Case Report

Interstitial granulomatous dermatitis presenting as erythematous plaque over both ankles

A 10-year-old girl had persistent asymptomatic erythematous plaques over both ankles for two years. Routine blood tests and fungal-related investigations were unremarkable. Clinical and histopathological findings suggested diagnosis of Interstitial Granulomatous Dermatitis (IGD) – a relatively new entity.

Keywords: Interstitial granulomatous dermatitis

Introduction

Interstitial granulomatous dermatitis is an uncommon condition with no known cause. It is a histopathologic diagnosis with variable clinical appearance and may be associated with autoimmune systemic disease.

Case report

A 10-year-old girl came to us because of erythematous rash over both ankles for two years. She enjoyed good health. Family or birth history was unremarkable. She was not on any drug. According to her mother, the lesions were unprovoked and grew without pain or itchiness. She was otherwise very well. Physical examination showed erythematous patches over both lateral malleoli. They were mildly indurated, non-scaly, distinct in border and normal sensation. No erosion or ulceration was noted (Figure 1). Buccal mucosa, scalp and nail were unremarkable. No arthropathy and lymphadenopathy were found. The ankle plaques slowly grew in size and became scaling (Figure 2). Most of the blood tests were normal (CBP, RFT, LFT, ANA, rheumatoid factor, ENA, C3, C4, TSH) except borderline elevated ESR ↑ 28 mm/1 hr (0-16 mm/1 hr). Skin scraping for
Fungal element and culture were negative. Radiography of chest was normal.

The brevity of characteristic dermatological change coupled with almost completely normal blood profiles have made this skin lesion difficult to diagnose. Common differentials such as chronic eczema, psoriasis and tinea infection are entities cannot be overlooked. However, all of the above are not in total conformity with what our patient had. For instance, it is usually itchy in eczema. Absence of silver scaling does not support
psoriasis. Tinea infection is usually annular in morphology and negative fungal culture does not support tinea infection.

This lesion could not be classified within the scope of any common disease categories and likely to be something unusual. The rare counterparts include granulomatous dermatitis such as inflammatory dermatosis associated with connective tissue disease, necrobiosis lipoidica diabetorum, granuloma annulare in erythematous or patch variant, mycosis fungoides and inflammatory stage of morphea.

An incisional biopsy from lesion of the right ankle was performed. It showed loosely organised aggregates of histiocytes both in the superficial dermis and deep dermis. There was also non-specific interstitial inflammatory infiltrate. Small area of karyorrhexis associated with neutrophils was also noted. There was no evidence of fungal infection (Figures 3 & 4). These were compatible with interstitial granulomatous dermatitis.

**Discussion**

Interstitial granulomatous dermatitis (IGD) was first described by Ackerman in 1983 and was associated with cutaneous cord and arthritis. It is an uncommon histopathologic diagnosis with variable clinical appearance. IGD with plaques describes the salient clinical and histopathologic features common to a distinctive group of patients, mostly women, who have rheumatoid polyarthritis along with abnormal serologic findings, often related to systemic autoimmune disease. Chu et al and Sangueza et al favor unifying IGD and redefining it as palisaded neutrophilic granulomatous dermatitis (PNGD). However, other authors disagree. IGD is a separate entity because of its polymorphic clinical presentation in comparison with PNGD and there is no evidence of leukocytoclastic vasculitis at histopathologic assessment of IGD.

The pathogenesis is mainly unknown and is postulated that the inciting event is the deposition of immune complexes in the dermal vessels with activation of complement and neutrophil which lead to damage to dermal collagen and eventually the granulomatous inflammation.

The clinical signs in the skin are variable, consisting of asymptomatic multiple erythematous plaques with indistinct borders, mainly involving the lateral chest wall and medial thighs in a bilateral and somewhat symmetric distribution. Cutaneous cord or subcuaneous linear band that resembles a rope
situates on the trunk is pathognomonic. Majority are asymptomatic, expand slowly in terms of weeks to months. In a study conducted by Tomasini and Pippione, 17 patients with IGD and plaque were recruited. It was found that patients with IGD and plaque were often associated with rheumatoid polyarthritis or rheumatoid arthritis which could occur before, overlapping, or after cutaneous eruption. Other associations included systemic lupus erythematosus, thyroiditis, diabetes mellitus etc. It was also associated with positive autoimmune marker (ANA, RF, DNA, antimicrosomial & antithyroglobulin Ab) and elevated ESR. None of them required systemic therapy because of mild form of disease. More than half had spontaneous resolution with post-inflammatory hyperpigmentation.

The optimal therapy for IGD is difficult to establish because the available information is anecdotal. Treatment strategies are mainly depend on the severity of underlying systemic disease. Majority do not require systemic therapy in view of limited involvement and asymptomatic nature of the lesions. Most relief is achieved with nonsteroidal anti-inflammatory drugs (NSAID) and topical corticosteroid or prednisolone. In some cases, the course is ebb and flow. Recommendations regarding treatment, and its exact relation to collagen vascular disease, await further studies.

References