Clinical Case Report

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Ulcerative colitis in an adult patient mimicking Henoch–Schönlein purpura

A case report

Bin Lu, MD*, Li-Li Niu, MD, Xi-Guang Xu, MD, Shu-Lan Yao, MD, Xing-You Tan, BS

Abstract

Rationale: Ulcerative colitis (UC) is one of the chronic inflammatory diseases of the intestinal tract. UC being misdiagnosed as Henoch–Schönlein purpura (HSP) in the elderly has seldom been reported about.

Patient concerns: A 64-year-old man was admitted to the hospital with petechiae and palpable purpura in lower limbs and abdominal pain for about 1 month.

Diagnoses: Colonoscopy demonstrated severe inflammation in the colon, mucosal congestion, and edema, and multiple hemorrhages and ulcerations, with purulent adhesions. A histopathologic examination of the colon biopsies revealed extensive infiltration of immune cells and mucosal ulcerations in the intestine. UC was diagnosed.

Interventions: The patient was treated with prednisone (1.0 mg/kg/d) with progressive dose reduction.

Outcomes: The skin lesions were healed within 4 weeks, and his abdominal pain was alleviated remarkably. He is currently under follow-up.

Lessons: As the treatment used for patients with HSP was not effective, it was advised that UC should be taken into consideration.

Abbreviations: HSP = Henoch–Schönlein purpura, UC = ulcerative colitis.

Keywords: Henoch–Schönlein purpura, inflammatory bowel disease, ulcerative colitis

1. Introduction

Ulcerative colitis (UC) is a chronic autoimmune disorder of the colonic mucosa, which generally affects the rectum and may extend proximally to other parts of the colon.[1,2] It is characterized by relapsing and remitting mucosal inflammation, starting in the rectum and extending to the proximal segments of the colon.[1] UC most commonly presents with blood in the stool and diarrhea. Almost 15% of patients can initially present with severe disease.[3] The symptoms can include urgency, incontinence, fatigue, increased frequency of bowel movements, mucus discharge, nocturnal defecations, and abdominal discomfort (cramps).[4] Extraintestinal manifestations can occur in about a third of patients with UC, and about a quarter might have extraintestinal manifestations before inflammatory bowel disease diagnosis.[5,6] Cutaneous manifestations are one of the uncommon extraintestinal symptoms.

Henoch–Schönlein purpura (HSP) is the most common type of vasculitis. It is characterized by cutaneous purpura, arthritis, abdominal pain, gastrointestinal bleeding, and nephritis.[7] HSP is more prevalent in children. Gastrointestinal tract involvement is the third most common manifestations of HSP.[8,9] It was reported that 70% of patients with HSP have gastrointestinal tract symptoms,[10] and approximately two-thirds of patients with HSP develop abdominal pain, usually diffuse, increasing after meals, and sometimes associated with nausea and vomiting. These symptoms are caused by submucosal hemorrhage and edema of the bowel wall, predominantly affecting the proximal small bowel. The most severe gastrointestinal complication is intussusception, affecting 3% to 4% of patients with HSP. In 60% of these cases, it is limited to the small bowel. Clinical presentation of intussusception is characterized by severe abdominal pain, often colicky in nature, and vomiting. Abdominal symptoms occur before the purpura in nearly 20% of those afflicted by HSP, leading to unnecessary morbidity through work-up if not recognized as the transient symptomatology of HSP.[8,9]

As HSP and UC share some symptoms, UC mimicking HSP[11] and HSP mimicking inflammatory bowel disease[12] have been reported, especially in children. In contrast, seldom such cases have been reported in the adult population. Here, we present an unusual case of UC in a 64-year-old man that was misdiagnosed as HSP.

Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal. This case report involves retrospective observations of a patient. Thus, ethical approval was not needed.

2. Case presentation

A 64-year-old man was admitted to our hospital with petechiae and palpable purpura in lower limbs and abdominal pain for about 1 month. He had a previous history of gastritis more than 1

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Department of Dermatology, Affiliated Hospital of Jining Medical University, Guhuai Road, Jining, Shandong Province, China.

* Correspondence: Bin Lu, Department of Dermatology, Affiliated Hospital of Jining Medical University, Guhuai Road, Jining, Shandong Province, China. (e-mail: lubinn@hotmail.com).

