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Case Report

A PEDIATRIC CASE OF SYSTEMIC LUPUS ERYTHEMATOSUS: CLINICAL AND DERMOSCOPIC FINDINGS

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ABSTRACT
A 14 years old girl applied to our clinic with complaint of facial erythematous plaques of two years duration. Photosensitivity was positive. She had erythematous scarring scaly plaques on the face; cicatricial alopecia on the scalp; eroded areas on the palate; and periungual telangiectatic erythema. Dermoscopic examination of facial lesions revealed follicular keratotic plugs, perifollicular whitish halo, polymorphous telengiectatic vessels, white scales, and structureless whitish areas. Irregularly dilated and tortuous, ramified/bushy capillaries were determined in periungual region. Histopatological and serological findings were consistent with SLE. Perifollicular whitish halo, follicular keratotic plugs, polymorphous telengiectatic vessels, white scales, pigmentation, structureless whitish areas and, follicular red dots are the dermoscopic findings reported in DLE. We observed all those findings other than follicular red dots and pigmentation, which are relatively uncommon. Nail fold dermoscopy findings were consistent with connective tissue disease. DLE in conjunction with SLE was suggested. Histopathological and serological findings confirmed the diagnosis. Dermoscopy is a useful tool in the diagnosis of DLE. DLE should not be overlooked in pediatric age because of the high risk of SLE.

Key words: SLE, pediatric, dermoscopy

ÖZET

On dört yaşında kız çocuğu kliniğimize iki yıldır var olan fasiyal eritematöz plak yakınması ile başvurdu. Fotosensitivite pozitifti. Yüzde eritematöz skatrisyel skuamlı plaklar; skalpte skatrisyel alopesi; damakta erode alanlar; ve periungual telenjiektatik eritem vardı. Yüz lezyonlarının dermoskopik muayenesinde foliküler keratotik tıkaçlar, perifolliküler beyazımsı halo, polimorfik telenjiektatik damarlar, beyaz skuamlar, ve beyaz yapısız alanlar görüldü. Periungual bölgede düzensiz olarak genişlemiş ve kıvrıntılı, dallı/çalı gibi kapiller damarlar tespit edildi. Histopatolojik ve serolojik bulgular sistemik lupus eritematozus (SLE) ile uyumluydu. Diskoid lupus eritematozus (DLE)'de bildirilen dermoskopik bulgular, perifoliküler beyazımsı halo, foliküler keratotik tıkaçlar, polimorfik telenjiektatik damarlar, beyaz skuamlar, pigmentasyon, beyaz yapısız alanlar ve foliküler kırmızı noktalardır. Nispeten nadir görülen foliküler kırmızı noktalar ve pigmentasyon dışındaki tüm bulguları hastamızda gözledik. Tırnak kıvrımı dermoskopik bulguları bağ dokusu hastalığı ile uyumlu idi. Hastada SLE ile birlikte DLE düşünüldü. Histopatolojik ve serolojik bulgular tanıyı doğruladı. Dermoskopi, DLE tanısında yararlı bir araçtır. Çocukluk çağında yüksek SLE riski olması nedeniyle DLE göz ardı edilmemelidir.

Anahtar kelimeler: SLE, pediatrik, dermoskopi

INTRODUCTION

Cutaneous lupus erythematosus (CLE) is classically subdivided into “specific” and “non-specific” skin eruptions. Specific skin eruptions are further divided as chronic CLE (CCLE), subacute CLE and acute CLE. The most frequent subset of CCLE is discoid lupus erythematosus (DLE). CLE is uncommon in pediatric age. Skin is the most commonly involved organ after joints in systemic lupus erythematosus (SLE). The risk of SLE in children with DLE has been found to be higher than in adults.[1,2]

Dermoscopy is a non-invasive diagnostic technique permitting visualization of structures, patterns and colors in skin lesions that are not evident to the naked eye. The contribution of dermoscopy in the diagnosis of facial inflammatory skin diseases has been demonstrated.[3] Herein, a pediatric case of SLE and the dermoscopic findings of DLE lesions are described.

