Decades of clinical research have yielded only one useful subset in systemic lupus erythematosus (SLE), which consists of those patients where lupus nephritis is the principal manifestation. Genetic studies have revealed numerous lupus-associated polymorphisms, but none of any practical use. In contrast to these disappointments, multiple molecular and cellular studies in recent years have unexpectedly begun to converge on the pivotal finding that SLE consists of a small number of distinct immunopathological entities, and that these can be identified by standard methods alone or in combination. Moreover, the same subsets can be identified in patient groups with different clinical diagnoses, such as Sjogren's syndrome, systemic sclerosis, or even rheumatoid arthritis. The results of these studies are not yet fully consistent with one another but gain in importance as multiple targeted therapies for lupus emerge. Thus, I will argue that we are standing on the threshold of a new taxonomy for the systemic autoimmune diseases, based on molecular and cellular analyses, and that such a taxonomy will facilitate more personalised therapeutic approaches.

Learning Objectives

- Explain the limitations of classifications based on clinical criteria
- Discuss recent research demonstrating the clustering of SLE and other systemic autoimmune diseases into distinct subgroups based on molecular and cellular markers
- Describe how a new taxonomy of the systemic autoimmune diseases will faciliate personalised therapeutic approaches

11 SHOULD PROS BE INCORPORATED IN THE RESPONSE EVALUATION?

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10.1136/lupus-2022-la.11

Health related quality of life (HRQOL) improvement in patients with SLE is defined as one of the treatment goals in the Treat to Target (T2T) recommendations and the 2019 EULAR recommendations for the management of SLE.[1 2] However, the definitions of remission and low disease activity (LLDAS) do not address the health-related quality of life or disease burden. In fact, the physicians' view on lupus dominated the development of remission criteria and it was postulated that a control of disease activity would improve QOL in patients with SLE.

The relationship between activity, organ damage, and HRQOL, however, remains complex and controversial, and the value of activity and damage indices as predictors of patient quality of life continues to be debated.[3] The attainment of remission in SLE represents the main treatment target, but QOL and fatigue are still insufficiently controlled in the state of remission and, despite improvement of disease activity, QOL can remain unchanged over several years.

A patient's perspective is still not accepted as equivalent to the physician's perspective in treatment decisions. HRQOL is neither directly nor indirectly captured by disease activity instruments. Therefore, a better understanding of the patients' experiences with the disease is crucial.[4]

Looking at the evidence of patient reported outcomes (PROs) as treatment targets for SLE, it is important to consider that, in clinical trials, the target response is mostly defined by changes in disease activity instruments and physician global assessments, while PROs were never used as the primary endpoint.

Studies to integrate the patient's perspective with the physician's definition of remission and low disease activity are needed.

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Learning Objectives

- Discuss quality of life in SLE, its determinants, discordance between physician's outcomes measures and PROs and the significance of PROs in clinical trials

Interactive case study workshops

12 MANAGEMENT OF LUPUS SKIN MANIFESTATIONS

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10.1136/lupus-2022-la.12

Case 1: A 19-year-old patient with bullous systemic lupus erythematosus

A 19-year-old patient presented with single, disseminated, erythematous papules and sharply demarcated, partly urticarial plaques on his face. Over the past few weeks, the skin manifestations had spread to the décolleté and blisters had developed on his forearms after sun exposure. A skin biopsy of one of the blisters showed a subepidermal vesicle containing neutrophils and scattered eosinophils. A perivascular and interstitial infiltrate of lymphocytes and neutrophils was seen in the upper and mid dermis, as well as formation of neutrophil microabscesses. In the direct immunofluorescence test (lupus band test), linear immunofluorescence was shown along the basement membrane zone with anti-IgA and anti-IgG antibodies. In addition, the patient showed a moderately diffuse alopecia, Raynaud's phenomenon, polyarthritis, synovitis, tendovaginitis splenomegaly, lymphadenopathy, fatigue, and night sweats. Laboratory analysis demonstrated anemia, leukopenia, hypocomplementemia, and autoantibodies (ANA and anti-dsDNA antibodies).

Bullous systemic lupus erythematosus (BSLE) is a rare disease associated with subepidermal blistering and, in most cases, severe systemic manifestations. The vesiculobullous skin changes can occur after sun exposure and can be associated with activation of SLE. Dapsone is the mainstay of systemic treatment in this disease and systemic corticosteroids and antimalarials have shown minimal improvement.[1–3]

Discussion Points: Different forms of skin lesions and treatment options in patients with cutaneous lupus erythematosus.

