IMAGES IN DERMATOLOGY

Disseminated necrobiotic xanthogranuloma: A cutaneous manifestation of monoclonal gammopathy

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Figure 1: Confluent infiltrated plaques ranging in color from yellowish-brown to erythematous-violaceous, with irregular, polycyclic, and arcuate borders, distributed over the breasts and thoracoabdominal region. Areas of central clearing are evident, along with a rounded lesion with a crusted surface on the lower abdomen.

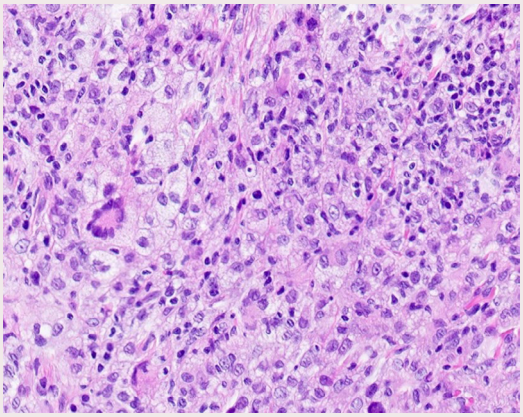


Figure 2: Skin biopsy showing a dermal xanthogranulomatous infiltrate composed of foamy histiocytes and multinucleated giant cells, associated with collagen necrobiosis (hematoxylin and eosin stain).

A 67-year-old woman presented with an 8-month history of multiple cutaneous lesions. Dermatological examination revealed firm, infiltrated plaques and nodules ranging in color from yellowish-brown to erythematous-violaceous, with dark brown areas, irregular, polycyclic, and arcuate borders, and a tendency to coalesce. The lesions predominantly involved the breasts and thoracoabdominal region, with additional lesions on the arms, neck, and periocular area. Figure 1 shows extensive confluent plaques with areas of central clearing and a rounded lesion with a crusted surface on the lower abdomen. This chromatic variability is consistent with necrobiotic xanthogranuloma, whose lesions do not invariably exhibit the characteristic yellow-orange discoloration.

Laboratory investigations revealed an IgG kappa monoclonal gammopathy, with a monoclonal protein (M-protein) level of 2.8 g/dL. Skin biopsy demonstrated a dermal xanthogranulomatous infiltrate composed of foamy histiocytes, multinucleated giant cells, and extensive areas of collagen necrobiosis (Figure 2). Congo red staining showed no evidence of amyloid deposition, and special stains for microorganisms were negative. Correlation of the clinical presentation, histopathological findings, and paraproteinemia established the diagnosis of necrobiotic xanthogranuloma.[1] Recently, consensus diagnostic criteria have been validated against its major clinicopathological mimickers.[2]

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Necrobiotic xanthogranuloma is a rare, chronic non-Langerhans cell histiocytosis that is strongly associated with monoclonal gammopathies and plasma cell dyscrasias. Multidisciplinary management involving both dermatology and hematology, together with long-term hematologic surveillance, is essential because the underlying clonal disorder may precede, coincide with, or develop after the onset of the cutaneous manifestations.[3-5] The differential diagnosis includes necrobiosis lipoidica, generalized granuloma annulare, xanthomas, and other non-Langerhans cell histiocytoses.

Treatment should be individualized according to the extent of cutaneous involvement, extracutaneous manifestations, and the underlying hematologic disorder. Current evidence is derived primarily from case series and systematic reviews. Intravenous immunoglobulin (IVIG) and systemic corticosteroids are among the therapies with the strongest supporting evidence, together with plasma cell clone-directed therapies when indicated.[6-7]

INFORMED CONSENT

Written informed consent was obtained from the patient for the publication of her clinical information and images. Patient confidentiality was maintained, and no identifying information was included.

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