CASE Reports

Colonoscopic and Capsule Endoscopic Findings in a Child with Henoch-Schönlein Purpura Manifesting as Severe Gastrointestinal Bleeding without Skin Involvement

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ABSTRACT

We reported the gastrointestinal (GI) endoscopic findings in a 12-year-old boy with Henoch-Schönlein purpura (HSP). The patient presented with massive rectal bleeding and abdominal pain, requiring multiple blood transfusions. The colonoscopic examination revealed colonic lymphonodular hyperplasia with mild mucosal erythema. The biopsies showed erosive colitis with vascular fibrinoid necrosis. Capsule endoscopy, performed 10 days after cessation of the bleeding, demonstrated petechial-like lesions on the gastric mucosa and mucosal inflammation in the terminal ileum. Although, there was no skin involvement, the patient was diagnosed as HSP on the basis of GI bleeding and the age less than 20 years old, using the criteria of the American College of Rheumatology 1990. The diagnosis was further supported by the evidence of vascular fibrinoid necrosis in the colonic biopsy, intermittent clinical history, clinical response to steroid therapy during the first admission, and extensive exclusion of possible significant patholgy, using panendosocpy, capsule endoscopy, upper GI study, angiography, and even surgery. He has been closely monitored for the recurrent symptoms, in which steroid therapy is planned.

Key words : Gastrointestinal bleeding, vasculitis, pathology, endoscopy, Henoch-Schönlein purpura, diagnosis

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INTRODUCTION

Henoch-Schönlein purpura (HSP) is one of common vasculitic disease in childhood. Aside from skin rash, gastrointestinal symptoms, including abdominal pain, vomiting, and gastrointestinal hemorrhage are the second most common of the clinical manifestation[1,2]. Sometimes, it is very difficult to make the diagnosis if there is no rash at the time of presentation; thereby rendering unnecessary investigations and treatments. There have many studies reporting on the utility of gastrointestinal endoscopy with biopsy in this condition[3-7]. Herein, we reported colonoscopic and capsule endosocpic findings on a 12-year-old boy with HSP manifesting as massive lower gastrointestinal hemorrhage without skin involvement.

CASE REPORT

A 12-year-old boy had presented with intermittent massive bleeding per rectum for five months. He was hospitalized twice before referral to our institute.

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The patient was symptomatically treated with multiple blood transfusions, in which the lowest hemoglobin level was 4.3 g/dl. A barium enema was done with a normal result. During each episode of gastrointestinal (GI) bleeding, he also experienced mild cramping periumbilical pain. The symptoms were not associated with vomiting or hematemesis. He denied a history of recent medication, immunization, and insect bites.

At the first admission to our hospital, the patient had active bleeding. On physical examination, only a moderate pallor was noted. The abdomen was soft without tenderness. Neither signs of arthritis nor skin rash was observed. The complete blood count showed Hb 9.4 g/dl, WBC count 8,300/mm^3 (N 64%, L 33%, Mono 3%), and platelet count 235,000/mm^3. Coagulograms were normal. The urinalysis revealed mild proteinuria (+1) with red blood cell of 1-2/HPF. The BUN was 12 mg/dl, creatinine was 0.6 mg/dl, and sedimentation rate was 9 mm/hr. Because the radionuclear scan was not available during that period, a colonosocpy was initially performed, revealing only patchy erythema and lymphonodular hyperplasia in the cecum and ascending colon. (Figure 1A and 1B) The terminal ileum appeared normal with lymphonodular hyperplasia. Neither ulcer, mass nor vascular lesion was noted during the colonoscopy. The histopathology confirmed lymphonodular hyperplasia and revealed inflammatory cells infiltration in the lamina propria with evidence of erosive colitis. The study also presented vascular fibrinoid necrosis in the colonic biopsies. (Figure 1C and 1D) With a diagnosis of colonic lymphonodular hyperplasia, the patient was initially treated with 0.7 mg/kg/day of prednisolone for two weeks, although the endoscopic findings could not explain his clinical presentation. The bleeding gradually stopped. He was discharged home and scheduled for four-weeks follow up. After discontinuing medication, the bleeding recurred, leading to re-hospitalization and blood transfusions. At the second admission, the esophagogastroduodenoscopy and repeated

