The Diagnostic Usefulness of Video Capsule Endoscopy in Adolescent Immunoglobulin A Vasculitis (Henoch-Schönlein Purpura)

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Abstract

Immunoglobulin (Ig) A vasculitis (IgAV), previously known as Henoch-Schönlein purpura, is a systemic IgA-mediated leukocytoclastic vasculitis that usually affects children. We report the usefulness of video capsule endoscopy in 2 adolescent patients with IgAV having gastrointestinal involvement. Both patient 1, a 15-year-old girl, and patient 2, a 14-year-old boy, presented with purpuric rash and abdominal pain. Video capsule endoscopy showed multiple areas of purpuric erythema throughout the small bowel in both patients and showed multiple ulcers with bleeding in patient 2. Patient 1 responded well to oral prednisolone at a dose of 0.5 mg/kg/day. However, in patient 2, prednisolone at a dose of 0.5 mg/kg/day failed to control the symptoms; therefore, the dose was increased to 1 mg/kg/day to provide relief. Video capsule endoscopy was safe in both cases and produced no side effects. In conclusion, video capsule endoscopy is a useful tool for evaluating small bowel lesions in patients with IgAV and provides valuable information for the treatment of IgAV with gastrointestinal involvement. (J Nippon Med Sch 2014; 81: 114–117)

Key words: IgA vasculitis, Henoch-Schönlein purpura, video capsule endoscopy, gastrointestinal involvement

Introduction

Immunoglobulin (Ig) A vasculitis (IgAV)¹, previously known as Henoch-Schönlein purpura, is a systemic IgA-mediated leukocytoclastic vasculitis that usually affects children². Clinical manifestations include palpable purpura unrelated to any underlying coagulopathy, abdominal pain, arthritis, and renal involvement². Abdominal pain occurs in 60% to 75% of patients, and gastrointestinal (GI)

bleeding, manifesting as melena and hematemesis, has been reported in up to 31% to 33% of children with $\rm IgAV^{34}$.

Any segment of the GI tract can be involved, but small bowel involvement is most common⁵. The second part of the duodenum is characteristically involved rather than the bulb⁵. Other GI manifestations can include bowel infarction and perforation and, in children, acute intussusception⁶. Occasionally, massive hemorrhage and shock occur and necessitate surgical intervention⁷.

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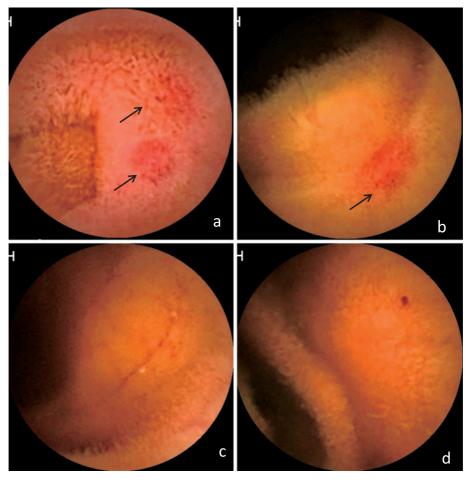


Fig. 1 A capsule endoscopic image from before treatment in case 1 showing areas of purpuric erythema throughout the jejunum (a) and in the ileum (b). The disappearance of the previously identified lesions in the jejunum (c) and the ileum (d) after treatment with prednisone at a dose of 0.5 mg/kg/day for 4 weeks.

Conventional endoscopy techniques for evaluating the small bowel are limited by its extensive length, and enteroscopy is technically complex and poorly tolerated⁸. Video capsule endoscopy is an accurate, noninvasive diagnostic technique for visualizing the small bowel and is safe for children older than 10 years⁹.

In the present study, we performed video capsule endoscopy to evaluate small bowel lesions in 2 adolescent patients with IgAV.

Case 1

A 15-year-old girl presented with a 2-month history of melena and recurrent purpura on the legs and buttocks. The initial symptom had been abdominal pain, which had improved by the time

she was referred to our hospital. A skin biopsy revealed leukocytoclastic vasculitis with IgA deposits in the walls of small vessels, consistent with a diagnosis of IgAV. Laboratory tests revealed a normal white blood cell count, hemoglobin level, platelet count, and serum IgA level and a coagulation factor XIII activity of 54% (normal, >70%). Urinalysis showed mild microscopic hematuria. Upper GI endoscopy showed erythema and erosions in the second part of the duodenum, whereas the findings of colonoscopy were negative.