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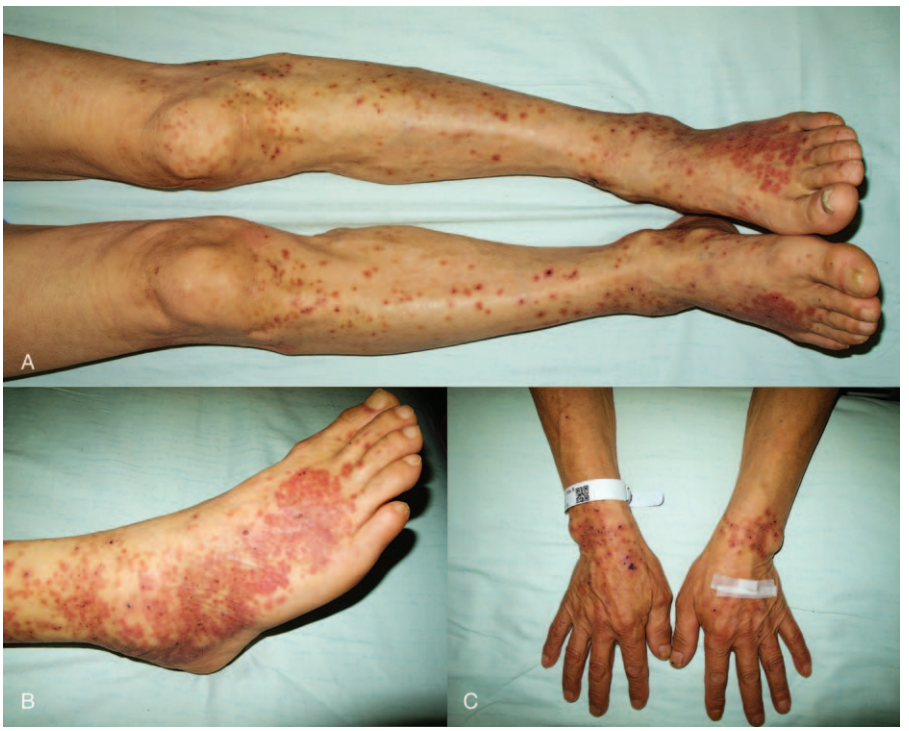

Figure 1. Numerous petechiae and palpable purpura ranging in size from pinpoint to several millimeters were symmetrically distributed on the lower extremities and the dorsum of the hand.

year ago. About 1 month before hospitalization, he had abdominal pain without hematochezia. Three days later, he complained of petechiae in his lower limbs. Cutaneous lesions began as erythematous macules or urticarial papules that evolved into palpable purpura. He previously was diagnosed with HSP and treated accordingly more than once. However, the treatment was not effective, and he was admitted for further treatment. He denied a history of bleeding or coagulation disorders, high blood pressure, or diabetes. The family history revealed nothing about both his parents and children. The allergic history was negative for any specific medication or food. He was married and had a son at the age of 25. He had no fever. He was still presenting with abdominal pain. Bowel frequency was about 5 times in 1 day.

It was revealed that both knees joints had pain and tenderness. Complete abdominal tenderness and rebounding pain were apparent, accompanied by bowel cries phonic hyperfunction. Numerous petechiae and palpable purpura ranging in size from pinpoint to several millimeters were symmetrically distributed on the lower extremities and the dorsum of the hand (Fig. 1).

Laboratory tests performed at admission revealed the erythrocyte sedimentation rate (ESR) was 23 mm/h; C-reactive protein (CRP) was 88.39 mg/L. All autoimmune antibodies, including antinuclear antibody, antismooth muscle antibody, anti-SSA antibody, anti-SSB antibody, perinuclear antineutrophil cytoplasmic antibody, anti-Scl-70 antibody, anti-Jo-1 antibody, anti-PM-Scl antibody, anti-double-stranded DNA antibody, antinucleosome antibody, antihistone antibody, and antimitochondrial M2 antibody, were negative. Renal and liver function tests were unremarkable. Stool microbial cultures were also negative. Bacterial toxins from *C difficile*, *E coli* O157, *Salmonella*, and *S aureus*, as well as lipopolysaccharide and peptidoglycan in serum, were undetectable. He was diagnosed with HSP and treated according to the European League Against Rheumatism/Pediatric Rheumatology International Trials Organization (EULAR/PRINTO) classification criteria,[13] but the effect was not as well as expected, as abdomen tenderness and rebounding pain were not relieved. Thus, he was advised to undergo a colonoscopy.

