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Nail changes in lupus erythematosus

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Abstract
Systemic lupus erythematosus is an autoimmune disease with a multifactorial etiology. It is characterized by the formation of autoantibodies and immune complexes, which frequently affect the skin. However, there is a paucity of literature that broadly describes nail changes associated with this disease. We present the clinical case of a young male patient with recently appeared skin lesions and nail changes, as well as arthralgia, myalgia, and general malaise. Therefore, a study approach was conducted, which included immunological studies. The patient was initially diagnosed with discoid lupus and subsequently diagnosed with systemic lupus erythematosus, which prompted the initiation of treatment. This case study presents the patient's nail alterations associated with this disease. A comprehensive nail evaluation can serve as a valuable initial diagnostic tool in patients suspected of having this disease.

Keywords: Lupus erythematosus, systemic, lupus erythematosus, discoid, nail, nail diseases, autoimmune diseases

Introduction
Systemic lupus erythematosus is an inflammatory, chronic illness, prototype of autoimmune disease, that affects the skin and provokes systemic conditions. It is more prevalent in females and, although its etiology remains unclear, research has identified several key factors that contribute to its development and progression, these include: genetic predisposition, hormonal influence, ultraviolet light, viral infections, and certain drugs. The most prevalent manifestations of systemic lupus erythematosus include joint pain, general symptoms, skin lesions, and muscular conditions. In terms of cutaneous manifestations, lesions are most frequently observed in areas exposed to light, including the face, V-neckline, and the external side of the arms and forearms [1].
Discoid lupus erythematosus represents a subset of cutaneous lesions observed in individuals with systemic lupus erythematosus; however, it is noteworthy to mention that only 20% of cases have it. Periungual lesions are observed in 4 to 30% of patients and antinuclear antibodies (ANA) are detected in 75 to 100% of cases, but these antibodies lack specificity. As part of the diagnostic protocol, a skin biopsy may be performed, and the treatment regimen is typically based on oral steroids [2]. The following case study presents a male patient with systemic, skin, and nail conditions. Immunological tests were conducted, leading to the diagnosis of discoid lupus erythematosus and systemic lupus erythematosus.

Case presentation
The subject is a 29-year-old male of Mexican origin and residence in Mexico City. There is no family history of connective tissue disorders, nor is there a personal pathological history. He exhibited symptoms of polyarthralgia in both hands, myalgia, and general malaise, which prompted a referral to Rheumatology for further evaluation. During the physical examination, additional findings included arthritis and hair loss. Up until that point, the provisional diagnosis was undifferentiated arthritis and the patient initiated treatment with leflunomide 20 mg every 24 hours from Monday to Friday and celecoxib 200 mg every 24 hours. Immunological tests were conducted and the patient was instructed to return in three months with the results. However, due to the patient's clinical improvement, follow-ups with Rheumatology were discontinued.
Subsequently, the patient exhibited the development of an erythematoedematous and pigmented plaque in the posterior aspect of the nose and the cheeks, which led to a referral to Dermatology. A biopsy was taken during the consultation and the results indicated the

presence of hypergranulosis, scarce melanophages, and follicular mononuclear inflammatory infiltrate in the superficial and profound dermis, with evidence of broken corneal cysts, granulomas, vascular congestion and the formation of small granulomas with foamy macrophages. In light of these findings, a diagnosis of discoid lupus erythematosus was established and the patient received topical treatment.

In a subsequent dermatological consultation, the patient reported the onset of erythematous lesions on his nails. Upon physical examination, the presence of nail erythema was observed on his hands (Figure 1). Dermoscopy showed periungual telangiectasia, and capillaroscopy showed dilated capillaries and erythema in the proximal nail fold, as well as a brown spot on the nail bed and an avascular area in the nail matrix (Figure 2). Furthermore, the patient reported a recurrence of general symptoms, specifically arthralgia in both hands.

In order to arrive at a diagnosis, laboratory tests were necessary, whose most relevant results are shown in the Table. The patient was again referred to Rheumatology, where the clinical picture and the biochemical results led to the diagnosis of systemic lupus erythematosus (SLEDAI score 2). The patient began a course of treatment consisting of prednisone (50 mg every 24 hours) and mycophenolic acid (500 mg every 12 hours). Additionally, the patient was referred to Endocrinology for further evaluation, given the possibility of subclinical hypothyroidism, which may be secondary to Hashimoto thyroiditis (another autoimmune condition).

