The relevance of complete imaging investigations in lymphomatoid papulosis

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In July 2020, a 47 years old women with silent past medical history referred because of the abrupt onset of grouped, small, flesh-colored papules and nodules localized mostly on the extremities and less on the trunk. The majority of the lesions were crusted and showed a necrotic evolution. Coexistence of lesions in different stages was noticed (Figure 1). Blood tests showed no relevant alterations, with CRP and ESR within normal ranges. A skin biopsy of a nodule was performed. Histopathology showed an irregular epidermal hyperplasia with pseudocarcinomatous features with focal parakeratosis and neutrophils. An edematous dermis was infiltrated by a mix of medium-large atypical partly pleomorphic, partly anaplastic CD3+ CD4+ CD30+ ALK- MUM-1+/- lymphocytes, neutrophils and eosinophils (Figure 2a-d,g). Staging investigations resulted negative for nodal involvement and the diagnosis of type C lymphomatoid papulosis (LyP) was made based on clinicopathological correlation.

The total body CT scan revealed a thickened and hyperemic wall gut at the level of the descending colon/proximal sigma and few lymphadenopathies in the gut. 18F-FDG PET/CT disclosed intense tracer uptake corresponding to a lesion of descending intestine (Figure 2e,f). Colonoscopy showed an obstructive malignant-looking polyp at 30cm from the anus, and biopsies from the lesion confirmed the diagnosis of ulcerated mucinous adenocarcinoma of the sigma, infiltrating the hole gut wall up to the perivisceral adipose tissue. The patient did not report gastrointestinal symptoms and denied familiar history for colon cancer. She underwent laparoscopic left hemicolectomy with sampling of sigmoidal lymph nodes, permitting a final staging pT3 pN1b M0. After multidisciplinary discussion, 4 cycles of adjuvant chemotherapy with oxaplatin and capecitabine was administrated from December 2020 to March 2022 with no severe side effect. Skin lesion progressively resolved and at last follow-up visit in September 2022 LyP is still in remission.

LyP belongs to the spectrum of primary cutaneous CD30+ lymphoproliferative disorders. Lyp presents with recurrent, self-healing papulonecrotic or nodular skin lesions typically in different evolution stages and a relapsing course that can affect patients of any age, with an incidence peak during the fifth decade. The main differential diagnosis is with primary cutaneous anaplastic large lymphoma (ALCL) and other cutaneous T-cell lymphomas, and it is upon clinic-pathologic correlation. Histopathology of LyP is extremely variable; in addition to the 3 original subtypes (LyP types A, B, and C), the 2018 update of the WHO-EORTC classification also recognizes the more recently described types D, E and with DUSP-IRF4 rearrangements.[1]

It has been demonstrated that patients with LyP have a lifelong increased risk (RR,11.9; 95% CI, 8.3–15.5) of developing hematological malignancies (HM). A recent Dutch multicenter cohort study of 504 patients with LyP showed that an associated HM was reported in 15.5% of cases (6.2% of cases associated with micosis fungoides, 5.8% with ALCL, 3% malignancies of B-cell and myeloid origin).[2] Moreover, non-HM were observed in 32% of patients with intestinal cancer in 3% of cases and a RR of 2.4 (CI, 1.2-3.7). After an extensive review of the English literature, we were able to find only one case report of Eber-positive LyP and colon carcinoma in a heart transplant recipient.[3]

Our experience underlines the relevance of performing accurate staging investigations in all LyP patients at the time of diagnosis, even in those that are asymptomatic, like our patient, not only for complete staging and therefore correct classification but also for cancer screening. Clinicians must be aware of the increased risk of Lyp patients to develop HM and non-HM compared to general population at the time of

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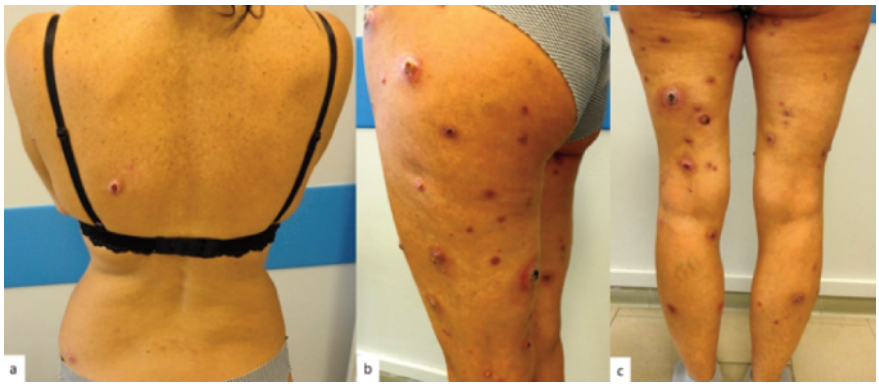

Figure 1. Coexistence of papules and nodules with and without necrotic center and in different stages of evolution.

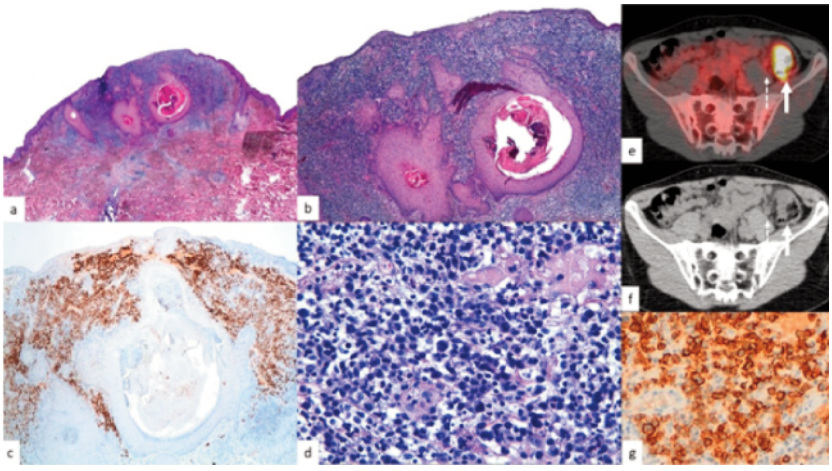

Figure 2. a, b) Irregular epidermal hyperplasia with pseudocarcinomatous features and dense nodular lymphoid infiltrate in the dermis (HE, 2x and 20x). c) Diffuse staining for CD30 (x20). d) medium-large atypical lymphocytes with partly pleomorphic, partly anaplastic morphology e)-f) 18F-FDG PET/CT (axial images) showed intense tracer uptake (e, arrow) corresponding to a lesion of descending intestine and a small area of faint uptake (e, dotted arrow) corresponding to 10 mm locoregional lymph-node (f, dotted arrow) g) medium-large atypical lymphocytes stained for CD30 (x40).

diagnosis and at follow-up and therefore Lyp patients must regularly follow cancer screening programs.

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