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CASE REPORT | INFLAMMATORY BOWEL DISEASE

Aseptic Splenic Abscesses With Concomitant Sweet Syndrome as Extraintestinal Manifestations of New-Onset Crohn's Disease

Mary McGrath, MD[1], Calvin Geng, MD[1], Anthony Rainho, MD[2], and Esteban Figueroa, MD[2]

[1]*Department of Internal Medicine, University of Virginia Health, Charlottesville, VA*
[2]*Division of Gastroenterology and Hepatology, University of Virginia Health, Charlottesville, VA*

ABSTRACT

Splenic abscesses are typically infectious in nature but have rarely been reported as an extraintestinal manifestation of inflammatory bowel disease, particularly of Crohn's disease. In the United States, reported cases are even more scarce. We present a case of aseptic splenic abscess with concomitant Sweet syndrome in a middle-aged woman with newly diagnosed Crohn's disease. Extensive workup was required to reach final diagnosis, and she rapidly improved with corticosteroid therapy and has been maintained on risankizumab. We aim to contribute to limited data and heighten clinician awareness of these atypical extraintestinal manifestations.

KEYWORDS: aseptic abscess syndrome; Sweet syndrome; Crohn's disease; inflammatory bowel disease

INTRODUCTION

Extraintestinal manifestations of Crohn's disease (CD) are well described in the musculoskeletal, dermatologic, and hepatobiliary systems, whereas splenic involvement is rare. Infectious abscesses in CD typically result from fistulizing disease rather than noncontiguous inflammation. We describe a middle-aged woman with nonspecific gastrointestinal symptoms and underwent an extensive workup revealing a new diagnosis of CD manifesting with aseptic splenic abscess (AA) and classical acute febrile neutrophilic dermatosis (Sweet syndrome).

CASE REPORT

A 52-year-old woman with hyperlipidemia and prediabetes presented to an academic center's emergency department for progressive epigastric pain and daily fevers. She endorsed 2 weeks of symptoms including a 15-pound weight loss and abdominal bloating. During this time, she was evaluated twice at other hospitals; abdominopelvic computed tomography had demonstrated findings of nonspecific colitis for which amoxicillin-clavulanate was ineffective. Of note, the patient followed regularly at an outside facility's gastroenterology clinic for chronic diarrhea (baseline 7–9 daily bowel movements) and lower abdominal pain. Colonoscopy and esophagoduodenoscopy performed 8 months before admission showed gastritis and superficial ileal erosions with indeterminate pathology.

On arrival, the patient was febrile to 102.1°F (38.9°C), tachycardic to 111 beats/min, and hypertensive at 153/77 mm Hg. Initial laboratory work yielded leukocytosis to 19.58 $\times$ 10^9/L, anemia to 9.5 g/dL, normal liver enzymes, and C-reactive protein to 29.3 mg/dL (normal $<$0.5 mg/dL). She was started on piperacillin-tazobactam. Stool calprotectin, resulting later in admission, was elevated to 176 mcg/g (normal $<$120 mcg/g). Abdominopelvic computed tomography and magnetic resonance imaging revealed numerous splenic abscesses (Figure 1), mesenteric lymphadenopathy with central necrosis, and scattered colonic wall thickening including in the rectum. Per the radiologist's interpretation, these findings were highly concerning for infectious (typhoid/paratyphoid *Salmonella* species, *Yersinia enterocolitica*, and *Escherichia coli*), autoimmune, or neoplastic processes. Lymph nodes were not amenable for biopsy. Daily fevers continued despite antibiotics.

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Correspondence: Mary McGrath, MD (xua4yg@uvahealth.org).

