Neutrophilic dermatosis of dorsal hands

S. Kaur, D. Gupta[1], B. Garg[2], N. Sood[2]

Departments of Dermatology and Venereology, [1]Internal Medicine and [2]Pathology, Dayanand Medical College and Hospital, Ludhiana, Punjab, India

ABSTRACT

Sweet's syndrome is characterized by erythematous tender nodules and plaques over face and extremities. Fever, leukocytosis with neutrophilia, and a neutrophilic infiltrate in the dermis are characteristic features. Neutrophilic dermatosis of dorsal hands is a rare localized variant of Sweet's syndrome occurring predominantly over dorsa of hands. Various degrees of vascular damage may be observed on histopathology of these lesions. Both Sweet's syndrome and its dorsal hand variant have been reported in association with malignancies, inflammatory bowel diseases, and drugs. We report a patient with neutrophilic dermatoses of dorsal hands associated with erythema nodosum. He showed an excellent response to corticosteroids and dapsone.

Key words: Neutrophilic dermatosis of dorsal hands, Sweet's syndrome, vasculitis

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.148940

Quick Response Code:

Address for correspondence:
Dr. Sukhjot Kaur, Department of Dermatology and Venereology, Dayanand Medical College and Hospital, Ludhiana - 141 001, Punjab, India.
E-mail: sukhjotgoraya@gmail.com

INTRODUCTION

Sweet's syndrome, originally described by Dr. Robert Douglas Sweet in 1964, is characterized by an acute onset of fever, leukocytosis, and tender erythematous plaques infiltrated by mature neutrophils.[1] The condition may be idiopathic or may occur after nonspecific respiratory or gastrointestinal tract infection.[2] Lesions usually involve the face and extremities and show an excellent response to corticosteroids.[2] Recently, several new variants have been described including neutrophilic dermatosis of the dorsal hands (NDDH), histiocytoid Sweet's syndrome, Sweet's syndrome associated leukemia cutis, neutrophilic panniculitis, and necrotizing Sweet's syndrome.[2,3]

Neutrophilic dermatosis of the dorsal hands is rare, localized variant of Sweet's syndrome that was originally described in 1996.[4,5] Skin lesions present as pustular nodules or boggy red plaques with ulceration over dorsa of hands.[4,5] NDDH is often initially misdiagnosed as an infective disorder, and may result in inappropriate antibiotic treatment, surgical debridement, and even amputation.[6] Prompt clinical recognition and treatment with corticosteroids is helpful to avoid unnecessary medical or surgical therapy, and to exclude associated diseases. We report a patient with NDDH with a brief review of the condition and its pathologic features.

CASE REPORT

A 50-year-old male was admitted with sudden onset of markedly painful erythematous nodules on bilateral lower limbs since 3 days. There was a history of generalized weakness and malaise but no complaints of fever, loose stools, joint pains, or sore throat. On the second day of admission, an erythematous plaque was observed over the dorsal aspect of the right hand and radial aspect of the index finger. These lesions gradually progressed, and similar plaques subsequently developed over the index finger of the left hand. He was on telmisartan and metformin for hypertension and diabetes mellitus, respectively, since five years. Examination revealed multiple tender, erythematous nodules over the anterior aspect of both lower legs and knees [Figure 1]. There was a large, erythematous, boggy, tender plaque on the dorsum of the right hand extending over to the dorsum of right index finger [Figure 2]. There were large bullous and pustular lesions on an erythematous base over the radial aspect of right index finger [Figure 3]. General physical and systemic examination was normal.

Investigations revealed a total leukocyte count of 12.4 × 10^9/L with 61% neutrophils, 30% lymphocytes. His erythrocyte sedimentation rate was 65 mm in first hour, and C-reactive protein was 104 mg/L (0–6 mg/L). Creatine