Discussion

Systemic lupus erythematosus is a disease that predominantly affects young women. However, in this case, the patient is a young male with skin lesions consistent with discoid lupus erythematosus and nail alterations. He was referred to Rheumatology due to a clinical picture and positive antibodies, which suggested the possibility of systemic lupus erythematosus. The rarity of this case lies in the combination of systemic lupus erythematosus and cutaneous lupus, which has only been observed in approximately 20% of patients. Aside from the patient in question being a male, he presents distinctive nail changes associated with systemic lupus, which are uncommon, and, as a result, have not been extensively documented.

In the United States of America, the annual incidence of systemic lupus erythematosus is 27.5 and 75.4 cases per million black and white women, respectively. The disease can manifest at any age, but it is more prevalent during the second to fourth decades of life, with a significant prevalence in females, with a ratio of 10:1, and across all racial groups [3]. It appears that genetic predisposition is polygenic, since the disease is associated with the HLA-DR2 and DR3, which results in a relative risk of 2 to 5 times higher for its development. The most prevalent haplotypes are HLA-B8, DR2, DQW1, and null alleles for C4. Furthermore, this condition has been associated with viral infections, such as measles, rubella, Epstein-Barr, and parainfluenza viruses. Some trigger drugs for the disease are hydralazine, procainamide, griseofulvin, phenytoin, penicillin, tetracycline, methyldopa, streptomycin, and terbinafine [4].

These predisposed factors result in the loss of immunological tolerance, as evidenced by the emergence of sensitized cells and the development of antibodies. Additionally, there is a disruption in the ability to eliminate cellular degradation products (Apoptotic material), with the persistence of numerous autoantigens that form immune complexes capable of damaging various tissues [4]. The most common symptoms of systemic lupus erythematosus are: joint symptoms (95%), general symptoms (90%), and fatigue (80-100%), as well as fever (83%). Skin lesions are present in 80% of cases, with alopecia occurring in 70% of cases. Other common manifestations include malar erythema (40-60%), photosensitivity (50%), buccal mucosa lesions (27-41%), discoid lupus erythematosus (20%), and muscular conditions (>60%), nephropathy (60%), heart disease (50%), lung and pleural disease (40%), and neurological disease (25%).

Discoid lupus erythematosus is defined by the presence of one or more plaques, which are circular or oval in shape and well-defined. These plaques vary in size and are characterized by a follicular spot, formed by a triad of phenomena: erythema, scale (Thin and adherent), and atrophy. The scale is difficult to remove and leaves a prolonged cornea, which is referred to as the "tack sign." When the condition extends to the cheeks and the posterior portion of the nose, without involvement of the nasolabial folds, it is referred to as malar erythema or "butterfly or bat (Vespertilio) wings", whose color can range from pink to purple. In the buccal mucosa, the appearance of white plaque in the inner aspect of the cheeks is common and, in some cases, these plaques may ulcerate, causing significant discomfort when the patient attempts to eat.

Alopecia, as a symptom of this disease, is characterized by diffuse hair loss, with the remaining hair exhibiting a thin, short length, and broken structure. On the neck and trunk, a poikilodermatous aspect may be observed, characterized by erythema, mottled pigmentation, telangiectasias, and atrophy. Atrophic lesions, lenticular and scaly in nature, may also manifest at the elbow and knees, as well as on the back of the phalanges or at the interphalangeal joints [6]. In patients with lupus, alterations are observed in the nail apparatus. Capillaroscopy is a technique that enables direct observation of the red capillary at the proximal nail fold. The majority of the changes observed in patients are unspecified, manifesting as the disease progresses or as a first symptom of lupus. The most frequently encountered alterations are telangiectasias, hyperpigmentation or depigmentation in the proximal nail fold, longitudinal ridges on the nail bed, and cuticle thickening [7].

The antibodies identified in systemic lupus erythematosus include antilymphocyte antibodies, anti-erythrocyte antibodies, antiplatelets, and anti-Ro. It is also notable that antinuclear antibodies (ANA) are detected in 75 to 100% of active cases and 80% of inactive cases. Similarly, anti-double stranded DNA antibodies are found in over 80% of cases, while anti-Smith antibodies represent the most specific marker, appearing in 30% of cases. Moreover, laboratory findings may include anemia (Normocytic normochromic, iron deficiency, or autoimmune hemolytic), leukopenia, lymphopenia, and thrombocytopenia. In 20% of cases, lupus anticoagulants yield positive results [8].