CASE REPORT

A 14 years old girl applied to our clinic with complaint of facial erythematous plaques of two years duration. Photosensitivity was positive. Erythematous scarring scaly plaques on the face; cicatricial alopecia on the scalp; eroded areas on the palate; and periungual telangiectatic erythema were detected on dermatological examination (Figure 1). Dermoscopic examination of facial lesions revealed follicular keratotic plugs, perifollicular whitish halo, polymorphous telengiectatic vessels, white scales, and structureless whitish areas. Irregularly dilated capillaries and tortuous, ramified/bushy capillaries were established on nail fold (Figure 2).

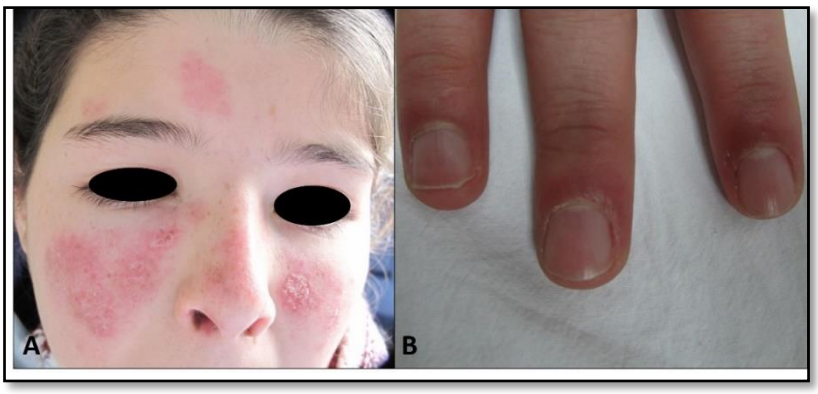

Figure 1. A) Erythematous-squamous plaques on face and B) periungual telangiectatic erythema on dermatological examination.

Histopathologic examination showed atrophy and occasional hypertrophy of the epidermis under the orthokeratotic lamellar keratin layer, vacuolar degeneration of the basal layer in addition to dermal perivascular edema and preifollicular mononuclear inflammatory infiltration. Immunfluorescence was positive in dermoepidermal junction for immunglobulin M, C1q and C3 in granular pattern. Histopathologic and direct immunofluorescence findings were consistent with DLE (Figure-3). Antinuclear antibodies, anti-dsDNA antibodies and, anti-Sm antibodies were found to be positive. Serum C3 and C4 levels were decreased. According to the clinical, dermoscopical, histopatological and serological findings the patient was diagnosed with SLE.

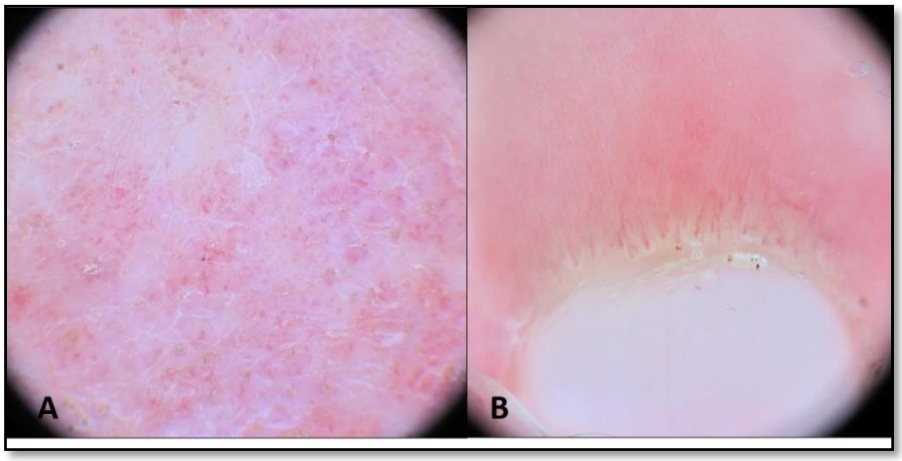

Figure 2. A) Mild disorganization of the capillary architecture and B) enlarged/giant capillaries on dermoscopic examination (X10).