kinase, rheumatoid factor, and antinuclear antibody were negative. His urine analysis, serum electrolytes, liver and renal function tests, and thyroid profile were normal. Serology for HIV, HBs Ag, and hepatitis C virus (HCV) was nonreactive. No abnormality was detected on chest X-ray and electrocardiogram. Blood cultures and pus culture from the finger lesion showed no growth. A skin biopsy taken from the hand lesion revealed a dense infiltrate of mature neutrophils in the papillary dermis associated with leukocytoclastic debris. Within this area, vessel wall dilatation and fibrinoid necrosis were also seen. Few inflammatory cells were seen infiltrating the vessel wall; however, there was no extravasation of erythrocytes [Figures 4 and 5]. No organisms were identified on hemotoxylin and eosin stain. Microbiologic cultures were negative. Based on the clinicopathologic features, diagnosis of neutrophilic dermatoses of dorsal hands was made. The patient did not consent for the skin biopsy from the leg lesions; these were presumptively diagnosed as erythema nodosum. During his hospital course, papulopustular lesions were seen to develop at sites of venepunture which were interpreted clinically as positive pathergy phenomena. Treatment with prednisolone 1 mg/kg/day was initiated, and remarkable improvement was observed over the next few days. The leg lesions were observed to become ecchymotic and then yellowish before fading. Corticosteroids were gradually tapered and dapsone 100 mg/day was initiated. Complete resolution of all lesions was observed over the next four weeks. Dapsone was safely continued, with no recurrence over a follow-up of 10 months.

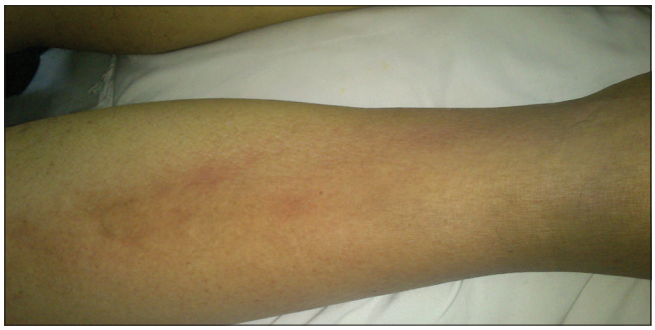

Figure 1: Erythematous nodules over leg

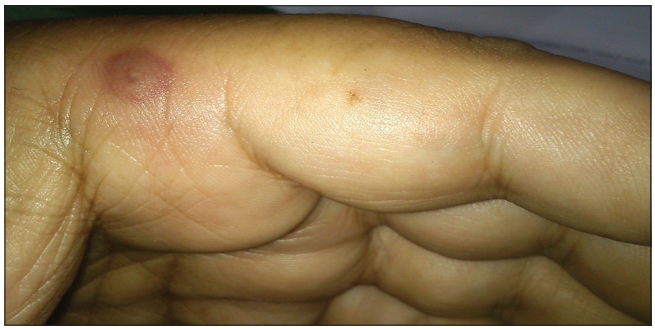

Figure 3: Targetoid erythematous nodule on the radial aspect of finger

DISCUSSION

Neutrophilic dermatosis of dorsal hands is a recently described, localized variant of Sweet's syndrome. Strutton *et al*. in 1996 reported six females presenting with violaceous papulonodules on the radial aspect of the dorsal hands with a histopathologic picture of neutrophilic dermatosis and leuckocytoclastic vasculitis showing good response to steroid therapy. They proposed the term pustular vasculitis of the hands for these lesions.[4] Subsequently, in 2000, Galaria *et al*. described three clinically and histologically similar cases limited to the hands, but without systemic symptoms having a therapeutic response similar to that of Sweet's syndrome.[5] The lesions lacked a true vasculitis and were limited to hands, and thus they proposed the term NDDH for these lesions.[5] Several case series of similar patients with lesions predominantly on hands showing a variable degree of vascular endothelial damage were subsequently reported.[7,8] Violaceous edematous plaques, ulcerated plaques with violaceous undermined borders, bullous, hemorrhagic, and ulcerated lesions on dorsal hands, necrotic pyoderma like lesions with pseudovesiculation, and atypical pyoderma gangrenosum like lesions with hemorrhagic bullous

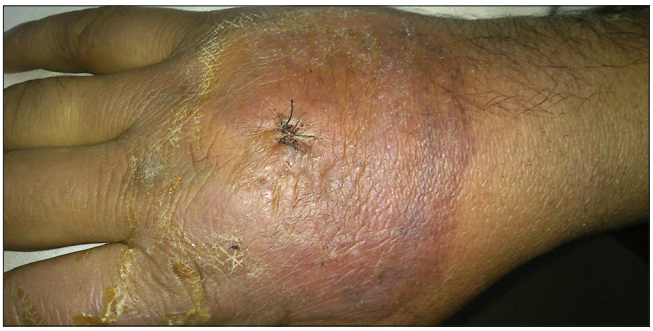

Figure 2: Boggy erythematous plaque over dorsum of the hand, with sutures at site of skin biopsy in the center

