Sweet syndrome secondary to inflammatory bowel disease

Malcolm M Wells BSc(Hons) MSc MD[1], William Stecho MD[2], Bret Wehrli MD FRCPC[2], Nitin Khanna MD FRCPC[1]

CASE PRESENTATION

A 64-year-old man presented to hospital with a three-month history of progressively worsening mucousy bloody diarrhea, polyarthritis and a rash covering his lateral malleolus. His history was significant for previous quadriceps tendon rupture and supraventricular tachycardia. His only medication was a short course of prednisone initiated shortly before his hospitalization. He was a nonsmoker and nondrinker, with no significant family history.

Physical examination was significant for a fever of 38.2°C as well as a warm swollen left knee, ankle and foot. A bullous lesion 5 cm in size was present on the medial aspect of the left malleolus (Figure 1), with an erythematous base and draining serosanginous fluid. He exhibited multiple oral ulcers. The remainder of the examination was noncontributory.

Laboratory investigations revealed neutrophilia (14.5×10^9/L) and a normocytic anemia (1×10^5 g/L). Stool was negative for ova and parasites, and cell culture was negative for *Clostridium difficile*.

DIAGNOSIS

Colonoscopy demonstrated patchy inflammation and ulceration in the rectum, sigmoid and descending colon (Figure 2). The transverse and ascending colon, as well as the terminal ileum were within normal limits.

Pathology of the colonic biopsies diagnosed inflammatory bowel disease (IBD). The patient ultimately required a hemicolectomy and the pathology of the resected colon confirmed IBD. Skin biopsies demonstrated diffuse inflammatory neutrophil-predominant infiltrate of the dermis, consistent with Sweet syndrome.

There are several features of the present case that are characteristic of Sweet syndrome. The patient presented with fever, leukocytosis and characteristic lesions. Arthralgias and oral ulcers can be extraintestinal manifestations of IBD, but are also apparent in 30% of patients with Sweet syndrome (1-4). Finally, the biopsy was characteristic of the diagnosis. Following bowel resection, the skin lesions, oral ulcers and arthralgias remained, all of which eventually responded to treatment with steroids.

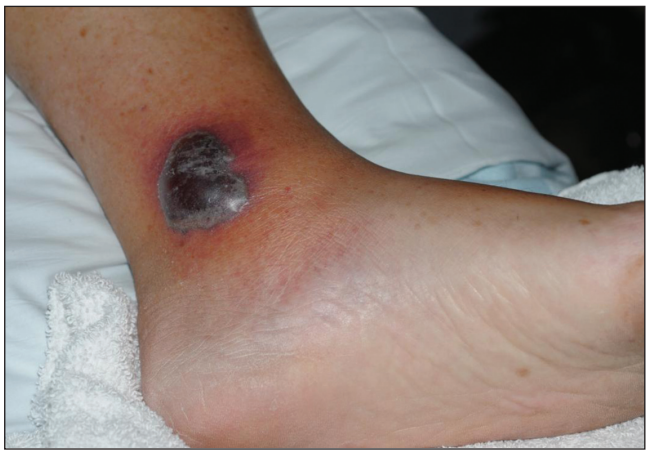

Figure 1) *Bullous lesion characteristic of Sweet syndrome*

DISCUSSION

Originally described by Robert Douglas Sweet in 1964 (5), Sweet syndrome is characterized by fever, leukocytosis and erythematous tender skin lesions with neutrophilic infiltration of the upper dermis (6,7). Several types of Sweet syndrome have been described including classical/idiopathic, malignancy-associated (8), drug-induced, pregnancy-related (9), parainfectious (10) and parainflammatory (1,2,6,7,11-13). Usually only the skin is involved but extracutaneous manifestations have been reported in bones, intestines, liver, aorta, lungs and muscles (1-4,14).

Differential diagnosis includes cellulitis, erythema multiforme, erythema nodosum, leukocytoclastic vasculitis, pyoderma gangrenosum, erysipelas and, rarely, disseminated *Fusarium* infection (7,15). While history, physical examination and other investigations can aid in the diagnosis, skin biopsy remains the gold standard.

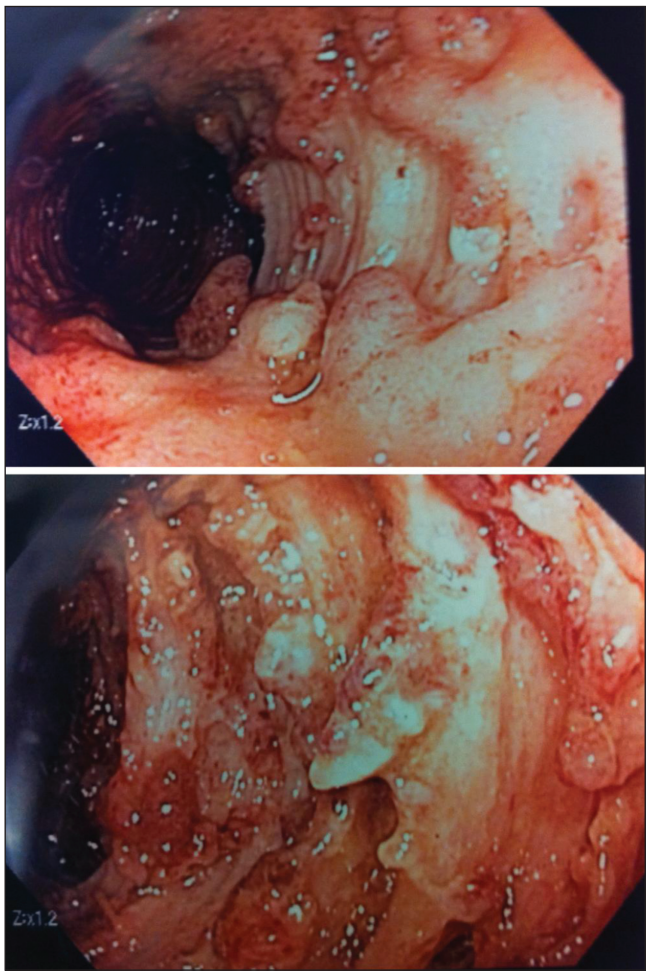

Figure 2) *Endoscopic images of severe ulcerated inflammatory bowel disease in the sigmoid colon*

[1]*Department of Gastroenterology;* [2]*Department of Pathology, Schulich School of Medicine and Dentistry, University of Western Ontario, London, Ontario*
Correspondence: Dr Malcolm M Wells, Schulich School of Medicine, University of Western Ontario, Room M106, Medical Science Building, London, Ontario N6A 5C1. Telephone 519-685-8500, e-mail malcolm.wells@gmail.com
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First-line treatment options, based on uncontrolled case series and reviews, are systemic corticosteroids (6), potassium iodide (6), colchicine (16) or topical corticosteroids (6). Second-line medications include indomethacin (17), clofazimine (1), cyclosporine (18-20), dapsone (21-23) and doxycycline (24). Prognosis is generally good, with the rashes usually responding well to systemic steroids.

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