CLINICAL VIGNETTE

Blastic Plasmacytoid Dendritic Cell Neoplasm

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A 73-year-old Vietnam War veteran with a history of diabetes mellitus type 2, hypertension and chronic kidney disease stage 3 presented to his Primary Care Physician with "ringworm." Two weeks prior to presentation, the patient noticed non-pruritic crescent-shaped purplish lesions on his arm, shoulder and upper back (Figure 1; images provided by the patient), and within a week more lesions appeared on his chest and back (Figures 2 and 3). Review of the patient's laboratory tests was most notable for an approximately 50% decline in platelets within the preceding year, and development of giant thrombocytes and elevated lymphocytes, with no new medications started within the prior year. Additional laboratory tests showed a mild normocytic anemia and platelets stable around 110, 000 White blood cell count, basic metabolic panel and hepatic panel were within normal limits, except for a stable eGFR consistent with chronic kidney disease stage 3.

The patient felt some of the lesions started to resolve, but within a month, he returned with new lesions on his face which later started to coalesce and worsening of his "rash" (Figure 4). On exam, the patient had multiple non-tender, non-blanching, reddish-purplish macules and nodules of his face, and some enlarging lesions on his arms, chest and back. He also had bilateral non-tender enlarged cervical lymph nodes. He denied any fevers, weight loss, or any family history of leukemia, lymphoma and carcinoma.

Initial shave biopsy of a shoulder lesion showed granulomatous dermatitis, with negative stains for microorganisms, and CD68 immunohistochemistry positive with highlights for histiocytes within granulomas. A subsequent shave biopsy of a forehead lesion showed cutaneous lymphoma, most consistent with blastic plasmacytoid dendritic cell neoplasm (BPDCN), with strong BCL2 positivity. A bone marrow biopsy showed hypercellularity with scattered and small clusters of immature mononuclear cells co-expressing CD4, CD43, CD56 and CD123, consistent with BPDCN.

Given the patient's age and co-morbidities, he was not considered a bone marrow transplant candidate, and was referred to oncology for clinical trial evaluation.

Discussion

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare (less than 0.7 percent of primary cutaneous skin lymphomas), clinically aggressive hematologic malignancy, derived from plasmacytoid dendritic cells (type II dendritic cells).[1] It most commonly presents with cutaneous lesions, with or without bone marrow involvement and leukemic dissemination. However, cases of BPDCN have been reported without cutaneous involvement.[2] Patients most often present with nodular lesions, but can also present with brown to violaceous "bruise-like" macules, patches and plaques. In addition to cytopenias, hematologic/hematopoetic organ involvement can also include lymphadenopathy, splenomegaly and hepatomegaly. The prognosis is often poor, given its aggressive clinical course.[1] The median age at diagnosis is 65 to 67 years-old, with a male predominance, but BPDCN can present in all age groups, and one literature review of pediatric BPDCN concluded that younger age is an independent prognostic indicator that predicts more favorable outcomes.[3]

Another feature of this care was the patient's 3-year service in the Vietnam War with stated exposure to Agent Orange. Some of the approximately 3 million Americans who served in the Vietnam War from the early 1960's to mid-1970's were exposed to Agent Orange, an herbicide defoliant (which contains dioxins). More than 10 million gallons of Agent Orange was sprayed from airplanes, helicopters, boats, ground vehicles and backpack sprayers.[4] The U.S. Department of Veterans Affairs website reports, Agent Orange exposure has been associated (although not necessarily caused) with soft tissue sarcomas (other than osteosarcoma, chondrosarcoma, Kaposi's sarcoma, or mesothelioma), chronic B-cell leukemias, Hodgkin's Disease, non-Hodgkin's lymphoma, multiple myeloma, porphyria cutanea tarda, respiratory cancers (includes cancers of the lung, larynx, trachea, and bronchus), prostate cancer, AL amyloidosis, and other medical conditions (such as diabetes mellitus type 2, ischemic heart disease, Parkinson's disease, and early-onset peripheral neuropathy).[5,6] Although not all neoplasms and malignancies are associated with Agent Orange exposure, a lesson from this vignette is the importance of a thorough patient history (as well as physical exam). Not only is a good history and physical exam a tenet of medicine, identification of Vietnam War veterans with exposure to Agent Orange and the associated medical conditions can potentially qualify them for health care and other benefits through the Department of Veterans Affairs.[1]

The treatment options for our patient was limited by his age and co-morbidities, including CKD Stage 3. Although he has had some progression of his disease, the patient has also had periods of improvement with chemotherapy, which has given him the

opportunity to maintain his daily activities and spend time with family and friends. On the numerous conversations with the patient, he attributes maintenance of his health to the attentive care of his Primary Care clinic (especially at the time of presentation), timely evaluations and biopsies by Dermatology and Pathology, and ongoing dedication of the Hematology/ Oncology service.

