Rare Case of Necrobiotic Xanthogranuloma on the Scalp

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PRACTICE **POINTS**

- In skin of color, necrobiotic xanthogranuloma can appear orange or brown compared to its yellow appearance in lighter skin types.
- When necrobiotic xanthogranuloma is suspected, a thorough malignancy workup should be conducted.

To the Editor:

Necrobiotic xanthogranuloma (NXG) is classified as a cutaneous non–Langerhans cell histiocytosis, often seen with monoclonal gammopathy of undetermined significance or multiple myeloma.[1] Clinically, it appears as a red or yellow plaque with occasional ulceration and telangiectasias, most commonly seen periorbitally and on the trunk. On pathology, NXG appears as necrobiosis, giant cells, and various inflammatory cells extending into the subcutaneous tissue.[2] In this article, we describe a rare presentation of NXG in location and skin type.

A 52-year-old woman with a history of systemic lupus erythematosus (SLE) presented with alopecia and a tender lesion on the scalp of 5 years' duration (Figure 1). The patient had no history of a similar lesion, and no other lesions were present. A biopsy performed at an outside clinic a few weeks to months prior to the initial presentation to our clinic showed NXG (Figure 2). Evaluation at our clinic revealed a 4x4-cm orange-brown annular plaque on the left parietal scalp. Serum and urine protein electrophoresis studies were negative. The patient reported she was up to date with recommended screenings such as mammography and colonoscopy.

We started the patient on topical triamcinolone and topical ruxolitinib and administered intralesional triamcinolone. She was already taking hydroxychloroquine and leflunomide for SLE. Three weeks later, she returned with improved symptoms and appearance (Figure 1). She

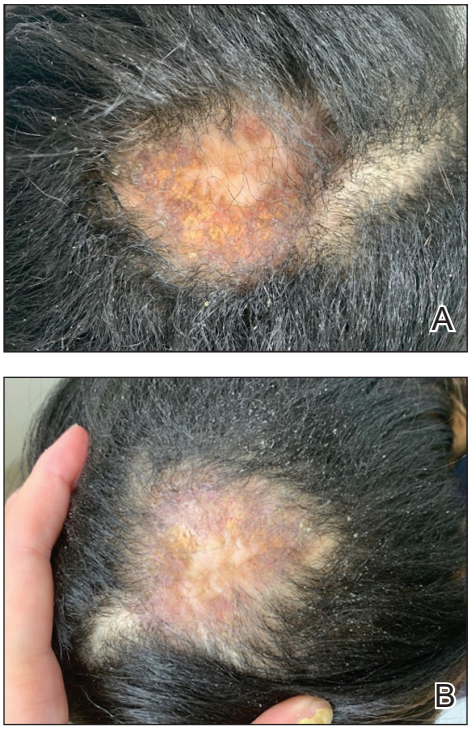

FIGURE 1. A and B, Necrobiotic xanthogranuloma of the scalp at baseline and 7 weeks after treatment with intralesional triamcinolone and topical ruxolitinib.

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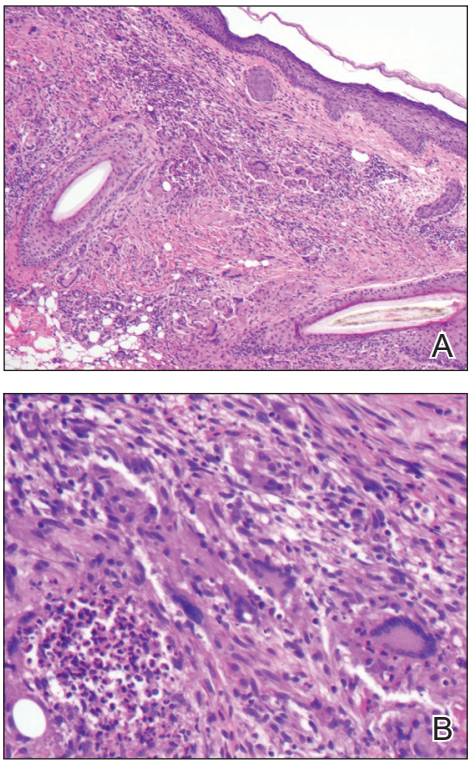

FIGURE 2. A and B, On H&E staining, the histologic sections demonstrated an uninvolved epidermis with marked necrobiosis and foci of xanthogranulomatous infiltration throughout the dermis with extension to subcutaneous fat. The xanthogranulomatous infiltration was comprised of epithelioid to ovoid foamy histiocytes in association with Touton-type giant cells. There was a background of lymphocytes, plasma cells, and neutrophils.

remained on intralesional triamcinolone and ruxolitinib and continues to experience improvement.

Necrobiotic xanthogranuloma is rare and typically is associated with monoclonal gammopathy.[2] In one study, 83 of 100 of patients with NXG presented with or were found to have a monoclonal gammopathy.[2] In another study, paraproteinemia was detected in 82.1% of patients.[3] The majority of case reports and systematic reviews detail periorbital or thoracic lesions.[4] The location on the scalp and lack of association with paraproteinemia make this a rare presentation of NXG. Studies may be warranted to explore any association of SLE with NXG if more cases present.

In a multicenter cross-sectional study and systematic review of 235 patients with NXG, 87% were White, 12% were Asian, and only 1% were Black or African American.[3] The limited representation of skin of color raises concern for the possibility of missed diagnoses and delays in care.

Treatment of NXG often is multimodal with use of intravenous immunoglobulin, oral steroids, chlorambucil, melphalan, and other alkylating agents, and response is variable.[3-6] Recent studies show treatment effectiveness with Janus kinase inhibitors in granulomatous dermatitides.[7-9] As our patient was not responding to prior treatments, we decided to try ruxolitinib, and she has continued to improve with it.[10,11] Interestingly, the patient experienced continued improvement with intralesional triamcinolone, which is not often reported in the literature.[2-6] Overall, NXG is an extremely rare condition that requires special care in workup to rule out paraproteinemia and a thoughtful approach to treatment modalities.

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