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Title

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Journal

Dermatology Online Journal, 31(3)

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Publication Date

2025

DOI

10.5070/D331365360

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Peer reviewed

Case Report

Histiocytoid Sweet syndrome-like presentation of mature plasmacytoid dendritic cell proliferation

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Keywords: dermatology, histiocytoid, mimic, pathology

Dermatology Online Journal

Vol. 31, Issue 3, 2025

Abstract

Mature plasmacytoid dendritic cell proliferation is a condition associated with myeloid neoplasms, most commonly chronic myelomonocytic leukemia. Plasmacytoid dendritic cells can resemble lymphocytes and histiocytes morphologically and immunophenotypically. Mature plasmacytoid dendritic cell proliferation may therefore go unrecognized if the diagnosis is not suspected and appropriate stains for plasmacytoid dendritic cells are not performed. Herein, we present a case of mature plasmacytoid dendritic cell proliferation masquerading clinically and histologically as histiocytoid Sweet syndrome. The patient, who had previously been diagnosed with mature plasmacytoid dendritic cell proliferation that presented as pink, edematous, pruritic papules and plaques, had initially resolved following induction chemotherapy for acute myelomonocytic leukemia. However, he presented later with indurated purpuric plaques on the trunk within weeks of receiving filgrastim for neutropenia. Biopsies demonstrated marked dermal edema, interstitial, superficial, and deep infiltrate with histiocytoid appearing cells concerning for histiocytoid Sweet syndrome. Further work-up demonstrated that the infiltrate was predominantly composed of CD3-, CD4+, CD34-, CD123+, CD56-, CD68-, myeloperoxidase negative mononuclear cells consistent with mature plasmacytoid dendritic cell proliferation. This case demonstrates that MPDCP

should be considered in the differential diagnosis of eruptions that clinically and histologically look like histiocytoid Sweet syndrome but stain negatively for myeloperoxidase.

Introduction

Mature plasmacytoid dendritic cell proliferation (MPDCP) is a condition associated with myeloid neoplasms, most commonly chronic myelomonocytic leukemia and less commonly acute leukemia with monocytic differentiation and myelodysplasia.¹ MPDCPs share clonality with the associated myeloid neoplasms and portend a poor prognosis.¹ MPDCP can present as a pink maculopapular eruption with pruritus and lymphadenopathy.^{1,2} Plasmacytoid dendritic cells (pDCs) can resemble lymphocytes and histiocytes morphologically and immunophenotypically. MPDCP may therefore go unrecognized if the diagnosis is not suspected and appropriate stains for pDCs are not performed. Herein, we present a case of MPDCP masquerading clinically and histologically as histiocytoid Sweet syndrome.

Case Synopsis

A 61-year-old man with no significant medical history presented with a one-year history of an intermittent pruritic rash affecting the chest. The eruption occurred four times over the prior year with spontaneous resolution. Two months prior to presentation, the eruption returned, spreading throughout the abdomen, back, bilat-

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Figure 1. Clinical photographs demonstrating numerous light pink papules and plaques throughout **A)** the back, **B)** chest, and **C)** the face.

