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Case Report

A case of severe multifocal enteritis caused by unusual pattern of Henoch–Schönlein purpura

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Abstract

A 58-year-old male visited our hospital with a complaint of epigastric pain for 2 days. Physical examination showed no purpuric skin lesion. Abdominal computed tomography scan showed concentric segmental wall thickening of the duodenum. Esophagogastroduodenoscopy showed diffuse erythematous mucosa with exudates at the duodenum. On 4th hospital day, he developed hematochezia. colonoscopy showed circumferential erythematous mucosa with ulcerative hemorrhage at terminal ileum. The microscopic examination identified lots of neutrophil infiltration and deposition of immunoglobulin A (IgA) on vascular endothelium. Treatment with oral prednisone 40 mg was initiated. On 19th day, the patient developed hematuria and proteinuria. Histologic examination on renal biopsy showed focal proliferative glomerulonephritis and strongly positive IgA staining in the glomerular mesangium. Severe hemorrhagic mucosa on endoscopy, deposition of IgA on kidney and intestinal vascular endothelium suggests the diagnosis of Henoch–Schönlein purpura (HSP). We report severe multifocal enteritis caused by unusual pattern of HSP.

Key words

Duodenitis, ileitis, purpura, Schoenlein–Henoch

Introduction

Henoch–Schönlein purpura (HSP) is a systemic vasculitis involving small sized vessels of multiple systems, especially skin, joint, gastrointestinal tract, and kidney. Incidence of HSP with age above 20 has been reported to be 0.1-1.2/million.[1] One of the classic tetrads of HSP is bowel involvement and 50-85% of HSP cases develop abdominal symptoms, such as poorly localized colicky abdominal pain, nausea, or vomiting, due to bowel or the mesenteric involvement.[2] One-third of HSP develop gastrointestinal hemorrhage caused by vasculitis induced mucosal ischemia.[3] The most common site of HSP involvement in the gastrointestinal tract is a small intestine,[4] followed by esophagus, stomach and colon. Skin purpura occurs in HSP in most cases, but other presentation of HSP may precede skin purpura, making diagnosis of HSP difficult. We report a case of severe multifocal enteritis caused by unusual pattern of HSP for clinicians to be well-informed of atypical pattern of HSP.

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Case Report

A 58-year-old male visited our hospital with a complaint of epigastric pain for a day. The patient had past medical history of hypertension and hypercholesterolemia and was on medication for them. On the review of the system, the patient had abdominal pain, nausea, and vomiting. The vital signs were as follows: Blood pressure, 150/100 mm Hg, pulse rate, 81 per minute, body temperature, 36.8°C and respiration rate, 20 per minute. Physical examination showed soft abdomen with mild tenderness in epigastric and peri-umbilical abdominal area and no purpuric skin lesion and tenderness on joints. Laboratory findings were as follows: White blood cells, 12,030/mm^3, haemoglobin, 16.8 mg/dL, platelet 368,000/mm^3, erythrocyte sedimentation rate, 7 mm/H, C-reactive protein, 0.925 mg/dL, gamma-glutamyl transpeptidase, 712, alkaline phosphatase, 425, prothrombin time, 11.8 s and

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partial thromboplastin time, 34.8 s. Abdominal computed tomography (CT) scan showed concentric segmental wall thickening of the duodenum. Esophagogastroduodenoscopy showed whole circumferential erythematous mucosa covered with scattered ulcerative exudates from bulb to a third portion of the duodenum [Figure 1] and histologic findings of biopsy specimen showed nonspecific chronic inflammation. Urine analysis was normal. On the 4th hospital day, the patient developed hematochezia. The colonoscopy demonstrated circumferential erythematous mucosa with ulcerative hemorrhage at terminal ileum [Figure 2]. The microscopic examination identified predominantly neutrophilic infiltration and deposition of immunoglobulin A (IgA) on vascular endothelium, compatible with vasculitis [Figure 3]. Cytomegalovirus (CMV) PCR and immunohistochemical stain of CMV on biopsy tissue showed negative results. A follow-up CT scan demonstrated a long segmental wall thickening of terminal ileum. Additional laboratory findings were as follows: IgG, 929 mg/dL, IgA, 293 mg/dL and IgM, 62 mg/dL. Antinuclear antibody, antineutrophil cytoplasmic antibody, CMV IgG, CMV IgM, paragonimus IgG and sphagnum IgG were all negative. Stool study was negative for bacterial, parasite or clostridium difficile infection. Treatment with oral prednisone 40 mg/day was initiated due to the persistent hematochezia and abdominal symptoms. On the 15th hospital day, a follow-up endoscopy showed marked healing of the mucosa at the duodenum with scattered several mucosal scars. A follow-up colonoscopy demonstrated a slight improvement of terminal ileum lesion combined with a circumferential hyperemic mucosa with intervening ulceration, but there were multiple, 2-8 mm sized, discrete, newly developed ulceration with surrounding hyperemia on whole colon [Figure 4]. On the 19th day, the patient developed hematuria and proteinuria on urinalysis. Total protein of 24 h urine was 3986 mg/24 h. Histological examination on renal biopsy showed a focal proliferative glomerulonephritis with crescent formation, with strongly positive IgA staining, mainly in the glomerular mesangium [Figure 5]. On the 29th day, a follow-up colonoscopy illustrated a dramatic improvement of a hemorrhagic mucosa of small intestine and ulcerations of the colon [Figure 6].

