

# Tumid Lupus Erythematosus: A Rare Case Report

Dr. J Mirudula Josena<sup>1</sup>, Dr. Abinaya Kuberan<sup>2</sup>, Dr. Soundarya S<sup>3</sup>, Dr. Jayakar Thomas<sup>4</sup>

<sup>1</sup>Junior Resident, Department of Dermatology Venereology and Leprosy, Chettinad Hospital and Research Institute (Corresponding Author)

<sup>2</sup>Junior Resident, Department of Dermatology Venereology and Leprosy, Chettinad Hospital and Research Institute

<sup>3</sup>Assistant Professor, Department of Dermatology Venereology and Leprosy, Chettinad Hospital and Research Institute

<sup>4</sup>Emeritus Professor, Department of Dermatology Venereology and Leprosy, Chettinad Hospital and Research Institute

**Abstract:** *Tumid lupus erythematosus, (TLE) is a rare type of Lupus erythematosus. It is currently considered as a subtype of Cutaneous lupus erythematosus. TLE differs from other variants of CLE as its association with systemic lupus erythematosus (SLE) is rare. TLE classification and exact features remains a topic of debate. The common differentials based on the clinical and/or histopathological features include Lymphocytic infiltrate of Jessner, Subacute Cutaneous lupus erythematosus, polymorphous light eruption, granuloma faciale, pseudo lymphoma/lymphocytoma cutis of the skin. In this article, we present a case report of Tumid Lupus Erythematosus (TLE) and review the literature to differentiate it from the above - mentioned differentials.*

**Keywords:** Tumid lupus erythematosus, Lupus erythematosus tumidus, Cutaneous lupus erythematosus, Subacute cutaneous lupus erythematosus, Lymphocytic infiltrate of Jessner, Polymorphous light eruptions.

## 1. Introduction

Lupus erythematosus is an autoimmune disease with a wide clinical spectrum ranging from mild cutaneous lesions to life threatening systemic disease. The most affected organ is skin. It usually affects women between the age of 20 and 40 years [1]. Tumid lupus erythematosus is an uncommon and photosensitive inflammatory skin disorder. It clinically presents with an annular, indurated, erythematous, asymptomatic edematous plaques over photo - exposed areas. Clinically TLE may be a diagnostic challenge as it can be easily confused with other photo dermatoses. Histopathologically, the epidermis is usually spared associated with perivascular lymphohistiocytic infiltrate in the papillary and reticular dermis and interstitial mucin deposition [2]. Direct immunofluorescence is usually negative. The presence of photosensitivity, distribution of lesions, lack of systemic and serological abnormalities and absence of epidermal changes aids in the diagnosis. They usually heal without scarring or hyperpigmentation on treatment with antimalarials, topical steroids and strict photoprotection.

## 2. Case Report

A 58 - year - old, married woman came to our dermatology outpatient department with an erythematous tender nodule over the nose, left elbow and behind the ears for the past 1 month (Figure 1). These nodules appeared insidiously, and history of photosensitivity was present. She denied history

of similar lesions elsewhere in the body, oral ulcerations, fever, and joint pains. Systemic and mucocutaneous examination was insignificant. Regional lymph nodes were not enlarged. A clinical differential diagnosis of Sweet Syndrome (acute febrile neutrophilic dermatosis) Erythema elevatum diutinum, polymorphous light eruptions and Pseudo lymphoma/ lymphocytoma cutis were considered. A 4mm skin punch biopsy from the left elbow was taken and sent for microscopical examination which shows skin with follicular plugging, focal loss of rete ridges and pigment incontinence. Deeper dermis shows mucin deposition on staining with Alcian blue, necrobiosis of collagen fibers, perivascular and peri adnexal lymphohistiocytic infiltrate (Figure 2). Subcutaneous fat showed extensive areas of calcification. Further investigations of anti - nuclear antibody (ANA) titres were slightly increased 1: 80 (in low titre) and Negative for dsDNA.

Complete blood count, liver and renal functions and blood sugar levels were within normal limits. Chest x - ray, ECG, Urine routine, 24 - hour urine protein were unremarkable. Accordingly, a diagnosis of Tumid lupus erythematosus was made. The patient was started on Hydroxychloroquine sulphate at dose of 200mg daily after obtaining ophthalmology clearance. She was also administered with topical corticosteroid Mometasone furoate cream 0.1% and was advised to take adequate photoprotection including broad spectrum sunscreen. After 2 weeks of treatment patient improved symptomatically better (Figure 3)



Figure 1: Erythematous tender nodule over the nose (Left) and elbow (Right)

