CASE REPORT

Linear chronic discoid lupus erythematosus following the lines of Blaschko *

Lúpus eritematoso crônico discoide nas linhas de Blaschko

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Abstract: Linear chronic discoid lupus erythematosus is a rare manifestation of cutaneous lupus in which erythematous, atrophic, dyschromic lesions are located along the lines of Blaschko. This report describes the case of a 15-year old boy with a 2-year history of discoid, erythematous, hyper and hypopigmented lesions with central atrophy, situated along the lines of Blaschko on his right arm. Histopathology showed epidermal atrophy, hyperkeratosis, follicular plugging, thickening of the basement membrane zone, and superficial and deep chronic perivascular and periadnexal inflammatory infiltrate, with dermal mucin deposit, thus confirming diagnosis. A total of 14 cases have been described of this variant, the onset of which is often in childhood. There is no difference in incidence between genders. Lesions most commonly develop on the face. There have been no reports of any association with systemic disease.
Keywords: Lupus erythematosus, cutaneous; Lupus erythematosus, discoid; Mosaicism

Resumo: O lúpus eritematoso crônico discoide linear é manifestação rara da doença lúpica cutânea, em que lesões eritêmato-atrófico-discrômicas dispõem-se nas linhas de Blaschko. Descrevemos o caso de um menino de 15 anos, com dois anos de história de lesões discoides eritêmato-atróficas, hipo e hiperpigmentadas, dispostas nas linhas de Blaschko do membro superior direito. O exame histopatológico revelou atrofia da epiderme, hiperqueratose, rolhas córneas, espessamento da zona da membrana basal, infiltrado inflamatório crônico perianexial e perivascular superficial e profundo, depósito de mucina na derme, confirmando o diagnóstico. Há, no total, 14 casos descritos dessa variante que se inicia frequentemente na infância e que não apresenta predomínio quanto ao sexo. As lesões ocorrem preferencialmente na face. Não há relatos de associação com doença sistêmica.
Palavras-chave: Lupus eritematoso cutâneo; Lupus eritematoso discóide; Mosaicismo

INTRODUCTION

Lupus erythematosus (LE) is an inflammatory autoimmune disease with numerous clinical presentations that range from the localized form of the disease that is restricted to the skin to the systemic form. Lupus is classified into three major types: systemic lupus erythematosus (SLE), subacute cutaneous lupus erythematosus (SCLE) and chronic cutaneous lupus erythematosus (CCLE). The latter is subdivided into other disorders, including discoid lupus erythematosus (DLE), which is characterized by erythematous, atrophic, hyperkeratotic, hypochromic discoid lesions with a hyperpigmented halo. [1] The typical lesions found in DLE may occasionally follow the lines of Blaschko, constituting linear discoid lupus erythematosus (LDLE). [2] The case described here refers to a 15-year old boy with typical signs of LDLE.

CASE REPORT

A 15-year old male patient reported having had erythematous lesions on his right hand for the past two years that extended linearly along his right arm and scapula. He denied having had any prior diseases or any family history of autoimmune disease. The patient denied photosensitivity; however, at two con-

Received on 13.12.2009.
Approved by the Advisory Board and accepted for publication on 13.05.2010.
* Study conducted at the School of Medicine, Pontifical University of Campinas (PUC Campinas), Campinas, São Paulo, Brazil.
Conflict of interest: None / *Conflito de interesse: Nenhum*
Financial funding: None / *Suporte financeiro: Nenhum*

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sultations the lesions were found to have worsened following unprotected exposure to solar radiation while the patient was fishing.

Physical examination revealed atrophic, hypo- and hyperpigmented lesions with an erythematous halo, together with hyperkeratosis in some of the lesions. The lesions followed the lines of Blaschko, affecting the back of the patient's forefinger and extending as far as the right scapula and ipsilateral paravertebral region (Figures 1, 2 and 3). The patient also had dermographism.

Following a clinical suspicion of linear lupus or LDLE, histopathology was performed, revealing epidermal repair with hyperkeratosis, the formation of follicular plugging, hydropic degeneration of the basal cell layer, thickening of the basement membrane zone, telangiectasia, superficial and deep perivascular and periadnexal lymphocytic inflammatory infiltrate with mucin deposit in the reticular dermis (Figure 4), thus confirming the diagnosis of LDLE.

The following laboratory tests were requested: Antinuclear factor (ANF), with titers 1:80, homogenous, and no clinical correlation with greater activity in the lesions. Anti-Sm, anti-Ro and anti-La were non-reactive. Full blood count, erythrocyte sedimentation rate, complement, fasting glucose and creatinine levels were all normal.

Following ophthalmological examination, treatment was initiated with chloroquine diphosphate 25 mg/day and an improvement was seen in the patient's condition.

