Case Report

A rare co-occurrence of Sweet's syndrome and leukemia cutis as a presenting feature of B-cell chronic lymphoid leukemia: Probably a first reported case

Siddheshwar Virbhadraappa Birajdar, Sheshrao Sakharam Chavan, Sanjay Arun Mundhe, Ashish Kumar Jain

Department of General Medicine, S.R.T.R. Government Medical College and Hospital, Ambajogai, Maharashtra, India

ABSTRACT

Sweet's syndrome (SS), also known as acute febrile neutrophilic dermatosis, is characterized by tender, red inflammatory papules or nodules with fever that occur in association with infection, malignancy, connective tissue disease, or drug exposure. Malignancy-related SS has been described mainly in patients with acute myeloblastic leukemia. Leukemia cutis (LC) is the infiltration of neoplastic leukocytes or their precursors into epidermis, dermis, or subcutis, resulting in clinically identifiable cutaneous lesions. LC and its co-occurrence with SS have been described as presenting feature of acute myeloblastic leukemia. There are only few case reports describing SS in patients of chronic lymphocytic leukemia (CLL). However, through literature search, we could not find out cases describing co-occurrence of SS and LC in patients of CLL. Here, we report a rare co-occurrence of SS with LC in case of B-cell CLL, probably first reported case.

Key words: Chronic lymphocytic leukemia, hematological malignancy, leukemia cutis, neutrophilic dermatosis, Sweet's syndrome

INTRODUCTION

Sweet's syndrome (SS) is characterized by tender, erythematous papules or nodules with fever occurring in association with infection, malignancy, connective tissue disease, or drug exposure.[1] SS can be a sentinel cutaneous sign of undiagnosed malignancy or sign of cancer recurrence.[2] Rarely, SS can concurrently demonstrate leukemia cutis (LC) where dermal infiltrate consists of mature neutrophils (SS) and leukemic cells (LC).[3] SS is rare in chronic lymphocytic leukemia (CLL), and few cases are reported so far.[4,5] Although co-occurrence of SS and LC is reported with acute myeloblastic leukemia, we could not find such reported cases with CLL.

Address for correspondence: Dr. Ashish Kumar Jain, Department of General Medicine, S.R.T.R. Government Medical College and Hospital, Ambajogai - 431 517, Maharashtra, India.
E-mail: dr.ashish31.jain@gmail.com

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CASE REPORT

A 60-year-old woman was admitted to our hospital with fever, joint pains, fatigue and multiple painful reddish lesions over face, neck, trunk, and palms of 1-week duration. The patient had no significant past medical history and was not using any drugs or herbal products prior or during illness. Examination of the patient revealed multiple erythematous, tender papulonodular lesions over face, back, and chest along with pustular lesions over palms and multiple enlarged lymph nodes in neck which were firm, mobile, and nontender [Figure 1]. Physical examination revealed hepatosplenomegaly along with pallor. Peripheral smear showed total leucocyte count - 90,000/cumm with prolymphocytes 5%, clefted and atypical lymphocytes 65%, mature

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lymphocytes 15%, and polymorphs 15% with reduced platelets and plenty of smudge cells s/o CLL [Figure 2]. Skin biopsy of the lesions showed predominantly neutrophilic infiltration in reticular dermis and adjacent epidermis forming pustules along with linear and focally nodular aggregate of small lymphoid cells with coarse clumped chromatin interspersed with prolymphocytes and exhibiting immunoreactivity for CD5/CD20/CD23 and immunonegative for Tdt/CD10/Bcl6/cyclin D1/CD43 and immunopositivity for ki67 seen mainly in prolymphocytes and ki67 proliferation rate of approximately 30% suggestive of LC with neutrophilic dermatosis [Figures 3 and 4]. Flow cytometry of peripheral blood confirmed the diagnosis of B-cell CLL. The diagnosis of SS was made considering clinical picture of fever with typical skin lesions having neutrophilic dermatosis on skin biopsy in the presence of CLL and other supportive laboratory findings. The patient was started on steroid and skin lesions responded to 2 weeks of prednisolone therapy.

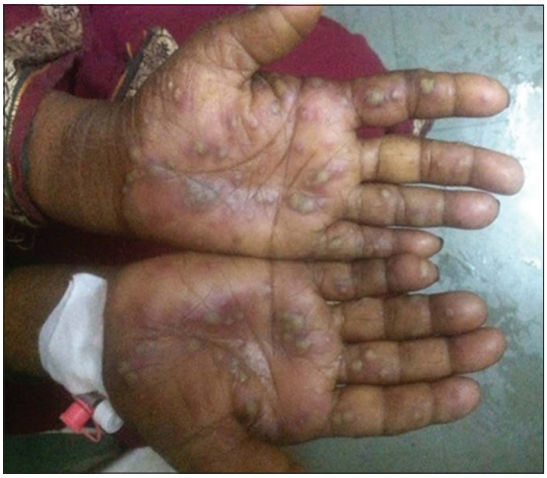

Figure 1: Multiple erythematous papulonodular and pustular lesions over both palms

