

Erythematous Annular Plaques in a Woman with New Onset Arthritis

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Case Report

A 55 year old Caucasian woman presented with a several month history of multiple erythematous plaques over her bilateral thighs, which gradually progressed to include multiple subcutaneous nodules of her face and palmar hands. Two days prior to her visit, she developed swelling and pain in her bilateral hands, in particular the fingertips. In addition, the patient had a history of Raynaud's with scaling and healing erosions of her fingertips. On examination, multiple erythematous, indurated, annular plaques were symmetrically distributed over her bilateral thighs and buttocks along with multiple subcutaneous nodules of the nasal bridge, forehead, and palmar surface of her hands were observed (Figure 1). The patient had appreciable swelling of her second and third digits bilaterally. She also had prominent nail fold telangiectasias (Figure 2). Laboratory analysis revealed an ANA of 1:640 (speckled, nucleolar), however ENA, RF, ESR and anti-dsDNA antibodies were all negative or within normal limits.

Histopathologic examination revealed palisaded histiocytes in the reticular dermis, surrounding areas with degenerated collagen and mucin, along with a perivascular lymphocytic infiltrate. Occasional scattered small nodular collections of histiocytes (including multinucleated histiocytes) were also present.

Based on the combination of clinical and histological findings, the diagnosis of Interstitial Granulomatous Dermatitis was made. The patient was initially treated with fluocinonide ointment and methotrexate, and the lesions persisted although with some improvement. Thus, hydroxychloroquine 200 mg BID was added to the regimen with marked improvement.

Interstitial Granulomatous Dermatitis (IGD) was first described in 1993 by Ackerman et al. [1]. The lesions were originally described as indurated, linear bands that were symmetrically distributed over the lateral chest wall, termed the rope sign [1]. With the increasing recognition of this disease, the literature reports a progressively greater variability of it's presentation [2,3]. The most common presentation reported is erythematous annular plaques, symmetrically distributed



Figure 1: Annular plaques were symmetrically distributed over her bilateral thighs and buttocks.



Figure 2: Prominent nail fold telangiectasias.

over the lateral chest and/or proximal extremities, particularly the thighs [2,4]. Other morphologies have also been observed such as papules and nodules [2-5]. IGD is typically seen in middle aged females and commonly lesions are asymptomatic [2,3].

One of the key features of Interstitial Granulomatous Dermatitis is the association with autoimmune diseases, abnormal immunologic serologies, and arthralgias or arthritis [2-6]. Serologic abnormalities commonly reported include positive rheumatoid factor, ANA, anti-ds DNA, or even elevated CRP or ESR [2-6]. A common feature, even in the absence of a specific autoimmune disease, are arthralgias or even frank arthritis [2-5].

The exact pathogenesis of IGD is unclear. It is thought to be immune mediated and possibly related to immune complexes given the association of IGD with autoimmune disease [2-5]. Treatment has been primarily anecdotal and includes medications such as topical or systemic steroids, NSAIDs, dapsone, methotrexate, hydroxychloroquine, phototherapy, IVIG, and TNF α inhibitors [2-7]. However, caution must be exhibited with use of biologics as there are case reports of IGD occurring in association with initiation of TNF α inhibitors [8].

Diagnosis requires a clinical-pathological correlation. Histologic examination reveals a typically "bottom-heavy" infiltrate of histiocytes, primarily interstitially [5,6]. The foci of palisading histiocytes surrounds degenerated collagen bundles [2,5]. Eosinophils and neutrophils are occasionally observed [2-4]. While mucin is typically not seen, there are reports of mild to moderate mucin occurring in IGD [2,3]. Vasculitis

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is never seen with IGD, which is important to distinguish it from the Palisaded Neutrophilic and Granulomatous Dermatitis spectrum including Churg Strauss granuloma, rheumatoid papules, and ulcerating rheumatoid necrobiosis [2-5,9]. Other differential diagnoses to consider are Interstitial granuloma annulare and granulomatous reactions to drugs [2-4]. However, granuloma annular is not seen in association with rheumatologic abnormalities and granulomatous reactions to drugs typically have a vacuolar infiltrate on histology [2,3].

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