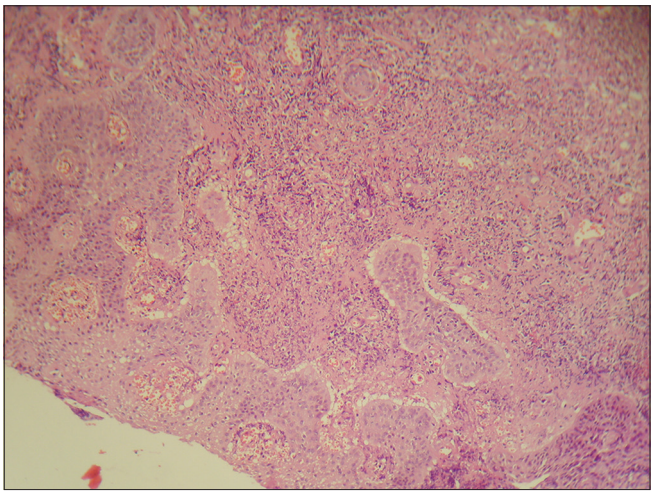

Figure 4: Histopathology of a skin biopsy showing dermal edema and a dense neutrophilic infiltrate in the dermis (H and E, ×40)

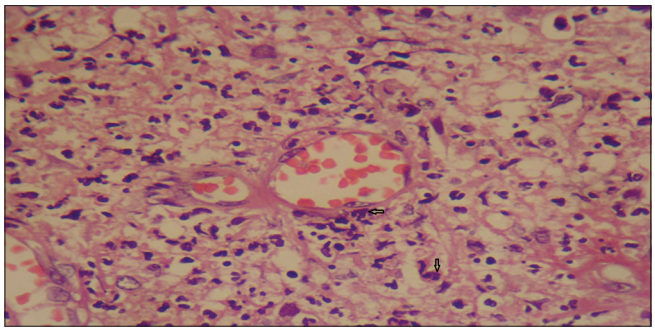

Figure 5: Section from the skin showing dense neutrophilic infiltrate (vertical arrow), along with vasculitis (horizontal arrow) (H and E, ×100)

lesions are the diverse morphologic patterns observed in patients with NDDH.[5-9]

Histopathology of lesions of NDDH reveals a dense dermal infiltrate of neutrophils usually associated with subepidermal edema and leukocytoclastic debris.[5,6,7] A number of reports describe the evidence of vasculitis in these patients presenting with erythematous nodules limited to the dorsal hands.[7] According to Malone *et al*., the vascular damage in these cases was probably a secondary event related to the intensity of the neutrophilic infiltrate and the time of evolution of the lesions and did not represent true vasculitis.[10] Cohen referred to the vasculitis seen in skin lesions of Sweet's syndrome and its dorsal hand variant as an epiphenomenon.[11] The term pustular vasculitis of hands was revised after recognizing that vasculitis was an inconsistent finding and of secondary importance in its diagnosis.[5]

Diagnosis and treatment of NDDH is similar to that of Sweet's syndrome. Infectious causes should be ruled out by special stains and cultures. Other neutrophilic dermatoses such as pyoderma gangrenosum, pustular drug reactions, rheumatoid neutrophilic dermatosis, bowel associated dermatosis arthritis syndrome, and erythema elevatum diutinum need to be considered in the differential diagnosis.[6,7]

Sweet's syndrome has been linked to various drugs, inflammatory bowel disease, and several solid tumors and hematologic malignancies.[2,3,9] Similarly, a number of important associations have also been observed in patients of NDDH including myelodysplasia, leukemia, inflammatory bowel disease, seropositive arthritis, sarcoidosis, lymphoma, lenalidomide, thalidomide, vaccinations, fertilizer exposure and HCV infection.[6,7]