DISCUSSION

In 1978, Umbert and Winkelmann reported the first case of LDLE with concomitant characteristics of scleroderma and for this reason referred to the condition as a mixed cutaneous syndrome, an association of cutaneous lupus with scleroderma. [2] Two similar cases in which these two pathologies with a linear pattern of involvement overlapped have been described previously. [3]

Linear involvement in cutaneous LE is rare and may present as LDLE, deep LE, tumid LE or SCLE. [2] A case of linear bullous LE has been described in a patient with SLE. [4] The present case is included within the context of linear DLE (LDLE). A total of 14 cases of LDLE have been described, including the case reported here. The face is the most commonly affected site (9/14 cases). Involvement of the upper limbs has only been described once, in the case reported by Umbert and Winkelmann, in which the patient had lesions on the forefinger and forearm. [5,6] In the pres-

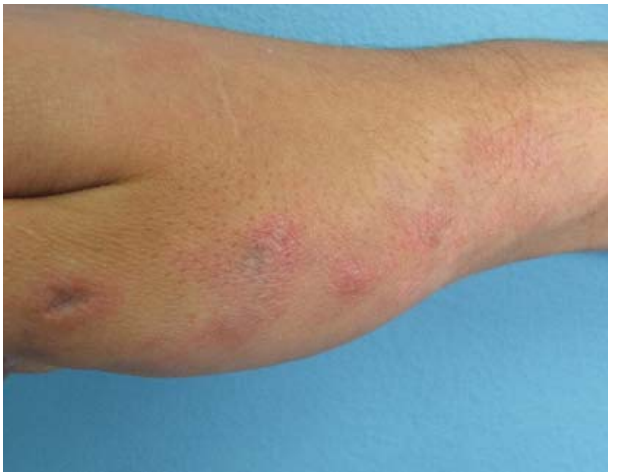

Figure 2: Discoid lesions with an atrophic, purplish center and erythematous halo, situated linearly on the patient's right hand

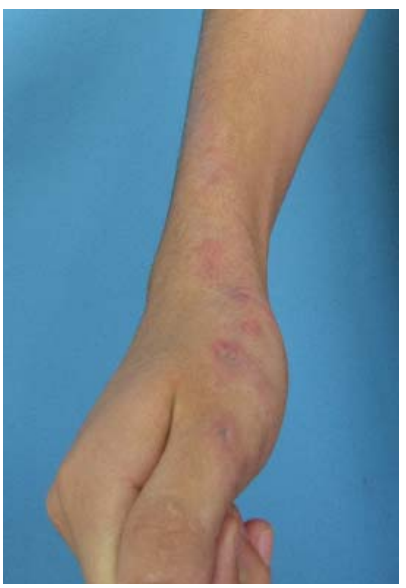

Figure 1: Erythematous atrophic lesions situated along the lines of Blaschko on the patient's right upper limb

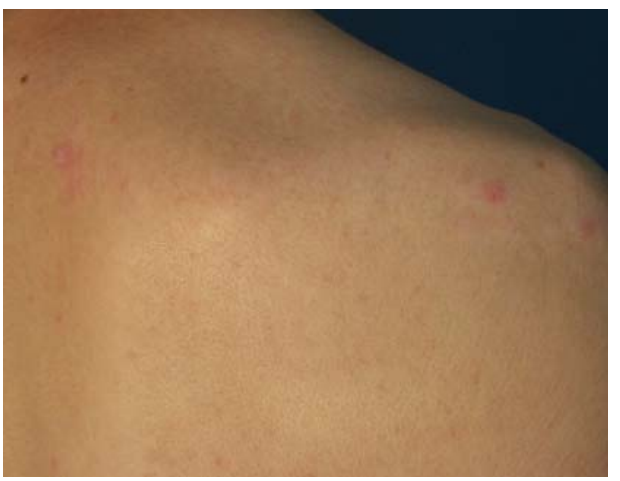

Figure 3: Erythematous, hypochromic, atrophic lesion in the right paravertebral region. Note the presence of two erythematous lesions on the posterior surface of the shoulder, at the right scapula. The lesions are arranged in a linear pattern