Video capsule endoscopy was performed and showed multiple areas of purpuric erythema throughout the jejunum and ileum (**Fig. 1a and b**). After the patient had been treated with oral prednisone (0.5 mg/kg/day) for 2 weeks, the rash and melena resolved. A second examination with

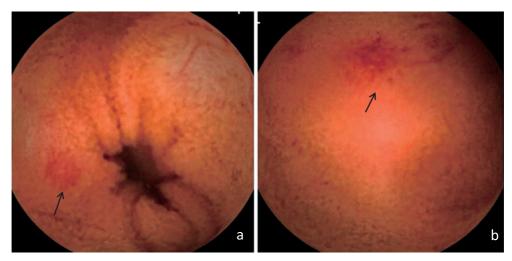


Fig. 2 A capsule endoscopic image from before treatment in case 2 showing areas of purpuric erythema (a) and ulcers with bleeding (b) in the jejunum.

video capsule endoscopy, performed 4 weeks after the start of treatment, showed that the previously identified lesions had completely resolved (**Fig. 1c and d**). The steroid does was tapered, and the total duration of steroid therapy was 9 months.

Case 2

A 14-year-old boy was admitted to our hospital with a 1-week history of colicky pain in the periumbilical area and palpable purpura on the lower extremities. The abdominal pain occurred in waves. A skin biopsy revealed leukocytoclastic vasculitis with IgA and C3 deposits in the walls of small vessels, which are findings consistent with IgAV. Laboratory tests revealed a normal white blood cell count, hemoglobin level, and platelet count; slightly elevated levels of serum C-reactive protein (0.99 mg/dL) and IgA (547 mg/dL); and decreased coagulation factor XIII activity (64%). Urinalysis showed microscopic hematuria (3+) and proteinuria (1+).

Upper GI endoscopy showed erythema, erosions, ulcerations, and hemorrhagic lesions in the second part of the duodenum, whereas the findings of colonoscopy were negative. Video capsule endoscopy revealed multiple areas of purpuric erythema and ulcers with bleeding in the jejunum (Fig. 2a and b) and the ileum.

The abdominal pain became severe on the sixth

day of treatment with oral prednisone (0.5 mg/kg/day); melena appeared, and the purpuric skin lesions spread. A computed tomography scan revealed bowel-wall thickening and luminal fluid accumulation. Treatment was changed from oral prednisone to intravenous prednisolone (1 mg/kg/day), and 1 week later, the abdominal symptoms and rash had fully resolved.

Discussion

Video capsule endoscopy has enabled noninvasive imaging of the small bowel. The indications for video capsule endoscopy include obscure small-bowel disorders undiagnosed with conventional endoscopic and radiological methods9. There have been, to our knowledge, 4 previous reports about the diagnostic role of video capsule endoscopy in patients with IgAV¹⁰⁻¹³. Video capsule endoscopy was used to detect erythema throughout the small bowel in all 8 of the previously reported cases and severe erosions and ulcers of the small bowel in 2 cases; the effects of therapy with steroids and cyclophosphamide were confirmed11,12. Video capsule endoscopy also provided information that aided in the decision to taper the steroid dose¹², as in our first case.

Coagulation factor XIII activity was decreased in both our cases. A correlation between decreased activity of coagulation factor XIII and the severity of abdominal symptoms has been shown in children and adults with IgAV^{14,15}. The severity of abdominal pain is also consistent with the findings of video capsule endoscopy^{10–12}. In our patient 2, the abdominal pain was more severe, and video capsule endoscopy showed multiple ulcers with bleeding in the small bowel which were not found in patient 1. This difference may be a reason why symptoms could be controlled only with intravenous prednisolone at a dose of 1 mg/kg/day rather than with oral prednisolone at a dose of 0.5 mg/kg/day. Video capsule endoscopy was safe, with no side effects in either case.

In conclusion, video capsule endoscopy is a useful tool for evaluating small-bowel lesions and provides valuable information for the treatment of IgAV with gastrointestinal involvement.

Conflict of Interest: None.

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