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Multifocal stenosing ulceration of the small intestine

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Abstract

Several reports have described an apparently uncommon clinicopathological disorder that is characterized by multifocal stenosing small-intestinal ulceration. Compared to Crohn’s disease, the ulcers are not transmural and typically remain shallow, and involve only the mucosa and submucosa. The disorder seems to be localized in the jejunum and proximal ileum only, and not the distal ileum or colon. Only nonspecific inflammatory changes are present without giant cells or other typical features of granulomatous inflammation. Most patients present clinically with recurrent obstructive events that usually respond to steroids, although occasionally, surgical resection is required. The location of ulceration and strictures in the more proximal small intestine, along with the absence of any associated granulomatous inflammatory changes in resected material are believed to differentiate this entity from Crohn's disease, which usually is localized in the distal ileum and the colon. CMUSE appears to be an entirely distinct disorder from other small-bowel disorders, which is characterized by multifocal ulceration with stricture formation. With new imaging modalities increasingly becoming available, particularly double-balloon enteroscopy, further appreciation of this entity and its natural history should result.

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Key words: Ulcer; Stenosis; Intestinal diseases; Small intestine; Crohn’s disease

INTRODUCTION

A syndrome that is characterized by intermittent episodes of small-intestinal obstruction caused by benign multifocal ulcerated stenosis has been described, largely, but not entirely, in the French literature [1-10]. This disorder has also been termed “cryptogenic multifocal ulcerous stenosing enteritis” (CMUSE) and has been reported to localize largely within the jejunum or proximal ileum. CMUSE typically is associated with shallow, rather than deep transmural ulceration, and has also been noted to be usually responsive to steroids, although occasionally, surgical resection is required. The location of ulceration and strictures in the more proximal small intestine, along with the absence of any associated granulomatous inflammatory changes in resected material are believed to differentiate this entity from Crohn's disease, which usually is localized in the distal ileum and the colon [11]. CMUSE appears to be an entirely distinct disorder from other small-bowel disorders, which is characterized by multifocal ulceration with stricture formation. With new imaging modalities increasingly becoming available, particularly double-balloon enteroscopy, further appreciation of this entity and its natural history should result.

DIFFERENTIAL DIAGNOSIS

A number of other entities may cause multifocal small-bowel ulcers (Table 1). Idiopathic ulcerative jejunoileitis, described elsewhere in detail [12], is a small-intestinal ulcerative disorder, often associated with celiac disease, or at least, with concomitant flattening of the villous architecture of the small bowel. Some believe that the disorder may represent a specific complication of celiac disease, but ulceration of the small intestine in this setting may also be caused by a difficult-to-diagnose focal lymphoma with lymphomatous cells localized at the ulcer edges [13].

A variety of infectious agents, some common, may also cause small bowel ulceration [7], but most of these (e.g. Campylobacter, Shigella, Yersinia and Salmonella) seem to resolve completely without stricture formation. Of course, the natural history of many infections in the small intestine are not particularly well defined (e.g. tuberculosis and cytomegalovirus infection), especially if immune
TABLE 1  Other causes of multifocal small-bowel ulceration and stenosis

| Cause                                                                 |
|----------------------------------------------------------------------|
| Ulcerative jejunoileitis with celiac disease or sprue-like intestinal disease |
| Lymphoma (especially mucosa only), including T-cell enteropathy and α-chain disease (Mediterranean type) |
| Crohn’s disease involving the small intestine                        |
| Infections (e.g. Campylobacter and Shigella)                         |
| Drug-induced type (especially with NSAIDs)                           |
| Zollinger-Ellison syndrome (gastrinoma) or other hypersecretory disorders |
| Traumatic injury (e.g. endoscopic or surgical treatment, seat-belt injury) |
| Ischemia related to vasculopathy (e.g. collagen vascular disease, coagulopathy, or inflammatory vasculitis) |

NSAIDs: Nonsteroidal anti-inflammatory drugs.

TABLE 2  Differentiation of CMUSE from Crohn’s disease

| Differentiation                                                                 |
|--------------------------------------------------------------------------------|
| Absence of clinical or laboratory features of an inflammatory syndrome        |
| Absence of small-intestinal transmural inflammatory process or ulceration    |
| Absence of small-intestinal giant-cell granulomatous inflammatory process    |
| Absence of small-intestinal fistula formation despite recurrent chronic disease |
| Absence of disease in other parts of gastrointestinal tract (i.e. stomach or colon) |
| Absence of most extraintestinal features of Crohn’s disease (e.g. skin manifestations) |

CMUSE: Cryptogenic multifocal ulcerous stenosing enteritis.

FUTURE DIRECTIONS

CMUSE needs to be defined carefully and more precisely. Its etiology and pathogenesis are unknown. Diagnosis of CMUSE should be considered only after exclusion of each entity listed in the differential diagnosis above, especially Crohn’s disease and drug-induced ulceration with stenosis. Nonspecific small-intestinal ulceration without stricture formation should be considered a separate entity. With the development of better imaging methods for the small intestine, especially double-balloon enteroscopy, the opportunity now may be present to further explore this intriguing entity.

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