A RARE DIAGNOSIS OF CRYPTOGENIC MULTIFOCAL ULCEROUS STENOSING ENTERITIS FOLLOWING PARTIAL SMALL BOWEL RESECTION FOR A RETAINED CAPSULE ENDOSCOPE

Samuel C. Purkey, DO[1], Katherine Cochrane, MD[2], Andrew Russ, MD[3]

Department of Medicine, University of Tennessee, Knoxville TN[1]
Department of Pathology, University of Tennessee, Knoxville TN[2]
Department of Surgery, University of Tennessee, Knoxville TN[3]

Introduction

Small bowel disease has long been a challenge to diagnose given the limitations in visualization and tissue biopsy. Advances have been made, including capsule endoscopy, however, making the diagnosis remains difficult. We describe a rare case of cryptogenic multifocal ulcerous stenosing enteritis (CMUSE) following partial small bowel resection secondary to a retained capsule endoscope.

Case Presentation

- Our patient is an 80-year-old male presenting to colorectal surgery clinic with concern for impacted capsule endoscope following a work-up for chronic anemia.
- Computed tomography imaging revealed a metallic foreign body within the small intestine.
- The patient underwent laparoscopic assisted removal of the capsule with unsuccessful attempts to manipulate due to strictures or intraluminal webs entrapping the capsule.
- Subsequently, he required laparoscopic assisted small bowel resection with side-to-side functional end to end anastomosis.
- Intraoperative findings revealed short segment inflammatory changes, creeping fat, and antimesenteric fat consistent with Crohn's Disease.
- Interestingly, pathology demonstrated segments of strictures, shallow fissuring ulcers involving mucosa and submucosa, and focal pyloric metaplasia; compatible with multifocal stenosing ulcerations of the small intestine.
- Given the presence of disease primarily in the jejunum rather than ileum, shallow mucosal and submucosal ulcers without architectural distortion of the mucosa immediately adjacent, and normal mucosa between ulcers and strictures, other etiologies for the multifocal stenosing ulcerations of the small intestine were considered leading to the diagnosis of CMUSE.
- The patient was discharge on post-operative day two without complications.

Pathology/Images

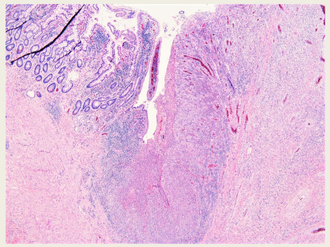


Figure 1. Small bowel demonstrating ulceration of the mucosa and submucosa with associated acute inflammation and without much architectural distortion of the adjacent mucosa (H&E, 40x).

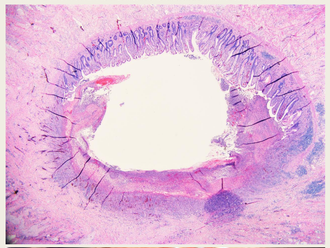


Figure 2. Stricture and ulceration of the small bowel mucosa (H&E, 20x).



Figure 3: Intra-operative findings concerning for erythema, creeping fat, and stricture formation.



Figure 4: Capsule endoscpe retrieved following partial small bowel resection.

Discussion

- CMUSE is a chronic or relapsing disease exclusively of the small intestine resulting from multiple strictures with mucosal and submucosal ulcerations presenting with an array of vague abdominal symptoms.
- First described in 1964, CMUSE is a rare diagnosis with less than 100 cases reported in literature.
- Common presenting symptoms include abdominal pain, nausea, ileus, and hematochezia.
- Patients commonly present with an associated anemia.
- Often, patients get misdiagnosed as irritable bowel disease, Crohn's disease, NSAID enteropathy, or intestinal tuberculosis with Crohn's disease being the most common.
- A key differentiating factor with Crohn's disease involves the sparring of the terminal ileum seen in CMUSE.
- Onset is commonly in adolescent and middle-aged populations, making our case unique.
- Initial therapy involves corticosteroids with surgery remaining a last resort given high rates of disease recurrence.
- Further investigation is warranted to better understand the underlying pathophysiology, etiology, and treatment; however, it remains important to keep on your differential as treatment may vary from other small bowel diseases.

References

1. Chen, D., Liu, W., Zhou, W. *et al.* Retrospective study of the differential diagnosis between cryptogenic multifocal ulcerous stenosing enteritis and small bowel Crohn's disease. *BMC Gastroenterol* **20,** 252 (2020). https://doi.org/10.1186/s12876-020-01389-7.
2. De Schepper H, Macken E, Van Marck V, Spinhoven M, Pelckmans P, Moreels T. Infliximab induces remission in cryptogenic multifocal ulcerous stenosing enteritis: First case. World J Gastroenterol. 2013;19(10):1661–4.
3. Debray C, Besancon F, Hardouin JP, Martin E, Marche C, Khoury K. [Cryptogenetic plurifocal ulcerative stenosing enteritis.]. Arch Mal Appar Dig Mal Nutr. 1964;53:193–206.
4. G.Perlemuter,L.Guillevin,P.Legman,L.Weiss,D.Couturier,andS.Chaussade,"Cryptogeneticmultifocalulcerousstenosingenteritis: an atypical type of vasculitis or a disease mimickingvasculitis,"Gut,vol.48,no.3,pp.333–338,2001
5. T. Matsumoto, M. Iida, T. Matsui, and T. Yao, "Chronicnonspecific multiple ulcers of the small intestine: a proposal ofthe entity from Japanese gastroenterologists to Western entero-scopists,"Gastrointestinal Endoscopy,vol.66,no.3,supplement,pp. S99–S107, 2007.

THE UNIVERSITY OF TENNESSEE MEDICAL CENTER

THE UNIVERSITY of TENNESSEE GRADUATE SCHOOL OF MEDICINE