Introduction

A 69-year-old man with a 20-year history of chronic lymphocytic leukemia (CLL) and a six-year history of mantle cell lymphoma (MCL) presented with a solitary lesion on his left leg. The patient had received multiple courses of chlorambucil for treatment of CLL and rituximab as well as CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) for treatment of MCL. Both conditions were in remission at the time of presentation.

Examination

Physical examination revealed a 4x4 cm red to violaceous dermal nodule on the left anterior lower leg.

Course and Therapy

The patient was referred to oncology for systemic treatment. No surgical excision was performed.

Histopathology

Biopsy of the lesion demonstrated a sheet-like proliferation of highly atypical lymphoid cells in the dermis and subcutaneous fat. The tumoral cells were blastoid in appearance and possessed high nuclear to cytoplasmic ratios, vesicular chromatin, and large, irregular and prominent nucleoli. Multiple apoptotic bodies and mitoses were evident. Immunohistochemical staining for CD20, BCL-2, BCL-6, MUM-1, and cyclin D1 were positive in lesional cells. Sox-11, TDT, and CD10 were negative. Break-apart FISH for cyclin D1 was positive for the cyclin D1 rearrangement (11q13).

Blastoid Mantle Cell Lymphoma

Discussion

Mantle cell lymphoma (MCL) is a rare, aggressive variant of non-Hodgkin’s lymphoma (NHL). MCL is named based on its involvement of lymphocytes from the mantle zone of lymph nodes. MCL typically occurs in middle-aged males and represents 2-10% of all non-Hodgkin’s lymphomas. The disease is often identified at later stages, with involvement of multiple lymph nodes and/or the spleen. Patients commonly present with constitutional symptoms including fever, chills, weight loss, night sweats, as well as generalized lymphadenopathy, splenomegaly and hepatomegaly.

Skin involvement in MCL is rare, found in only 2-6% of patients. Cutaneous disease typically occurs as a progression of the common lymphoid form, but rarely may be the primary manifestation of MCL. Clinically, lesions appear as solitary or multiple non-descript erythematous papules and nodules.

MCL presents with a blastoid histological morphology in 10-20% of cases, a feature which is more commonly associated with cutaneous manifestations. Histopathologic examination reveals a dense dermal proliferation of atypical lymphoid cells displaying lymphoblastic morphology, which stain positively for CD20, BCL-2, BCL-6, MUM-1, and cyclin D1. MCL is associated with the chromosome translocation t(11;14)(q13;q32). This translocation results in overexpression of cyclin D1, a protein involved in cell cycle regulation, specifically the progression of cells from G1 phase to S phase. Immunohistochemical stains are positive for cyclin D1 in 98% of patients.