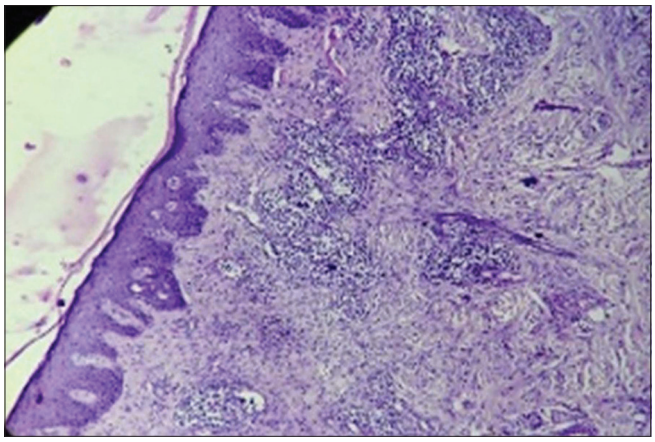

Figure 3: Reticular dermis and epidermis shows neutrophilic infiltration (pustules) with aggregates of lymphoid cells in dermis (H and E, ×40)

DISCUSSION

In 1964, Robert D. Sweet described a new entity called "acute febrile neutrophilic dermatosis" with cardinal features of fever, leukocytosis, tender erythematous skin papules, nodules or plaques with biopsy of these skin lesions showing dense, neutrophilic dermal infiltrate into the upper or papillary dermis.[1] Dr. Sweet himself preferred the disease be called as Gomm-Button disease in honor of the first two patients afflicted with the condition

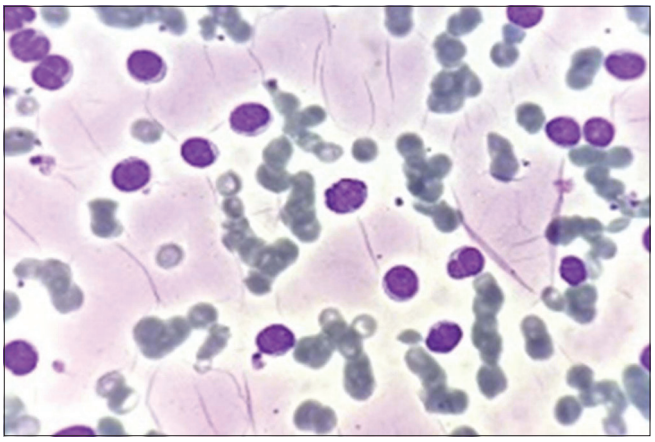

Figure 2: Peripheral smear shows raised thin layer chromatography with prolymphocytes, clefted atypical lymphocytes, polymorphs with smudge cells (Leishmann's stain, ×100)

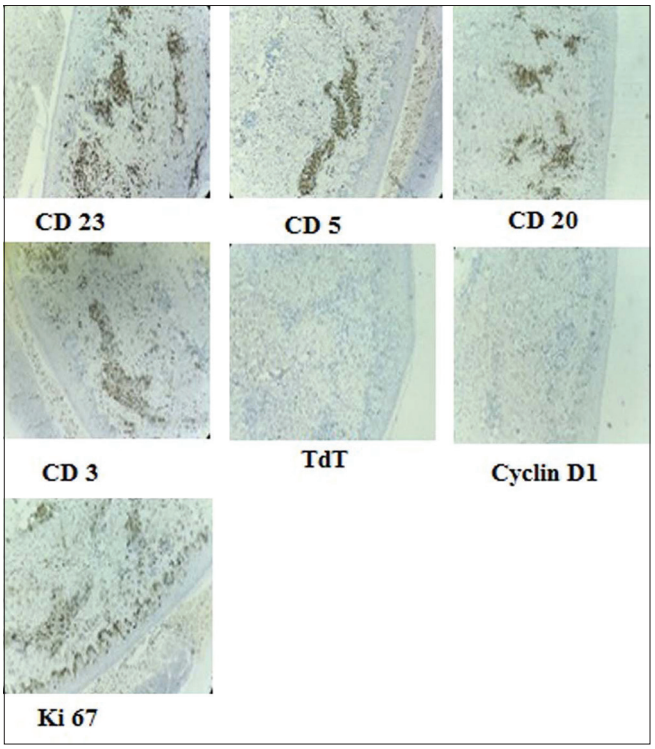

Figure 4: Immunohistochemistry of skin biopsy shows lymphoid cells immunopositive for CD20, CD5, CD23, CD3, and immunonegative for CD10, Bcl 6, cyclin D1, CD43, TdT