Rarely Sweet's syndrome and erythema nodosum may occur simultaneously in the same individual.[12] The simultaneous presence of the two reactive dermatoses in the same patient is intriguing. It is not clear whether erythema nodosum-like lesions accompanying Sweet's syndrome are a true erythema nodosum or a manifestation of Sweet's syndrome itself.[13] These two entities may be different clinical manifestations of a common underlying mechanism in different anatomic locations.[13] A subcutaneous Sweet's syndrome has also been proposed recently.[13] Formation of immune complexes has been proposed as the initiating factor for both disorders.[12] Cohen *et al*. hypothesized that the causative agent of Sweet's syndrome or erythema nodosum may stimulate the production of various cytokines such as interleukin (IL-1), IL-8 or granulocyte colony-stimulating factor, leading to the lesions of Sweet's syndrome if they were produced in the dermis or erythema nodosum if they were produced in the subcutaneous tissue.[12]

Our patient presented with development of typical lesions of NDDH over hands and possible erythema nodosum over the legs. Limitation of the current report is a lack of histopathological confirmation of clinical lesions of erythema nodosum over the legs. Though erythema nodosum is mostly diagnosed clinically, biopsy is often required to support the diagnosis.[14] Leukocytosis and neutrophilia are commonly observed on laboratory evaluation; however, they may not always be present, as in the current case.[9]

Prompt clinical recognition and appropriate treatment of this rare and localized variant of Sweet's syndrome is essential to improve outcomes and to prevent unnecessary surgical intervention.

REFERENCES

1. Sweet RD. An Acute febrile neutrophilic dermatosis. Br J Dermatol 1964;76:349-56.
2. Anzalone CL, Cohen PR. Acute febrile neutrophilic dermatosis (Sweet's syndrome). Curr Opin Hematol 2013;20:26-35.
3. Neoh CY, Tan AW, Ng SK. Sweet's syndrome: A spectrum of unusual clinical presentations and associations. Br J Dermatol 2007;156:480-5.
4. Strutton G, Weedon D, Robertson I. Pustular vasculitis of the hands. J Am Acad Dermatol 1995;32:192-8.
5. Galaria NA, Junkins-Hopkins JM, Kligman D, James WD. Neutrophilic dermatosis of the dorsal hands: Pustular vasculitis revisited. J Am Acad Dermatol 2000;43:870-4.
6. Cook-Norris RH, Youse JS, Gibson LE. Neutrophilic dermatosis of the hands: An underrecognized hematological condition that may result in unnecessary surgery. Am J Hematol 2009;84:60-1.
7. Del Pozo J, Sacristán F, Martínez W, Paradela S, Fernández-Jorge B, Fonseca E. Neutrophilic dermatosis of the hands: Presentation of eight cases and review of the literature. J Dermatol 2007;34:243-7.
8. Walling HW, Snipes CJ, Gerami P, Piette WW. The relationship between neutrophilic dermatosis of the dorsal hands and Sweet syndrome: Report of 9 cases and comparison to atypical pyoderma gangrenosum. Arch Dermatol 2006;142:57-63.
9. Cohen PR. Sweet's syndrome – a comprehensive review of an acute febrile neutrophilic dermatosis. Orphanet J Rare Dis 2007;2:34.
10. Malone JC, Slone SP, Wills-Frank LA, Fearneyhough PK, Lear SC, Goldsmith LJ, *et al*. Vascular inflammation (vasculitis) in Sweet syndrome: A clinicopathologic study of 28 biopsy specimens from 21 patients. Arch Dermatol 2002;138:345-9.
11. Cohen PR. Skin lesions of Sweet syndrome and its dorsal hand variant contain vasculitis: An oxymoron or an epiphenomenon? Arch Dermatol 2002;138:400-3.

12. Waltz KM, Long D, Marks JG Jr, Billingsley EM. Sweet's syndrome and erythema nodosum: The simultaneous occurrence of 2 reactive dermatoses. Arch Dermatol 1999;135:62-6.
13. Tabanlioglu D, Boztepe G, Erkin G, Gököz O, Karaduman A. Sweet's syndrome and erythema nodosum: A companionship or a spectrum? – a case report with review of the literature. Int J Dermatol 2010;49:62-6.
14. Cox NH, Jorizzo JL, Bourke JF, Savage CO. Vasculitis, Neutrophilic dermatoses and related disorders. In: Burns T, Breathnach H, Cox N, Griffiths C, editors. Rook's Textbook of Dermatology. 8th ed. Oxford: Wiley-Blackwell; 2010. p. 50.1-90.

Cite this article as: Kaur S, Gupta D, Garg B, Sood N. Neutrophilic dermatosis of dorsal hands. Indian Dermatol Online J 2015;6:42-5.

Source of Support: Nil, **Conflict of Interest:** None declared.