Blastic Plasmacytoid Dendritic Cell Neoplasm

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Introduction

Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare, aggressive hematologic tumor derived from plasmacytoid monocytes (2). BPDCN typically presents in the seventh to eighth decade with a higher prevalence in men. Most cases present with red to violaceous, indurated plaques and nodules and eventually involve the bone marrow, peripheral blood, lymph nodes and visceral organs (4,5). Diagnosis is made via biopsy of the skin, lymph node or bone marrow and will reveal a dense dermal infiltrate of atypical mononuclear cells extending to the subcutaneous fat. A immunohistochemistry panel is strongly positive for CD123, CD4, CD56, and TCL1 and will differentiate the tumor from mature lymphoid malignancies (1,5). BPDCN has a poor prognosis even with conventional chemotherapy. Emerging therapies have improved prognosis. Here we present a case of BPDCN in an 83-year-old male.

Case Presentation

An 83-year-old male with a history of throat cancer presented to the dermatology clinic with a gradually expanding lesion on the right distal thigh present for three months. Physical examination revealed an 8.5 cm asymptomatic, firm, violaceous plaque on the right distal thigh as seen in figures 1 and 2. The patient denied fever, weight loss, or night sweats at the time of presentation. A biopsy was taken by punch technique for H&E evaluation. Pathology revealed a diffuse atypical hematolymphoid infiltrate with an associated grenz zone and positive staining for CD4, CD33, CD56, TCL1 and CD123 (Representative images in figures 3-4). The findings were consistent with blastic plasmacytoid dendritic cell neoplasm.

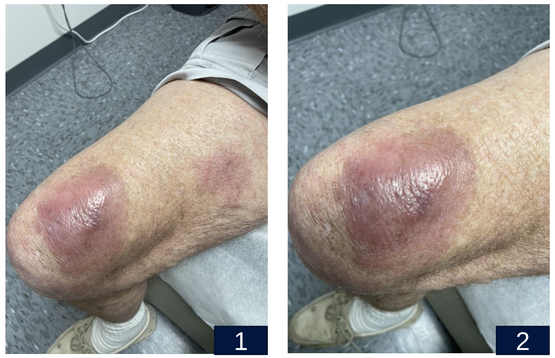

Figure 1 and 2. Initial clinical presentation of an 8.5 cm violaceous, firm plaque on the right anterior distal thigh.

Pathology

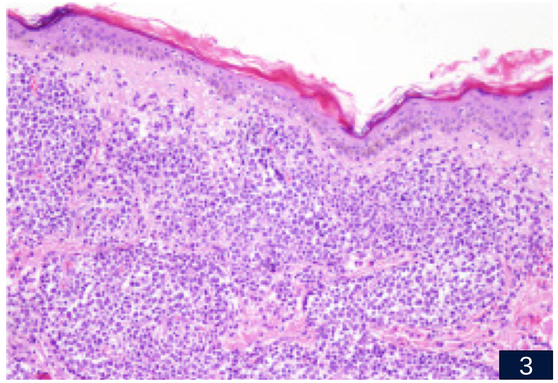

Figure 3. Representative H&E stain revealing a dense dermal infiltrate of atypical mononuclear cells, extending to to the subcutaneous fat; cells resemble lymphoblasts with irregular shaped nuclei and minimal cytoplasm. The tumor does not involve the superficial dermis. Adnexal structures completely obliterated by tumor

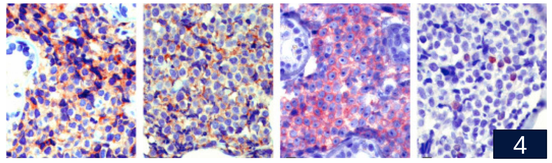

Figure 4. Representative immunohistochemical CD4, CD56, CD123, and TdT stains, respectively.

Management and Follow Up

Our patient was immediately referred to hematology/oncology for treatment. However, prior to evaluation by hematology/oncology, the patient returned to the dermatology clinic with eruptive, violaceous plaques on the trunk and upper extremities. This presentation was indicative of disease progression and we again stressed the importance of oncology evaluation. The patient is now being seen by a multidisciplinary team at Moffitt Cancer Center.

Discussion

- Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare entity comprising only 0.44% of all hematologic malignancies (1,6).
- BPDCN primarily affects the elderly, however there have been documented pediatric cases. There is a male predominance of 3:1 to 5:1 (5).
- Initial presentation commonly involves the skin in the form of solitary to multifocal violaceous nodules and plaques. Eventual spread to the bone marrow, peripheral blood, lymph nodes, and visceral organs is expected (2,5).
- Diagnosis of BPDCN via cutaneous biopsy can be difficult due to overlapping histopathological findings with various hematologic malignancies necessitating comprehensive immunohistochemical analysis (1,6).
- Collaboration with dermatology, hematology/oncology and pathology (dermatopathology and hematopathology) needed to make accurate diagnosis. Further workup may include a CBC, flow cytometry of bone marrow aspirate, lymph node biopsy, and PET scan (2,4).
- BPDCN has a poor prognosis despite conventional therapies, with median overall survival of 8 to 16 months (5,6).
- Conventional chemotherapy is the mainstay of treatment, even in patients with only cutaneous disease. Hematopoietic stem cell transplantation may be of benefit for younger patients (2,3)
- Tagraxofusp is a novel therapy targeting CD123, expressed in BPDCN malignant cells. However, tagraxofusp has not shown efficacy in all cases. Trials are ongoing for other targeted therapies (3,6)

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