A skin biopsy may reveal a range of findings, including varying degrees of skin atrophy, the presence of corneal plugs in the infundibulum, vacuolization of the dermoepidermal interface, and alterations in basement membrane thickness. In addition, the dermis may exhibit an

inflammatory infiltrate comprising perivascular lymphocytic superficial and profound cells, as well as periadnexal infiltrates. Direct immunofluorescence identifies IgG, IgM, IgA, C1, C3, and properdin deposits in the dermoepidermal junction and surrounding the vessels. These deposits are not exclusive to the affected skin (90%), but are also present in apparently healthy skin [9]. The treatment plan is primarily based on the administration of prednisone (0.5 to 1 mg/kg/day, administered over a period of 4 to 6 weeks and gradually reduced until the treatment is discontinued or the dosage is reduced to the minimum). For severe cases, the administration of methylprednisolone pulses and cyclophosphamide may be indicated. In instances of refractory cases, the treatments include azathioprine, cyclosporine, plasmapheresis, intravenous immunoglobulin, and mycophenolate mofetil [10].

Fig 1: Nail erythema.

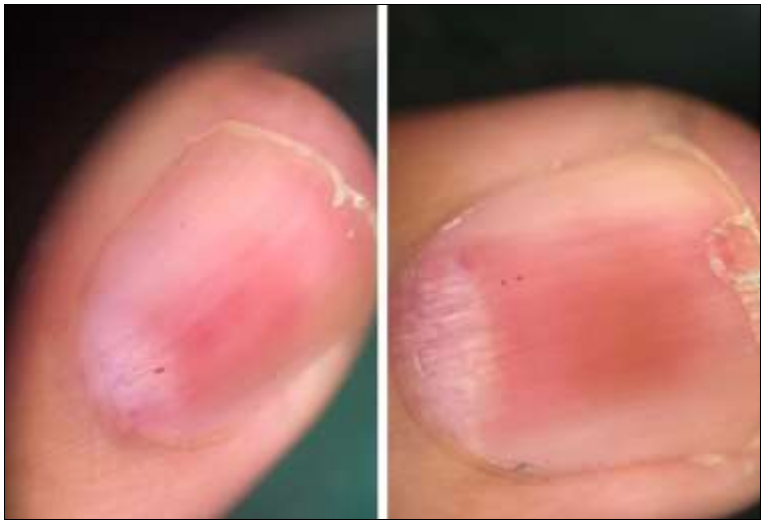

Fig 2: Dilated capillaries and erythematous spot on the nail bed

Table: Main results in laboratory testing

Parameters	Result	Normal values
C-reactive protein	8.07 mg/dl	<5 mg/dl
Sedimentation rate	26 mm/hour	0-15 mm/hour
Anti-double stranded DNA antibodies	378.3 U/ml	<85 U/ml
Antinuclear antibodies (ANA)	Positive, patrón nuclear homogéneo AC-1, dilución 1:640	Negative
Thyroid stimulating hormone (THS)	6.41 µU/ml	0.2-4 µU/ml
Free T4 thyroxine	1.29 ng/dl	0.71-1.85 ng/dl
Total triiodothyronine T3	1.4 ng/ml	0.8-2 ng/ml
Antithyroglobulin antibody	1235 ng/dl	0-40 ng/dl
Antithyroperoxidase antibody	1125 ng/ml	0-34 ng/ml
Rheumatoid Factor	6 U/ml	40-60 U/ml
Complement C3 fraction	83 mg/dl	86-206 mg/dl
Complement C4 fraction	<6 mg/dl	8-55 mg/dl

Conclusions

The objective of this case study is to describe the nail changes that can appear in systemic lupus erythematosus (since the condition is relatively uncommon and has been barely described in the literature, we considered it necessary to conduct further research), in the context of a rare case of a male patient with discoid lupus associated with systemic lupus erythematosus. The aim is to establish nail evaluation as a routine component of the initial assessment for patients with suspected systemic lupus erythematosus. In light of these considerations, we emphasize the crucial role of the dermatologist and the value of a multidisciplinary approach

in facilitating the early detection of associated systemic conditions.

Conflicts of interest statement
No conflicts of interest.

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Consent
No identifying markers are included as part of medical images. No consent is required.

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