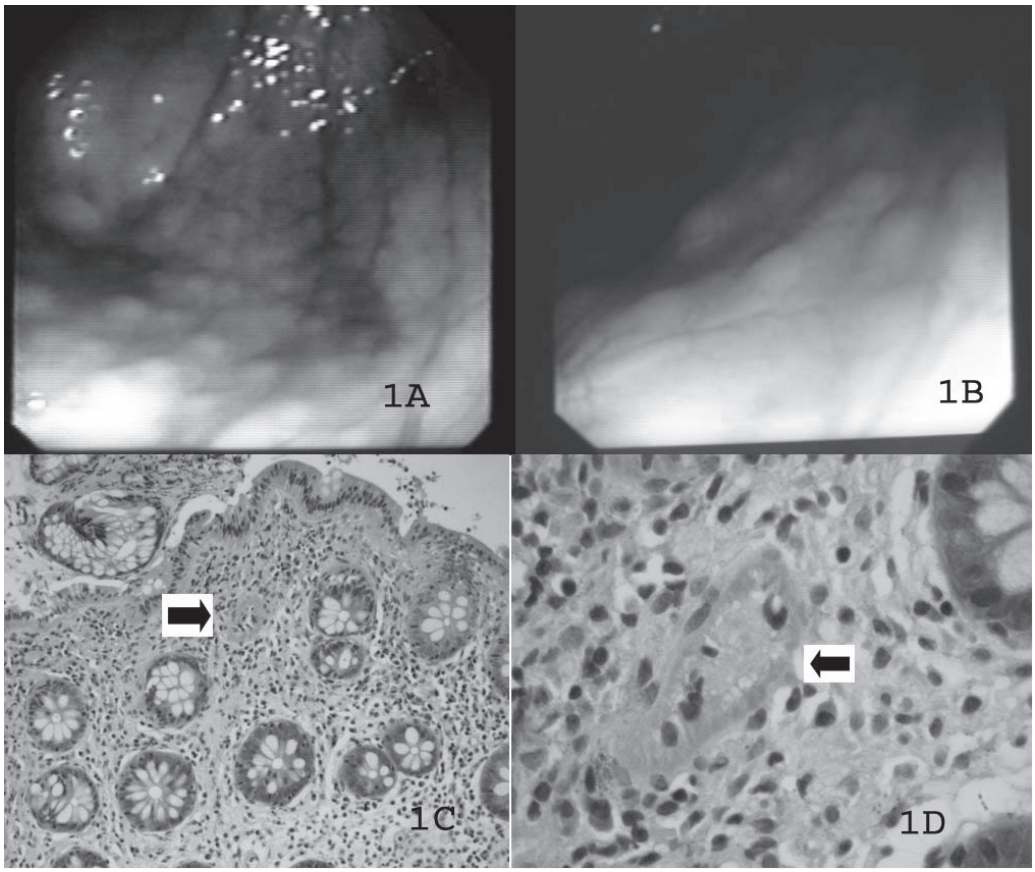

Figure 1 The colonoscopic picture shows lymphonodular hyperpalasia with mild mucosal erythema in the cecum (1A and 1B). There is evidence of acute inflammatory cell infiltration in the lamina propria of the colonic biopsy (1C). Two areas of vascular fibrinoid necrosis (arrows) are also observed (1C and 1D).

colonoscopy were performed. The upper gastrointestinal tract was normal, whereas the colonic lymphonodular hyperplasia markedly improved compared to the previous study. Duodenal biopsies appeared normal. The patient ultimately underwent exploratory laparotomy. Unfortunately, the operative finding was normal. Neither Meckel's diverticulum, mass, polyp, nor vascular abnormality was noted in the small bowel.

