LUPUS ERITEMATOS TUMID – DISCUȚIE PE UN CAZ CLINIC

TUMID LUPUS ERYTHEMATOSUS – DISCUSSION ON A CLINICAL CASE

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Rezumat

Lupusul eritematos tumid (TLE) este o afecțiune inflamatorie rară a pielii, caracterizată prin plăci eritematoase, edematoase și fotosensibilitate ridicată. Astăzi, deși clasificarea este încă controversată, TLE este considerat un subtip separat de lupus eritematos cutanat (CLE) cu un curs clinic benign și intermitent (CLE intermitent, ICLE) și doar rar asociat cu lupus eritematos sistemic (LES) [1].

În această lucrare, prezentăm cazul unui pacient în vârstă de 67 de ani care a fost internat în secția noastră cu erupții pruriginoase recurente eritemato-violacee cu aspect urticarian și edematos, localizate în zonele foto-expuse în urma expunerii prelungite și repetate la soare. Pe baza corelației dintre prezentarea clinică, constatările histopatologice și evaluările imunologice, a fost stabilit un diagnostic de lupus tumidus, o variantă rară a lupusului eritematos cutanat.

Cuvinte cheie: *lupus eritematos tumidus, forme rare de lupus, lupus intermitent.*

Summary

Tumid lupus erythematosus (TLE) is a rare inflammatory skin condition characterized by erythematous, edematous plaques and high photosensitivity. Today, although the classification is still controversial, TLE is considered a separate subtype of cutaneous lupus erythematosus (CLE) with a benign and intermittent clinical course (intermittent CLE, ICLE) and only rarely associated with systemic lupus erythematosus (SLE) [1].

In this paper, we present the case of a 67-year-old patient who was admitted to our department with recurrent pruritic erythematous-violaceous eruptions exhibiting a urticarial and edematous appearance, localized to photo-exposed areas following prolonged and repeated sun exposure. Based on the correlation between clinical presentation, histopathological findings, and immunological assessments, a diagnosis of lupus tumidus was established, a rare variant of cutaneous lupus erythematosus.

Keywords: *lupus erythematosus tumidus, rare forms of lupus, intermittent lupus.*

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Introduction

The term *tumid lupus erythematosus* (TLE) was first introduced in the scientific literature by Gougerot and Burnier in 1930 [2] to describe erythematous, smooth, infiltrated lesions without scaling, atrophy, ulceration or other superficial alterations. However, the condition received little attention in the following years, with only a few isolated case reports published, and it was not included in the classification of cutaneous lupus erythematosus (CLE) proposed by Gilliam in the 1970s.[3].

Over the past decade, several researchers have demonstrated a growing interest in TLE and have undertaken efforts to characterize the condition. Consequently, TLE has been defined as a distinct subtype of CLE with its own clinical, prognostic and microscopic features. Additionally, it has become evident that the prevalence of TLE is likely underestimated. In fact, there is a widely held view that TLE may be even more common than classic discoid lupus erythematosus (DLE) [4]. Nevertheless, several unresolved issues remain, including its classification, certain histopathological characteristics and its differential diagnosis from other dermatologic conditions.

TLE is considered a rare disease. Its exact prevalence and incidence remain unknown, as its benign nature and clinical course, marked by spontaneous relapses and remissions, may contribute to underreporting. [5].

Epidemiological data indicate an equal prevalence between sexes, although some studies have reported a slight female predominance. For instance, a 2013 study conducted by the European Society of Cutaneous Lupus Erythematosus (EUSCLE) found that women accounted for 60% of cases. [6].

The mean age of disease onset ranges between 36.4 and 38.5 years, which is comparable to that of discoid lupus erythematosus (DLE) [7], [8]. Notably, although rare, cases have also been reported in pediatric patients, presenting with the same clinical and histopathological characteristics as those observed in adults.[9]

Case-report

We present the case of a 67-year-old male patient from a rural environment suffering from essential hypertension (maximum systolic blood pressure of 180 mmHg, under treatment with indapamide 1,5 mg, candesartan 16 mg), coronary ischemic heart disease under treatment with clopidogrel 75 mg and trimetazidinum 70 mg/day, well as dyslipidemia treated using statins. Regarding lifestyle and hygienic-dietary habits, it is important to note that the patient is a smoker with a 40 pack-year history, denies alcohol consumption and does not work in a toxic environment. However, it is relevant to mention that the patient is employed in agriculture and is continuously exposed to solar radiation. The patient denies any significant hereditary or collateral medical history.