Colonoscopy demonstrated severe inflammation in the colon, mucosal congestion, and edema, and multiple hemorrhages and ulcerations, with purulent adhesions (Fig. 2). A histopathological examination of colon biopsies revealed extensive infiltration of

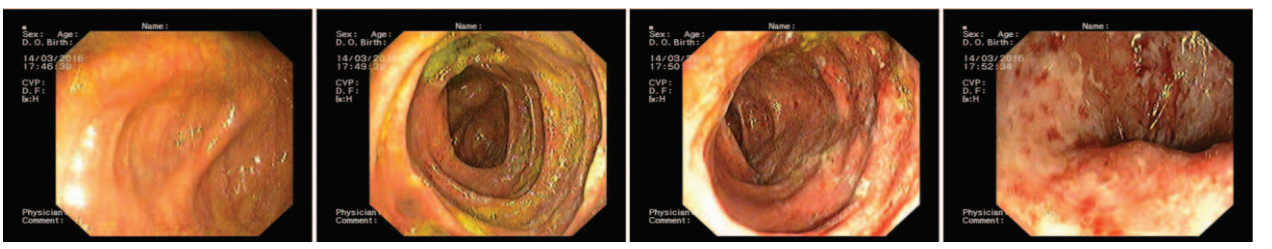

Figure 2. Colonoscopy demonstrated severe mucosal congestion and edema, multiple hemorrhages and ulcerations with purulent adhesions in the colon.

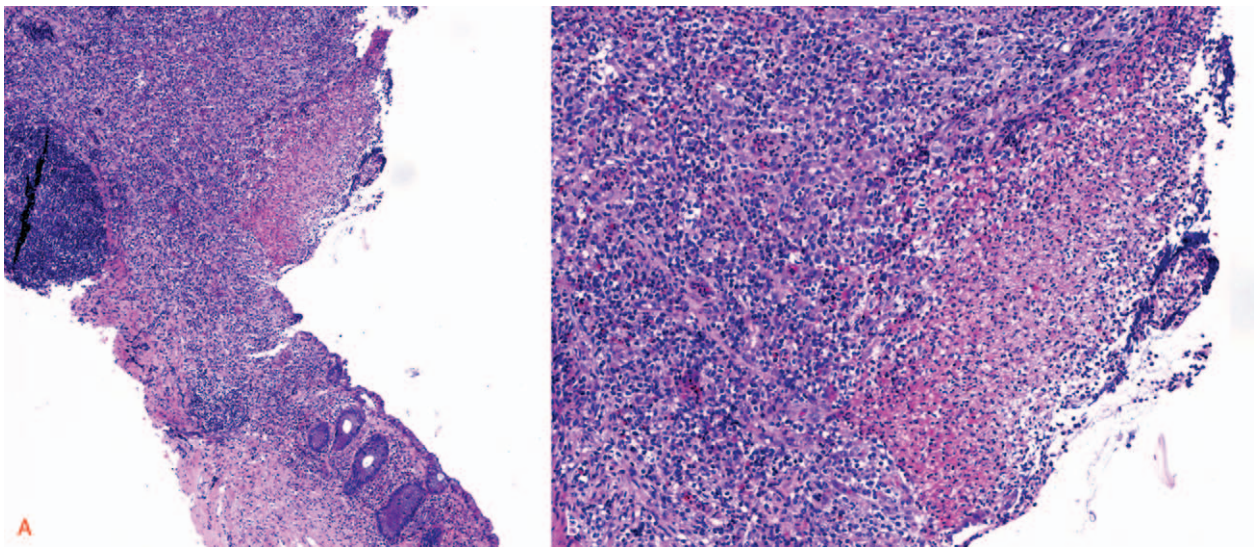

Figure 3. A histological section revealed extensive infiltration of immune cells and mucosal ulcerations (A, ×40). Magnification showing the infiltration of immune cells and mucosal ulcerations (B, ×100).

immune cells and mucosal ulcerations in the intestine (Fig. 3). Thus, a diagnosis of UC was confirmed. He was treated with prednisone (1.0 mg/kg/d) with progressive dose reduction. The skin lesions healed within 4 weeks, and his abdomen pain was alleviated remarkably. He is currently under follow-up.