Discussion

Bowel involvement by HSP illustrates diffuse mucosal hyperemia, small coin like petechiae, ecchymotic lesions or hemorrhagic erosions.[2,5,6] Differential diagnosis of gastrointestinal involvement by HSP are Crohn's disease, eosinophilic gastroenteropathy, Zollinger Ellison syndrome, Yersinia enterocolitis, malignancy, Bechet`s syndrome, radiation or medication induced gastroenteropathy and ischemic enterocolitis. In our case, repetitive stool culture showed negative results and history taking, laboratory and radiologic examination ruled out disease that need differential diagnosis. Lanzkowsky *et al*. reported that symptoms

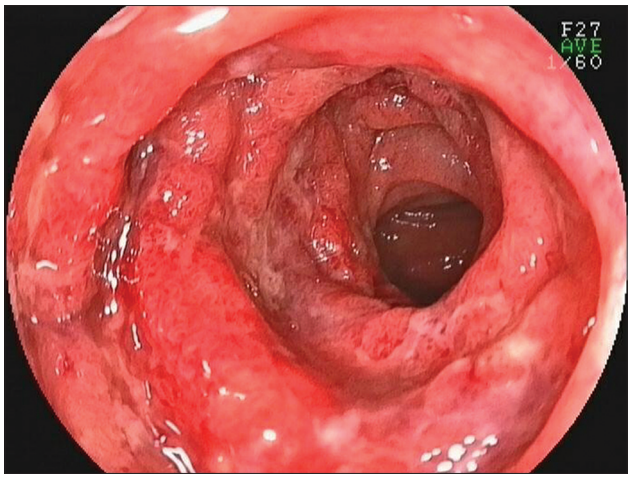

Figure 1: Esophagogastroduodenoscopy finding: Diffuse segmental erythematous mucosa covered with scattered overlying ulcerative exudates from bulb to the third portion of duodenum

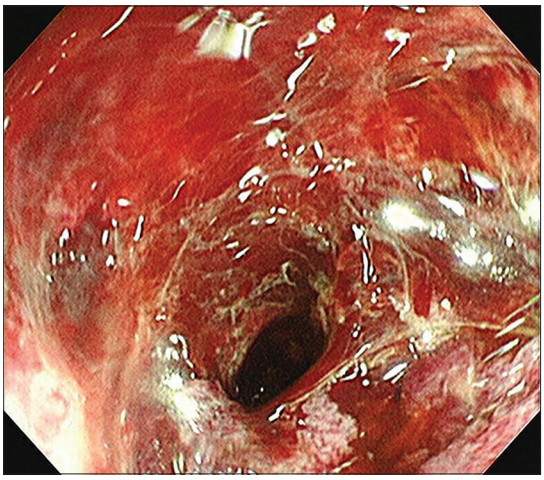

Figure 2: Colonoscopy finding: Diffuse erythematous and friable mucosa covered with scattered ulcerative hemorrhage

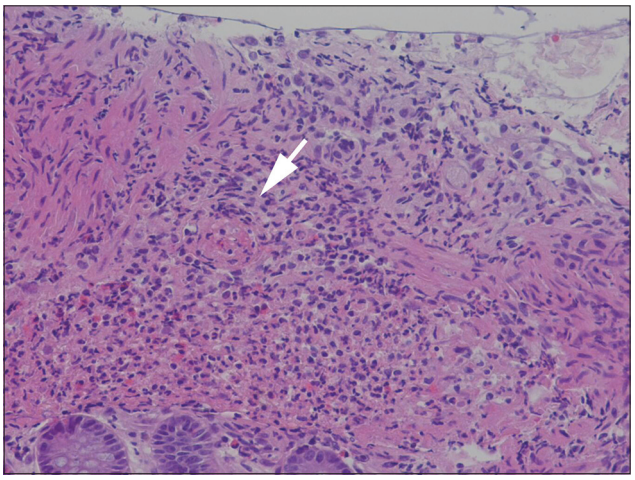

Figure 3: Histologic findings of endoscopic biopsy: Neutrophilic infiltration of vascular endothelium (white arrow head), suggesting vasculitis of small vessels (H and E, ×200)

