The Sweet syndrome as a cutaneous manifestation of Crohn's disease

Syed A. Huda, MBBS^a , Sara Kahlown, MBBS^b, Muhammad Hashim Jilani, MBBS^a, Syed H. Alam, MD^a, Sana Riaz, MBBS^a , Mostafa Vasigh, MD^a , and Bashar Sharma, MD^c

^aDepartment of Internal Medicine, State University of New York Upstate Medical University, Syracuse, New York; ^bDepartment of Internal Medicine, UHS Wilson Medical Center, Johnson City, New York; ^cDepartment of Internal Medicine, Syracuse Veterans Affairs Medical Center, Syracuse, New York

ABSTRACT

Crohn's disease is an inflammatory bowel disease associated with many extraintestinal manifestations involving multiple organs, including the skin, eyes, liver, and joints. Classic Sweet's syndrome is a cutaneous manifestation of Crohn's disease, characterized by sudden onset of painful plaques or nodules of the skin associated with fever and neutrophilia. We present a case of classic Sweet's syndrome in a patient with well-controlled Crohn's disease.

KEYWORDS Crohn's disease; extraintestinal manifestations; inflammatory bowel disease; Sweet's syndrome

Inflammatory bowel disease (IBD) primarily includes ulcerative colitis and Crohn's disease (CD). CD is characterized by inflammatory skip lesions involving any part of the gastrointestinal tract, but most commonly targeting the small intestine and/or colon.¹ In addition to systemic and intestinal manifestations, CD can present with extraintestinal involvement, including ocular, cutaneous, dermatologic, and joint manifestations. The classic cutaneous manifestations are erythema nodosum and pyoderma gangrenosum. Sweet's syndrome is one of the rarer cutaneous presentations of CD.² We present a case of the Sweet syndrome in a patient with well-controlled CD.

CASE DESCRIPTION

A 29-year-old man with known CD involving the ascending colon that was well controlled on mesalamine and infliximab presented to his primary care physician with a new skin rash. Given his history of using a hot tub at the gym, he was diagnosed with hot tub folliculitis and sent home on a 10-day course of oral ciprofloxacin. He continued to have new eruptions, however, so went to the emergency room. At presentation, his blood pressure was 124/76 mm Hg; heart rate, 85 beats/minute; and temperature, 38°C. He had numerous erythematous papules on the chest and back

(*Figure 1*) and a few pustules around hair follicles on the arms. There was no rash on the lower extremities. The white blood cell count was 15,000/μL of blood with neutrophilic predominance (absolute neutrophil count of 12,300). The sedimentation rate was 63 mm/h (reference 0 to 20 mm/h), and C-reactive protein was 11.2 mg/L (reference 0 to 0.8 mg/L). Varicella infection was diagnosed, and he was started on intravenous valacyclovir while viral serology was pending. He continued to be febrile with new lesions appearing on the face and torso. The varicella zoster virus and herpes simplex virus polymerase chain reaction, viral cultures, fungal cultures, hepatitis panel, human immunodeficiency virus, and rapid plasma reagin tests were negative.

Since the patient had no clinical improvement on antiviral drugs, a punch skin biopsy of the chest lesion was performed and showed diffuse infiltration of the epidermis and dermis by a neutrophil-rich inflammatory cell infiltrate without any signs of leukocytoclastic vasculitis (*Figure 2*). Periodic acid–Schiff stain was negative for fungal organism and no viral cytopathic changes were seen. These histopathological findings were consistent with the Sweet syndrome. His antiviral medications were discontinued, and he was started on 1 mg/kg/day prednisone with dramatic improvement in the rash and normalization of his white blood cell count and inflammatory markers. He was then discharged

Corresponding author: Syed A. Huda, MBBS, Department of Medicine, Upstate University Hospital, Room 5142, 750 E. Adams Street, Syracuse, NY 13210 (e-mail: hudas@upstate.edu)

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Figure 1. Erythematous papules on the chest and back.

Figure 2. Epidermis with mild spongiosis and diffuse neutrophilic infiltration of the dermis. Dermal neutrophil-rich inflammatory cell infiltrate with pustule formation (hematoxylin and eosin staining, $\times 20$).

home on 1 month of oral prednisone taper. He had complete resolution of the rash when he was seen in the outpatient clinic 1 week after discharge.

DISCUSSION

The Sweet syndrome was first described in 1964 by Dr. Robert Douglas Sweet as an acute febrile neutrophilic dermatosis. It can be associated with malignancy, induced by a drug, or classic (idiopathic), as present in our patient. Classic Sweet's syndrome is the most common type and is characterized by fever, leukocytosis, sudden-onset painful papules, plaques, or nodules of the skin, and a diffuse infiltrate consisting predominantly of mature neutrophils typically located in the upper dermis. Classic Sweet's syndrome occurs more in women than men at a ratio of 4:1 and is most common in those from 30 to 60 years. It recurs in one-third of patients.³ It is most frequently associated with infections (particularly upper respiratory tract

infections and gastrointestinal infections), IBD, and pregnancy. Less common associations include other infections (e.g., human immunodeficiency virus, tuberculosis, chlamydia), primary immunodeficiencies, and autoimmune diseases (e.g., Behcet disease,⁴ relapsing polychondritis,⁵ rheumatoid arthritis,⁶ sarcoidosis,⁷ autoimmune thyroid disease,⁸ and connective tissue disorders including systemic lupus erythematosus and dermatomyositis³).

The diagnostic criteria for classic Sweet's syndrome were first introduced by Su and Liu in 1986⁹ and were modified by von den Driesch in 1994.¹⁰ Classic Sweet's syndrome can be diagnosed if both major criteria and two out of four minor criteria are present. Major criteria include (1) abrupt onset of painful erythematous plaques or nodules and (2) histopathologic evidence of a dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis. Minor criteria are (1) pyrexia $>38^{\circ}\text{C}$; (2) association with an underlying hematologic or visceral malignancy, inflammatory disease, or pregnancy, preceded by an upper respiratory infection, gastrointestinal infection, or vaccination; (3) excellent response to treatment with systemic glucocorticoids or potassium iodide; and (4) abnormal laboratory values at presentation (three of four of the following: erythrocyte sedimentation rate >20 mm/h, positive C-reactive protein, >8000 leukocytes, or $>70\%$ neutrophils).

Sweet's syndrome is a rare extraintestinal manifestation of CD. There are <50 case reports of Sweet's syndrome related to CD.¹¹ The occurrence of Sweet's syndrome in IBD usually correlates with gastrointestinal activity; however, in up to 25% of cases, it precedes the diagnosis of IBD.¹² First-line treatments include systemic corticosteroids, potassium iodide, colchicine, and topical corticosteroids. Second-line medications include indomethacin, clofazimine, cyclosporine, dapsone, and doxycycline.¹³ Our patient had uncontrolled CD for 2 years prior to starting infliximab infusions.

ORCID

Syed A. Huda <http://orcid.org/0000-0002-2605-8273>

Sana Riaz <http://orcid.org/0000-0001-5634-0037>

Mostafa Vasigh <http://orcid.org/0000-0003-2148-8407>

Bashar Sharma <http://orcid.org/0000-0001-8160-2211>

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