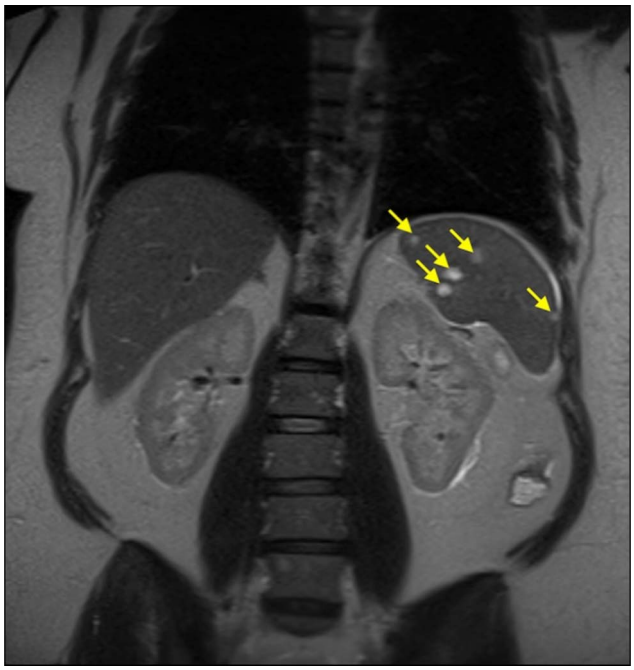

Figure 1. Splenic abscesses were seen in a T2-weighted magnetic resonance imaging. Yellow arrows point to the 5 lesions observed in this particular image, though numerous abscesses were seen throughout the patient's spleen.

Upper and lower endoscopies were performed on hospital day 3. Esophagoduodenoscopy showed Los Angeles Grade A esophagitis, diffuse gastropathy with white "mucosal specks," gastric ulcers, and erythematous duodenopathy. Colonoscopy showed multiple erosions in terminal ileum and diffuse colitis with rectal involvement (Figure 2). Serologic and stool testing for community gastrointestinal pathogens, *Clostridioides difficile*, *Coxiella burnetii*, *Tropheryma whipplei*, *Bartonella henselae*, *Francisella tularensis*, tuberculosis, and streptococcal and fungal pathogens resulted negative, as did blood cultures.

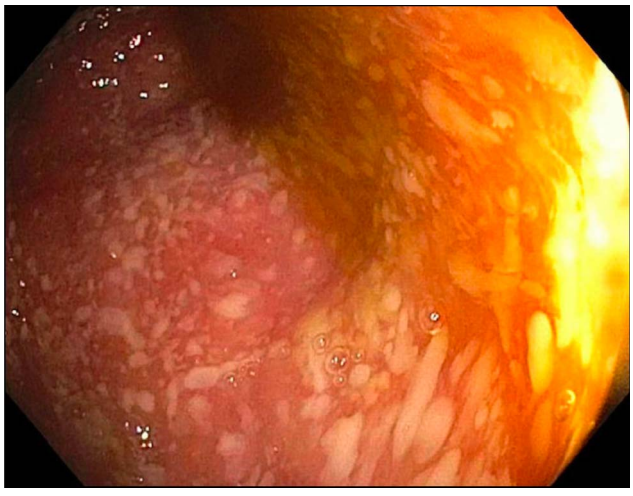

Figure 2. Rectal ulcers were seen endoscopically. During colonoscopy, findings such as this raised suspicion for a diagnosis of inflammatory bowel disease.

Antinuclear antibody and peripheral flow cytometry were unrevealing. On hospital day 7, the patient developed erythema nodosum as well as vesicular lesions, which were found to be consistent with acute febrile neutrophilic dermatosis (Sweet syndrome) by tissue biopsy (Figure 3).

Pathology from endoscopy showed both active and chronic gastritis, active duodenitis with foveolar metaplasia with giant cells, and diffuse acute and chronic inflammation in all colon biopsies; terminal ileum was normal. The pathologist's report specifically suggested CD. In combination with clinical and imaging data despite the less classic finding of rectal involvement, this confirmed a new diagnosis of nonstricturing, nonpenetrating CD. Given negative infectious workup, the patient was initiated on high-dose steroids and experienced rapid resolution of diarrhea, epigastric pain, fevers, and all cutaneous findings.

After discharge, shared decision-making was used to initiate risankizumab, which provided continued symptomatic relief. On follow-up imaging one month after discharge, splenic abscesses and lymphadenopathy were nearly resolved. She has since remained in both clinical and biochemical remission.

