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Cutaneous manifestations of pediatric Systemic Lupus Erythematosus

Abstract

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune inflammatory disease that affects both adults and children, but with different degrees of severity. Indeed, pediatric Systemic Lupus Erythematosus (pSLE) is more serious than that of adults and the management is heavier with significant comorbidity. The clinical manifestations are polymorphic. The skin is one of the target organs most diversely affected by the disease. Indeed the cutaneous manifestations are frequent, they are almost constant during the evolution reflecting the activity of the disease. Long considered relatively benign, they are sometimes very debilitating and can take several forms, some of which can lead to functional, aesthetic and psychic disabilities. Therapeutic management depends on the types of skin lesions, the depth, the extent of the lesions and the aesthetic impact. Therapeutic management uses several molecules, in particular synthetic antimalarial and other treatments ranging from sun protection products, to topical and systemic corticosteroids, to immunosuppressants and immunomodulators.

Keywords: systemic lupus erythematosus, children, skin, aesthetic, psychological disabilities

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Introduction

Systemic Lupus Erythematosus (SLE) is a chronic, non-organ-specific autoimmune inflammatory disease characterized by an immune response directed against nucleolar material and the presence of autoantibodies, particularly native anti-DNA antibodies.[1] It is a heterogeneous disease, characterized by its great clinical polymorphism, which can affect almost all the organs, and by its difficult to predict evolution with flare-ups interspersed with multiple remissions.[2] Pediatric SLE is a rare entity, affecting children under the age of 16, and whose diagnosis is not always easy and the course is generally more severe than that of adults according to most of the few pediatric series. Published.[3,4] More than 80% of patients with SLE develop mucocutaneous involvement over the course of the disease. Long considered the hallmark of lupus, skin involvement is one of the most well-known and identified clinical features.[1] Indeed, it is the earliest of the clinical manifestations and the most constant. Certain cutaneous manifestations allow, by the simple clinical character and even in the absence of other signs of the disease, to evoke an SLE and to quickly make the diagnosis. The diagnosis of cutaneous lupus is not always easy; it is in fact based on a bundle of arguments; taking into account the clinical appearance of dermatological lesions, their topography, their evolution, compatible histology and the immunological context.

The different classifications of adult SLE, used and validated in children, in particular, the American College of Rheumatology ACR 1997 classification, the Lupus International Collaborating Clinics SLICC-2012 classification and that of the European League against Rheumatism EULAR/ACR -2019, all included several dermatological criteria. Indeed, the signs of acute and sub-acute cutaneous lupus, mouth ulcers and non-scarring alopecia were well defined and the rating assigned to them contributed strongly to the diagnosis of SLE.[5] Cutaneous manifestations observed in pediatric systemic lupus erythematosus can be schematically classified into three groups,[6] lupus lesions, vascular lesions and non-lupus non-vascular lesions.

Lupus lesions

Among these lesions, acute lupus erythematosus is by far the most frequent:

Acute lupus erythematosus: It is clinically characterized by a more or less edematous or scaly erythematous aspect, more rarely papular. In the localized form, the lesions sit mainly on the cheeks and the bridge of the nose; it is a butterfly-shaped rash or vespertilio, relatively respecting the nasolabial folds and the eyelids, often extending to the forehead, neck, and sun-exposed areas (Figure 1). The butterfly rash is seen in 60 - 85% of children with SLE.[7] In the diffuse form, the lesions usually predominate on the photo-exposed areas, producing a morbilliform, papular, eczematous, or bullous rash. On the back of the hands, lupus lesions mainly affect the inter-articular areas. The mucosal lesions of acute lupus are erosive and especially oral (Figure 2).[8, 6,1]

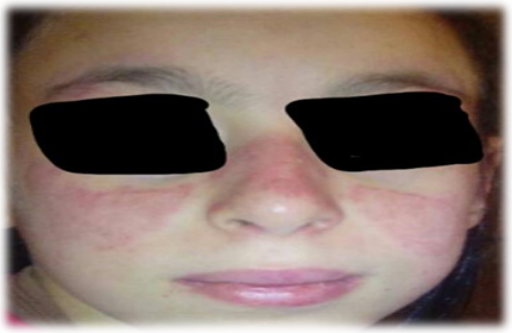

Figure 1 Butterfly rash or vespertilio.

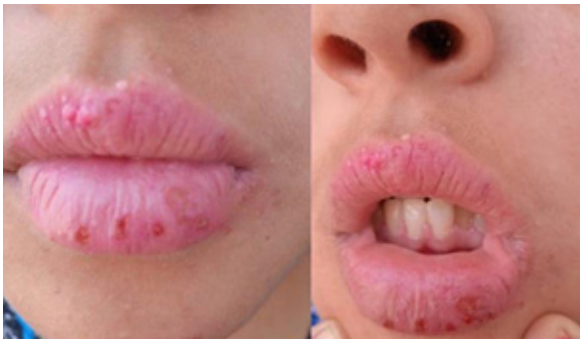

Figure 2 Erosive mucosal oral lesions.