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Learning Objectives

- Recognise the specific (i.e. ACLE, SCLE, CCLE, ICLE) and non-specific skin manifestations in cutaneous lupus
- Explain the RCLASI as validated activity and damage score of cutaneous lupus
- Discuss the therapeutic guidelines of cutaneous lupus
- Describe the preventive strategies in cutaneous lupus, including photoprotection
- Discuss the topical and systemic treatment options in cutaneous lupus

13 MANAGEMENT OF LUPUS SKIN MANIFESTATIONS

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10.1136/lupus-2022-la.13

Case 1. A 75-year-old woman with erythematous-scaling plaque of the nose

A 75-year-old woman presented with a unique erythematous-scaling plaque of the nose. A skin biopsy showed an interface dermatitis, consistent with the diagnosis of localised cutaneous lupus. Based on clinical and laboratory findings, systemic lupus was revealed. According to guidelines, hydroxychloroquine 400 mg/day was introduced in association with topical treatment with resolution of skin lesions in a few weeks with no relapses in the follow-up period.

Case 2. Recalcitrant skin lesion in a 62-year-old woman with systemic lupus erythematosus

A 62-year-old-woman with a 16 year-history of systemic lupus, presented with erythematous-infiltrated and partially hyperkeratotic skin lesions on the trunk, arms and face. Lesions were extremely itchy. A skin biopsy confirmed cutaneous lupus. All systemic treatments, including hydroxychloroquine, quinacrine and belimumab failed. The patient was re-evaluated to exclude the presence of external factors associated to the exacerbation of the disease. Finally, methotrexate 15 mg weekly improved the lesions.

Discussion Points: We discuss two cases focusing on the importance of correct diagnosis and treatment of specific lupus skin lesions. Antimalarials remain the first-line therapeutic option, but choice of other drugs should be taken into consideration in cases of refractory forms of cutaneous lupus.

Learning Objectives

- Discuss the therapeutic guidelines of lupus erythematosus
- Discuss the therapeutic options in recalcitrant cutaneous lupus erythematosus
- Explain the role of itching in cutaneous lupus erythematosus
- Describe factors predicting exacerbation

14 MANAGEMENT OF LUPUS NEPHRITIS

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10.1136/lupus-2022-la.14

Case 1: A 27-year-old female with lupus nephritis

A 27-year-old Caucasian female was diagnosed with systemic lupus erythematosus (SLE) in 2016 based on arthralgias, high fever, malar rash, leukopenia, anemia, ANA, anti-SM, anti-RNP, anti-dsDNA, and anti-C1q positivity. Renal function and urinalysis were normal. She was treated with prednisone 8 mg/day, hydroxychloroquine 300 mg/day and mycophenolate mofetil (MMF) 1.5 g/day.

In April 2018, urinalysis showed proteinuria 1.1 g/day and active urinary sediment with dysmorphic erythrocytes and erythrocyte casts, and normal renal function.

The patient underwent kidney biopsy. Despite the mild renal lab' alterations, kidney biopsy showed a severe intra- and extra-capillary glomerulonephritis Class IV ISN/RPS with an activity index of 15 and chronicity index of 1. Due to the severity of the histology, cyclophosphamide was suggested, but the patient refused and was treated with the methylprednisolone pulses and rituximab 2 g 15 days apart, and MMF at higher dose than originally prescribed. Twelve months later urinary manifestations were clearly improved, however, to prove that also histological lesions had also improved, a second kidney biopsy was performed. The second biopsy revealed the persistence of active lesions though of lesser severity compared to previous biopsy but an increase in the chronicity index. Based on this result, immunosuppressive therapy was strengthened.

Learning Objectives

- Describe the discrepancies between clinical and histological data
- Explain the importance of kidney biopsy and of activity and chronicity indexes particularly in cases of mild clinical renal presentation.
- Explain why the approach to lupus nephritis cannot be standardised
- Describe the new therapeutic approaches of lupus nephritis
- Explain of the importance of repeated kidney biopsy to evaluate the response to therapy

Case 2: A 28-year-old pregnant Caucasian woman

This is the case of a 28-year-old Caucasian woman at her second pregnancy. The patient experienced a deep venous thrombosis in the lower limbs at 19 years and a miscarriage at 26 years. ANA:1/160 was found during an immunological screening performed after the miscarriage.

At the 32nd week of the second pregnancy the patient developed arterial hypertension, severe proteinuria and 15 kg body weight increase. Preeclampsia was diagnosed, C-section was performed, giving birth to a 2.45 kg male child.

Six months later proteinuria persisted and reached nephrotic range (proteinuria 8 g/24 h, serum protein 5.1 g/dl, albumin 2.8 g/dl). Renal function was normal and urinary sediment showed only lipid casts and fat oval bodies. Immunological screening confirmed ANA positivity 1/160 only. A