After the surgery, the bleeding continued. The angiography was carried out, in which there was no arteriovenous malformation, abnormal vessels, or tumor identified. During the angiogram, the bleeding site could not be demonstrated. An upper GI study was also performed. Only thickened folds of the jejunal loop was seen and suggestive of mucosal inflammation. Because the investigations performed earlier could not disclose the underlying pathology and the bleeding was extremely severe, therefore a wireless capsule endoscopy (M2A: Yoqneam, Israel) was used to discover any possible pathologic lesion in the small bowel. Additional laboratories were obtained, including serum IgE 92.6 IU/L, IgA 147.1 mg/dl, albumin 4.4 g/dl, globulin 2.6 g/dl, alkaline phosphatase 104 IU/L, cholesterol 125 mg/dl, and AST/ALT 15/4 IU/L, in which all were considered normal for age. At the time of capsule endosocopic study, the patient had not bled for 14 days. Small petechial-like lesions were noted on the gastric mucosa and pyloric opening. (Figure 2A and 2B) The lymphonodular hyperplasia was noted in the terminal ileum, which could be normal for his age; however, there was also evidence of patchy erythema and mild mucosal inflammation seen in this area. (Figure 2C and 2D) Neither hemangioma, telangiectasia, angiodysplasia, ulcer, nor mass was noted in the small bowel. Although we have tried very hard to recognize the skin rash, it has not occurred in this patient. Due to the intermittent clinical history of gastrointestinal bleeding accompanying with abdominal pain, age (<20 years old), gender, and extensive exclu-

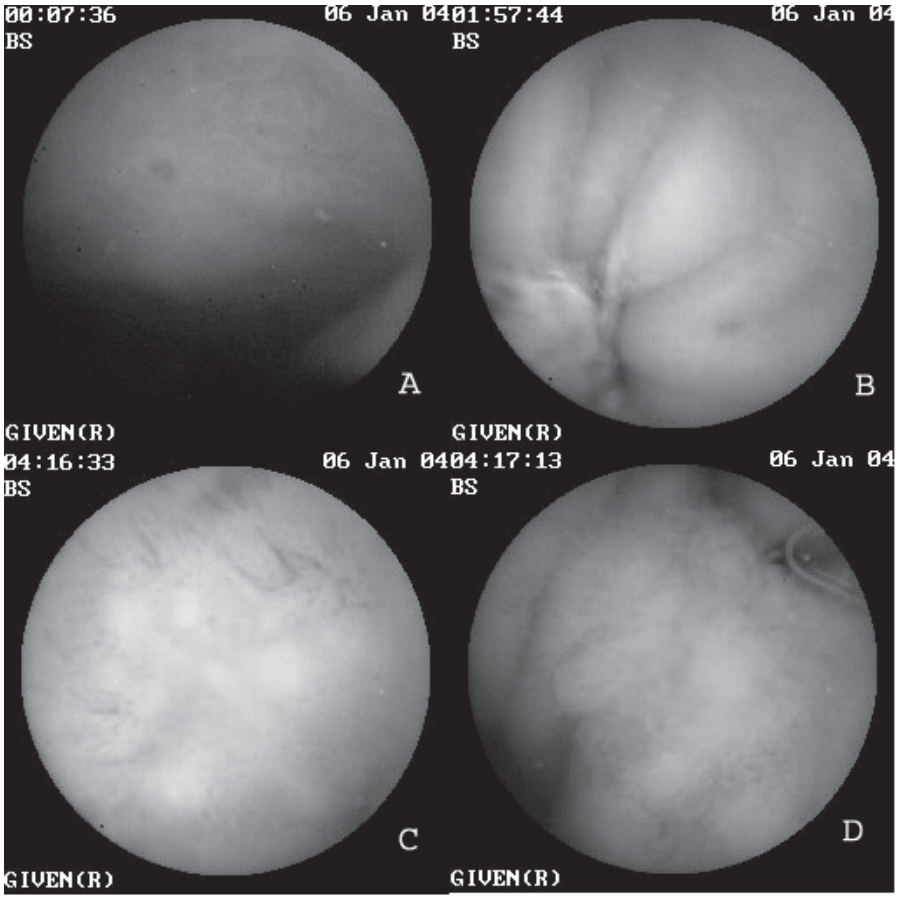

Figure 2 Pictures of capsule endosocpy reveal petechial-like lesion on the gastric mucosa and pyloric opening (2A and 2B). The remaining small bowel looks normal. There is lymphonodular hyperplasia in the terminal ileum with mild patchy erythema (2C and 2D).

sion of significant pathology, Henoch-Schönlein purpura was the most likely diagnosis. The patient was later on discharged home and closely monitored for the recurrent symptoms, in which steroid therapy was planned.

