Irregular Yellow-Brown Plaques on the Trunk and Thighs

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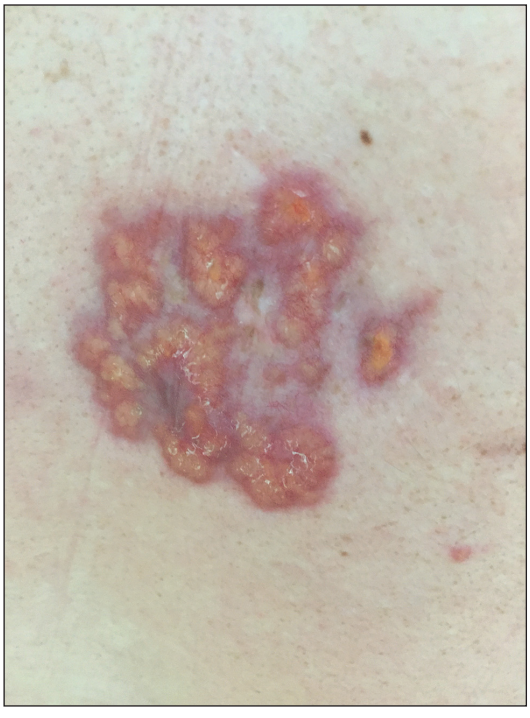

A 40-year-old man presented with tender lesions on the back, abdomen, and thighs of 10 years' duration. His medical history was remarkable for follicular lymphoma treated with chemotherapy and a monoclonal gammopathy of uncertain significance diagnosed 5 years after the onset of skin symptoms. Physical examination revealed numerous irregularly shaped, yellow plaques on the back, abdomen, and thighs with overlying telangiectasia. A single lesion was noted to extend from a scar.

WHAT'S THE **DIAGNOSIS?**

a. gout
b. Langerhans cell histiocytosis
c. necrobiotic xanthogranuloma
d. Rosai-Dorfman disease
e. sarcoidosis

PLEASE TURN TO PAGE 15 FOR THE DIAGNOSIS

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THE **DIAGNOSIS:** Necrobiotic Xanthogranuloma

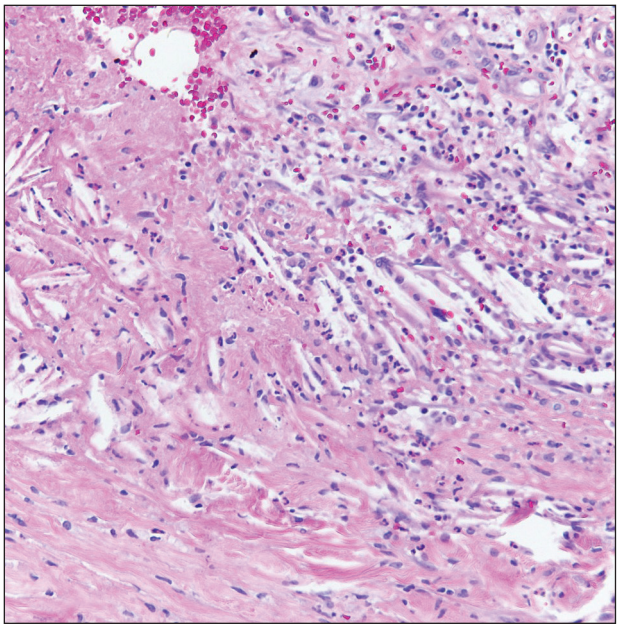
A 4-mm punch biopsy was performed for routine stain with hematoxylin and eosin. The differential diagnosis included sarcoidosis, necrobiosis lipoidica, xanthoma disseminatum, and multicentric reticulohistiocytosis. Histopathologic examination demonstrated a dermal infiltrate of foamy histiocytes and neutrophils (Figure). There were surrounding areas of degenerated collagen containing numerous cholesterol clefts. After clinical pathologic correlation, a diagnosis of necrobiotic xanthogranuloma (NXG) was elucidated.

The patient was referred to general surgery for elective excision of 1 or more of the lesions. Excision of an abdominal lesion was performed without complication. After several months, a new lesion reformed within the excisional scar that also was consistent with NXG. At further dermatologic visits, a trial of intralesional corticosteroids was attempted to the largest lesions with modest improvement. In addition, follow-up with hematology and oncology was recommended for routine surveillance of the known blood dyscrasia.

Necrobiotic xanthogranuloma is a multisystem non–Langerhans cell histiocytic disease. Clinically, NXG is characterized by infiltrative plaques and ulcerative nodules. Lesions may appear red, brown, or yellow with associated atrophy and telangiectasia.[1] Koch et al[2] described a predilection for granuloma formation within preexisting scars. Periorbital location is the most common cutaneous site of involvement of NXG, seen in 80% of cases, but the trunk and extremities also may be involved.[1,3] Approximately half of those with periocular involvement experience ocular symptoms including proptosis, blepharoptosis, and restricted eye movements.[4] The onset of NXG most commonly is seen in middle age.

Characteristic systemic associations have been reported in the setting of NXG. More than 20% of patients may exhibit hepatomegaly. Hematologic abnormalities, hyperlipidemia, and cryoglobulinemia also may be seen.[1] In addition, a monoclonal gammopathy of uncertain significance is found in more than 80% of NXG cases. The IgG κ light chain is most commonly identified.[2] A foreign body reaction is incited by the immunoglobulin-lipid complex, which is thought to contribute to the formation of cutaneous lesions. There may be associated plasma cell dyscrasia such as multiple myeloma or B-cell lymphoma in approximately 13% of cases.[2] Evaluation for underlying plasma cell dyscrasia or lymphoproliferative disorder should be performed regularly with serum protein electrophoresis or immunofixation electrophoresis, and in some cases full-body imaging with computed tomography or magnetic resonance imaging may be warranted.[1]

Treatment of NXG often is unsuccessful. Surgical excision, systemic immunosuppressive agents, electron beam radiation, and destructive therapies such as cryotherapy may be trialed, often with little success.[1] Cutaneous regression has been reported with combination treatment of high-dose dexamethasone and high-dose lenalidomide.[5]



Punch biopsy results demonstrated a dermal infiltrate of foamy histiocytes and neutrophils surrounding areas of degenerated collagen containing numerous cholesterol clefts (H&E, original magnification ×100).

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