Case Report

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# Palisaded Neutrophilic Granulomatous Dermatitis Associated with Adult Onset Still's Disease

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## Abstract

A 66-year-old male presented with a new onset of rash. The rash was erythematous, macular and localized to the shoulders and neck. Biopsy showed palisading neutrophilic granulomatous dermatitis with features of atypical granuloma annulare tissue. Improvement was noted with steriods. To our knowledge, there has only been one reported case of PNGD in a patient with AOSD. It is diagnosed with skin biopsy and historically glucocorticoids have been the mainstay of treatment. However, clinicians should have a high suspicion of PNGD in patients with AOSD who present with features of atypical rash, as treatment may be required. This association is extremely rare, since this is only the second reported case. We strongly believe that to further delineate different forms of PNGD and rare instances such as in patients with AOSD, further case reports are needed. Additional research into the pathophysiology of PNGD is also warranted, as this would optimize medical management.

**Keywords:** Dermatitis; Still's disease; Steroids

## Introduction

Adult onset Still's disease (AOSD) is an inflammatory disorder, which is a very uncommon disease, with occurrence rate of 0.16 cases per 100,000 people [1]. It is characterized by fever, arthralgia, rash, and occasionally lymphadenopathy and splenomegaly. The rash involves the trunk and extremities usually. It is a non-pruritic, evanescent, salmon-colored, macular or maculopapular type of rash [1]. Palisaded neutrophilic granulomatous dermatitis (PNGD) is also an inflammatory disorder and it is characterized by skin-colored to erythematous papules or plaques on the extremities [2]. It most commonly presents in the upper extremities, particularly the fingers and elbows, however it can also be found in the lower extremities. It is commonly associated with connective tissue diseases and can be seen with rheumatoid arthritis, systemic lupus erythematous, sarcoidosis, and vasculitis [3-5]/ Drug-induced PNGD has also been reported, particularly with methotrexate and anti-TNF inhibitors [4,6]. We report a rare case of a patient with adult onset Still's disease who develops PNGD.

## Case Report

A 66-year-old male with past medical history significant for AOSD presented with a new onset of rash. The rash was erythematous, macular and localized to the shoulders and neck. The patient denied a prior history of similar rashes. Vitals were remarkable for blood pressure of 131/92 mm Hg, pulse of 91 beats per minute, temperature of 97.5°F, respiratory rate of 18 breaths per minute. Physical exam was significant for erythematous granular rash localized to the shoulders along with minimal cervical lymphadenopathy. Lab work was remarkable for elevated CRP of 12.8 mg/L, ESR of 49 mm/hr, and ferritin of 29.671 ng/ml. The CEA level and CA19-9 levels were within normal range. Immunological workup revealed a negative nucleolar pattern ANA, rheumatoid factor, ANCA, anti-CCP, and immunoglobulin levels. An infectious disease panel including HIV, Babesiosis, Hepatitis B and C were negative as well. He had a normal ACE level and negative HLA-B27, quantiferon and PPD testing. The rash was biopsied and showed palisading neutrophilic granulomatous dermatitis with features of atypical granuloma annulare tissue reaction along with overlapping features with necrobiosis lipoidica as a manifestation of a certain underlying systemic disease. Solumedrol was administered and the patient was started on Prednisone 50 mg daily PO, with a tapering dose. His rash predominantly resolved at his next follow up visit.

The diagnosis of PNGD is made based on clinical and histological findings. Skin biopsy shows neutrophilic dermal infiltrate, leukocytoclastic vasculitis, palisaded granulomas, and collagen degeneration [3]. PNGD is believed to result from the deposition of immune complexes in dermal vessels leading to

ischemic injury and fibrosis, however the pathophysiologic mechanisms have not been fully elucidated [4,7].

PNGD is usually associated with systemic disease such as rheumatoid arthritis, systemic lupus erythematous, inflammatory bowel disease and it resolves once the underlying etiology is treated. To our knowledge, there has only been one reported case of PNGD in a patient with AOSD [8]. It is diagnosed with skin biopsy and historically glucocorticoids have been the mainstay of treatment while varied success has been achieved with colchicine, cyclosporine, cyclophosphamide, hydroxychloroquine, triamcinolone, NSAIDS, and dapsone [4,6]. In approximately 20% of cases, self-resolution of PNGD has been observed [9,10]. However, clinicians should have a high suspicion of PNGD in patients with AOSD who present with features of atypical rash, as treatment may be required. This association is extremely rare, since this is only the second reported case. We strongly believe that to further delineate different forms of PNGD and rare instances such as in patients with AOSD, further case reports are needed. Additional research into the pathophysiology of PNGD is also warranted, as this would optimize medical management.

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