Case Report

A Steroid-resistant Cryptogenic Multifocal Ulcerous Stenosing Enteritis

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Abstract

Cryptogenic multifocal ulcerous stenosing enteritis (CMUSE) is a rare chronic and recurrent disease with unknown etiology. It characterized by shallow ulcers, submucosa thickness, and no signs of systematic inflammation. Now, steroids are the principle therapy for CMSUE. However, still, there were few patients who did not response to it. Here, we present a young woman who diagnosed with CMUSE. She underwent a surgery because of capsule endoscopy retention. After relapse, she had received budesonide and methylprednisolone separately while both of them did not stop her disease. Till now, 24 years after her initial symptom, there are no signs of carcinoma yet.

Keywords: *Cryptogenic multifocal ulcerous stenosing enteritis, intestinal ulceration, steroid-resistant*

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Introduction

Cryptogenic multifocal ulcerous stenosing enteritis (CMUSE) is a rare idiopathic disease, which was first reported in 1964.[1] It featured by unexplained superficial ulcers, thickened submucosa of small intestine, and no biological signs of systematic inflammation.[2,3] Most of the reported cases did not affect colon. Anemia, abdominal pain, and tarry stool are the main symptoms of CMUSE. There is no big difference of clinical manifestation between pediatric and adult patients, but those developed at early age may present with retardation. Differential diagnosis includes Crohn's disease,[3] nonsteroidal anti-inflammatory drugs (NSAIDs)-related enteritis,[4] infections, vascular disease, and neoplasms. Main treatment for CMUSE consists of nutrition supplement, systemic steroids, intestinal resection, or balloon dilation. Steroids are effective in most cases, but it is not true of a young woman who was resistant to budesonide and methylprednisolone.

Case Report

A 29-year-old woman with remittent abdominal pain and tarry stool for 16 years was referred to our hospital in 2009. On examination, she had lower limbs edema. Her vital signs and chest and abdominal examinations were normal. She denied NSAIDs taking. Fecal occult blood test was positive. A laboratory test confirmed hypoalbuminemia (20.8 g/L), iron deficiency anemia (hemoglobin at 66 g/L), and slightly increased C-reactive protein level (8.43mg/L). Her renal and liver functions, erythrocyte sedimentation rate, immunoglobulin level, complement level, and autoimmune antibodies were within normal range. (PPD) purified protein derivative skin test test was negative. Gastroscopy showed mild gastritis, and colonoscopy was normal. Abdominal computed tomography revealed thickened small intestinal wall. On capsule endoscopy, multiple shallow ulcers and stenosis were found in the small intestine. Exploratory laparotomy was performed for capsule retention. In operation, several strictures located between 130 and 50 cm proximal to ileocecal valve were found and the minimal distance between two strictures was about 3 cm. The stricture lesions were removed. Postoperative pathology showed nonspecific chronic inflammatory, superficial ulcers restricted in the mucosa, and submucosa fibrosis. No granulomas, lymphadenopathy, or vasculitis was found [Figure 1]. She received symptomatic treatment and discharged without further therapy.

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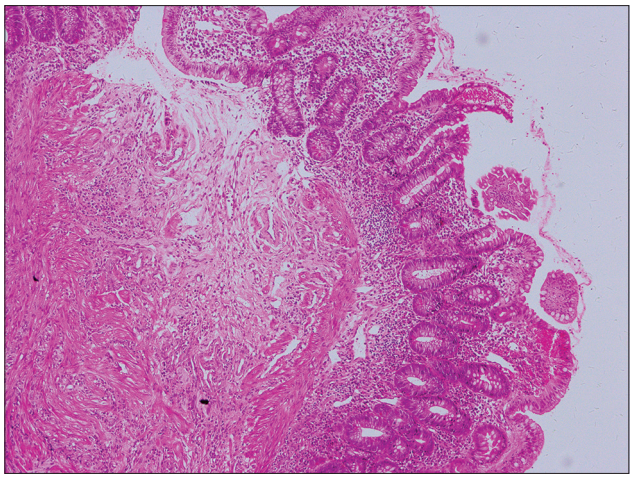

Figure 1: Pathology of the small intestine showing superficial ulcers of small intestine and submucosa fibrosis (H and E, ×100)

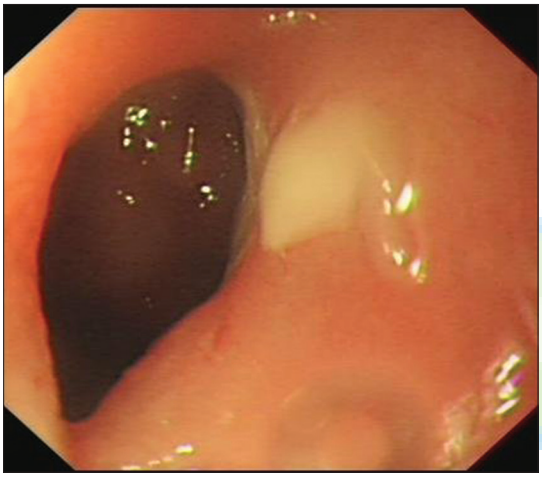

Figure 2: Enterosc]opy showing shallow ulcers and stenosis, 55cm proximal from ileocecal valve

From then on, she was readmitted to our hospital three times for the same reason and along with low albumin, anemia. Following enteroscopy found circular and irregular superficial ulcers with multiple stenosis 60 cm to ileocecal valve [Figure 2]. Biopsy still showed nonspecific moderate chronic inflammation and focal erosion. Acid-fast stain and polymerase chain reaction for tuberculosis were negative. CMUSE was diagnosed. We first prescribed budesonide (9 mg/day) for 1 month; she had lasted abdominal pain and melena. Then, methylprednisolone (40 mg/day) was tried and did not improve her symptom. Steroids were discontinued. She refused further therapy, except symptomatic treatment. Now 1 year after her last hospitalization, she still had continued tarry stool.

Discussion

The etiology of CMUSE is still unclear. Vasculitis[2] and gene mutations[5] had been suspected. Two siblings onset at early ages involved gastric and duodenum, who had a mutation in cytosolic phospholipase A2-α. It catalyzes arachidonic acid metabolism. Steroids are the principle therapy and can improve or cure the disease, even some patients will develop steroids-dependence. Other drugs such as 5-aminosalicylates, azathioprine, and methotrexate have been proved to be ineffective. However, not all patient's respond to steroids. Our patient underwent a surgery before. After relapse, no remission was got by budesonide and methylprednisolone.

Till now, four steroid-resistant CMUSE patients had been reported. Three of them had undertaken at least operation. One had a recurrent ulcerated stenosis after 3 years.[6] Another presented with hypoalbuminemia again just 1 month after surgery.[5] The third one received four surgeries, but her long-term outcome was not reported.[7] To avoid surgery, the last one received infliximab and maintained clinical remission 6 months after starting treatment.[8] How to cope with those steroid-resistant CMUSEs remains a challenge. Now, the patients do not have symptom of obstruction yet. After failure above therapies, she chose to receive only symptomatic treatment, including nutrition, albumin, and iron supplement. Despite taking medicine for long time is not that pleasant, the disease itself does not have big influence on her daily life.

Conclusion

We reported a steroid-resistant CMSUE patient with continued tarry stool for almost 24 years. More studies are needed to find the cause of CMUSE and the therapy for steroid-resistant ones. Fortunately, no CMUSE-related cancer or death has been reported. In this situation, symptomatic treatment may be a choice for noninvasive treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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