthralgia, and skin rash. The gastrointestinal tract was viewed by upper endoscopy and colonoscopy. We found characteristic endoscopic findings in the stomach, cecum and sigmoid colon, the combination of which has rarely been demonstrated in one patient. Histologic examination of skin biopsy specimens revealed leukocytoclastic vasculitis with positive staining for IgA in the capillaries. Endoscopy appears to have substantial diagnostic utility in patients suspected of having HSP, especially when abdominal symptoms precede the cutaneous lesions.

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Key words: Henoch-Schönlein purpura; Purpura; Endoscopy

Chen MJ, Wang TE, Chang WH, Tsai SJ, Liao WS. Endoscopic findings in a patient with Henoch-Schönlein purpura. *World J Gastroenterol* 2005; 11(15): 2354-2356
<http://www.wjgnet.com/1007-9327/11/2354.asp>

INTRODUCTION

Henoch-Schönlein purpura (HSP) is a systemic vasculitic disorder of the small vessels of the skin, joints, GI tract, and kidney. The clinical manifestations usually include arthralgia, a purpuric skin rash, nephritis, abdominal pain, and GI bleeding. It commonly affects young children, with the peak age of onset between 4 and 7 years. The pathologic changes are mediated by IgA deposition^[1], complement activation, granulocyte recruitment, and destruction of endothelial cells. The American College of Rheumatology published diagnostic criteria for HSP in 1990^[2] including (1) age less than or equal to 20 years at disease onset, (2) the presence of palpable purpura, (3) GI bleeding, and

(4) a biopsy showing granulocytes in the walls of small arterioles or venules. The diagnosis is made when three or more of those criteria are fulfilled.

CASE REPORT

A 60-year-old man presented with three days of colicky abdominal pain with bloody diarrhea, arthralgia in the knees, and ankle edema. His medical history was unremarkable except for a self-limited upper respiratory infection one week previously. Physical examination revealed diffuse lower abdominal rebound tenderness. There was no fever and no skin rash on admission. The initial leukocyte count was 17 830 /mm³, and the C-reactive protein was 12 mg/dL. A plain abdominal film showed generalized ileus and no evidence of free air. Stool cultures were negative for enteric pathogens, and no ova or parasites were found. Upper endoscopy revealed multiple discrete coin-like petechiae and hemorrhagic erosions in the gastric antrum and lower body (Figure 1). Colonoscopy revealed patches of hyperemic and ecchymotic lesions in the cecum (Figure 2), ileocecal valve, and sigmoid colon. The patient was managed with fasting and intravenous fluids. The day after admission, he developed numerous erythematous, purpuric lesions, predominantly on the lower extremities and buttocks. The lesions rapidly became palpable (Figure 3). A platelet count, prothrombin time and partial thromboplastin time were normal. Based on this constellation of findings, HSP was suspected. Tissue from a skin biopsy of the lesions was compatible with leukocytoclastic vasculitis with deposition of IgA, confirming the diagnosis of HSP. The urinalysis and renal function were normal. Parenteral steroid therapy (5 mg/(kg·d) of hydrocortisone) was instituted initially, resulting in relief of the abdominal pain and arthralgia and gradual resolution of the skin lesions within 5 d. The treatment was switched to oral prednisolone 20 mg/d with gradual tapering over 4 wk. The patient has remained well over 2 years of follow up.

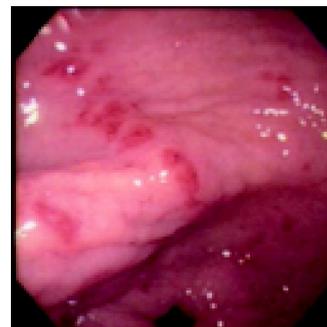


Figure 1 Upper endoscopic view of multiple discrete coin-like petechiae in the lower gastric body.



Figure 2 Colonscopic view of hyperemic and ecchymotic lesions in the cecum.



Figure 3 Numerous erythematous papules and palpable purpura on the lower extremities and buttocks.

