Sweet's Syndrome in a Case of Ulcerative Colitis-Case Report and Review of Literature

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Abstract

Sweet syndrome,also known as acute febrile neutrophilic dermatosis , is one of the rare cutaneous association of ulcerative colitis.Only few cases of Sweet syndrome associated with ulcerative colitis have been reported in literature. We herein describe a case of young female with acute exacerbation of ulcerative colitis associated with erythematous , papular skin lesions which on biopsy were consistent with Sweet syndrome. Treatment with intravenous steroids resulted in improvement of ulcerative colitis and disappearance of cutaneous lesions. Cutaneous lesions of Sweet syndrome in ulcerative colitis parallel the bowel disease activity in majority of the cases but sometimes may precede the intestinal symptoms and rarely may appear after procto-colectomy for acute severe ulcerative colitis.Introduction

Inflammatory bowel disease,that comprises ulcerative colitis and crohn's disease,is associated with variety of extra-intestinal manifestations involving different organ systems including the skin. Extra-intestinal manifestations are more frequent in ulcerative colitis. Overall prevalence of these manifestations is 21 % and more than one extra-intestinal manifestation occurs in about 25 % of patients. Extra-intestinal involvement occurs involving kidneys, eyes, joints, lungs and skin.

Cutaneous involvement occurs in 5% of the patients of ulcerative colitis. Skin involvement may occur during the acute exacerbation of bowel disease activity or sometimes it may precede the bowel involvement. Among the different cutaneous manifestations, the most common is erythema nodosum.

Sweet syndrome is one of the rare associations of ulcerative colitis. It is also known as acute febrile neutrophilic dermatosis. Its first description was given in 1964 by Robert Douglas Sweet.[1] Its association with ulcerative colitis was first reported in 1988. It is characterized by acute onset fever accompanied by leucocytosis and histology reveals neutrophilic infiltration of the upper dermis.

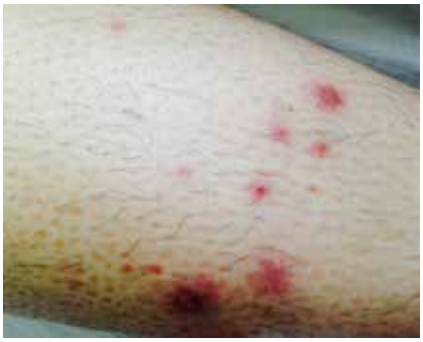

Fig. 1: Image showing maculo-papular erythematous eruptions on extensor aspect of leg

There have been only few cases of Sweet syndrome associated with ulcerative colitis reported in literature. We therefore report this rare association of ulcerative colitis.

What's known

1. Sweet syndrome, acute febrile neutrophilic dermatosis, is characterized by acute onset fever accompanied by leucocytosis and skin rash and histology of skin lesions histology reveals neutrophilic infiltration of the upper dermis.
2. It is one of the rare associations of ulcerative colitis.

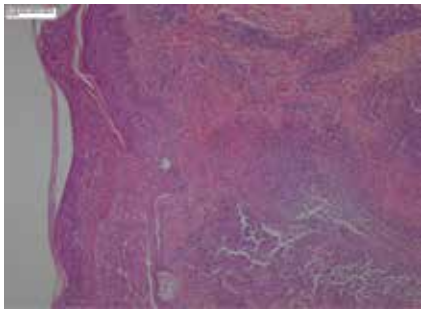

Fig. 2: Low power view of skin biopsy showing dense inflammatory infiltrate into dermis

Case Report

Our patient was 23 years old female,known case of ulcerative colitis,who was in remission on treatment with oral mesalamine and azathioprine. Presented with 3 days history of acute onset high grade fever, skin eruptions in the form of tender, well demarcated red colored lesions (Figure 1). These lesions were non-pruritic. She gave history of increased frequency of stools, 5-6 times, watery in consistency with blood and mucus. There was no abdominal pain, cough, chest pain, headache or joint pains. In the past, the clinical course of ulcerative colitis has been waxing and waning with few flares in last couple of years managed with oral steroids. Prior to current admission, her disease was in remission.

On examination, she was febrile, there were ulcers on the uvula, the skin lesions were tender, well demarcated papules with few showing pustular transformation. There were spread throughout the body including face, trunk and limbs. There were no genital ulcers. Abdominal examination was unremarkable.

She was admitted under gastroenterology unit. Complete blood counts showed leucocytosis. Her tests for malaria, dengue, leptospira, Weil Felix, Cytomegalovirus (CMV IgM) and Ebstein Barr Virus (EBV IgM) were all negative. Her Anti-nuclear antibody by immunofluorescence was negative.

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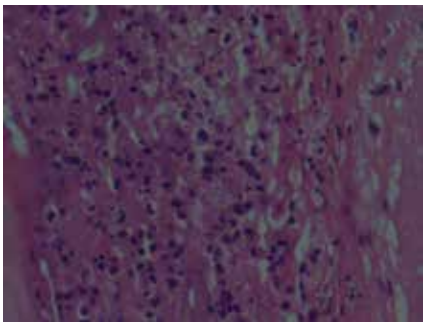

Fig. 3: High power view of skin biopsy showing polymorpho-nuclear cells in upper dermis

Her IgA and IgG levels were normal. Her CRP was 309 mg/dL and ESR was elevated at 78. Other antibodies against extractable nuclear antigens. These lesions first appeared on legs and then subsequently noticed on trunk and face over period of 3 days like nucleosome, dsDNA, histones, Sm, Sm/RNP, SSa, SSb, Scl-70, ku, Pm-SCL, mi, jo, SRP-54, ribosomes, CENP, PCNA, Sp-100, were negative. ANCA was also negative

Her colonoscopy showed loss of vascular pattern of mucosa, with mild ulcerations. There was no spontaneous oozing or friability. Colonic biopsy showed disorganization of crypt architecture, dilated and branched crypts, crypt destruction with cryptitis and crypt abscess. The lamina propria showed dense acute and chronic inflammatory infiltrate. All the findings were suggestive of moderate activity in a case of ulcerative colitis.