eral upper extremities, neck, and cheeks. Cutaneous examination was notable for widespread, light pink, edematous papules and plaques in the involved areas. In the following months, multiple biopsies demonstrated a dense superficial and deep perivascular mononuclear cell infiltrate involving adnexal structures, with a predominance of $CD4+$ cells. An antinuclear antibody test was negative. Six months later, the patient returned after noting increased itching and crusting of lesions on his face along with chills, and low-grade fevers. On examination, lesions demonstrated similar morphology as above but with new central hemorrhagic crusting (**Figure 1A-C**). Laboratory evaluation was notable for a leukocytosis of 18,000 WBC/ml with myeloblasts on the differential. Bone marrow biopsy was consistent with acute myelomonocytic leukemia with flow cytometry showing a population of myeloblasts expressing partial $CD4$, $CD13$, $CD33$, $CD34$, and $CD117$. Repeat skin biopsies demonstrated similar histologic findings as before, but the infiltrate included a dominant population of $CD3-$, $CD4+$, $CD34-$, $CD123+$ (>75%), $CD56-$, lysozyme-, myeloperoxidase- mononuclear cells consistent with MPDCP. The patient's skin began clearing soon after induction chemotherapy, but a few weeks later, after being treated with filgrastim for neutropenia, he developed new indurated purpuric plaques on the trunk (**Figure 2**). Biopsy demonstrated sparing of the epidermis, marked papillary dermal edema, and a dense, bandlike, interstitial infiltrate in the superficial and deep dermis composed of histiocytoid appearing cells consistent with the pathology findings of histiocytoid Sweet syndrome (**Figure 3A-B**).³ Notably, there was also the absence of histologic blastic features such as prominent nucleoli and scant cytoplasm. Given the recent diagnosis of acute myelomonocytic leukemia and MPDCP, additional stains were performed. Immunophenotypic studies showed a preponderance of $CD3-$, $CD4+$, $CD34-$, $CD123+$, $CD56-$, $CD68-$, lysozyme-, myeloperoxidase- mononuclear cells, again consistent with the diagnosis of MPDCP (**Figure 4A-E**).



Figure 2. Clinical photograph demonstrating new indurated purpuric plaques on the trunk.

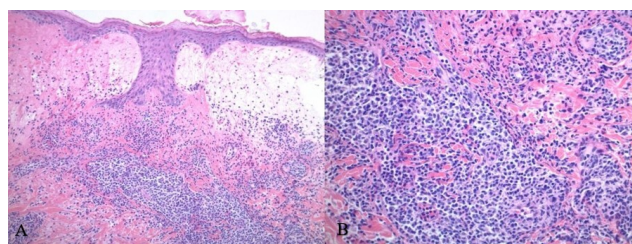


Figure 3. Skin biopsy demonstrating aggregates of cells with large, epithelioid, discohesive cells with abundant amphophilic cytoplasm noted in a perivascular and interstitial distribution in the superficial and deep dermis with papillary dermal edema as well as plasmacytoid cells with eccentric, heterochromatic nuclei (black arrows). Hematoxylin-eosin stain, original magnification **A)** 50X; **B)** 400X.

Case Discussion

Diagnosing MPDCP involves a high index of suspicion. MPDCP is a low-grade clonal proliferation of mature pDCs.⁴ In humans, pDCs derive from monocyte progenitor cells.¹ Given the shared clonality of MPDCP and