DISCUSSION

The above-mentioned patient fulfilled three of four criteria for the diagnosis of HSP. The only exception was his age. The diagnostic sensitivity and specificity of three out of the four criteria are 89.4% and 88.1%, respectively^[2]. Although the cause of HSP is unknown, immunizations, certain food allergies, insect bites, infection^[3], and some medications^[4] may play a role in the development of the disease. It may be that our patient's recent cold predisposed him to HSP. The final diagnosis of HSP was confirmed by histologic examination of a skin biopsy specimen demonstrating a vasculitis with leukocyte infiltration and deposition of IgA.

The diagnosis of HSP can be difficult, especially when abdominal symptoms precede cutaneous lesions. A skin rash is nearly always present, although not necessarily in the earliest stages. Lanzkowsky reported that 14% of patients had abdominal symptoms preceding the skin rash^[5]. As in our case, the skin rash is typically on the lower extremities and buttocks, less commonly on the face and trunk. It starts as erythematous papules that quickly become palpable purpura. Normal platelets counts, prothrombin time, and partial thromboplastin time exclude a platelet or hemorrhagic disorder as the cause of purpura. Other laboratory tests are usually not conclusive.

Abdominal symptoms reportedly occur in 50-85% of HSP patients, possibly secondary to edema and hemorrhage within the bowel wall and mesentery. The abdominal pain is always colicky and poorly localized. On physical examination the abdomen may be rigid or distended, occasionally mimicking an acute abdomen and resulting in unnecessary exploratory laparotomy. Signs suggestive of intussusception including an abdominal mass may be present in children. GI bleeding occurs in one-third of cases and may be overt, with hematemesis, melena, or bloody stools^[6,7]. While bloody diarrhea is common in HSP, it may also occur in eosinophilic gastroenteritis^[8], systemic lupus erythematosus, parasitic infection, and drug-induced vasculitis. Distinguishing between these different vasculitic disorders depends on clinical, serological, hematologic, and histologic findings. Arthralgia is usually symmetrical, with the ankles being the most commonly affected joints. Joint involvement is considered as an indication for systemic steroids. Proteinuria and hematuria indicate renal involvement and generally occur within the first 3 mo after the onset of the disease.

The hallmarks of HSP on computed tomography are multiple focal areas of bowel thickening with skip lesions, mesenteric edema, vascular engorgement, and non-specific lymphadenopathy^[9]. However, similar thickening may also be seen in eosinophilic gastroenteritis^[8], lymphoma, Crohn's disease, and granulomatous disease. It is thus not a specific sign for HSP. Ultrasound may reveal generalized thickening of the intestinal wall, ascites, and sometimes intussusception.

GI involvement in HSP is seen predominantly in the small bowel^[10,11] but may also affect the esophagus, stomach, terminal ileum^[12,13] and colon^[14]. Our patient had diffuse, poorly localized abdominal pain and bloody diarrhea, prompting us to visualize both the upper and lower GI tract. The endoscopic findings in our patient included discrete coin-like petechiae, hemorrhagic erosions, and skip hyperemic and ecchymotic lesions. These were seen in the gastric antrum, cecum, ileocecal valve, and sigmoid colon. It is important to recognize these characteristic endoscopic findings when a previously healthy patient presents with the sudden onset of an acute abdomen. When encountered, they should suggest the possibility of HSP. Although we did not biopsy the lesions in the GI tract, we expected to find histologic abnormalities similar to those found on skin biopsy. However, endoscopic biopsies often miss the submucosal vessels and only reveal nonspecific inflammation.

HSP is primarily a medical disease and requires only supportive treatment once other acute surgical conditions have been excluded. Simple analgesics or non-steroidal anti-inflammatory drugs are used as first-line therapy for relief of arthralgia. Parenteral steroids have been advocated for more severe abdominal or joint pains, or painful angioedema, but they should be used with caution if there is active GI bleeding. All patients with HSP should have their urine analyzed on several occasions during the initial stages of the disease. Proteinuria and hematuria indicate possible renal involvement which, if it progresses to renal insufficiency, has a poor long-term outcome^[15,16]. Our patient's urinalysis was normal, and his renal function has been remained normal over two years of follow up.

In conclusion, the majority of patients with HSP recover fully. The prognosis is best in the absence of renal involvement. The diagnosis of HSP may be difficult, especially when abdominal symptoms precede the characteristic palpable

purpura. Typical endoscopic findings may alert gastroenterologists to consider this disease early and thus avoid unnecessary laparotomy.