The general clinical examination showed the chest wall symmetric and without deformity, a bilaterally present vesicular murmur, without rales, rhytmic heart spunds without valvular murmurs, BP=145/86 mmHg, AV=65/min. The clinical examination of digestive system revealed a supple abdomen, mobile with breathing, not painful spontaneously and upon palpation, affirmative regular bowel habits. Otherwise, the general clinical examination was within normal limits.

Regarding laboratory analyses, the complete blood count showed a low creatinin clearence (59, 14 mL/min), leukocyturia, the presence of urobilinogen in urine and microalbuminuria 0,3 g/L.

Regarding the patient's medical history, the first presentation to our clinic occurred in 2019 due to the appearance of relatively extensive plaques on the face, neckline, anterior and posterior thorax and upper limbs. These lesions developed following repeated sun exposure without chemical or physical photoprotection, particularly on the upper body, as the patient worked in an agricultural setting.

During the anamnesis, it was noted that the patient had been experiencing intermittent erythematous-violaceous, mildly pruritic eruptions for approximately 10 years, without associated atrophy or scarring. The patient

reported a previous diagnosis of systemic lupus erythematosus (SLE) in 2004, based on a biopsy performed on the nasal pyramid, with immunological findings at that time indicating the presence of anti-double-stranded DNA (anti-dsDNA) antibodies. However, the patient was unable to provide any medical documentation to confirm this diagnosis and had not received any treatment for the condition until 2019.

The dermatological examination revealed the presence of extensive, mildly pruritic, erythematous-violaceous plaques on the face (Figure 1, Figure 2). Some areas exhibited a vaguely polycyclic contour, while others appeared slightly edematous, particularly on the face. The face and neckline displayed intense erythema, whereas the plaques on the thorax and upper limbs were pinkish and mildly infiltrated (Figure 3).

A skin biopsy was performed on erythematous lesions from the facial region. Histopathological analysis revealed an epidermis with subtle keratotic plug formation and minimal ballooning degeneration at the basal layer, along with a moderate perivascular and peri-adnexal chronic lymphomononuclear inflammatory infiltrate (Figure 4), findings suggestive of tumid lupus. Immunological investigations showed the absence of antinuclear antibodies (ANA) and anti-double-stranded DNA (anti-dsDNA) antibodies.

By integrating clinical, histological, and immunological data, a diagnosis of *tumid lupus erythematosus* was established.

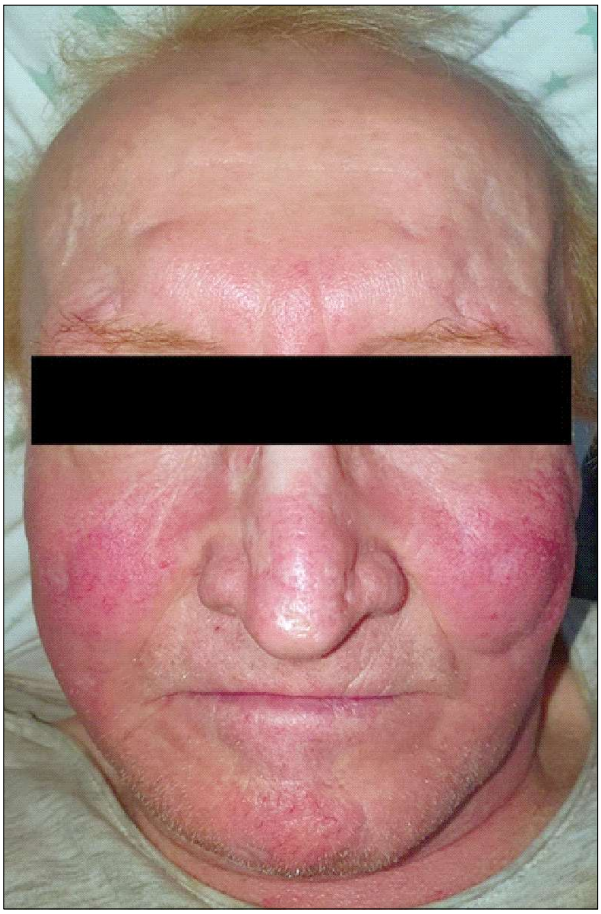

Figure 1. Erythematous-violaceous and edematous plaques on the most photoexposed areas of the face.