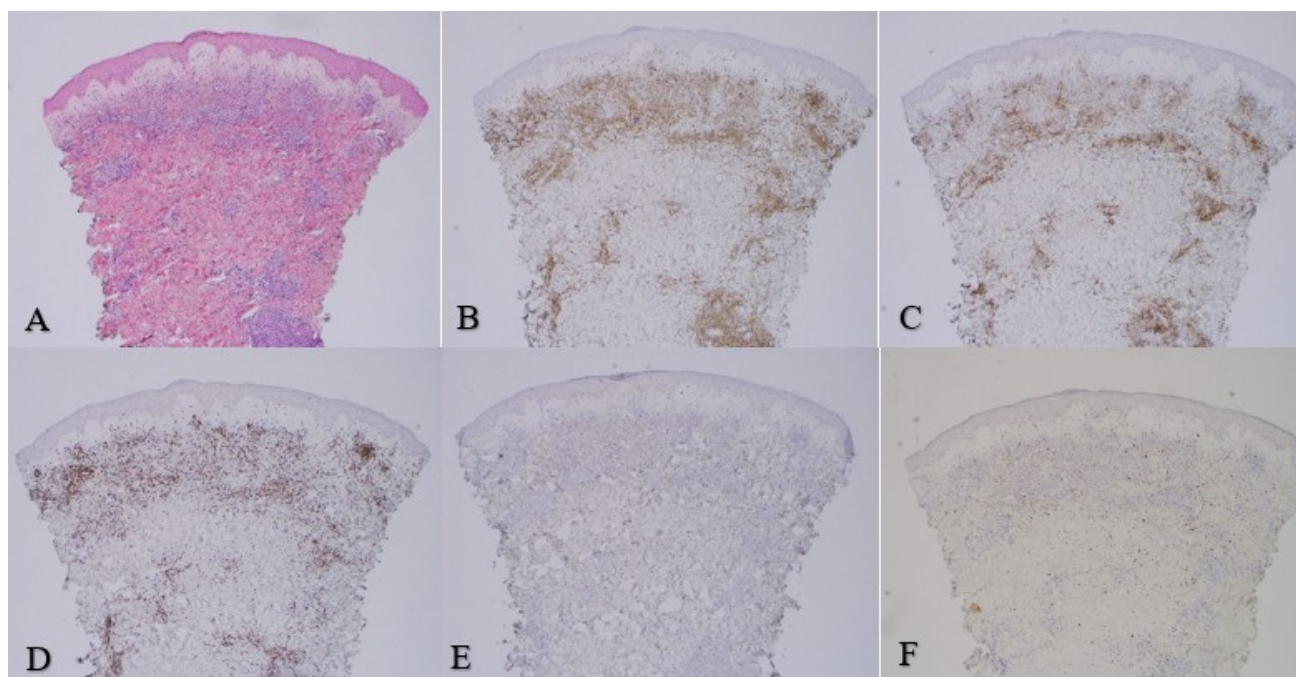


Figure 4. Skin biopsy demonstrating marked papillary dermal edema and a dense, bandlike, interstitial infiltrate in the superficial and deep dermis on **A)** hematoxylin-eosin stain, original magnification 4x. Staining was positive for **B)** CD4 and **C)** CD123, and negative for **D)** CD3 (with some background T-cell staining), **E)** MPO, and **F)** CD68.

the underlying neoplasm, MPDCP likely originates from the differentiation of the underlying neoplastic cells.^{1,5} Morphologically, pDCs can resemble histiocytes and lymphocytes, with recognition relying on immunophenotyping studies. The most reliable markers are *CD123*, *CD303*, and T-cell Leukemia/Lymphoma 1(*TCL1*).¹ Mature pDCs express the monocytic markers *CD4*, *CD15*, and *CD68* but are negative for lineage-specific T-cell and B-cell markers.¹ They differ morphologically and immunophenotypically from blastic plasmacytoid dendritic cell neoplasm (BPDCN), a type of acute leukemia in which the cells appear immature (blastic) and are typically positive for *CD4* and *CD56*. BPDCN is also positive for the blast markers *CD38* and HLA-DR and variable for *CD34* and terminal deoxynucleotidyl transferase.¹ Several prior case reports have documented MPDCP clinically and histologically mimicking lupus erythematosus, in which *CD4*+ pDCs as well as clusters of lymphocytes were found in the superficial and deep dermis surrounding blood vessels and adnexal structures.⁶ In our case, the clinical and histological presentation overlapped with histiocytoid Sweet syndrome. Histiocytoid Sweet syndrome was first described in 2005.³ Histiocytoid Sweet syndrome lesions are comprised of immature myeloid cells that resemble histiocytes morphologically but can be differentiated by the expression of immature myeloid markers such as

myeloperoxidase.³ Unlike MPDCP, these cells do not share clonality with the underlying neoplasm.³ Cases of histiocytoid Sweet syndrome with weak-to-no myeloperoxidase positivity have been described, but it was not reported whether stains for markers of MPDCP were performed.³

Conclusion

This case demonstrates that MPDCP should be considered in the differential diagnosis of eruptions that clinically and histologically look like histiocytoid Sweet syndrome but are myeloperoxidase negative.

Potential conflicts of interest

Dr. Goldfarb has participated in clinical trials with Abbvie, Pfizer, Chemocentrix, and DeepX Health, and has served on advisory boards and consulted for Novartis and Boehringer Ingelheim. The content of this article is solely the responsibility of the authors and does not necessarily represent the official views of any other companies or organizations.

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