Perforating Granuloma Annulare Appears as a Psoriasiform Lesion

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Abstract
We present a 54-year-old Caucasian female with a history of diabetes mellitus, multiple sclerosis, and ulcerative colitis who presented with nail dystrophy, polyarthralgia in her hands and hips, myalgia, proximal myopathy, dactylitis, and psoriasiform-appearing progressive, painful plaques on the dorsal aspect of her hands. Histologic examination of the lesion from the left index finger revealed an interstitial and necrobiotic granulomatous dermatitis with connective tissue mucin deposition associated with a perivascular lymphoid infiltrate. Additionally, there was focal transepidermal elimination of the necrobiotic material. She was diagnosed with perforating granuloma annulare (PGA). We are unaware of any other cases of PGA in the literature with this presentation.

Introduction

Perforating granuloma annulare (PGA) is a rare subtype of granuloma annulare that was first described in 1971 [1–3]. The etiology of this condition is unknown, and it typically has a chronic course [1]. PGA occurs more frequently in children and young adults. Lesions usually present as umbilicated papules that are commonly found on the extremities; however, generalized forms of PGA have been described [1, 4]. Although some palmar cases have been seen,
it is more commonly seen on the dorsal aspect of the hand [2]. In this case report, we describe an atypical presentation of PGA in an adult patient with multiple sclerosis.

Case Report

A 54-year-old Caucasian female with a history of diabetes mellitus, multiple sclerosis, and ulcerative colitis presented with nail dystrophy and psoriasiform-appearing progressive, painful plaques on the dorsal aspect of her hands. Her symptoms began 1–2 months after a hospitalization for inflammatory colitis determined to be an adverse reaction to teriflunomide, which she had begun taking for multiple sclerosis. Initial presentation showed scattered pink scaly plaques on the lateral and medial surfaces of her digits. The plaques were intermittently painful and burning. Six months later, she presented with painful and thickened nails with shape change to overcurvature. Additionally, she had well-demarcated scaly erythematous plaques on her left dorsal index finger and right upper extremity, mimicking psoriasis (Fig. 1). Furthermore, she also reported polyarthralgia in her hands and hips, myalgia, proximal myopathy, and dactylitis (Fig. 2) for which she is followed by rheumatology. Laboratory workup for an underlying rheumatologic disorder was negative.

The differential diagnosis included psoriasis, systemic lupus erythematosus, and giant cell reticulohistiocytoma. Punch biopsies of the lesions were taken. Histologic examination of the lesion from the left index finger revealed an interstitial and necrobiotic granulomatous dermatitis with connective tissue mucin deposition associated with a perivascular lymphoid infiltrate (Fig. 3a–c). Additionally, there was focal transepidermal elimination of the necrobiotic material. Direct immunofluorescence examination of the lesion from the right arm showed no specific immunoreactants. Based on her histopathologic findings, she was diagnosed with PGA. After biopsy, there was improvement of the biopsied plaques, which has been reported in cases of granuloma annulare [5]. She was treated with a 2-week taper of prednisone, prescribed by her rheumatologist. This provided a few days of improved joint pain, with slight reduction of plaque erythema. However, 2–3 days after completing the prednisone course she experienced rebound joint pain, dactylitis, and worsening pain of existing plaques, with appearance of new crops of lesions. She did not return to our clinic for planned intralesional triamcinolone and was lost to follow-up.

Discussion

Although PGA was first discovered over 40 years ago, it remains a poorly understood disease. Similar to non-PGA subtypes, the etiology of PGA is unknown [6]. Cases have been reported that were potentially associated with diabetes mellitus, tuberculosis, rheumatoid arthritis, HIV/AIDS, and herpes zoster infection [7, 8]. Madan et al. [7] found PGA after application of a tattoo on the dorsum of the hand, with the lesion confined to the location of the tattoo. Furthermore, a familial case has been described as well as a higher incidence of PGA in the Hawaiian Islands [2, 6, 9]. It is proposed that PGA is the result of an abnormal, delayed helper T cell hypersensitivity reaction in response to exogenous antigens [2, 10].

