A Rare Cause of Ileocolitis: Henoch-Schonlein Purpura: Case Report

İleokolitin Nadir Bir Nedeni: Henoch-Schönlein Purpurası

Enver ÜÇBİLEK, MD,[a]
Engin ALTINTAŞ, MD,[a]
Orhan SEZGİN, MD,[a]
Tülin DOĞANER, MD,[b]
Refik OLMAZ, MD,[b]
Ayşe POLAT, MD[c]

Departments of
[a]Gastroenterology,
[b]Internal Medicine,
[c]Patology,
Mersin University Faculty of Medicine, Mersin

Geliş Tarihi/*Received:* 20.10.2008
Kabul Tarihi/*Accepted:* 29.01.2009

Yazışma Adresi/*Correspondence:*
Enver ÜÇBİLEK, MD
Mersin University Faculty of Medicine,
Department of Gastroenterology,
Mersin,
TÜRKİYE/TURKEY
enucbilek@hotmail.com

ABSTRACT Henoch-Schonlein Purpura (HSP) is a disease which may frequently involve the gastrointestinal tract and may cause gastrointestinal bleeding. Terminal ileum involvement of HSP has been rarely reported. Here, we will report ultrasonographic and endoscopic findings of a case of ileocolic HSP presenting with hematochezia. Abdominal ultrasonography showed an increase in the wall thickness of the terminal ileum. On colonoscopy, the terminal ileum was hyperemic and edematous and had small ulcers and the colon had very small hemorrhagic ulcers. Biopsy obtained from the terminal ileum showed vasculitis. Steroids were administered and hematochezia and the signs of Henoch-Schönlein Purpura detected on ultrasonography relieved.

Key Words: Purpura, Schoenlein-Henoch; Crohn disease

ÖZET Henoch-Schönlein Purpurası (HSP) sıklıkla gastrointestinal kanalı tutan ve gastrointestinal kanamaya yol açabilen bir hastalıktır. HSP'de terminal ileum tutulumu nadir bildirilmiştir. Biz, burada hematokezya ile başvuran ileokolik tutulumlu HSP olgusunu ultrasonografik ve endoskopik bulguları ile sunuyoruz. Abdominal ultrasonografi, terminal ileum duvarında kalınlaşmayı göstermiştir. Kolonoskopide terminal ileumda hiperemi, ödemli olup küçük ülserler izlenirken, kolonda çok küçük hemorajik ülserler görülmüştür. Terminal ileumdan alınan biyopside vaskülit izlenmiştir. Hastaya steroid tedavisi verilmiş ve hematokezya ve Henoch-Schönlein Purpurasına ait ultrasonografik bulgular gerilemiştir.

Anahtar Kelimeler: Henoch-Schönlein purpurası; Crohn hastalığı

Turkiye Klinikleri J Gastroenterohepatol 2010;17(1):44-7

Henoch-Schönlein purpura (HSP) is a rare cause of bleeding in the lower gastrointestinal tract although it frequently involves the gastrointestinal tract. HSP involving the terminal ileum has been even more rarely reported.[1,2] In the light of endoscopic and ultrasonographic findings and the relevant literature, we will report a case of HSP involving both the ileum and the colon, predominantly the terminal ileum and presenting with hematochezia.

CASE REPORT

An eighteen-year-old female presented to our clinic with hematochezia continuing for two days. On history, she had upper respiratory tract infection one month ago and purpura on her legs and, mild, widespread abdominal pa-