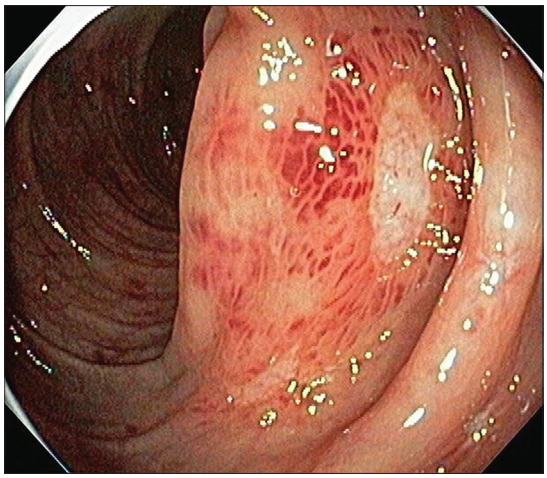

Figure 4: Follow-up colonoscopic finding: Newly developed multiple 2-10 mm sized discrete ulcer with hyperemic halo on whole colon

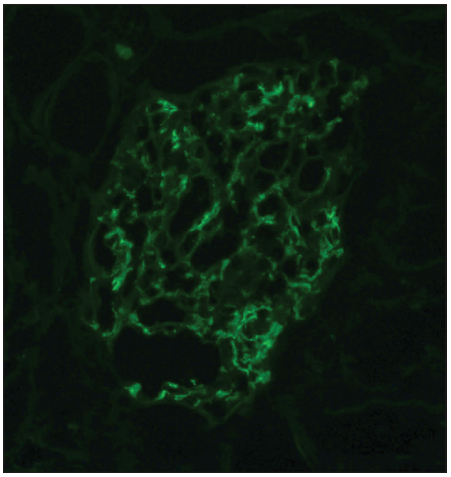

Figure 5: Immunofluorescence study of renal biopsy: Strong immunoglobulin A positivity in the glomerular mesangium (immunofluorescence, ×200)

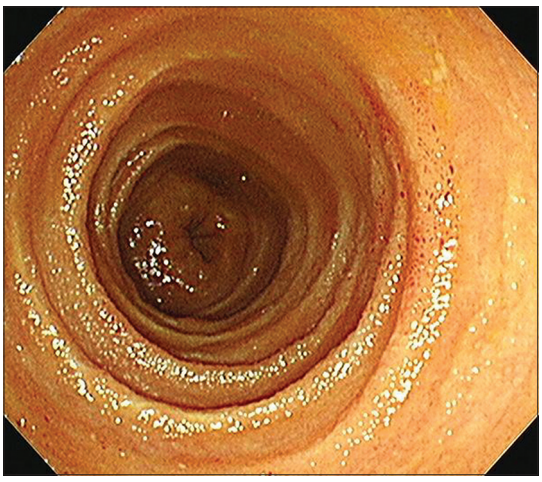

Figure 6: Follow-up colonoscopy finding after oral steroid treatment: Marked mucosal improvement with focal scars at terminal ileum

associated with bowel involvement of HSP may precede presence of purpuric skin lesion in 14% of HSP[7] and Saulbury reported that gastrointestinal symptoms and sign could precede onset of purpuric skin lesions up to 2 weeks.[8] In our case, purpuric skin lesion was not observed during admission and follow-up period after discharge. Gunasekaran *et al*. reported three cases of HSP without purpura and skin purpura was not seen during the whole period of follow-up and segmental involvement of HSP at different organs suggested diagnosis HSP without purpura.[9] HSP can be pathologically confirmed by biopsy of involved organ showing leukocytoclastic vasculitis with IgA deposition in blood vessel wall. In our case, pathologic examination of endoscopic biopsy specimen from terminal ileum revealed neutrophilic infiltration in vessel wall of intestine with IgA deposition, suggesting involvement by HSP. In our case, although purpuric skin lesion was not seen, patchy involvement of small vessels of intestine and kidney with IgA deposition suggests systemic involvement of HSP and suggests diagnosis of HSP. In our case, serial follow-up of abdominal CT scan and endoscopy was done which showed interesting findings, such as migration of intestinal involvement site from duodenum to terminal ileum and colon. Although there was an improvement of terminal ileum lesion, the newly developed colonic ulcerations were observed during treatment with oral prednisone. Reported cases of endoscopic finding of colon with HSP involvement showed numerous ecchymosis.[10] In our case, endoscopic finding of colon involvement showed discrete various sized ulcers with surrounding hyperemia and this has not been reported previously.

Severe hemorrhagic mucosa at duodenum and terminal ileum on endoscopy, deposition of IgA on vascular endothelium of intestine and kidney shows systemic involvement of HSP, and although characteristic skin purpura was not seen, these findings suggest the diagnosis of HSP.

We report a case of severe duodenoileitis caused by unusual pattern of HSP for clinicians to be well informed of atypical pattern of HSP.

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