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An Osseous Enigma: Intravascular Large B Cell Lymphoma

Tamana Ebrat

Keywords

Lymphoma, Large B-Cell, Diffuse; Fever of Unknown Origin; Splenomegaly; Biopsy; Headache

Abstract

Intravascular Large B Cell Lymphoma (IVLBCL) is a rare subtype of Diffuse Large B Cell Lymphoma, characterized by B cell proliferation within blood vessel lumens without apparent extravascular masses. IVLBCL often manifests with nonspecific signs such as fever of unknown origin, as well as highly variable proportions of neurological, hematologic, and cutaneous findings, rendering it diagnostically challenging. A 66-year-old woman presented to the ED with worsening hypotension, nausea, and malaise over the past year. Despite multiple admissions, prior workups were inconclusive, except for splenomegaly. On admission, her blood pressure was 82/69, and physical examination revealed tenderness over the left upper abdomen, splenomegaly, and pustules on the lower abdomen. Laboratory results showed hemoglobin of 11.3 g/dl and Na 130 mEq/L. Over the subsequent week, she developed fever of 102.9F, tachycardia, hypotensive episodes, vivid dreams, tremors, headache, and new onset cough. Infectious and malignancy workup 9 months antecedent failed to reveal the source of her symptoms. Her fevers, skin rash, and markedly elevated ferritin (1650 mg/L) led to early presumptive diagnoses of Still's or Sweet's, however, corticosteroid therapy failed to produce durable improvement. Additional labs including ESR (49 mm/hr), CRP (89 mg/L), IL-2R (4955 pg/mL), and LDH (759 U/L) were elevated despite non-revealing rheumatologic and infectious workups. Imaging revealed splenomegaly and diffuse bone marrow uptake, but biopsies were initially negative. A repeat PET-CT identified osseous lesions, confirmed by right tibial biopsy unveiling small aggregates of atypical cells within the marrow space, expressing CD20, CD5, and BCL6, highly suspicious for B-cell lymphoma. A review of prior abdominal skin biopsies also confirmed IVLBCL expressing similar markers. The patient commenced R-CHOP chemotherapy and intrathecal methotrexate. After three cycles of treatment, a PET scan showed resolution of previous lesions and no evidence of new or suspicious lesions.