REFERENCES

- 1 **Uchiyama K**, Yoshida N, Mizobuchi M, Higashihara H, Naito Y, Yoshikawa T. Mucosal IgA deposition [correction of depositon] in Henoch-Schonlein purpura with duodenal ulcer. *J Gastroenterol Hepatol* 2002; **17**: 728-729
- 2 **Mills JA**, Michel BA, Bloch DA, Calabrese LH, Hunder GG, Arend WP, Edworthy SM, Fauci AS, Leavitt RY, Lie JT. The American College of Rheumatology 1990 criteria for the classification of Henoch-Schonlein purpura. *Arthritis Rheum* 1990; **33**: 1114-1121
- 3 **Gimenez-Esparza Vich JA**, Arguelles BF, Martin IH, Gutierrez Fernandez MJ, Porras Vivas JJ. Recurrence of Henoch-Schonlein purpura in association with colitis. *J Clin Gastroenterol* 2002; **34**: 492-493
- 4 **Borras-Blasco J**, Enriquez R, Amoros F, Cabezuelo JB, Navarro-Ruiz A, Perez M, Fernandez J. Henoch-Schonlein purpura associated with clarithromycin. Case report and review of literature. *Int J Clin Pharmacol Ther* 2003; **41**: 213-216
- 5 **Lanzkowsky S**, Lanzkowsky L, Lanzkowsky P. Henoch-Schoenlein purpura. *Pediatr Rev* 1992; **13**: 130-137
- 6 **Nathan K**, Gunasekaran TS, Berman JH. Recurrent gastrointestinal Henoch-Schonlein purpura. *J Clin Gastroenterol* 1999; **29**: 86-89
- 7 **Lippl F**, Huber W, Werner M, Nekarda H, Berger H, Weigert N. Life-threatening gastrointestinal bleeding due to a jejunal lesion of Henoch-Schonlein purpura. *Endoscopy* 2001; **33**: 811-813
- 8 **Chen MJ**, Chu CH, Lin SC, Shih SC, Wang TE. Eosinophilic gastroenteritis: clinical experience with 15 patients. *World J Gastroenterol* 2003; **9**: 2813-2816
- 9 **Jeong YK**, Ha HK, Yoon CH, Gong G, Kim PN, Lee MG, Min YI, Auh YH. Gastrointestinal involvement in Henoch Schonlein syndrome: CT findings. *AJR Am J Roentgenol* 1997; **168**: 965-968
- 10 **Esaki M**, Matsumoto T, Nakamura S, Kawasaki M, Iwai K, Hirakawa K, Tarumi K, Yao T, Iida M. GI involvement in Henoch-Schonlein purpura. *Gastrointest Endosc* 2002; **56**: 920-923
- 11 **Pore G**. GI lesions in Henoch-Schonlein purpura. *Gastrointest Endosc* 2002; **55**: 283-286
- 12 **Ortego-Centeno N**, Callejas-Rubio JL, Lopez-Manas JG, Troncoso-Garcia E, de la Higuera Torres-Puchol J, Ileitis terminalis in a patient with Henoch-Schonlein purpura. *Dig Dis Sci* 1999; **44**: 1590-1593
- 13 **Kim HS**, Lee DK, Baik SK, Kwon SO. Ileal vasculitis in Henoch-Schonlein purpura. *Gastrointest Endosc* 2001; **54**: 493-494
- 14 **Nakasono H**, Hokama A, Fukuchi J, Makishi T, Yamashiro T, Sakugawa H, Kinjo F, Saito A. Colonoscopic findings in an adult patient with Henoch-Schonlein purpura. *Gastrointest Endosc* 2000; **52**: 392
- 15 **Pillebout E**, Thervet E, Hill G, Alberti C, Vanhille P, Nochy D. Henoch-Schonlein Purpura in adults: outcome and prognostic factors. *J Am Soc Nephrol* 2002; **13**: 1271-1278
- 16 **Garcia-Porrúa C**, Gonzalez-Louzao C, Llorca J, Gonzalez-Gay MA. Predictive factors for renal sequelae in adults with Henoch-Schonlein purpura. *J Rheumatol* 2001; **28**: 1019-1024

Science Editor Guo SY Language Editor Elsevier HK