

QUIZ SECTION

Self-healing Eyelid Nodule: A Quiz

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A 53-year-old woman presented with an isolated and non-tender necrotic nodule on her left eyelid, which had begun as a small rapid-growing inflammatory papule 3 weeks earlier. Physical examination revealed a 15 mm, erythematous, firm but mobile nodule with a crusted and necrotic core (Fig. 1A). No other cutaneous or mucosal lesions were identified. There was neither regional lymphadenopathy nor systemic involvement and ophthalmological findings were otherwise unremarkable. All laboratory and microbiological studies showed normal or negative results.

The patient had a previous 6-month history of disseminated and self-healing crops of similar cutaneous lesions affecting her upper trunk and extremities, which were biopsied months ago (Fig. 1B, C).

The eyelid nodule resolved itself spontaneously within 2 weeks. There was no recurrence during the following year.

What is your diagnosis?

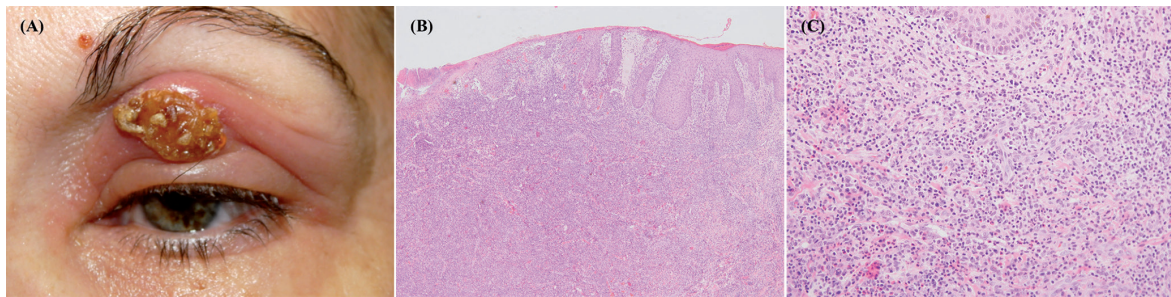


Fig. 1. (A) Isolated erythematous and crusted nodule on the upper left eyelid. (B) The microscopic examination of an incisional skin biopsy found the presence of a diffuse wedge-shaped dermal infiltrate (H&E stain, $\times 40$), (C) containing large atypical lymphocytes within a dense inflammatory background of smaller lymphocytes, histiocytes, neutrophils, and abundant eosinophils (H&E stain, $\times 200$).

ANSWERS TO QUIZ

Self-healing Eyelid Nodule: A Comment

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Diagnosis: Lymphomatoid papulosis (LyP), type A

Immunohistochemical analysis of the skin biopsy showed the anaplastic lymphocytes to be CD4⁺ and CD30⁺ (Fig. 2), i.e. consistent with LyP. Along with cutaneous anaplastic large cell lymphoma (cALCL), LyP belongs to the continuous spectrum of cutaneous CD30⁺ lymphoproliferative disorders, the second most common group of primary cutaneous T-cell lymphomas (CTCL). Originally described by Macaulay (1) as a “self-healing, rhythmic and paradoxical eruption, histologically malignant but clinically benign”, it is currently well known that LyP is a relapsing but indolent skin disease that may be histologically indistinguishable from aggressive cutaneous lymphocytic proliferations (2–10). Achieving an accurate clinicopathological correlation is therefore absolutely mandatory for a conclusive diagnosis.

Clinically, LyP is characterised by outbreaks of papular or nodular lesions that usually become necrotic and typically heal spontaneously within a few weeks leaving an atrophic or hypopigmented scar (1–10). Although this entity may be easily confused with other skin diseases such as arthropod-bites, prurigo, pyodermas, pityriasis lichenoides, or even cutaneous malignancies, its chronic recurrent course and especially its self-healing nature are both the differential clinical clues. Widespread cutaneous forms consisting of papulonecrotic cutaneous lesions scattered along the trunk and extremities are the most common clinical appearance of LyP. Atypical single presentations on unusual locations such as the scalp, face, or mucose membranes have occasionally been reported. However, isolated eyelid involvement by LyP seems to be extremely rare. Since Lemagne et al. (5) described the first patient with a rapidly growing ulcerated tumour of the outer canthus as the presenting feature of this condition in 1987, a total of 6 cases of eyelid LyP have been reported worldwide (5–10). Three of them showed simultaneous lesions over their head, neck, trunk, and limbs (6, 9, 10) while the rest consisted of solitary eyelid LyP lesions characteristically accompanied by a previous or subsequent history of several waxing and waning necrotic papulonodules (5, 7, 8), as we noticed in our patient. This important finding is probably the most helpful clinical trait to consider LyP when atypical single presentations appear, differentia-

ting it from cALCL and other much more common and aggressive papulonecrotic eyelid proliferations, such as keratoacanthoma and squamous cell carcinoma (2).

As proposed by Macaulay (1), the diagnosis of LyP may be also quite challenging for dermatopathologists. To date, 5 histopathological subtypes of LyP, respectively named from A to E, have been defined (2–10). Overlapping patterns in the same specimen or in the same patient are observed in up to 10% of skin biopsies generally between the types A and C, the most common variants of LyP. The histiocytic pattern (type A) is characterised by a wedge-shaped mixed inflammatory infiltrate containing a variable number of large atypical CD30⁺ lymphocytes. In contrast, type C LyP usually shows a predominant population of monomorphic large CD30⁺ cells mimicking ALCL. The least frequent, but most questioned variant, type B LyP, is considered as a histologic simulator of mycosis fungoides (MF) due to its epidermotropism and its typical band-like infiltrate of cerebriform pleomorphic lymphocytes. Especially among those patients with CD30⁻ type B LyP, a detailed clinical history is required to differentiate it from a papular variant of MF. The recently described patterns D and E may resemble highly aggressive primary CTCL, such as the epidermotropic CD8⁺ cytotoxic and the angi-destructive variants (3, 4).

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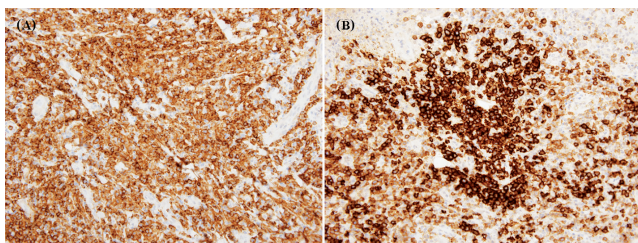


Fig. 2. Immunohistochemical staining of the biopsy shown in Fig. 1 demonstrated (A) anaplastic lymphocytes strongly positive for T-cell antigens (CD4 stain, ×200), and (B) for the lymphoid activation antigen (CD30 stain, ×200).