Chronic Cutaneous Lupus: A Case with Histopathological Mystery

Kronik Kutanöz Lupus: Histopatolojisi Gizemli Bir Vaka

ABSTRACT: Lupus erythematosus tumidus (LET) is a form of chronic cutaneous lupus which presents with erythematosus urticaria like plaques on sun-exposed sites. Histologically, lack of epidermal alterations differentiates the LET from other cutaneous lupus variants. Perivascular and periadnexal lymphocytic infiltration and interstitial mucin deposition are the main microscopic characteristics. However, in the literature there are few reports that assert variation of the histopathological findings. From this point of view, we report here a male patient who is an interesting example of cutaneous lupus erythematosus with clinical behaviour of lupus tumidus but histological characteristics of discoid lupus.

Keywords: Lupus erythematosus, discoid; pathology, clinical


Anahtar Kelimeler: Lupus eritematozus, discoid; patoloji, klinik

Erythematous and edematous plaques without scaling or hyperkeratosis affecting primarily sun-exposed areas are the most common clinical presentation of lupus erythematosus tumidus (LET). Histological analysis is the key to confirm the diagnosis and one of the diagnostic criteria however some controversy regarding histopathological characteristics still remains. Absence of epidermal involvement, follicular plugs or atrophy are usual histological features which provide differentiating the disorder from other lupus erythematosus (LE) subtypes. We describe here a case with facial plaque which presents clinical features of LET and exhibits microscopic characteristics of discoid lupus erythematosus (DLE).

CASE REPORT

A 44-year-old otherwise healthy non-smoker man, hairdresser, was admitted to our clinic for a 6 month history of progressive asymptomatic swelling
on his face. Past medical history and family history were unremarkable. On dermatologic examination, there was indurated erythematous plaque on edematous background over the right cheek which causes slight facial asymmetry (Figure 1). He was previously treated with antibiotics and topical corticosteroids and emollients. Other skin and mucosa findings were unremarkable. Lymphadenopathy was not detected. Physical examination revealed no evidence of systemic involvement. Laboratory tests including complete blood count, blood chemistry, urinalysis with microscopy, complement levels, hepatitis and human immunodeficiency virus (HIV) serology and imaging diagnostic procedures including chest X-ray and echocardiography were all in the normal range. Superficial soft tissue ultrasonography revealed diffuse inflammatory enlargement of subcutaneous adipose tissue. Positive serology with antinuclear antibody titer of 1/320, strong positive anti-Ro (SS-A), elevated erythrocyte sedimentation rate were noted. C-reactive protein and rheumatoid factor levels were normal. The presumptive diagnoses of B cell lymphoma, pseudolymphoma, lupus tumidus and polymorphous light eruption were noted. A lesional skin biopsy specimen revealed parakeratosis and focal hydroptic degeneration in the epidermis. Dermis showed dense infiltration of lymphocytes and plasma cells around follicular structures and vessels. Immunohistochemistry revealed mix type infiltration including CD3+ T cells and CD20+ B cells. CD38 and CD138 highlighted plasma cells. Since significant number of plasma cells in the infiltrate, kappa lambda immunohistology showed polyclonal infiltration with 3/2 kappa/lambda ratio (Figure 2). Alcian blue staining did not reveal interstitial mucin deposition.

The patient did not present any constitutional, musculoskeletal, gastrointestinal, psychiatric signs of systemic lupus erythematosus. His neurological and ophthalmological examinations were normal. Given his clinical presentation, serologic profile and histopathological findings a diagnosis of chronic cutaneous lupus was made. The patient was treated with hydroxychloroquine 400 mg/day and sunscreen with SPF 50. We observed complete remission after 2 month course of treatment with hydroxychloroquine (Figure 3). The plaque disappeared without leaving scars or dyspigmentation. After this clinical improvement, initial dosage of hydroxychloroquine was reduced to 200 mg/day. Owing to maintenance of clinical improvement for a month following the dosage taper, the therapy was discontinued. Since then, no recurrences have been observed for three months.

DISCUSSION

To date any definite criteria for diagnosis of LET has not been commonly accepted. In the literature extreme photosensitivity, distinct histopathologic features and favourable response to antimalarial drugs are described the main diagnostic clues of LET. The clinical picture of our patient was compatible with LET. We could not detect clear relationship with sun-exposure in our patient. However existence of swollen urticaria like plaque, absence of epidermal involvement and absence annular lesions and collarette scaling were the main clues differentiating the disease clinically from DLE and the other differential diagnoses. Furthermore, in contrast to DLE, dramatic response to an-
timalarial therapy and regression of the lesion without scarring and hypo-hyperpigmentation supported our clinical diagnosis.

Even though histopathologic heterogeneity of all lupus variants are evident, lymphocytic and periangnexal infiltrate with dermal mucin deposition are commonly accepted histopathological features by majority of authors. While some authors highlighted that existence of minimal epidermal and dermal-epidermal junction alterations are exclusion findings, the others considered these are acceptable.\(^3\)-\(^5\) Moreover Sontheimer included LET in the most variable histopathological form of lupus.\(^6\) But the exact mechanisms leading this histomorphological variation are still unclear yet.

In the series of Choonhakarn et al. two of the 15 patients exhibited perifollicular infiltration and one had focal vacuolar degeneration like as in our case.\(^7\) Interestingly, the case presented here had some unique histopathological aspects including epidermal involvement, dermo-epidermal alteration, dense plasma cell infiltration without aberrant mucin deposition. These microscopic findings were more consistent with DLE rather than LET. Unfortunately direct immunofluorescence test
could not be performed because of health insurance issues. Beyond the scope of this presentation the discrepancy between the clinical and histological presentation may support the arguments regarding that LET is a histological non-specific manifestation of lupus rather than a distinct subtype.

In conclusion as because LET is a rare reported disease and presents different serologic and microscopic characteristics, diagnostic challenge still remains. Obviously there is a need for future large reports to provide better understanding of histopathological mystery.

**Conflict of Interest**

Authors declared no conflict of interest or financial support.

**Authorship Contributions**

**Idea/Concept:** Constructing the hypothesis or idea of research and/or article: Pınar İncel Uysal, Emine Tamer; **Design:** Planning methodology to reach the conclusions: Pınar İncel Uysal, Başak Yalçın; **Control/Supervision:** Organizing, supervising the course of progress and taking the responsibility of the research/study: Başak Yalçın, Önder Bozoğan; **Data Collection and/or Processing:** Taking responsibility in patient follow-up, collection of relevant biological materials, data management and reporting, execution of the experiments: Pınar İncel Uysal, Emine Tamer, Önder Bozoğan; **Analysis and/or Interpretation:** Taking responsibility in logical interpretation and conclusion of the results: Pınar İncel Uysal, Önder Bozoğan; **Literature Review:** Taking responsibility in necessary literature review for the study: Pınar İncel Uysal, Başak Yalçın; **Writing the Article:** Taking responsibility in the writing of the whole or important parts of the study: Pınar İncel, Başak Yalçın; **Critical Review:** Reviewing the article before submission scientifically besides spelling and grammar: Başak Yalçın, Önder Bozoğan; **References and Fundings:** Providing personnel, environment, financial support tools that are vital for the study: Pınar İncel Uysal, Önder Bozoğan; **Materials:** Biological materials, taking responsibility of the referred patients: Pınar İncel Uysal.

**REFERENCES**