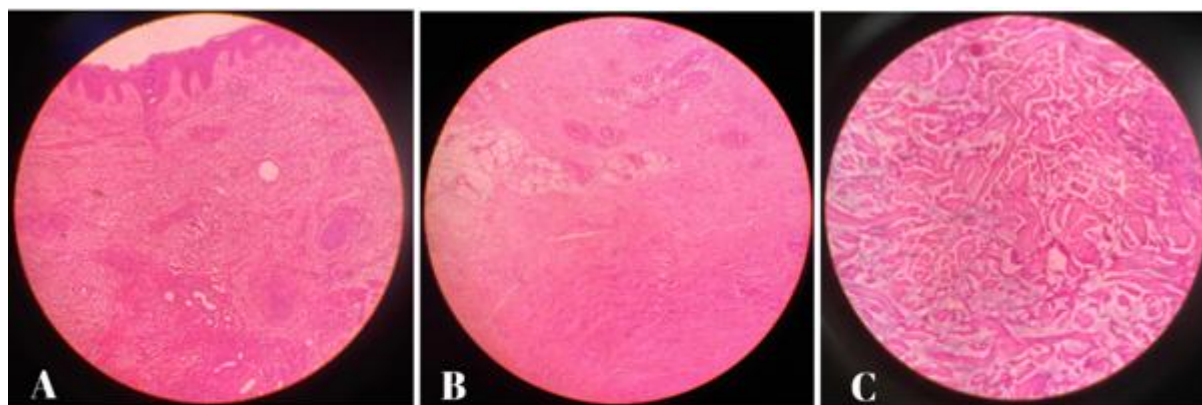


Figure 2:

(A) Skin with follicular plugging, focal loss of rete ridges and pigment incontinence under 40x magnification,

(B) Deeper dermis shows mucin deposition, necrobiosis of collagen fibres perivascular and periadnexal lymphohistiocytic infiltrate under 40x magnification ,

(C) Dermis showing mucin deposition under oil immersion 100x magnification  
(Hematoxylin- Eosin Stain)



Figure 2: Resolved lesions of nose (left) and elbow (right) after 2 weeks of treatment

### 3. Discussion

Lupus tumidus was first described by Erich Hoffmann in 1909 followed by a report by Gougerot and Bournier in 1930 with subsequent reports in 1950 [3]. Cutaneous manifestation of lupus can be classified into various types which includes Acute lupus erythematosus (ALE), subacute lupus erythematosus (SLE) and chronic cutaneous lupus erythematosus (CCLE) and some nonspecific lesions such as panniculitis, vasculitis and tumid lesions [4].

Based upon the classification that was originally proposed by Dr James Gilliam and Sontheimer, Tumid Lupus Erythematosus is a subtype of chronic cutaneous LE [5]. Because of its weak association with SLE and a relative lack of serological abnormalities, it is now considered as a separate entity by some. There has been a modification and extension of the Gilliam classification recently which suggested an intermittent CLE (ICLE) subtype, including Tumid LD as a separate entity of the disease. The diagnosis of lupus tumidus is usually delayed as it can be confused with other dermatoses due to the lack of systemic manifestations.

The Diagnostic criteria for TLE as proposed by Kuhn et al (2000) includes: clinical and histopathological results, reproduction of lesions after exposure to UVA and/or UVB, quick and effective response to anti-malarial drugs [6]. The clinical features include annular, indurated, erythematous, asymptomatic edematous plaques without epidermal involvement, over photo exposed areas of the skin that leave no scar on healing. Histological signs are perivascular and peri adnexal lymphocytic infiltrate, interstitial mucin deposition and in some cases diffuse lymphocytes or absence of epidermal involvement such as hyperkeratosis and follicular plugging which is characteristic of discoid LE; and any change in dermo-epidermal junction which are hallmark of most of the cutaneous forms of LE. In contrast to some types

of LE, LET is characteristically not associated with the presence of anti-nuclear antibody (ANA). There are very few case reports with ANA positive titers and one it being 1:640 [7]. However, a positive ANA test does not rule out LET just like in our case [8].

Tumid lupus erythematosus has a better prognosis compared to other forms. Spontaneous resolution of the rash is possible, although tumid lupus erythematosus is known to react well to a combination of photoprotection, topical corticosteroids and oral antimalarials as the first-line treatment. Second-line treatments include methotrexate and mycophenolate mofetil, while third-line treatments are thalidomide and lenalidomide. The rash of tumid lupus erythematosus is known to heal without plugging, atrophy, scaling, skin disfiguration, scarring, and dyspigmentation. Complete resolution of the rash might take up to three months with treatment.

### 4. Conclusion

In conclusion, TLE is a rare clinical condition reported hence we emphasize the importance of detailed clinical examination along with histopathological study. Since isolated examination may lead to underdiagnosis of a disease which when diagnosed and treated has a very good prognosis.

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