in started two weeks ago. Based on the clinical signs, HSP was diagnosed. Since the clinical signs of the disease were so evident that we did not need to perform a skin biopsy. The patient was administered a single dose of 40 mg methylprednisolone. Three days before her presentation to our clinic, abdominal pain became more severe and prednisolone 40 mg/day po was initiated. One day later, bloody diarrhea started. She had bloody stool seven-eight times a day and bright rectal bleeding only twice a day. Physical examination revealed palpable purpura 3-8mm in diameter on the lower extremities. There was bright red blood on the gloves during digital examination of the rectum. Laboratory investigations revealed normal results except for a high white cell count (24.000/mm^3). Abdominal ultrasonography (USG) demonstrated an increase in the thickness of the terminal ileum wall (Figure 1). On colonoscopy, the terminal ileum was hyperemic, edematous and granular. There were occasional small ulcers covered with white exudate. Throughout the colon a few very small hemorrhagic ulcers were present (Figure 2). Biopsy obtained from the terminal ileum revealed edema in the lamina propria, intensive neutrophil infiltration, swollen endothelium in few vessels with a small diameter, neutrophil leukocyte and cell infiltration in the vessel walls and fibrinoid necrosis in the walls of a few vessels (Figure 3). Treatment with prednisolone 40 mg/day (0.8 mg/kg/day) was

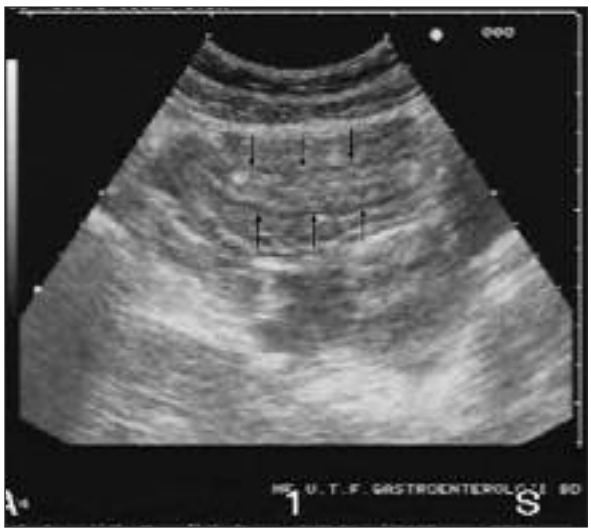

FIGURE 1: Abdominal ultrasonography demonstrated an increase in wall thickness of the terminal ileum.

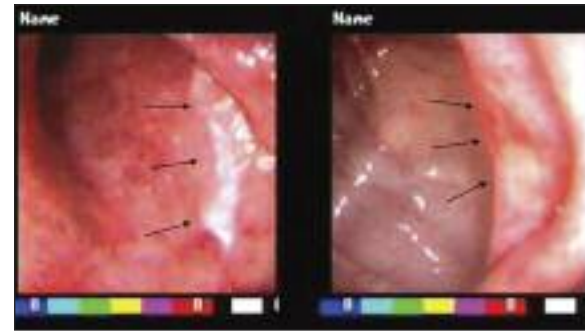

FIGURE 2: On colonoscopy, the terminal ileum was hyperemic, edematous and granular. There were occasional small ulcers covered with white exudate.

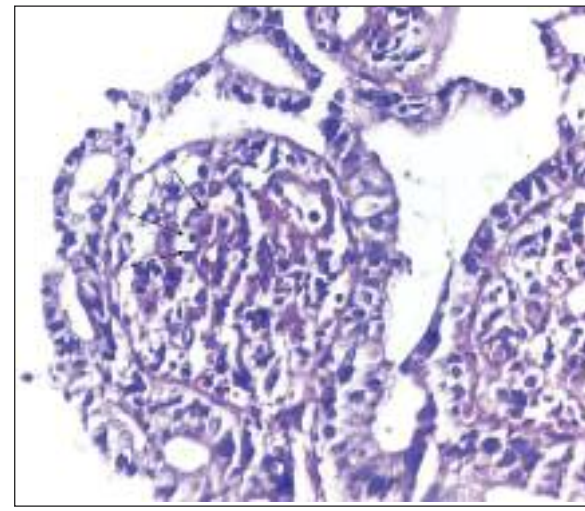

FIGURE 3: Biopsy obtained from the terminal ileum revealed edema in the lamina propria, intensive neutrophil infiltration, swollen endothelium in few vessels with a small diameter, neutrophil leukocyte and cell infiltration in the vessel walls and fibrinoid necrosis in the walls of a few vessels.

continued. Within two days of treatment, palpable purpura and diarrhea relieved, abdominal USG demonstrated an improvement in the ileal wall thickening. Two weeks later, the dose of the steroid was begun to gradually decrease and then it was discontinued. Two months after the end of the treatment, the patient did not have any complaints and physical examination and laboratory investigations were found normal.