Sub-acute cutaneous lupus: Most often presents as a papulosquamous, non-scarring rash. Typically, they are characterized by annular or polycyclic scales with an erythemato-scaly, vesicular or crusty border, with a grayish, clear or psoriasis form center. Rarely, they may look like erythema multiform (Rowell's syndrome). The rash is typically symmetrically distributed on the sun-exposed sites of the neck, the upper trunk, and the outer arms. Lesions resolve with post-inflammatory hypopigmentation; normal pigmentation recovers over time.[9]

Chronic cutaneous lupus: Include several varieties of types: discoid lupus, tumidus lupus, chilblains lupus and lupus erythematosus panniculitis or lupus profundus panniculitis. Among these chronic lesions, some are more frequent and aesthetically marked in this case:

Discoid lesions: These are large, scaly, erythematous, coin-shaped papules or plaques with prominent follicular plugging occurring in fewer than 10% of patients.[7] These lesions, often multiple and symmetrical, are found in sun-exposed areas located on the face, the bridge of the nose and the cheekbones; sometimes taking on a butterfly shape. They can affect the eyebrows, eyelids and scalp.[10,8,6] Discoid lupus can be disseminated, the lesions affect the whole body, in this case the limbs, especially the elbows, the palmoplantar region. Over time, discoid lupus may heal with discolored scarring and even hair loss when the scalp is involved (pseudopelades). Sometimes patients may feel pain or itch.[11]

Lupus tumidus: Its exact incidence in children is unknown. These are infiltrated erythematous plaques, characterized by erythema and bright urticarial erythematous-violaceous lesions that leave no scars after regression. They are mainly located on the face and on the photo-exposed areas of the upper part of the trunk (Figure 3).[12,11,9]

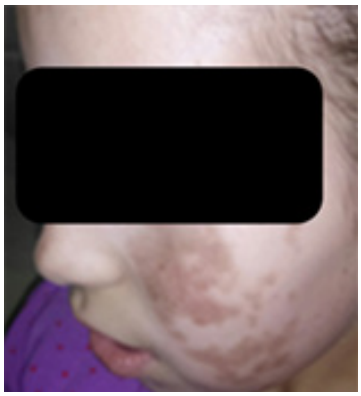

Figure 3 Lupus tumidus.

Chilblain erythematosus: It has a clinical aspect of banal chilblains persisting beyond the cold season. The most frequent topographies are the fingers and toes (Figure 4), but it is possible to observe lesions on the ears, nose, calves, heels, elbows and knees.[9]

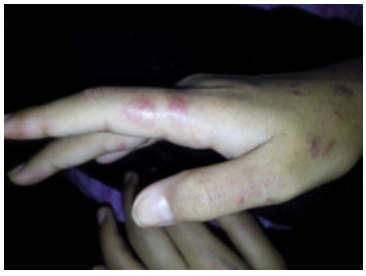

Figure 4 Chilblain Erythematosus Lupus.

Lupus panniculitis or profundus lupus: Begins with infiltrated erythematous, painful subcutaneous nodules of variable size. It happens that the skin next to these lesions is the seat of lesion of discoid lupus. These lesions regress giving way to a characteristic cupuliform depression, allowing a retrospective diagnosis (Figure 5).[9]

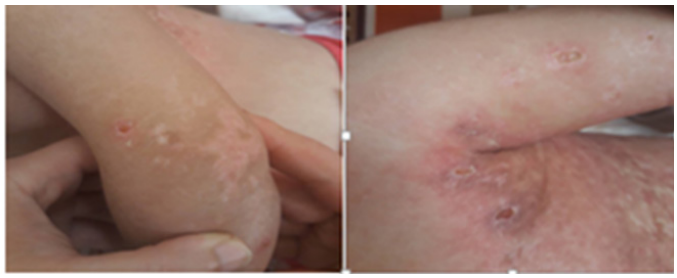

Figure 5 Profundus lupus.

Vascular lesions: Among these lesions, Raynaud's phenomenon is the most common: it is present in 15 to 45% of patients who may precede the onset of systemic lupus.[6,1] As to livedo reticularis, it can be associated with lupus but remains uncommon compared to other skin disorders. It is characterized by a reticular reddish-blue discoloration of the skin, which turns white on pressure. Livedo reticularis is permanent, not disappear with warming. It can take several aspects; it is termed regular, when the lesions are annular, complete, with unbroken circles unlike livedo racemosa when the circles are incomplete.[13] Most often these lesions are associated with lupus anticoagulants, and antiphospholipids. It can be serious in the presence of arterial or venous thrombosis which must always be checked.

Non-vascular non-lupus cutaneuos manifestations

Form a heterogeneous group of dermatological manifestations preferentially observed during lupus. Some are common, such as alopecia and Photosensitivity, while others are rare, such as bullous lupus, mucinosis or amicrobial pustulosis.

