Plasmablasticlike lymphoma arising within chronic pyoderma gangrenosum

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INTRODUCTION

Pyoderma gangrenosum (PG) is an uncommon neutrophilic dermatosis characterized by the rapid development of painful, ill-defined ulcerations with undermined gun-metal gray borders. Although half of the cases are idiopathic, PG may be associated with inflammatory bowel disease, connective tissue disease, and hematologic disorders. Here we describe the first case, to our knowledge, of plasmablastic lymphoma (PBL) arising within a chronic PG ulcer.

CASE REPORT

A 69-year-old healthy, HIV-negative man presented with a 9-year history of recalcitrant idiopathic PG involving his shins and ankles bilaterally. His disease was unresponsive to pulse methylprednisolone, dapsone, cyclosporine, and infliximab; however, it stabilized with a combination of prednisone, mycophenolate mofetil (1 g twice a day), and intravenous immunoglobulin (2 g/kg/mo). Two months before his presentation, he had a 4 × 3-cm fungating nodule within a chronic PG ulcer over his right shin (Fig 1).

A 4.0-mm punch biopsy was performed to exclude the clinical diagnosis of squamous cell carcinoma. The histologic findings showed a diffuse dermal infiltrate of plasmacytoid malignant cells with numerous mitoses and surrounding necrosis (Fig 2). Immunohistochemistry found CD3, C138, CD38, and MUM1 positivity, whereas stains for CD45RB, CD20, CD56, CD79a, CD117, HHV8, PAX5, and ALK were negative. Ki-67 expression was seen in 90% of cells. Light chains were λ+ and κ−. Epstein-Barr virus-encoded RNA in situ hybridization showed diffusely positive cells. Based on these findings, a diagnosis of PBL was favored.

A staging workup, including bone marrow biopsy and computed tomography imaging of the chest, abdomen, and pelvis, was unremarkable. Treatment for his stage 1A cutaneous PBL entailed localized radiotherapy followed by 3 cycles of CHOP (cyclophosphamide, doxorubicin, vincristine) chemotherapy and prednisone. Two years after diagnosis, the patient remains in remission. His PG showed a slow, but gradual response to treatment with prednisone and intravenous immunoglobulin.

DISCUSSION

PBL is an aggressive, rapidly disseminating non-Hodgkin’s lymphoma. It is typically associated with HIV infection (CD4+ <200 cells/mm3) where it presents as an ulcerating or necrotizing mass in the oral cavity. Primary extranodal PBL may present as a polymorphous eruption on the skin. Treatment includes CHOP or CHOP-like chemotherapy with possible adjuvant radiotherapy; however, prognosis is often poor.

The histologic differential diagnosis for PBL is broad and includes immunoblastic diffuse large...
B-cell lymphoma (DLBCL), ALK+ DLBCL, primary effusion lymphoma, anaplastic myeloma, Castleman disease, extramedullary plasmablastic neoplasms, and Kaposi’s sarcoma—associated lymphoproliferative disorders. Sheets of atypical plasmablasts with a high Ki67 expression (>80%); positive plasma cell markers (CD38, CD138, MUM1); c-MYC rearrangement; weak-to-negative staining of CD56, ALK, CD45RB, and B-cell markers (CD20, CD79a, PAX5); and variable immunoglobulin expression help differentiate PBL from other histologic considerations.

The development of PBL in the context of idiopathic PG is unique. To date, only 13 cases describe a B-cell lymphoma, including hairy cell leukemia, DLBCL, and follicular lymphoma, occurring in conjunction with PG. In these reports, PG developed at the onset or shortly after the diagnosis of malignancy; in contrast, our patient’s PG preceded malignancy by 9 years.

In HIV-negative patients, PBL is most commonly observed in the context of iatrogenic immunosuppression (ie, organ transplantation). These patients are less likely to have oral involvement (ie, 40%) or systemic involvement (ie, 25%) compared with their HIV-positive counterparts. A review of 114 HIV-negative PBL cases identified EBV negativity, immunosuppression, Ann Arbor stage IV disease, and treatment resistance as poor prognostic factors.

Our patient’s favorable course may be attributed to early diagnosis, lack of bone marrow involvement, and Epstein-Barr virus status. Although the role of Epstein-Barr virus in PBL remains unclear, the degree and duration of immunosuppression likely contribute to tumor formation. Further studies are required to elucidate the mechanisms that underlie carcinogenesis in PG patients.

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