Skin biopsy (Figures 2 and 3) was taken from one of the lesions which showed inflammatory exudate covering the epithelium. There were neutrophilic abscesses in epithelium as well as sub epithelium, mainly concentrated around vessels and adnexal tissues. There were perivascular inflammatory cell infiltrates composed of lymphocytes and neutrophils. There was no evidence of granulomas or dysplasias. On immunohistochemistry, IgA was negative. All the above staining for findings were suggestive of Acute Neutrophilic Dermatoses.

She was started on intravenous hydrocortisone (100 mg thrice daily). Her clinical symptoms including skin lesions, responded well by day 5 of treatment. She was subsequently discharged on tapering dose of oral steroids.

Discussion

Sweet syndrome, also known as acute febrile neutrophilic dermatoses is one of the rare cutaneous manifestation of ulcerative colitis. Other conditions reported to be associated with Sweet syndrome are- Sarcoidosis, rheumatoid arthritis, subacute thyroiditis, Behcet's disease, ankylosing spondylitis, Sjogren's syndrome, and malignancies specially the haematological malignancies like lymphoma.[2] It has also been reported in association with some medications like trimethoprim, nitrofurantoin, sulfamethoxazole, diclofenac and ofloxacin.[3,4]

In majority of the cases, Sweet syndrome occurs during the periods of acute exacerbation of the intestinal disease but it may precede the bowel disease activity and sometimes it may occur even after procto-colectomy.[5] In a retrospective analysis of 29 cases of Sweet syndrome, underlying disease could be identified only in four cases(lymphoma, polycythemia, sarcoidosis and ulcerative colitis),[6] while another study of 16 cases of Sweet syndrome by Ginarte, M. et al, one had ulcerative colitis and another two had underlying malignancy.[7] Neutrophils infiltration occurs in upper dermis in Sweet syndrome but the factors that stimulate the neutrophil migration into skin have not been elucidated. Sweet syndrome was reported in patients with T cell immunodeficiency[8] and in patients receiving G-CSF treatment.[9] Endogenous G-CSF might play role in mediating neutrophil migration to the dermis. In patients of inflammatory bowel disease, Sweet syndrome is more frequently seen with colonic disease and it is more common in females. Idiopathic Sweet Syndrome can present as photo-dermatoses.[10]

Treatment of Sweet syndrome is the immunosuppression mainly with systemic steroids. Other drugs that have been tried like Cyclosporin, indomethacin, dapsone, colchicine and clofazimine[11] also been reported for successful treatment. There have been few case reports of spontaneous remission of Sweet syndrome.

What's new

Sweet syndrome is one of the rare cutaneous association of ulcerative colitis. Skin lesions mostly appear during the periods of acute exacerbation of the disease and respond to steroid treatment.

References

1. Sweet RD. An acute febrile neutrophilic dermatosis. *Br J Dermatol* 1964; 76:349-56.
2. Kalmus Y, Kovatz S, Shilo L, Ganem G, Shenkman L. Sweet's syndromen and subacute thyroiditis. *Postgrad Med J* 2000; 76:229-230.
3. Cohen PR, Kurzrock R. Sweet's syndrome revisited: A review of disease concepts. *Int J Dermatol* 2003; 42:761-78.
4. Gupta SK, Bajpai M, Uraiya D. Diclofenac-induced sweet's syndrome. *Indian J Dermatol* 2015; 60:424.
5. Carpels W, Mattelaer C, Geboes K, Coremans G, Tack J. Sweet's syndrome in a patient with Crohn's disease. *Acta Gastroenterol Belg* 1999; 62:372-374.
6. Hommel L, Harms M, Saurat JH. The incidence of Sweet's syndrome in Geneva. A retrospective study of 29 cases. *Dermatol* 1993; 187:303-305.
7. Ginarte M, Garcia I, Toribio J. Sweet's syndrome: a study of 16 cases. *Med Clin (Barc.)* 1997; 109:588-591.
8. Lipp KE, Shenefelt PD, Nelson RP, Messina JL, Fenske NA. Persistent Sweet's syndrome occuring in a child with a primary immunodeficiency. *Am Acad Dermatol* 1999; 40:838-841.
9. Arbetter KR, Hubbard KW, Markovic SN, Gibson LE, Phyliky RL. Case of granulocyte colony-stimulating factor induced Sweet's syndrome. *Am J Hematol* 1999; 61:126-129.
10. Verma R, Vasudevan B, Pragasam V, Mitra D. Unusual Presentation of Idiopathic Sweet's Syndrome in a Photodistributed Pattern. *Indian J Dermatol* 2014; 59:186–189.
11. Burrall, B. Sweet's syndrome (acute febrile neutrophilic dermatosis). *Dermatol Online J* 1999; 5:8.
12. Rahier JF, Lion L, Dewit O, Lambert M. Regression of Sweet's syndrome associated with Crohn's disease after anti-Tumour Necrosis Factor therapy. *Acta Gastroenterol Belg* 2005; 68:376-9.