in Dr. Sweet's practice.[1] However with time, the eponym "SS" is being increasingly used to describe this condition. However, manifestations of SS are not limited to the skin, and various extracutaneous manifestations of SS have been described.[6] SS has been described in three clinical settings: classical or idiopathic SS, malignancy-associated SS and drug-induced SS.[6] LC is the infiltration of neoplastic leukocytes or their precursors into the epidermis, the dermis, or the subcutis, resulting in clinically identifiable cutaneous lesions. Less commonly, it may occur as a sole presenting feature in the absence of simultaneous marrow involvement.[7] Malignancy-related SS commonly accompanies myeloid malignancies. In 1993, a review of 15 studies of patients with SS found that ~21% of patients newly diagnosed with SS were subsequently diagnosed or already diagnosed with either a hematologic (15%) or solid cancer (6%).[3] Approximately, 85% of reported cases of malignancy-associated SS had underlying hematological malignancies, most commonly acute myeloblastic leukemia. Other hematological malignancies include myeloproliferative neoplasm, diffuse large B-cell lymphoma, Hodgkin's lymphoma, myelodysplastic syndrome, and myelofibrosis.[3] The most common solid malignancies reported with SS are adenocarcinomas (57%) of the genitourinary organs, breast, and gastrointestinal tract.[2,3,8] LC is a relatively rare condition and may manifest in a variety of leukemia subtypes. Leukemia cutis usually presents concomitantly with systemic leukemia or after diagnosis of leukemia. While acute monocytic, myelomonocytic, and the T-cell leukemia show the highest incidence (50%–70%) of LC, it is an uncommon manifestation of chronic myelocytic leukemia (2%–8%), and when present points towards blastic transformation of the disease.[9] Only a small number of patients with SS can also present with skin lesions concurrently demonstrating LC.[3] Acute myelocytic leukemia and acute promyelocytic leukemia are the most frequent hematological malignancies associated with concurrent LC and SS.[2,3,8] Small lymphocytic lymphoma/CLL (SLL/CLL) is a relatively rare entity accompanying SS, and a few cases have been reported so far.[4,5] Although the co-occurrence of SS and LC has been reported with acute myeloblastic leukemia and we could not find any description of such co-occurrence in patients of CLL. Our patient presented to us with the dermatological and clinical features of SS and diagnosis was made as per the modified diagnostic criteria for malignancy associated SS described by von den Driesch[3,10] and was subsequently diagnosed to have B-cell CLL considering peripheral blood picture and immunophenotyping of blood and LC by demonstrating the lymphoid precursor cells in dermis with immunohistochemistry. Our patient responded to 2 weeks of prednisolone therapy.

CONCLUSION

Sweet's syndrome is rare in patient with SLL/CLL along with leukemia cutis. It should be kept in mind that this syndrome and leukemia cutis can occur together in patients with CLL. This report emphasizes the co-occurrence of SS with leukemia cutis in chronic lymphocytic leukemia as with acute myeloblastic leukemia.

Acknowledgment

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Sweet RD. AN Acute febrile neutrophilic dermatosis. Br J Dermatol 1964;76:349-56.
2. Paydas S. Sweet's syndrome: A revisit for haematologists and oncologists. Crit Rev Oncol Hematol 2013;86:85-95.
3. Raza S, Kirkland RS, Patel AA, Shortridge JR, Freter C. Insight into Sweet's syndrome and associated-malignancy: A review of the current literature. Int J Oncol 2013;42:1516-22.
4. Cholongitas E, Pipili C, Dasenaki M, Kaklamanis L. Piperacillin/tazobactam-induced Sweet syndrome in a patient with chronic lymphocytic leukaemia and autoimmune cholangitis. Am J Dermatopathol 2008;30:203-4.
5. Thompson MA, Dyson SW, Faderl S. Sweet's syndrome in chronic lymphocytic leukaemia associated with neutropenic fever and granulocyte colony stimulation factor. Am J Hematol 2006;81:703-5.
6. Cohen PR. Sweet's syndrome – A comprehensive review of an acute febrile neutrophilic dermatosis. Orphanet J Rare Dis 2007;2:34.
7. Singhal M, Singh S, Kumar R, Raina V. Extensive cutaneous manifestations: Presenting feature of chronic myelocytic leukemia in second blast crisis. Indian J Dermatol 2010;55:265-7.
8. Paydas S, Sahin B, Zorludemir S. Sweet's syndrome accompanying leukaemia: Seven cases and review of the literature. Leuk Res 2000;24:83-6.
9. Cho-Vega JH, Medeiros LJ, Prieto VG, Vega F. Leukemia cutis. Am J Clin Pathol 2008;129:130-42.
10. von den Driesch P. Sweet's syndrome (acute febrile neutrophilic dermatosis) J Am Acad Dermatol 1994;31:535-56.