DISCUSSION

HSP is a systemic vasculitis involving small vessels and characterized by palpable purpura, artralgia, abdominal pain, gastrointestinal bleeding and nephritis.

It usually occurs in young children. An immunocomplex deposition plays a role in its pathogenesis.[3,4] The diagnostic criteria for HSP published in 2006 were palpable purpura (a mandatory criterion) in the presence of at least one of the following: (1) diffuse abdominal pain; (2) any biopsy showing predominant IgA deposition; (3) arthritis or arthralgia; and (4) renal involvement (hematuria and/or proteinuria). Most studies, however, used the old criteria, or variations thereof where at least two of the following are required: (1) age less than 20; (2) palpable purpura; (3) acute abdominal pain usually with hematochezia; and (4) granulocytic infiltration of arteriolar or venular walls.[5] Although diagnostic criteria include gastrointestinal involvement, gastrointestinal symptoms appear before typical clinical signs occur. This causes diagnostic confusion and makes it difficult to determine whether bleeding is due to HSP or another condition in 10-15% of the cases.[1]

HSP is often associated with upper respiratory tract infections or tumors. It may coexist with or mimic Crohn's disease. Various drugs and food and insect bites are implicated in HSP.[6,7] The disease may involve the gastrointestinal tract from the esophagus to the rectum.[2] Among gastrointestinal symptoms there are abdominal pain (86%), severe gastrointestinal bleeding (20%), occult gastrointestinal bleeding (20%) and diarrhea (20%). Abdominal pain worsens with meal from bowel angina. It has been reported that 3-5% of the cases might have intestinal infarct, perforation and invagination. Sometimes gastrointestinal symptoms may precede skin rash.[8] In the case presented here, skin rash and abdominal pain started concurrently and two weeks later bloody diarrhea and hematochezia developed.

In the present case, USG showed an increase in the thickness of the terminal ileum wall. This was the sign that helped us to determine the involved organ. At first, the thickening suggested the diseases involving the terminal ileum such as Crohn's disease, infections (Yersinia enterocolitica, Campylobacter jejuni, Entamoeba histolytica, Salmonella and tuberculosis etc.), Behçet's disease and lymphoproliferative diseases. However, based on the marked clinical signs, we easily made the diagnosis of HSP. HSP may coexist with Crohn's disease or may mimic its findings with ileitis or colitis. Compared to HSP, Crohn's disease does not have IgA deposition, rarely affects the duodenum and jejunum, more likely results and fibrosis and stricture formation, and usually does not manifest as thumbprinting from edema and intramural hemorrhage. In addition, Crohn's disease usually does not resolve as quickly as HSP and is more prone to relapse.[5] In our case, there was no endoscopically typical findings such as cobble-stone and fissure-like ulcers and stricture for Crohn's disease. Additionally, rapid resolution of the sonographic ileal wall thickness was determined. There have been few cases of HSP involving the terminal ileum and there is little information about endoscopic and ultrasonographic views of HSP involving the ileum.[2,8-10] Akdamar et al. were the first to report views of HSP obtained through gastric and duodenal endoscopy.[10] Endoscopic signs of HSP involving the ileum were first reported by Kawasaki et al. in 1997. They noted that a 43-year-old patient had marked edema in the terminal ileum accompanied by hemorrhagic and ulcerated mucosa, but a normal colorectum.[2] In the case presented here, there was ileocolonic involvement and a more severe involvement of the terminal ileum, with edema, granularity and ulcers covered with white exudate in the mucosa of the terminal ileum. There were also a few very small hemorrhagic ulcers in the entire colon.

