

CASE REPORT

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A case report of intestinal obstruction caused by cryptogenic multifocal ulcerous stenosing enteritis

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Abstract

Background: Cryptogenic multifocal ulcer stenosing enteritis (CMUSE) is a rare disease characterized by multiple superficial ulcers, stenosis, and obstruction of the small intestine of unknown origin, and the course can recur.

Case presentation: We encountered a 62-year-old male patient with intestinal obstruction. The patient was admitted to the hospital for surgical treatment due to intestinal obstruction, and was diagnosed with cryptogenic multifocal ulcer stenosis enteritis due to comprehensive surgery and postoperative pathological considerations.

Conclusion: In the future, we will continue to follow up the patient. The present study aims to remind clinicians of this disease, and reduce the incidence of misdiagnosis.

Keywords: Cryptogenic multifocal ulcer stenosing enteritis, Intestinal obstruction, Small intestinal ulcer, Small intestinal stenosis

Background

Cryptogenic multifocal ulcerous stenosing enteritis (CMUSE) is a rare idiopathic disease. At present, there are only over 60 cases of CMUSE reported in the world [1], this was first reported in 1964 [2]. This is featured by unexplained small bowel multiple superficial ulcers, stenosis, obstruction, and no biological signs of systematic inflammation, which are prone to recurrence. Hormone therapy is effective for some patients [3]. We encountered a patient with intestinal obstruction, which was very severe, and this patient was admitted to the hospital for immediate surgical treatment. Combined with the intraoperative findings and postoperative pathological diagnosis, the patient was diagnosed as CMUSE.

Case presentation

A 62-year-old male patient was admitted to the hospital due to “abdominal pain and abdominal distension for one month, with aggravation for five days”. The patient presented with abdominal pain in the previous one month without obvious inducement, mainly around the umbilicus, paroxysmal, accompanied by abdominal distension, with a small amount of exhaust and defecation. The patient continued to eat without paying attention to this. Five days ago, these symptoms aggravated, and the patient stopped venting and defecating. The patient visited our hospital for treatment and underwent abdominal CT, which revealed small intestinal obstruction (Fig. 1). Physical examination: abdominal distention, intestinal type, total abdominal tenderness, obvious periumbilical, and bowel sounds hyperactive. Laboratory examination: WBC count: 19.61×10^9 /L, hemoglobin: 91 g/L, serum albumin 22 g/L, and the remaining assay examination was normal. Previous history: good health, no history of abdominal surgery, and no history of

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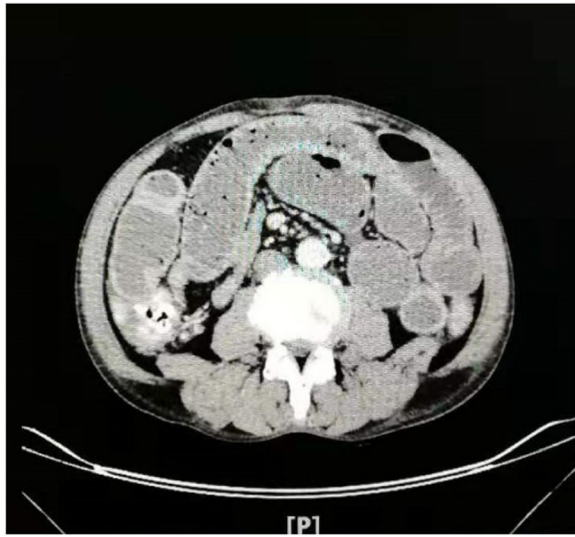


Fig. 1 Preoperative abdominal CT examination showed small bowel obstruction

medication. Since the obstruction was long and severe, emergency laparotomy was performed.

Observations during the operation: there was no adhesion in the abdominal cavity, there was no twist and compression of the intestine, no tumor was detected, and the obstruction was located in the small intestine at 70 cm away from the ileocecal area. The small intestine above the obstruction was highly dilated, and the diameter of the intestine was 8–10 cm. The appearance of the following small intestine was normal. The intestinal canal was longitudinally dissected at the obstruction, a small amount of food fiber was expelled from the intestinal canal at the obstruction site, and a “stenosis ring” was formed in the small intestinal cavity. Merely pores with a diameter of 0.3–0.4 cm were left, and no obvious abnormality was observed in the intestinal mucosa (Fig. 2). The distal intestine was longitudinally opened to the ileocecal region, and multiple “stenosis rings” were found in the intestinal lumen at approximately 40 cm from the distal obstruction, presenting with the same shape (Fig. 3). The shortest spacing between the “stenosis ring” was 1.0 cm, and the longest spacing was 6 cm. The diseased bowel was removed, and the patient recovered well without discomfort, such as diarrhea. The patient was followed up for two months, and no discomfort was found.

