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

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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, necrobiotic 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. Koch et al 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. Approximately half of those with periocular involvement experience ocular symptoms including proptosis, blepharoptosis, and restricted eye movements. 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. 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. 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. 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.

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. Cutaneous regression has been reported with combination treatment of high-dose dexamethasone and high-dose lenalidomide.

REFERENCES