It has been reported that among ultrasonographic findings of HSP there are thickening in the intestinal wall, mesenteric edema, peritoneal fluid and non-specific lymphodenopathy.[11-14] In the case presented here, USG demonstrated an increase in the thickness of the terminal ileum wall. The thickening regressed following the treatment with corticosteroids. This shows that USG can be useful in diagnosis and follow up examinations of HSP.

HSP is treated with analgesics and non-steroidal anti-inflammatory drugs. Steroids are recommended for severe abdominal and joint pain and severe renal involvement.[11] In the case presented here, steroids relieved gastrointestinal and skin symptoms. To conclude, HSP may involve the ileocolon, though rarely, and abdominal USG is a valuable diagnostic tool for the gastrointestinal involvement of HSP.

REFERENCES

1. Agha FP, Nostrant TT, Keren DF. Leucocytoclastic vasculitis (hypersensitivity angiitis) of the small bowel presenting with severe gastrointestinal hemorrhage. Am J Gastroenterol 1986;81(3):195-8.
2. Kawasaki M, Hizawa K, Aoyagi K, Kuroki F, Nakahara T, Sakamoto K, et al. Ileitis caused by Henoch-Schönlein purpura. An endoscopic view of the terminal ileum. J Clin Gastroenterol 1997;25(1):396-8.
3. Uchiyama K, Yoshida N, Mizobuchi M, Higashihara H, Naito Y, Yoshikawa T. Mucosal IgA deposition [correction of depositon] in Henoch-Schönlein purpura with duodenal ulcer. J Gastroenterol Hepatol 2002;17(6):728-9.
4. Zengin G, Lenk N, Artüz F, Sönmezer S, Allı N. [Henoch-schonlein purpura]. Turkiye Klinikleri J Dermatol 1994;4(2):91-3.
5. Ebert EC. Gastrointestinal manifestations of Henoch-Schonlein Purpura. Dig Dis Sci 2008;53(8):2011-9.
6. Giménez-Esparza Vich JA, Argüelles BF, Martin IH, Gutierrez Fernández MJ, Porras Vivas JJ. Recurrence of Henoch-Schönlein purpura in association with colitis. J Clin Gastroenterol 2002;34(4):492-3.
7. Borrás-Blasco J, Enriquez R, Amoros F, Cabezuelo JB, Navarro-Ruiz A, Pérez M, et al. Henoch-Schönlein purpura associated with clarithromycin. Case report and review of literature. Int J Clin Pharmacol Ther 2003;41(5):213-6.
8. Al-Toma AA, Brink MA, Hagen EC. Henoch-Schönlein purpura presenting as terminal ileitis and complicated by thrombotic microangiopathy. Eur J Intern Med 2005;16(7):510-2.
9. Ortego-Centeno N, Callejas-Rubio JL, López-Mañas JG, Troncoso-García E, de la Higuera Torres-Puchol J. Ileitis terminalis in a patient with Henoch-Schönlein purpura. Dig Dis Sci. 1999;44(8):1590-3.
10. Akdamar K, Agrawal NM, Varela PY. The endoscopic appearance of anaphylactoid purpura. Gastrointest Endosc 1973;20(2):68-9.
11. 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):2354-6.
12. Conolly B, O'Halpin D. Sonographic evaluation of the abdomen in Henoch-Schonlein purpura. Clin Radiol 1994;49(5):320-3.
13. Shirahama M, Umeno Y, Tomimasu R, Dohmen K, Miyamoto Y, Shimoda Y, et al. The value of colour Doppler ultrasonography for small bowel involvement of adult Henoch-Schönlein purpura. Br J Radiol 1998;71(847):788-91.
14. Chang WL, Yang YH, Lin YT, Chiang BL. Gastrointestinal manifestations in Henoch-Schönlein purpura: a review of 261 patients. Acta Paediatr 2004;93(11):1427-31.