Pathological findings included: intestinal stenosis was obtained, shallow intestinal segmental ulcer formation, mucosal erosion and inflammatory granulation tissue formation, the mucosal muscle layer disappeared, submucosal fiber hyperplasia, and longitudinal extension to the mucosal layer, causing mucosal uplift and intestinal wall stenosis. A large amount of neutrophil infiltration, and vasodilatation and congestion were observed in the

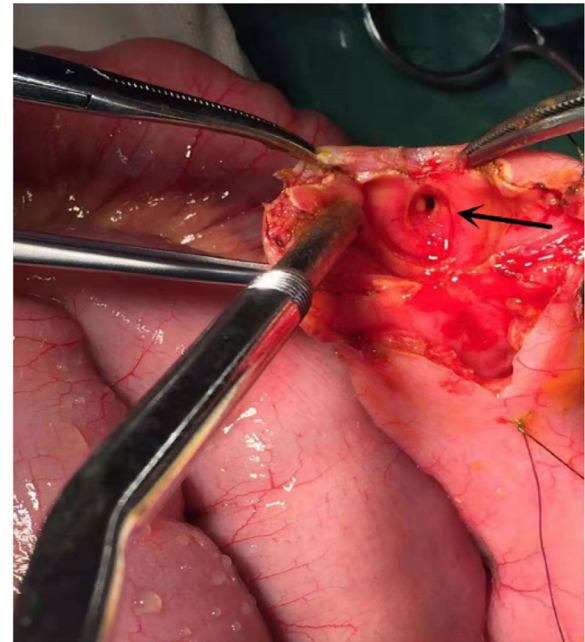


Fig. 2 Intraoperative photograph. The black arrow shows “stenosis ring”

serosa, but no obvious structural abnormalities were found in the muscle layer and serosa layer (Fig. 4).

Discussion and conclusion

The etiology and pathogenesis of CMUSE remains unknown. Some scholars speculate that autoimmune

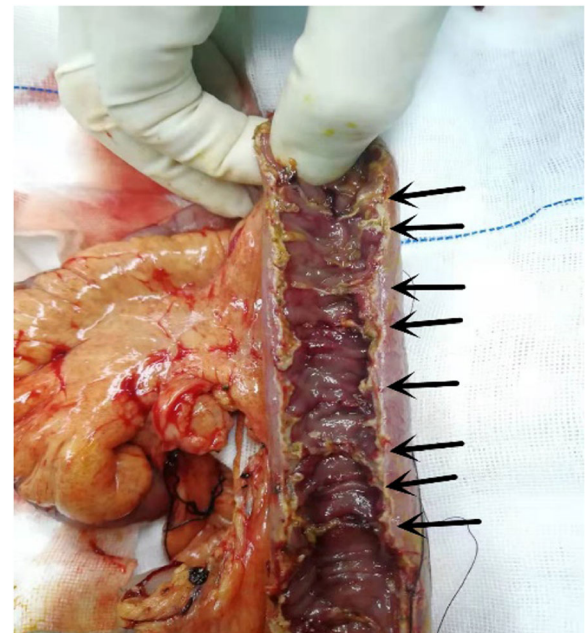


Fig. 3 Intraoperative photograph. The black arrow shows “stenosis ring” (Has been opened)

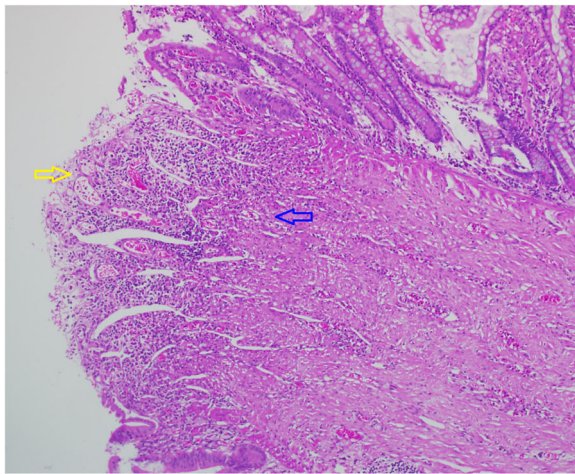


Fig. 4 Postoperative pathological photos. HE stain, 100 times magnification. Yellow arrow: shallow intestinal mucosa ulcer. Blue arrow: muscle fibers growing longitudinally below the ulcer.

abnormalities, excessive formation of fibrous tissue, vasculitis, and mutations in the gene *PLA2G4A* encoding cytosolic phospholipase A2-a are correlated to its pathogenesis [3, 4].