DISCUSSION

We present a case of new CD with aseptic splenic abscesses and Sweet syndrome. Aseptic splenic abscesses are an atypical extraintestinal manifestation of CD that has been infrequently described in the literature.[1–3] These abscesses result from AA syndrome, an inflammatory condition causing neutrophil-predominant sterile lesions and constitutional symptoms including fever. This syndrome carries an incidence of $<$1%, involves the spleen 71.8% of the time, and is associated with inflammatory bowel disease (IBD), particularly CD, in 42.2% of reported cases.[1,2] The first large case series on IBD-associated AA found a 70% correlation between AA and IBD, suggesting a stronger relationship.[4]

This patient's concomitant Sweet syndrome further adds to this unique case. Alternative associations for Sweet syndrome, such as drug-induced or malignancy-associated, were ruled out, thus linking it to her IBD. Sweet syndrome, a rare tender exanthem, occurs with IBD 0.07%–0.21% of the time.[5] This is significantly less frequent than IBD with pyoderma gangrenosum or erythema nodosum, the latter of which was also present in our patient.[5] Interestingly, cutaneous manifestations are seen in IBD-associated AA syndrome 20% of the time; Sweet syndrome itself could be categorized under AA syndrome.[6,7] Sweet syndrome and AA are perhaps commonly driven by systemic neutrophilic infiltration, although details are ill-defined.[7]

Bollegala et al's[3] literature review of IBD with AA syndrome reveals that our patient (52 years old) is older than the typical demographic of the second and third decades of life. Our patient's fever, abdominal pain, diarrhea, and weight loss

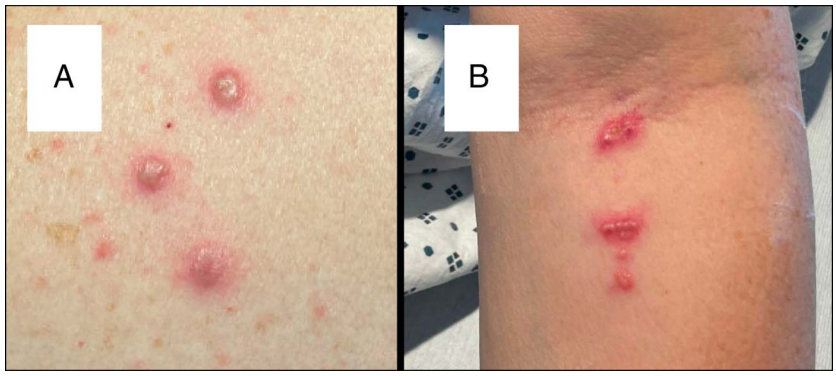

Figure 3. The patient developed a rash consistent with acute febrile neutrophilic dermatosis, or Sweet syndrome, on the back and arms as shown. Panel (A) shows lesions on the upper back. Panel (B) shows lesions on the left arm intravenous insertion site, representative of pathergy.

matched the 4 most common presenting features in the review. Thus, her prolonged fever was most likely a result of a combination of her aseptic splenic abscesses and Sweet syndrome, with perhaps less contribution from CD.

In about half (48.6%) of IBD with AA cases, IBD diagnosis precedes the development of such abscesses.[3] Although our patient's diagnoses were made simultaneously, her chronic diarrhea likely represented long-standing CD with previous diagnosis delayed because of atypical endoscopic findings. Prospective data are needed, but AA can be a relapsing-remitting condition and does not typically coincide with flares of underlying IBD, although Sweet syndrome *does* coincide with IBD.[4,8] Reassuringly, mortality from AA is low.[4]

Corticosteroids are the mainstay of treatment in IBD-associated AA. Reports show a 94.6% improvement rate with steroid induction, but initiation is understandably risky, given splenic abscesses' known association with bacterial infections.[3] In immunosuppression-refractory relapsing AA syndrome, colchicine has interestingly been protective, but splenectomy may be considered as definitive therapy and has been successful in rarely described cases.[2,9]

This case further contributes to the growing pool of knowledge surrounding IBD-associated AA by highlighting another rare extraintestinal manifestation of Sweet syndrome. Aseptic splenic abscesses induce significant morbidity, and prompt initiation of corticosteroids results in swift resolution. In this case, the patient maintains remission with risankizumab therapy.

DISCLOSURES

Author contributions: M. McGrath wrote the manuscript and reviewed the literature. C. Geng, A. Rainho, and E. Figueroa revised the work. E. Figueroa is the article guarantor. All authors read and approved the final manuscript.

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