

The Eyes Never Lie: A Rare Cutaneous Manifestation of Hematological Malignancy

Os Olhos Não Mentem: Uma Manifestação Cutânea Rara de Neoplasia Hematológica

Keywords: Multiple Myeloma; Necrobiotic Xanthogranuloma
Palavras-chave: Mieloma Múltiplo; Xantogranuloma Necrobiótico

A 92-year-old female patient was referred to our Dermatology department for symmetrical yellow plaques around the eyes, which were asymptomatic but had grown for over a decade (Fig. 1A). Her medical history was relevant for monoclonal gammopathy of undetermined significance, diagnosed approximately a decade before the cutaneous eruption first developed. A skin biopsy was performed that showed numerous foamy histiocytes occupying the dermis, as well as giant multinucleated Touton cells (Fig. 1B). This is consistent with the diagnosis of xanthogranuloma *necrobioticum* (NXG), a non-Langerhans cell histiocytosis, belonging to the mucocutaneous group. It manifests as yellowish indurated plaques or nodules (that can ulcerate), usually around the eyes, although they can also arise on the torso and limbs. It is notably associated with underlying systemic disorders, especially monoclonal gammopathies (up to 80% of cases).¹⁻⁴ Clinically, NXG should be distinguished from xanthelasma, which presents as multiple soft, flat, yellow papules and plaques, symmetrically located on the medial eyelids of adults, which differ from the bigger confluent lesions of NXG. The characteristic yellow colour of NXG occurs due to cholesterol deposition in macrophages, as a result of a complex interaction between immunoglobulins and lipoproteins.⁵ The histopathological features of NXG include foamy macrophages, multinucleated giant cells (Touton cells) and necrobiotic collagen degeneration (the latter two features are absent in xanthelasma).^{4,6} The diagnosis

of NXG requires two major criteria: 1) yellow/orange cutaneous papules, plaques or nodules; 2) suggestive histopathology and one of two minor criteria; 3) paraproteinemia/lymphoproliferative disease; 4) periorbital distribution of lesions – all four are present in our case.⁷

AUTHOR CONTRIBUTIONS

GAS: Conception and writing of the manuscript.

LS: Literature review, writing of the manuscript.

JA: Critical review of the manuscript.

All authors approved the final version to be published.

PROTECTION OF HUMANS AND ANIMALS

The authors declare that the procedures were followed according to the regulations established by the Clinical Research and Ethics Committee and to the Helsinki Declaration of the World Medical Association updated in October 2024.

DATA CONFIDENTIALITY

The authors declare having followed the protocols in use at their working center regarding patients' data publication.

PATIENT CONSENT

Obtained.

COMPETING INTERESTS

The authors have declared that no competing interests exist.

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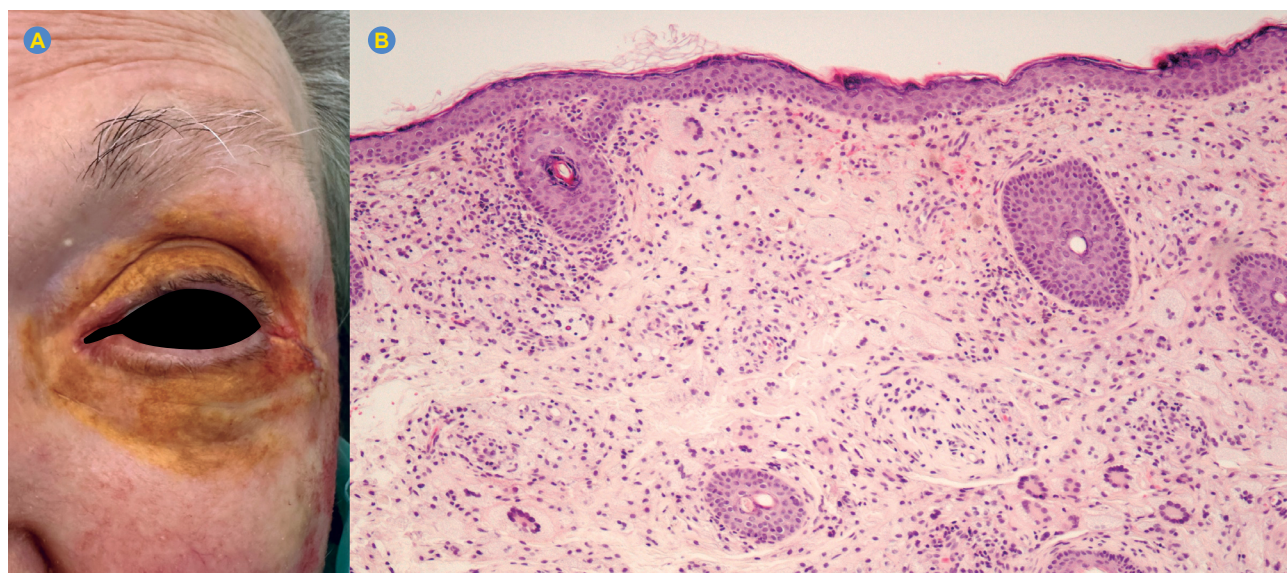


Figure 1 – Asymptomatic yellow plaques on the peri-orbital area (A). Foamy histiocytes occupying the dermis, as well as giant multinucleated Touton cells (B).

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