Yu Zhang et al. [5] summarized the diagnostic criteria for CMUSE: (1) Medical history: long course; persistent and occult blood loss from the gastrointestinal tract with severe anemia. (2) Clinical features: chronic and relapsing subileus episodes; chronic iron-deficiency anemia with fatigue, edema, or growth retardation, rarely with diarrhea; with normal inflammatory makers or other biological signs of systemic inflammation; normal colon and stomach; no extraintestinal manifestations. (3) Imaging manifestation: In enteroclysis: multiple persisting stenoses of the small intestine caused by brous strictures. (4) Endoscopic findings: multiple, pleiomorphous and superficial ulcers, and sharply demarcated from the surrounding normal mucosa; caution was taken during the capsule endoscopy examination, because this may become stranded in the strictures. (5) Histological findings: necrotic inflammatory ulcers not reaching the proper muscular layer; nonspecific inflammation, and erosion with submucosal fibrosis.

Differential diagnosis could include: (1) Crohn's disease (CD): CD is one of the most important differential diagnoses for CMUSE. The difference is that Crohn's disease is transmural inflammation, and the typical lesions are longitudinal and fissured ulcers, which are prone to fistula formation and intestinal perforation. Giant cell granulomas are observed in its pathology [6]. (2) NSAID-associated bowel disease: This disease was very similar to CMUSE under endoscopy, with superficial ulcers and intestinal stenosis [7], but NSAID-associated bowel disease has a long history of medication, and can be recovered after

withdrawal [8]. (3) Intestinal tuberculosis can occur with intestinal ulcers or stenosis. Its characteristics include the following: circular ulcers, which can be accompanied by extraintestinal tuberculosis; the histopathological findings of caseous necrotizing granulomatous or positive acid staining could be confirmed. (4) Vasculitis involves multiple ulcers in the intestine, such as Behcet's disease, granulomatous vasculitis, and systemic scleroderma [9]. However, these ulcers are deep, and may have perforations. Furthermore, multifocal stenosis is rare, and blood vessel inflammation is a systemic disease that often involves multiple systems.

Main treatment methods includes: (1) Hormones: Glucocorticoids are presently the first line of treatment, but these are likely to cause hormone dependence, relapse, and steroid resistance [7]. (2) Endoscopic balloon dilatation: This can alleviate the symptoms, and prevent small bowel resection [3, 4]. (3) Surgery: This can easily relapse after surgery, and repeated surgery can easily cause complications, such as short bowel syndrome [3]. (4) Nutritional support therapy: Enteral or parenteral nutrition and iron supplementation can improve the symptoms in the short term, and even obtain mucosal healing. However, since the patient resumed eating, the mucosal ulcer, anemia and hypoproteinemia rapidly relapsed [10].

CMUSE is a rare disease of unknown etiology. Multiple superficial ulcers and intestinal stenosis in the small intestine are its typical manifestations. Its diagnosis depends on the comprehensive judgment of clinical manifestations, imaging examinations, endoscopy and pathological examinations. The investigators will continue to follow the patient and monitor for its recurrence. The present study aims to remind clinicians of this disease. When encountering similar lesions, the possibility of this disease must be considered. This also provides a differential diagnosis for small bowel ulcers and stenotic diseases, reducing the occurrence of misdiagnosis.

Abbreviations

CMUSE: Cryptogenic multifocal ulcer stenosing enteritis; CT: Computer tomography; WBC: White blood cell count; cm: Centimeters; CD: Crohn's disease; NSAIDs: Non-Steroidal Anti-inflammatory Drugs

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Authors' contributions

CC and BF were involved in the treatment of the patient described in the case report, and initiated the case report. CC,BF,YYM and LLZ prepared the first draft of the manuscript. CJ provided the pathological information. All authors participated in further drafting and revision of the manuscript. All authors have read and approved the final manuscript.

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Availability of data and materials

This case report contains clinical data obtained from the electronic medical record in Qingdao Municipal Hospital (Group). Additional information is available upon request, but only in accordance with the privacy restrictions of the hospital.

Ethics approval and consent to participate

Not applicable.

Consent for publication

An informed consent was obtained from the patient. Verbal and written consents were obtained for the publication of the case report and the accompanying images. A copy of the written consent is available upon request.

Competing interests

The authors declare that they have no competing interests.

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