PGA is found more frequently in children and young adults and is primarily found on the extremities, especially on the dorsal aspect of the hand [2, 9]. Only 9% of patients with PGA will have a single lesion [11]. While it is uncommon for the first presentation of lesions to occur over the age of 50, the patient presented with multiple lesions of the characteristic
location on the dorsum of the hand. Additionally, a generalized form has been described with presentation on the ears [4]. Although there was a similar-appearing lesion on the patient’s right arm, it was histologically not determined to be PGA and thus cannot be classified as a generalized form.

Two different forms of PGA have been described: a papular perforating type (P type) and an ulcerative perforating type (U type), which is strongly correlated with diabetes [12]. Lesions also present demarcated papules that may or may not be umbilicated and are usually 1–5 mm in size. Centrally, they will have an area of perforation where mucoid material is eliminated [13]. This transepidermal elimination of mucoid material or collagen is thought to be due to the superficial location of necrobiotic granulomas [11]. Our patient did not have this classic presentation. We found an atypical presentation that appeared as progressive, well-marked pink plaques with scale similar to that of psoriasis. We are unaware of any other cases of PGA in the literature with this presentation.

As evident from this patient’s presentation, biopsy and histologic examination of lesions were necessary for making the diagnosis. The histopathologic features of PGA include the presence of granulomas with histiocytes organized in a palisading pattern, surrounding necrobiotic collagen, which is typically represented by deposition of mucin, and rarely fibrin [1]. Additionally, the location of the granulomas in the superficial reticular dermis results in perforation of the epidermis and expulsion of necrobiotic material. The histopathologic differential diagnosis includes granulomatous diseases and perforating dermatoses [1]. Granulomatous diseases include sarcoidosis and tuberculids. Perforating dermatoses include a variety of conditions, such as primary perforating and acquired perforating dermatoses, and other dermatologic conditions that exhibit transepithelial elimination as an incident histopathologic finding [1]. Histologic examination from the patient’s lesions was consistent with PGA and provided the correct diagnosis when the clinical presentation was atypical for PGA.

The treatment of PGA has been shown to be difficult as it typically responds poorly to current treatment options [11]. Some options that have been suggested have been high-potency topical steroids, intralesional triamcinolone acetonide, cryotherapy, and excision of isolated lesions [2]. Alternative agents that have been suggested are tacrolimus and imiquimod cream 5% [7, 14, 15]. The patient’s lesions improved with biopsy and systemic steroids, which were prescribed with the intention to treat her arthralgias. Although PGA is a rare condition, systemic steroids or other immunomodulatory agents may serve as viable treatment options.

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**Statement of Ethics**

The research in this case report was conducted ethically in accordance with the World Medical Association Declaration of Helsinki.
Disclosure Statement

The authors have no conflicts of interest to declare. They certify that they have no affiliations with or involvement in any organization or entity with any financial or nonfinancial interest in the subject matter discussed in this case report.

Author Contributions

B.M. Witkoff made substantial contributions to the design of the paper and the drafting of the work as primary author and gave final approval for this case report to be published. N.N. Ivanov made substantial contributions to the design of the paper and the drafting of the work as secondary author and gave final approval for this case report to be published. S.C. Trotter made substantial contributions to the design of the paper, revising it for important intellectual content, and gave final approval for this case report to be published. All authors agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work were appropriately investigated and resolved.

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Fig. 1. Scaly psoriasiform-appearing plaques and papules with constant nonpruritic, severe burning pain sensation and tenderness.

Fig. 2. Dactylitis ("sausage digits").
Fig. 3. a Hyperkeratotic surface with communicating epidermal perforation to underlying necrobiotic granulomatous inflammation of the dermis. H&E, ×40. b Palisading histocytes and granulomatous inflammation surrounding a zone of collagen degeneration with mucin accumulation. H&E, ×200. c Periphery of palisading inflammatory cells surrounding central collagenolysis with neutrophils and nuclear dust present. H&E, ×400.