Livedo reticularis: A helpful clue in the diagnosis of intravascular large B-cell lymphoma

Samuel Ecker DO; Andrew Jensen DO; László Kárai MD, PhD; Carlos Ricotti MD, FAAD; Francisco Kerdel MD, FAAD; Stanley Skopit DO, MSE FAOCD
1Dermatologist, Larkin Community Hospital, Miami, FL; 2Intern, Larkin Community Hospital, Miami, FL; 3DermDx Dermatopathology Services, Miami Lakes, FL; 4Florida Academic Dermatology Center, Coral Gables, FL; 5Program Director, Larkin Community Hospital, Miami, FL

INTRODUCTION

Intravascular large B-cell lymphoma (IVL; BCL) is a rare type of malignant, extranodal lymphoma characterized by the selective growth of neoplastic B-cells in the microvasculature. The disease is extremely aggressive and often rapidly fatal when diagnosis and treatment are delayed. This condition is often diagnosed postmortem given its rapid and aggressive clinical course. This disease can affect any organ system and can present with any myriad of nonspecific symptoms making the diagnosis difficult. Cutaneous findings are often an early manifestation therefore dermatologists need to be aware of its existence. Additional findings that may aid diagnosis include fever, cognitive impairment, stroke-like symptoms and elevated serum lactate dehydrogenase levels. Early diagnosis and treatment have been shown to improve survival outcome, however the decision to initiate chemotherapy can be difficult and requires definitive diagnosis.

CASE PRESENTATION

Presentation: 76 year-old female with livedo reticularis and painful subcutaneous nodules on her breasts, flanks, abdomen, buttocks, inguinal folds and upper thighs x 3 months. She reported generalized fatigue and muscle weakness in her lower extremities one month prior to the rash. She was previously seen by a rheumatologist and a dermatologist who performed an incisonal biopsy of her left thigh. She was referred to our care after she failed treatment with prednisone and plaquenil. Review of symptoms was significant for an unintentional twelve pound weight loss in the last month.

Medical history: MMIS, CHF, AFib, HLD

Medications: Prednisone, Plaquenil, Carvedilol, Ramipril, Spironolactone, Coumadin, Digoxin, Furosemide, Crestor

Clinical Examination: Livedo reticularis and painful subcutaneous nodules on the lateral aspects of the back, breast, abdomen, buttocks, inguinal folds and upper thighs (Figure 1).

Labs: CBC, CMP WNL, PT/INR therapeutic. ANA (+), anti-dsDNA (+), atypical pANCA (+), ESR 100, CRP 12.30, Hepatitis panel negative

Imaging: PET-CT and bone marrow biopsy were negative

Pathology: Intravascular atypical, hyperchromatic lymphocytes were seen within the lumen of scattered vessels and small capillaries of the fat leading to distention of the capillary network and fibrin deposition (Figure 2). The atypical intravascular cells stained strongly positive with CD20 and Mum-1 (Figures 3, 4). Molecular studies for B-cell gene rearrangement showed the presence of a clonal process. The previous biopsy was also reanalyzed and found to contain the same atypical intravascular cells. B-cell gene rearrangement results confirmed the presence of the same neoplastic clone in all biopsy specimens.

DIAGNOSIS AND TREATMENT

Diagnosis: Intravascular large B-cell lymphoma

Treatment: Patient was referred to an oncologist for combination chemotherapy with Rituximab, cyclophosphamide, etoposide, vincristine, prednisone (R-CHOP).

FOLLOW UP

Initially, the patient responded well to chemotherapy; she reported complete resolution of pain and nearly 100% clearance of her skin lesions a few days after her first treatment with R-CHOP. Soon after her second treatment, she developed several complications including severe bone pain, muscle weakness, pancytopenia, and was hospitalized for an acute exacerbation of CHF. The oncologist postponed further treatment and her symptoms slowly returned. Several weeks later, she was hospitalized a second time with sepsis and died four months after the date of her diagnosis.

DISCUSSION

Intravascular large B-cell lymphoma is a rare aggressive disease that carries a grim prognosis and death frequently occurs within two years. Diagnosis may rely on subtle histopathologic findings that require serial biopsies. Livedo reticularis warrants an increased level of suspicion as this clinical picture is often the result of occluded vascular channels.

Combination chemotherapy with R-CHOP is the treatment of choice and early intervention has been shown to offer the greatest chance of survival. A 2008 retrospective analysis of 106 IVL; BCL patients treated with chemotherapy alone versus chemotherapy plus rituximab found the 2-year survival rate was 46% in patients treated with chemotherapy alone, whereas the 3-year survival rate was 60% in patients who received combination chemotherapy with rituximab. Unfortunately, our patient was unable to tolerate treatment and her death illustrates the clinically frustrating nature of this malevolent disease.

REFERENCES