

REFERENCES

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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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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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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