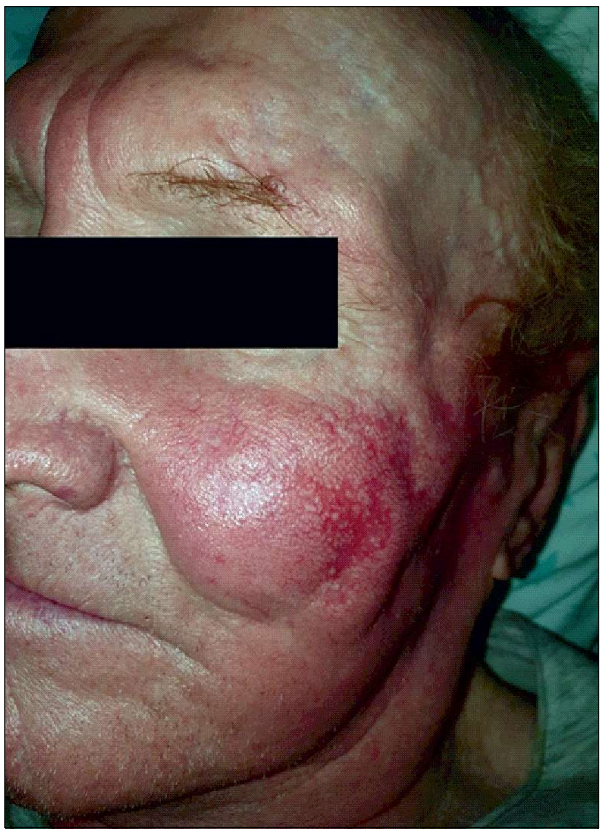

Figure 2. Erythematous-violaceous and edematous plaques on the most photoexposed areas of the face.

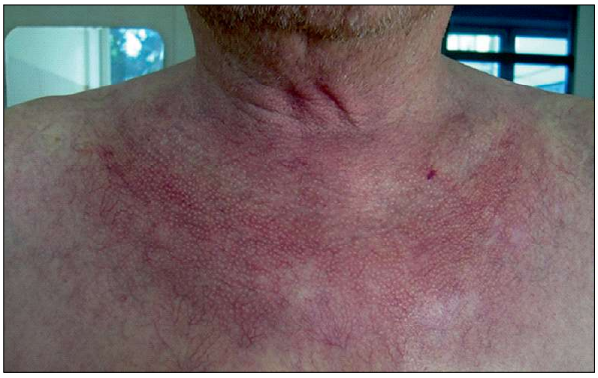

Figure 3. Erythematous, infiltrated plaques on the upper anterior thorax and neckline

The patient was initiated on topical treatment with a medium-potency corticosteroid (mometasone cream) and systemic therapy with hydroxychloroquine at a dose of 400 mg/day until lesion resolution. Additionally, strict photoprotection against UVA, UVB, and blue light was recommended.

The patient's evolution was favorable, with near-complete resolution of cutaneous manifestations after 10 weeks of treatment.

In 2024, the patient presented again to our department with an erythematous-violaceous, mildly pruritic, urticarial-like, edematous eruption on the face, without scaling or scarring. A well-demarcated, intensely red-violaceous plaque was observed on the neckline. Reassessment of immunological parameters revealed an equivocal ANA titer and weakly positive anti-mitochondrial antibodies (AMA-M2).

The patient underwent a course of corticosteroid therapy with prednisone at a dose of 0.5 mg/kg/day for seven days, followed by a gradual taper. Additionally, treatment with hydroxychloroquine (Plaquenil) at 400 mg/day and topical corticosteroids was continued. The cutaneous lesions showed a favorable evolution during hospitalization.

Subsequently, the patient remained compliant with treatment (Plaquenil 400 mg/day), and at the most recent dermatological examination, only mild facial erythema was observed, with no edema or associated symptoms.

