An unusual presentation of erythema elevatum diutinum with underlying hepatitis B infection

Jessica Vincent DO1, Adeline Kikam DO2, Kelly Tyler MD3, Sara Peters MD3, Ryan Kirkland MD4, Benjamin Kaffenberger MD3

1OhioHealth O’Brienness Hospital, Athens, Ohio, 2Firelands Regional Medical Center, Sandusky, Ohio, 3Ohio State University Medical Center, Columbus, Ohio, 4Dermatologists of Greater Columbus, Columbus, Ohio

Abstract

Erythema Elevatum Diutinum (EED) is a rare chronic cutaneous small vessel vasculitis of unclear pathogenesis. Classically, lesions present as symmetric red to purple plaques, papules and nodules overlying joints. First-line therapy is with dapsone. We report a case of EED with widespread lesions involving the hands, extensor extremities and trunk. Multiple biopsies showed concentric intradermal perivascular inflammation with dermal fibrosis and leukocytoclastic vasculitis suggesting EED in various stages of evolution. An extensive workup was positive for underlying hepatitis B infection. Our case represents the clinicopathologic spectrum on which EED can present and emphasizes the importance of searching for an underlying etiology.

Case Report

A 57-year-old white male presented complaining of burning and stinging red nodules on the dorsum of his hands for about 1 year. He also admitted to an episode rash over the lower legs and bilateral flanks of 7 years duration. He was briefly treated with an oral prednisone taper and topical corticosteroids including triamcinolone 0.1% cream and clobetasol 0.05% cream without improvement. On exam were deep red to violaceous discrete nodules and plaques with overlying hyperkeratosis involving all distal and proximal interphalangeal joints of the hands and extensor elbows (Figure 1). On the bilateral posterior arms, anterior legs and periumbilical area were deeply erythematous papules and plaques with background hyperpigmentation (Figure 3). Across his low back and bilateral flanks were erythematous papules with central hemorrhagic crusting (Figure 5).

Pertinent laboratory findings included a positive hepatitis B surface antigen with hepatitis B DNA value 4313876 IU/mL (reference range <10 IU/mL) and a HBV quantitative PCR value of 6.64 units (reference range <1.00 unit).

An additional infectious workup was negative for hepatitis C, streptococcus, syphilis, tuberculosis and HIV. A complete blood count, complete metabolic panel, urinalysis, complement, cryoglobulins and serum protein electrophoresis were within normal limits. Autoimmune serologies were negative including anti-nuclear antibody, rheumatoid factor, anti-Sjogren’s syndrome-related antigen A and B, anti-cyclic citrullinated peptide, anti-Smith, anti-neutrophil cytoplasmic antibodies. Peripheral blood immunophenotyping, lactate dehydrogenase, quantitative immunoglobulins, and age appropriate cancer screens did not demonstrate evidence for malignancy underlying his disease.

Three 4-mm punch biopsies were performed from the left 5th digit, left posterior arm, and left flank (Figures 2, 4 and 6, respectively). The constellation of clinical findings together with the histopathologic changes represented EED in various stages of evolution. The patient was started on dapsone 100mg daily and referred to the Infectious Disease service for treatment of the chronic hepatitis B, however, he was subsequently lost to follow up.

Discussion

Erythema elevatum diutinum represents a rare form of chronic cutaneous small vessel vasculitis. The disease classically presents as firm, fixed red-brown to violaceous papules, plaques and nodules affecting the extensor extremities. Less common locations have been described including palms and soles, face, trunk and periauricular region. Our patient was unique as in addition to typical lesions of EED, he presented with crusted papules on the flanks and violaceous plaques of the lower legs and periumbilicus.

Originally associated with Streptococcus as isolated from EED lesions,2,3 additional infectious etiologies include viral hepatitis, HIV-1 and HIV-1.4 Hepatitis B and C are well-known to be associated with EED, however, only previously reported in patients with concomitant HIV infection.

References