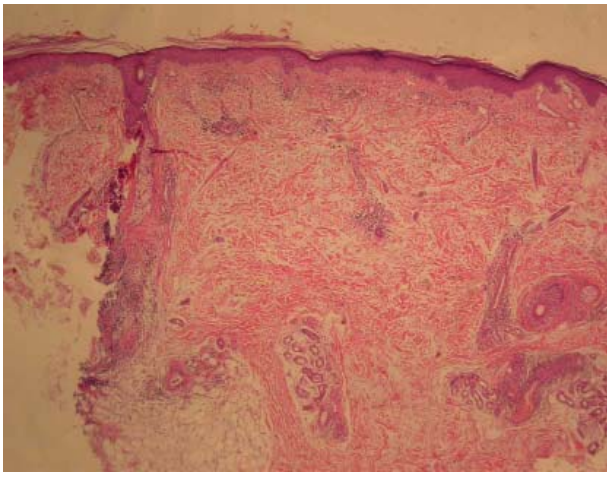

Figure 4: Epidermal atrophy associated with hyperkeratosis, with the formation of follicular plugging, telangiectasia, superficial and deep perivascular and periadnexal chronic inflammatory infiltrate. Dermal mucin deposit

ent case, the lesions were affecting the patient's right arm, extending along the lines of Blaschko onto the trunk. In the majority of cases, a single anatomical site was affected (12/14 cases). As previously mentioned, two sites were affected in the present case. Low antinuclear antibody titers were found in three patients, including the present one. In all three cases titers of 1/80 were found. [5,6] In six cases, direct immunofluorescence was performed, resulting in complement C3 and immunoglobulin deposits (IgM in 5 cases, IgG in 2 cases) along the basement membrane. [5] The duration of the symptoms of the disease prior to diagnosis ranged from 2 months to 5 years, with a mean of 1.5 years. In 8 cases, patients were under 15 years of age. Eight of the patients were male and 6 were female. [5,6] The patient in the present case was male. He had had a clinical history of symptoms for two years prior to diagnosis, with the onset of the lesions occurring at 13 years of age.

Diagnosis is made clinicopathologically. Histopathology reveals the typical characteristics of LE such as epidermal atrophy, hyperkeratosis, follicular plugging, thickening of the basement membrane, hydropic degeneration of the cells of the basal layer, telangiectasia, pigmentary incontinence, superficial, medium and deep perivascular and periadnexal chronic inflammatory infiltrate, and dermal mucin deposits. [1] These characteristics were all found in the present case.

DLE is rare in childhood. Onset of the disease occurs prior to 10 years of age in less than 2% of all cases. [7] Unlike adult DLE, childhood DLE does not predominantly affect girls and photosensitivity is rare; however, there is a higher rate of progression to the systemic form of the disease. [7,8] The onset of linear DLE occurs generally in childhood, although some cases have been described in adults. [5,6,8] Linear DLE in childhood, as well as non linear DLE, occurs equally in males and females. However, in the case of linear DLE, progression to SLE is low. [8,9] An association between linear DLE (LDLE) in the left submandibular region and ipsilateral submandibular myoepithelial sialoadenitis has been described in the absence of either Sjögren's syndrome or SLE. [10] Photosensitivity has not been reported previously in cases of LDLE; [8,9] however, in the present case, photosensitivity was found clinically on two occasions, both following exposure of the patient to sun while fishing without wearing a shirt or sunscreen, which resulted in reactivation of the lesions.

Abe et al. proposed classifying cases of linear LE as linear cutaneous lupus erythematosus, since the lesions were linear in nature and not discoid in the restricted sense of the term. [7,11] Nevertheless, other subtypes of LE that follow a linear pattern have already been described, including deep LE, subacute LE and tumid LE. Therefore, we believe that linear chronic discoid lupus erythematosus would be a more appropriate denomination, since this term emphasizes the subclassification of LE in which this case is included. In the case presented here, the lesions were both discoid and linear, with morphological characteristics typical of LDLE, as shown in Figure 2, which illustrates the affected region of the patient's forefinger.

Various acquired inflammatory pathologies that follow the Blaschko lines have already been described, including striated lichen, lichen planus, scleroderma and psoriasis. [12,13] These conditions are believed to be caused by genetic mosaicism. [2]

The explanation suggested is the occurrence of post-zygotic mutation in the keratinocytes, giving rise to abnormal cells that are cloned and migrate in a single direction during embryogenesis. [5,6,12] The cells arising from these mosaicisms express neoantigens capable of eliciting local immune response. [3] The trigger for the onset of the disease may be trauma, primary irritation or exogenous agents such as ultraviolet light, drugs, pesticides, heavy metals or other elements. [2,14]

The apoptosis of keratinocytes has been indicated as a key event in triggering cutaneous lupus lesions through various apoptotic pathways such as p53, tumor necrosis factor-alpha (TNF-α) and Fas/FasL. [5,15] It is speculated that the aberrant keratinocytes may be unable to express the essential proteins required to regulate apoptosis, and are hence unable to prevent the apoptosis induced by solar radiation, for example. Another mechanism that has been suggested is that these keratinocytes may have an anomalous major histocompatibility complex (MHC) or may release abnormal cytokines. [5] ❑

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How to cite this article/*Como citar este artigo*: Lazzarini R, Rotter A, Farias DC, Muller H. Linear chronic discoid lupus erythematosus following the lines of Blaschko. An Bras Dermatol. 2011;86(3):553-6.