DERMATOPATHOLOGY

A CASE OF ERYTHEMA ELEVATUM DIUTINUM WITH EARLY HISTOPATHOLOGICAL PRESENTATION OF NEUTROPHILIC DERMATOSIS

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Background: Erythema elevatum diutinum is a rare, chronic form of cutaneous vasculitis. Histopathologic features consist of leukocytoclastic vasculitis in early lesions, followed by fibrotic replacement of the dermis in older lesions. Dapsone can induce dramatic improvement.

Observation: A 20-year-old male patient presented with red papules on limbs over 3 months. The patient was well without fever and arthralgia. Physical examination revealed multiple red-violaceous papules on his elbows, hands, fingers, and knees. An initial skin biopsy from the right hand showed a dense infiltrate composed mostly of neutrophils with nuclear debris in the superficial dermis, but no fibrin deposition in small blood vessel walls. Direct immunofluorescence was negative for IgG, IgA, IgM and C3 deposition in the BMZ and vessel walls. Neutrophilic dermatosis with a possible diagnosis of Sweet's syndrome was initially suspected. A revisit after four months revealed multiple new papules and nodules on his hands and fingers, that constrained free movement of his finger joints. Another skin biopsy specimen taken from a nodular lesion revealed dense layered fibrosis throughout dermis, with focal areas of concentric fibrosis around small blood vessels and with foci of neutrophilic clusters with nuclear dusts, characteristic of late-stage lesions of erythema elevatum diutinum. Laboratory work-ups including peripheral blood count, urine analysis, CRP, ESR, RF, ANCA, HIV, HBV, HCV and RPR were all within normal range or negative except elevation of ASO:364 IU/L, IgA: 5.87g/L, and IgG:16.4 g/L. The patient responded well with oral dapsone, 50mg twice a day, and two months later, nearly all papular lesions resolved, however, nodular lesions persisted.

Key message: Histopathological diagnosis of early stage of erythema elevatum diutinum may be challenging. As in our case, it can be present as neutrophilic dermatosis without evidence of vasculitis. Clinicopathological correlation with follow-up is vital to correct diagnosis of such patients.