DISCUSSION

Henoch-Schönlein purpura is a clinical syndrome characterized by acute febrile illness, nonthrombopenic purpura, arthritis, nephritis, and gastrointestinal symptoms; in which the most two common clinical presentations comprises skin rash and GI symptoms, including abdominal pain, vomiting, hematemesis, and hematochezia[1,2]. It is not uncommon that a child with HSP can develop massive rectal bleeding[8]. However, the purpura might not be present in all patients[1,9, 10], as a result it is very difficult to establish a definite diagnosis. Like our patient, because he had severe gastrointestinal hemorrhage, we had to extensively investigate on him, including unnecessary operation. Up to date, there has been no single laboratory specific for the diagnosis of HSP, although elevation of serum immunoglobulin A and reduction of factor XIII level have been found in HSP[1,9-11]. Nonetheless, this association is not consistent. Serum IgA in our patient was within the normal range for age and the factor XIII level is not available in our hospital. The underlying pathology of this condition is allergic vasculitis of small arterioles and venules, in which the typical histopathology revealed fibrinoid necrosis of a small vessel wall as well as polymorphonuclear leukocyte infiltration and extravasation of red blood cells in the skin or gastrointestinal biopsies[1,4,7]. After retrospectively reviewing the colonic biopsies in our case, we demonstrated vascular fibrinoid necrosis which supported the diagnosis of HSP. However, the typical leukocytoclastic vasculitis was not seen, in which this might result from too superficial biopsy obtained from the colonic mucosa. According to the American College of Rheumatology 1990 criteria for the classification of Henoch-Schönlein purpura, our patient met at least two of four criteria (gastrointestinal bleeding and age less than 20 at disease onset, but no skin rash and leukocytoclastic vasculitis in the biopsy). Therefore the diagnosis of HSP was made with a sensitivity and specificity of 89.4% and 88.1%, respectively[1]. The diagnosis was strengthened by the evidence of vascular fibrinoid necrosis in the colonic biopsy, intermittent clinical history, clinical response to steroid therapy during the first admission, and extensive exclusion of possible significant pathology, using panendosocpy, wireless capsule endoscopy, upper GI study, angiography, and even surgery.

During the past two decades, there have been many reports on endoscopic findings in HSP[3-7]. The gastrointestinal involvement can be noted throughout the GI tract, in which the stomach, duodenum, and colon are the commonly affected sites. The gross endoscopic findings include petechial-like lesion, erythema, edema, erosion, and ulceration. As not previously reported, we additionally described colonic lymphonodular hyperplasia accompanying with mild mucosal erythema as endoscopic findings in a child with HSP. Apart from lymphoid aggregation, the biopsies also demonstrated erosive colitis and vascular fibrinoid necrosis, suggesting HSP. Although the underlying pathogenesis of gastrointestinal lymphonodular hyperplasia has still unknown, an up-regulated immune response to certain food antigens is speculated[12], whereas this is also one of suspected contributing factors in HSP[3]. In some cases, the colonic lymphonodular hyperplasia was related to inflammatory bowel diseases[13]. However, ulcerative colitis and Crohn's disease were excluded in our patient, based on absence of typical endoscopic and histopathology findings and normal sedimentation rate.

With the advent of a new technology, wireless capsule endoscopy, assessment of the small bowel has been increasingly investigated. Unfortunately, we did capsule endoscopy after the bleeding had stopped for 14 days and the clinical course of HSP could spontaneously remit and recur; therefore only readily neglected lesions were noted, including petechial-like lesions in the stomach and erythema in the terminal ileum. However, this study was still helpful to firmly exclude significant small bowel pathology in our patient.

Regarding treatments, immunosuppressive agents should be considered exclusively in the patient with severe gastrointestinal symptoms, in which it could result in serious complications, such as intussusception, massive gastrointestinal hemorrhage and bowel perforation. Among those, prednisolone has been the first-line therapy; whereas, in refractory cases, NSAIDs, IVIG, and factor XIII have been tried[14].

In conclusion, Henoch-Schönlein purpura should be considered in a child presenting with massive rectal

bleeding. Careful interpretation of gastrointestinal endoscopy and pathology are crucial to provide the proper diagnosis; thereby avoiding unnecessary operation. Although skin involvement is frequently noted simultaneously or subsequently during the clinical course, it is not uncommon that the patient could have a diagnosis of HSP without skin rash.

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