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Case presentation

Sporotrichoid granuloma annulare-like dermatitis associated with systemic B-cell lymphoma

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Abstract

Importance: Granuloma annulare is typically a benign, self-limited disease. Atypical presentations have been reported in association with systemic disease, including malignancy. Such patients may require additional diagnostic studies to assess for underlying malignancy. We report a patient with extensive sporotrichoid granuloma annulare-like dermatitis in association with systemic B-cell lymphoma.

Observations: An 83-year-old man with a three-year history of progressive sporotrichoid annular plaques and nodules on the arm developed ipsilateral retroauricular palpable lymphadenopathy, the latter consistent with B-cell lymphoma. Multiple skin biopsies of the plaques and nodules revealed granuloma annulare-like dermatitis. Lesions were unresponsive to intralesional and intramuscular corticosteroids, antibiotics, and antifungal agents, but rapidly improved following initiation of rituximab to treat his underlying lymphoma.

Conclusions and Relevance: Atypical presentations of granuloma annulare including granuloma annulare-like dermatitis warrant evaluation for systemic malignancy in a subset of patients.

Keywords: granuloma annulare; B-cell lymphoma; malignancy

Abbreviations: Granuloma annulare, GA; Acid-fast bacilli, AFB; Gomori methenamine silver, GMS; Periodic acid-Schiff-diastase, PAS-D

Case synopsis

Although granuloma annulare (GA) is typically a benign skin disease, atypical presentations associated with malignancy have been reported. We present a case of progressive sporotrichoid GA-like dermatitis as a harbinger of low-grade follicular B-cell lymphoma.

An 83-year-old previously healthy man presented with a three-year history of progressive non-healing arcuate plaques and nodules on the right arm. On physical examination, numerous firm pink and violaceous coalescing arcuate plaques and subcutaneous nodules extended in a sporotrichoid pattern from the extensor distal forearm and elbow to the extensor surface of the upper arm in the absence of axillary lymphadenopathy (Figure 1).



Figure 1. Right upper arm with coalescing arcuate indurated plaques and nodules at initial presentation

Several punch biopsies from the right upper arm revealed palisaded granulomatous dermatitis (Figure 2,3). Acid-fast bacilli (AFB), Fite, Gomori methenamine silver (GMS), Periodic acid-Schiff-diastase (PAS-D), and Gram stains were repeatedly negative for microorganisms. Three months later, the patient presented with ipsilateral retroauricular lymphadenopathy in the setting of progression of skin lesions. Excisional lymph node biopsy was remarkable for BCL2 and BCL6-positive low-grade systemic follicular B-cell lymphoma. The patient was referred to the oncology department for management of lymphoma. A PET scan revealed generalized lymphadenopathy, but bone marrow biopsy was negative for evidence of lymphoma. Rituximab was initiated with marked improvement of skin lesions following the first cycle of therapy (once weekly infusion for four weeks) (Figure 4). In total, he received three months of weekly rituximab with improvement of his lymphoma and near-resolution of the plaques and nodules on the right arm. Owing to residual presence of systemic lymphoma, he subsequently received a combination of rituximab and bendamustine, and is currently receiving maintenance rituximab.

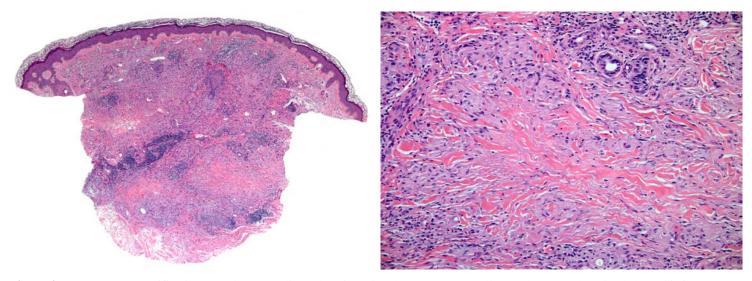


Figure 2. Low power magnification (4X, hematoxylin and eosin stain) demonstrates small granulomas and a mixed dermal inflammatory infiltrate composed of multinucleated histiocytes, lymphocytes, plasma cells, and neutrophils palisading around degenerated collagen. There are associated perivascular lymphocytes. **Figure 3.** High power magnification (40X, hematoxylin and eosin stain) shows interstitial mucin and palisaded histiocytes surrounding altered collagen fibers.



Figure 4. Dramatic improvement in cutaneous lesions 4 weeks after initiation of once weekly rituximab.

This case describes an unusual presentation of sporotrichoid GA-like dermatitis preceding the diagnosis of an indolent systemic lymphoma. Classically, GA presents with asymptomatic annular or arciform erythematous plaques. However, several reports document GA-associated solid tumors and hematologic malignancies, most commonly with lymphoma [1,2]. Lesions may appear several years before or after the diagnosis of lymphoma or may present concurrently with relapse of malignancy [3,4]. Patients with concurrent GA and malignancy are more likely to present atypically; for example, skin lesions may occur in older patients, involve unusual locations, have unique clinical distributions, have atypical histopathologic findings, and be symptomatically painful or pruritic [1]. The etiology of non-infectious granulomatous dermatitis in patients with malignancy is unclear, although it has been suggested to represent a paraneoplastic or immune-mediated phenomenon [5].

Conclusion

Given that atypical clinical or histopathologic presentations of GA may be indicative of an underlying malignancy, we recommend that clinicians consider a thorough and age-appropriate malignancy screening in patients with GA-like dermatitis, especially those who present unusually. These patients may be longitudinally monitored as dermatitis may precede clinically evident malignancy by several years.

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