Patient with Blastic Plasmacytoid Dendritic Cell Neoplasm and CMML

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Patient with Blastic Plasmacytoid Dendritic Cell Neoplasm and CMML

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History of Present Illness: The patient presents with a six-month history of an evolving eruption. Initially, the patient had pink scaly lesions covering most of his body including his dorsal MCP joints but sparing his palms and soles, He also had redness on the forehead and lateral cheeks. This improved with an oral prednisone taper. As these lesions resolved, he developed palpable lymph nodes in the neck, axillae, and inguinal creases. He then developed blisters on his hands and feet. The blisters were very itchy and he also had generalized pruritus.

Medical History: Osteoarthritis

Medications: CoQ10, aspirin, allopurinol

Previous Treatments: Triamcinolone 0.1% ointment

Current Treatment: Augmented betamethasone diproprionate 0.05% cream, gabapentin 100mg daily, deceased before starting tagraxofusp

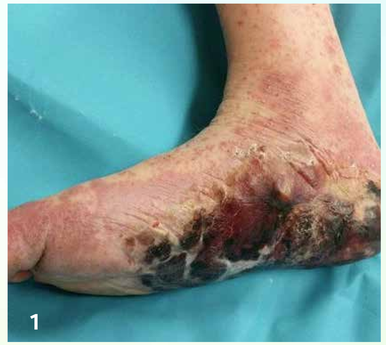
Physical Examination: Multiple violaceous vesicles and tense bullae on bilateral dorsal and palmar hands, plantar and medial feet. Several 1-2 cm palpable lymph nodes in the neck, axillae, and inguinal creases.

Laboratory Data: Uric acid 9.1 mg/dL (3.5-7.0), LDH 586 U/L (100-250), WBC 24.2 thou/cmm (4-10.5), platelets 44 thou/cmm (140-350), absolute neutrophils 16.2 thou/cmm (1.8-7.9), absolute monocytes 3.9 thou/cmm (0.2-1.4), absolute eosinophils 1.9 thou/cmm (0-0.7), absolute basophils 0.5 thou/cmm (0-0.2)

Studies: Tumoral analysis showed a stable microsatellite status with low tumor mutational burden. Atypical genetic mutations were found in KRAS, NRAS, TET2, ASXL1, RUNX1, and SRSF2.

Biopsy: *Memorial Sloan-Kettering Cancer Center* (H19-6023, 7/9/19 consult for AD19-06437 and AD19-3001) Left hand and left forearm: "Spongiotic dermatitis with clusters of plasmacytoid dendritic cells. PCR-TCR-negative."

Health Network Laboratories (S19-35688-A, B) Bone marrow aspirate: "Myeloid neoplasm with increased blasts (15%) and scattered plasmacytoid dendritic cells, consistent with CMML-2."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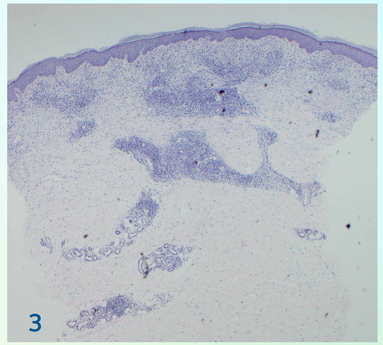
Figures 1 and 2: Multiple violaceous vesicles and eroded tense bullae within pink papules and plaques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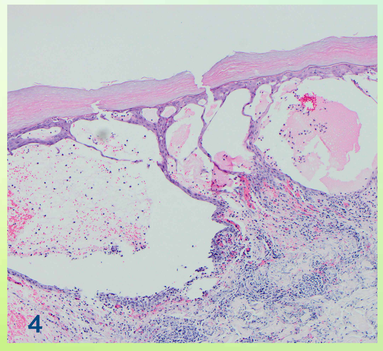
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Figures 3 and 4: Spongiotic dermatitis with clusters of plasmacytoid dendritic cells.

REFERENCES

[1] Zhang Y, et.al. Blastic plasmacytoid dendritic cell neoplasm: A case report and literature review. *Exp Ther Med* 2016;12:319-322.

[2] Sullivan J, Rizzieri D. Treatment of blastic plasmacytoid dendritic cell neoplasm. Hematology AM Soc *Hematol Educ Program* 2016;12:16-23.

[3] Jen E, et.al. FDA Approval Summary: Tagraxofusp-erzs for treatment of blastic plasmacytoid dendritic cell neoplasm. *Clin Cancer Res* 2019; doi: 10.1158/1078-0432.CCR-19-2329.

Diagnosis: Blastic Plamacytoid Dendritic Cell Neoplasm

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) was originally identified in 1994. It was originally classified with other acute myeloid leukemia-associated precursor neoplasms. However in 2016 was classified as its own category/entity. Although rare, BPDCN is most common in men in their 7th decade. It is an aggressive disease with a mean overall survival rate of 12–14 months.

Clinically, BPDCN manifests as violaceous macules, plaques, nodules, and occasionally bruise-like lesions. These may be single or multiple. Although it most commonly presents with cutaneous lesions, patients may develop subsequent lymph node, peripheral blood, and/or bone marrow involvement. It may be associated with thrombocytopenia, anemia, and/or leukocytosis.

Histopathologically, BPDCN shows diffuse infiltrates of monomorphic small-to-large-sized blastic cells with scant cytoplasm, round or oval nuclei with irregular contours, fine chromatin, and one to three small nucleoli. Immunohistochemical markers for the disease commonly stain positive for CD4, CD56, CD123 (almost universally), TCL1, and BDCA-2/CD303. The tumor should be negative for other B cell, T cell, and myeloid markers. However, some cases have demonstrated B cell marker overlap and negative CD4 and CD56, making BPDCN difficult to diagnose.

Due to the rarity of BPDCN, there is currently no standard treatment regimen. Historically, therapies and treatment protocols directed towards acute lymphoblastic leukemia and non-Hodgkin lymphoma have been used with a complete response rate ranging from 40-90%. However, relapse occurs in 50-90% of patients treated with this alone. There have been several variations of chemotherapy attempted with varied results and no clear best therapy. Hematopoietic stem cell transplants have also been used. This, when combined with chemotherapy, has shown the best long-term treatment response rates increasing the overall survival rate to 31.5 months. Recently, the FDA has approved tagraxofusp-erzs, a CD123 inhibitor, which has a response rate of 90% with unknown longevity. Many patients receiving tagraxofusp-erzs were bridged to stem cell transplant.

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