Alopecia: Scalp involvement is a frequent phenomenon in systemic lupus, and particularly non-sccaring alopecia with an incidence of 17-48% in children.[14] Most often, it is a significant loss of diffuse hair without alopecic plaque; it is characterized by fine, brittle hair that usually appears along the hairline. Patchy bald patches typical of alopecia are also seen, giving a sparse appearance to the scalp. Scalp attacks are most often acute or subacute, contemporaneous with flare-ups or occurring after, reversible after treatment; they generally reflect disease activity (Figure 6).[15]

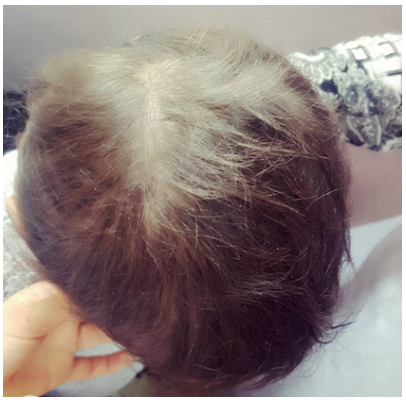

Figure 6 Alopecia in pediatrics lupus.

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Photosensitivity: Patients with SLE exhibit high photosensitivity and ultraviolet (UV) irradiation can lead to systemic flares. Photosensitivity is a frequent symptom that is one of the reasons for lupus disease consultation; It has an important diagnostic value, especially since it is part of the American Rheumatology Association (ARA) diagnostic criteria for SLE. 57% to 73% of patients with SLE describe photosensitivity. The frequency of triggering lupus lesions by sun exposure is very difficult to assess, because their appearance is delayed compared to exposure to the sun and the radiation is harmful even in the shade. However, it is estimated in SLE children between 16 and 50%.[14] The mechanism of its lesions is not very clear. It may be a matter of a systemic circulation of immunogenic photo-altered DNA or an effect of pro-inflammatory cytokines released by keratinocytes under the influence of Ultraviolet rays.[16,17]

Treatment of cutaneous lupus erythematosus

It is based on systemic and topical medication, designed to reduce disease activity and minimize aesthetics sequelae.[18]

Anti-malarial: They are the gold standard of pSLE therapy. Currently, the use of a synthetic antimalarial should be considered in children as a routine treatment whatever the clinical impairment. With their anti-inflammatory, immuno-modulatory and photo-protective activity, they are used to limit the risk of skin flare-ups induced by ultraviolet radiation. They have the property of concentrating in the epidermis and reducing the inflammatory reactions associated with UV irradiation.[18,9]

Hydroxychloroquine (HCQ): It is the most used molecule among anti-malarial in children and adolescents with a daily dose of 4 to 6.5 mg/kg and a maximum of 400 mg/day.[19] These dosages must therefore of course be adapted and modified in the event of weight variation in a growing being.

Systemic corticosteroids: New therapeutic recommendations in children try to reduce the use of systemic corticosteroids in order to avoid deleterious effects on growth, and serious complications, such as osteoporosis with the High risk of fractures and Cushing's syndrome. Short-term treatments are rather recommended with combinations of adjuvants (calcium, vitamin D and gastric protector). The generally recommended dosage is 0.5 to 1 mg/kg/day over 2 to 4 weeks followed by a gradual reduction.[20,21]

Topical corticosteroids: they can be used in well localized and limited lesions sparing their chronic use in the face to avoid unsightly repercussions in this case atrophy.[20]

Immunomodulators: Their immunosuppressive activity used with varying degrees of success during skin involvement in adults is long. Several molecules have been used such as methotrexate, azathioprine, cyclophosphamide, ciclosporin, mycophenolate mofetil, tacrolimus. Their use in children in cutaneous forms refractory to anti-malarial has not been evaluated by controlled clinical studies and therefore their use is discussed on a case-by-case basis in the face of refractory cutaneous lesions after ensuring optimal and regular intake anti-malarial by a plasma assay which can be assessed after 6 months of continuous use.

Oral retinoids: Use of vitamin A derivative is classified as second-line therapies for the treatment of cutaneous lupus-erythematosus by the American Academy of Dermatology guidelines. Retinoids are especially useful in patients with hypertrophic lesions on the palms and sole.

Physical therapy: The use of physical therapy in patients with cutaneous lupus erythematosus has been described in adults, with reported success with laser and cryotherapy. However, in children their use has not been well documented.

Effective sun protection : Sun protection and in particular the use of sun creams and sunscreen are important preventive measures which have been proven to minimize the exacerbations of lupus cutaneous attacks. Sometimes the use of topical corticosteroids or calcineurin inhibitors is essential.[20,21]

Biologics: Biotherapy is another therapeutic line to be used in cases of skin damage that is refractory to the usual treatments; although the results with certain biologics such as JAK inhibitors, are encouraging in adults, larger studies are needed to assess the effectiveness in children.[22,23]

Conclusion

The skin is a very affected organ during the pediatrics SLE. It is the real reflection of the activity of the disease and sometimes the annociator of a severe clinical presentation. However, the flip side of the coin is just as important, it can be the cause of disabling aesthetic damage, often poorly tolerated, with serious damage to self-image; which can have social and psychological repercussions. It is important to recognize the characteristics of cutaneous lupus in children in order to establish an appropriate and early diagnosis and initiate specific therapy.

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None.

Conflicts of interest

Authors declare there is no conflict of interest.

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