3. Discussion

Inflammatory bowel disease is a heterogeneous group of chronic inflammatory diseases of the intestinal tract. Crohn disease and UC are the 2 major phenotypic forms and both may present with extraintestinal manifestations.[2] UC is a chronic, idiopathic inflammatory disease that affects the colon, most commonly afflicting adults aged 30 to 40 years and resulting in disability.[1,2,14]

The diagnosis of UC relies on the identification of the typical phenotype of chronic inflammation of the colon upon colonoscopy and colonic biopsies, and the exclusion of both Crohn disease and infectious causes of colitis.[15] On the contrary, there is no single set of macroscopic or microscopic criteria that can accurately diagnose UC, and there are multiple atypical phenotypes that do not fit into this category.[15] Most patients are diagnosed with UC based on the clinical signs and symptoms, endoscopy, histology, and radiology.[16] The patient in our case was diagnosed with UC according to the symptoms, endoscopy, and histology.

HSP is a small-vessel vasculitis clinically characterized by arthralgia, abdominal pain, and renal involvement and presenting with palpable purpura on the extensor aspects of the limbs, thighs, and buttocks.[7] The etiology of HSP remains clouded with genetic, environmental, microbial, drug-related, and antigenic influences, all of which contribute to a multifactorial origin.[2,7] IgA plays a pivotal role in the pathogenesis of HSP.[7] There are 2 subclasses of IgA, IgA1, and IgA2, but only IgA1 is involved in the pathogenesis of HSP.[7] Infection as well as allergy may be associated with HSP, as confirmed by recent reports of its occurrence with pandemic influenza,[17] streptococcal pathogen,[7] and hepatitis B virus.[18,19] Environmental factors have also been implicated, and the so-called "hygiene hypothesis" as an etiological factor in inflammatory bowel disease is under active investigation.[20]

As there are no disease-specific laboratory abnormalities, HSP is currently diagnosed based on symptoms, signs, and histopathological findings. To date, several sets of diagnostic criteria for HSP have been proposed. EULAR and PRINTO have proposed a new criteria for HSP diagnosis in 2010.[13] According to these, a patient is classified with HSP in the presence of purpura or petechiae, particularly in the lower limb (mandatory criterion) and 1 out of 4 of the following findings: abdominal pain, arthritis or arthralgia, renal involvement (hematuria or proteinuria), and histopathology (leukocytoclastic vasculitis with predominant IgA deposits)[13] However, these criteria are limited to children, and do not apply to adults. Therefore, the first diagnosis of HSP in the patient in this case according to these criteria was incorrect.

The gastrointestinal and the cutaneous organ systems are closely linked. Cutaneous lesions associated with inflammatory bowel disease include erythema nodosum, pyoderma gangrenosum, vitiligo, and rarely, vasculitis.[2,16] The prevalence of cutaneous manifestations of UC is reported to be about 9% to 23%.[2,21,22] This variation in the rate can be attributed to the variation in the classification of the signs and symptoms comprising a cutaneous manifestation of UC.[2] HSP mimicking inflammatory bowel disease has been described, especially in children.[12]

Despite our patient's symptoms matching both the HSP and UC criteria, indicating a possible coexistence or overlap of the diseases, the analysis of the histopathology of intestinal biopsies confirmed the singular UC diagnosis and excluded intestinal vasculitis. However, in a genetically predisposed patient, an infective/allergic trigger activating the immune system with simultaneous occurrence of UC and HSP is a possibility.[23] Both diseases may, therefore, share the same environmental risk factors. We do not know whether one disease precedes the other, and the reasons for the simultaneous occurrence of the 2 diseases with different microscopic and macroscopic lesions remain unknown.

4. Conclusions

In summary, we reported a case of UC patient misdiagnosed as HSP. UC and HSP may share the same or similar clinical features.

UC patients presenting with cutaneous vasculitis (palpable purpura) may be misdiagnosed as HSP. As the treatment of our patients with HSP was not effective, colonoscopy and histopathologic examination of colon biopsies were advised. This confirms that UC should be taken into consideration in such cases.

Author contributions

Conceptualization: Bin Lu.
Data curation: Xi-Guang Xu, Shu-Lan Yao.
Investigation: Xi-Guang Xu, Shu-Lan Yao, Xing-You Tan.
Writing – original draft: Li-Li Niu.
Writing – review & editing: Bin Lu.
Bin Lu orcid: 0000-0002-9431-7169

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