Given the presence of weakly positive anti-mitochondrial antibodies (AMA-M2), the patient

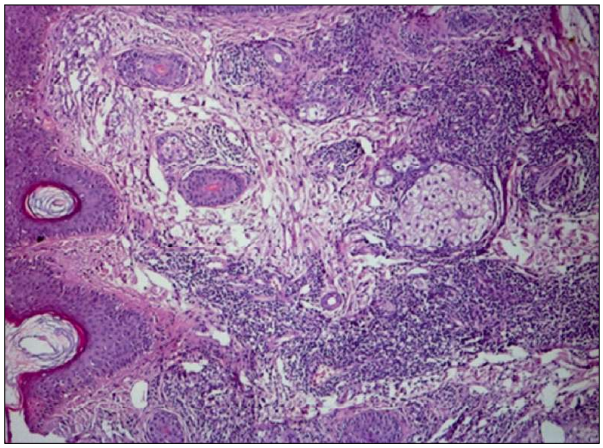

Figure 4. Histopathological examination Hematoxylin-eosin staining x 100- epidermis with subtle keratotic plug formation and minimal ballooning degeneration at the basal layer, along with a moderate perivascular and peri-adnexal chronic lymphomononuclear inflammatory infiltrate.

was referred for a specialized gastroenterology consultation. However, primary biliary cholangitis has been excluded. Additionally, due to urinary abnormalities, the patient was directed to nephrology for further evaluation, with additional investigations planned in the following period.

Discussions

The diagnosis of tumid lupus erythematosus (TLE) can be challenging, given that its lesions lack highly specific features and often overlap with other forms of lupus erythematosus.

It is important to consider the patient's status as a chronic smoker. Studies have reported an increased prevalence of smoking among patients with tumid lupus erythematosus (TLE) compared to the general population. [10]. Thus, the frequent recurrences observed in this patient may, to some extent, be attributable to smoking.

TLE is a subtype of cutaneous lupus erythematosus (CLE) characterized by a high degree of photosensitivity. The patient's occupational exposure to sunlight without adequate UVA/UVB physical and chemical photoprotection constitutes a significant predisposing factor for disease recurrence. Given that our recommendations were not fully adhered to, the

patient's disease exhibited multiple recurrences between 2019 and 2024.

Differential diagnosis posed a significant challenge. Three dermatological conditions closely resemble TLE both clinically and histopathologically, making distinction difficult: Jessner's lymphocytic infiltration, polymorphic light eruption, and reticular erythematous mucinosis.

Jessner's lymphocytic infiltration is described as a non-scarring dermatological condition that primarily affects the facial region. Histopathological examination under light microscopy reveals a periadnexal and perivascular lymphocytic infiltrate, without epidermal involvement.[11] Classically, patients do not exhibit photosensitivity and show no response to antimalarial therapy, which led us to exclude this condition as a possible diagnosis for our patient.

Polymorphic light eruption (PLE) is a photodermatosis that manifests with a broad spectrum of cutaneous lesions, which may present as vesicular, pseudo-vesicular, papular or plaque-like lesions, often resembling tumid lupus erythematosus (TLE). Unlike TLE, PLE lesions develop shortly after sun exposure and resolve within a few days in the absence of further ultraviolet exposure[7]. In contrast, our patient's lesions were persistent and occurred outside of the summer months.

Reticular erythematous mucinosis (REM) predominantly affects young women and typically presents as a reticulated macular or papular erythema. Histopathological examination of lesions reveals a periadnexal and perivascular lymphocytic infiltrate associated with interstitial mucin deposition. Marked photosensitivity is a common clinical feature, leading some authors to classify REM as a variant of chronic cutaneous lupus erythematosus (CCLE) or TLE [8]. In our case, histopathological findings were conclusive in excluding this diagnosis.

Conclusions

This case highlights the diagnostic complexity of lupus erythematosus tumidus (TLE), a rare form of cutaneous lupus erythematosus, whose clinical manifestations may closely mimic other dermatological conditions. In our patient, the clinical course necessitated histopathological reevaluation, ultimately confirming the diagnosis of TLE. This underscores the importance of continuous monitoring and treatment adaptation based on clinical progression. The differential diagnosis of TLE remains challenging due to significant clinical and histopathological overlap with other lupus subtypes and similar dermatological disorders. However, despite these challenges, the establishment of an accurate diagnosis and the implementation of an appropriate therapeutic regimen resulted in a favorable clinical outcome for our patient.

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Conflict of interest
NONE DECLARED

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