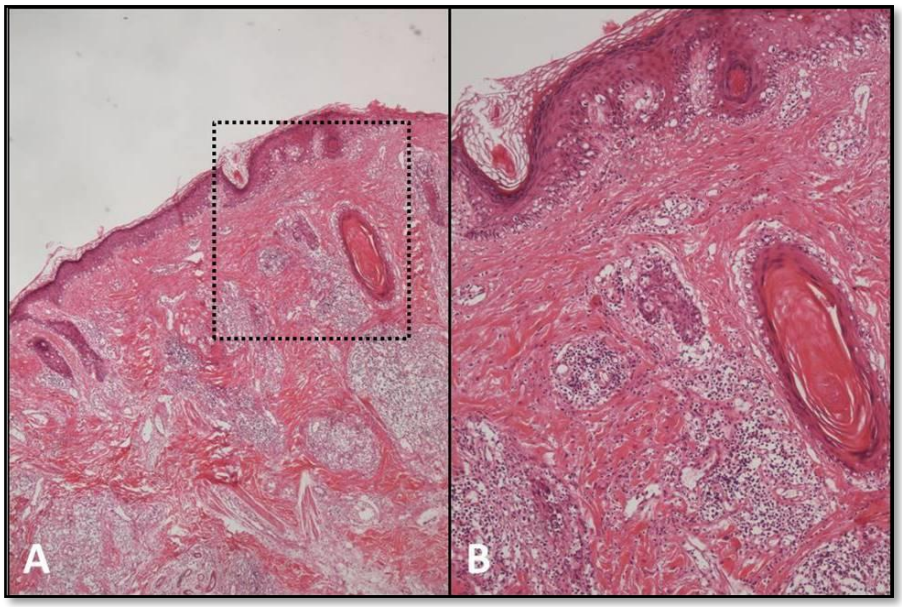

Figure 3. Perifollicular degeneration and periadnexial mononuclear inflammation A) H&EX10 B) H&EX4.

DISCUSSION

DLE is rare in children. The disease emerges before the age of 10 years in more than 50% of the pediatric patients. The rate of transformation to SLE in pediatric disease was found in 25% of the patients who were followed more than a year, which is higher than the rate reported in adult patients.[4] Clinical differential diagnoses of DLE includes tinea faciei, lupus vulgaris, sarcoidosis, psoriasis, and even actinic keratosis. Differentiating DLE from those disorders by only clinical aspect is generally hard. It has been demonstrated that dermoscopic examination may reveal clues for DLE. Perifollicular whitish halo, follicular keratotic plugs, polymorphous telengiectatic vessels, white scales, pigmentation, structureless whitish areas and, follicular red dots are the dermoscopic findings reported in DLE.[5,6] Perifollicular whitish halo and follicular keratotic plugs were observed in more recent lesions; however structureless whitish areas, telengiectatic vessels and honey comb pigment network in older ones.[6] The above mentioned dermoscopic findings can help to differentiate DLE from squamous cell carcinoma and actinic keratosis.[7,8] We observed all of the dermoscopic findings reported for DLE, other than follicular red dots and pigmentation, in our patient. Follicular red dots were first observed by Tosti et al. in the 5 of 13 patients with scalp DLE. They did not identify this finding in any other diseases causing cicatricial alopecia and thought follicular red dots to be a specific finding of DLE. In their patients, none of the facial DLE lesion demonstrated this finding.[9] Later study by Lallas et al. recognized follicular red dots in non-scalp DLE lesions; however, its frequency was lower than reported by Tosti et el. in scalp DLE lesions.[6] Honey-comb pattern pigmentation was also reported in scalp DLE lesions and thought to be related with sun exposure.[10] Based on aforementioned observations it is not unexpected that we did not detect follicular red dots and pigmentation.

Nail fold dermoscopy is also important in the diagnosis of connective tissue diseases. Nail fold capillaries changes are mostly seen in scleroderma spectrum disorders and dermatomyositis. Disorganization of the capillary architecture, enlarged/giant capillaries, capillary hemorrhages, avascular areas and ramified/bushy capillaries can be seen depending on the degree of the illness. Milder dermoscopic findings are generally seen in SLE.[11] We

identified mild disorganization in the capillary architecture in addition to enlarged, tortuous and ramified/bushy capillaries in the nail fold supporting systemic disease. At sum, DLE in conjunction with SLE was suggested. Histopathological and serological findings confirmed the diagnosis.

CONCLUSION

Although uncommon, DLE should be kept in mind in, also, pediatric age. Early diagnosis is important because of the high risk of SLE. There are various skin diseases that can simulate DLE. In this context, dermoscopy seems to be a useful tool in the diagnosis of DLE.

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