Figures

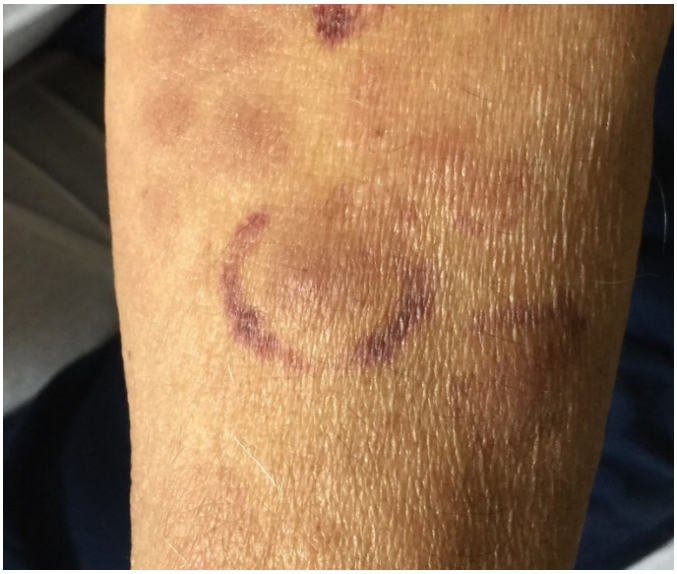

Figure 1

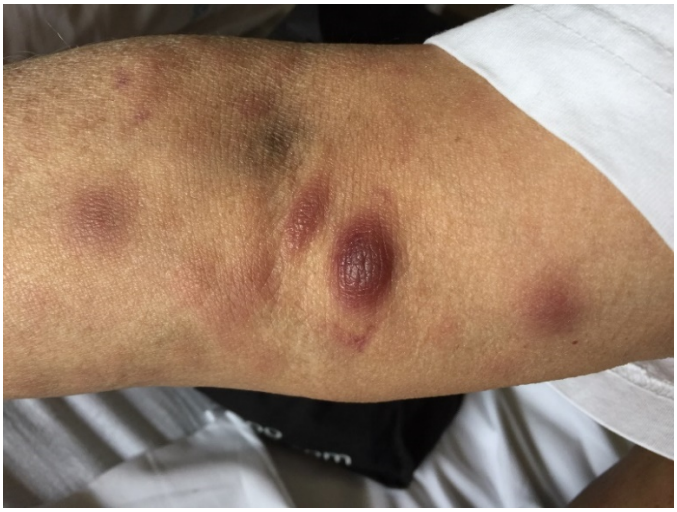

Figure 2

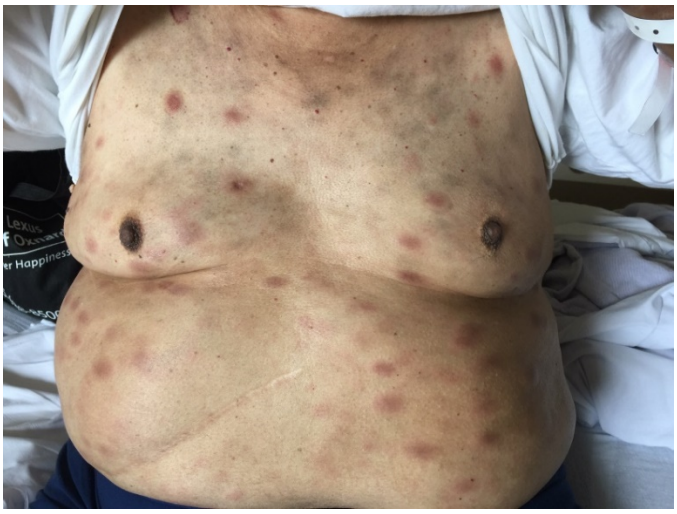

Figure 3

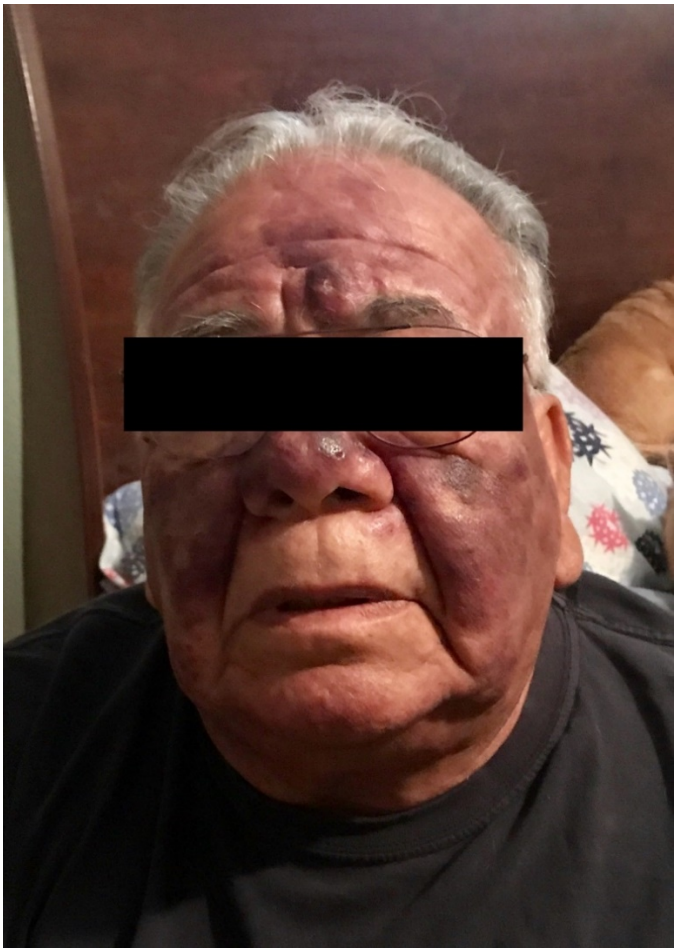

Figure 4

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