Case 2. Role of the imaging in lupus

A 34-year-old Caucasian patient was diagnosed with SLE in 2020 after presenting with inflammatory arthritis, Raynaud's phenomenon, chilblain lupus, and oral ulcers. She started treatment with hydroxychloroquine 200 mg/day plus prednisone 5 mg/day. Two months later she complained of joint pain, but no swollen joints were seen on physical examination. We performed a hand ultrasound assessment. Longitudinal examination at the level of the proximal interphalangeal joint indicates the presence of a discrete component of synovial effusion, irregularity, and prominence of the articular component of the distal epiphysis of the proximal phalanx. The Power Doppler evaluation showed a moderate increase in vascular uptake at the joint level of synovial capsular distention and periarticular soft tissues in the distal epiphysis of the proximal phalanx. Findings were suggestive of active synovitis with associated joint remodelling and active inflammatory and neovascular signs of the periarticular soft tissues. Treatment was changed. She started MTX treatment 15 mg/week initially oral and then SC. Eight weeks later she persisted with articular pain in MCP and proximal inter-phalangeal joints in both hands, but no evidence of synovitis was seen on clinical examination. She underwent magnetic resonance imaging of the hands and wrist - coronal STIR sequence. Mild synovitis was present in the second, third, and fourth metacarpophalangeal joints and the distal radioulnar joint, there was no structural or inflammatory bone damage.

Learning Objectives

- Discuss the treatment approach for patients with SLE and erosive arthritis (rhupus)
- Discuss the role of imaging in SLE and treatment approaches in patients with articular involvement

20 MANAGEMENT OF SKIN INVOLVEMENT IN SLE

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Topical calcineurin inhibitors in cutaneous lupus erythematosus

In the management of cutaneous lupus erythematosus (CLE), topical calcineurin inhibitors have recently been established as the first-line treatment according to the S2k guidelines for the treatment of CLE.[1 2] This recommendation is further supported by the 2019 European League Against Rheumatism (EULAR) recommendations, which state that topical calcineurin inhibitors should be considered as a primary choice for treating cutaneous lesions in patients with systemic lupus erythematosus (SLE).[3] A randomized, controlled trial demonstrated tacrolimus 0.1% ointment to be significantly more effective than placebo in treating CLE.[4] Additionally, facial lesions showed a better response to tacrolimus ointment 0.1% compared to lesions on the body, particularly when the lesions had been present for less than 6 months. In summary, tacrolimus ointment 0.1% is recommended primarily for treating facial lesions in CLE and can serve as an alternative to topical glucocorticoids. In cases where the disease is widespread and/or there is a risk of scarring, concurrent treatment with antimalarials is recommended.

Case: A 34-year-old female with SLE A 34-year-old female patient diagnosed with SLE presented with confluent erythematous, edematous papules and plaques, known as 'malar rash', on the left side of her face. She received treatment with antimalarial agents and mycophenolate mofetil; however, over the past month, she had developed skin lesions after sun exposure. After 28 days of treatment with tacrolimus ointment 0.1%, her skin lesions had completely resolved. No recurrence of skin lesions was observed after 84 days of treatment with 0.1% tacrolimus ointment.[4]

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Learning Objectives

- Discuss the topical and systemic treatment options in CLE
- Describe the preventive strategies in CLE
- Discuss the therapeutic guidelines of CLE
- Describe the RCLASI as validated activity and damage score of CLE

21 MANAGEMENT OF SKIN INVOLVEMENT IN SLE

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Case 1: Blisters in systemic lupus erythematosus

Skin lesions can be very heterogeneous in systemic lupus erythematosus (SLE). Clinicopathological correlation is crucial to identify the type of cutaneous lesion since it provides prognostic and therapeutic implications. Blistering lesions are rare in lupus erythematosus; however, they can be difficult to characterize and the cutaneous differential diagnosis is wide. The term 'bullous lupus' can be confusing since it may even reflect different immune mechanisms that need specific management.

A 42-year-old with a 10-month history of hand arthritis and positivity for antinuclear antibodies (ANA), anti-RNP and anti-Sm, presented with a 2-month history of cutaneous lesions affecting her upper trunk and upper limbs. Vesicle and blisters were observed on the aforementioned sites. She had been treated with non-steroidal anti-inflammatory drugs and short pulses of prednisone that improved partially her skin lesions. The lesions resolved without scarring but leaving hypopigmentation.

Learning Objectives

- Demonstrate the clinical differential diagnosis of blisters in SLE
- Describe the main immunopathologic findings of blistering eruptions in SLE
- Explain the main features of bullous systemic lupus erythematosus
- Discuss the extracutaneous associations of blistering eruptions in SLE

- Describe the treatment of bullous systemic lupus erythematosus

Case 2: Hand lesions in SLE

Hand lesions are relatively common in patients with SLE. Since these lesions are not always biopsied (especially those of the digits) a correct characterization may be lacking. These lesions can appear in patients with or without SLE. The differential diagnosis includes vasculitis, vasculopathy, chilblain lupus and palmoplantar lupus. Although localized, they are usually painful with functional impairment of loss of quality of life. Among them, the most common lesions are chilblain lupus. Response to topical treatment and antimalarials is typically poor. Chilblain lupus may not respond as well to systemic therapy compared to other visceral manifestations of SLE, hence being a treatment challenge both for rheumatologists and dermatologists.

A 40-year-old woman, with an 8-year history of cutaneous lesions in the face and arms (diagnosed of cutaneous lupus erythematosus by biopsy), arthritis and ANA+, treated with hydroxychloroquine and variable doses of prednisone, was admitted in the rheumatology department due to headache and visual loss with bilateral papilledema. Magnetic resonance imaging was normal but the ophthalmological exploration showed inferior branch retinal vein thrombosis in the left eye. The autoimmunity study showed ANA 1/1280 and positivity for anti-dsDNA, anti-RNP and anti-Sm. With the possible diagnosis of SLE-related pseudotumor cerebri, prednisone dose was increased, hydroxychloroquine was maintained and acetazolamide was added, with progressive vision recovery. Two months after discharge, she started to progressively present papulosquamous lesions on the back, neckline, back of the hands and fingertips, the latter being painful.

Learning Objectives

- Describe the differential diagnosis of hand lesions in SLE
- Understand the prognostic implications of hand lesions in SLE
- Discuss how cutaneous manifestations may not respond to systemic treatments aimed at controlling active SLE
- Explain chilblain lupus and its treatment
- Describe available treatments strictly directed to the cutaneous component of lupus erythematosus

Prime time session: (hybrid)

Part I: the patient at centre stage

22 THE PATIENT´S VOICE

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Systemic lupus erythematosus (SLE) is a very complicated and heterogeneous disease, and the popular saying is that no two lupus patients are the same. This of course also means that the treatment and planning of care can be very complicated and needs to be adjusted to the individual patient.

One of the best ways to achieve this goal is through shared decision making. If the patient feels like they have a voice in the treatment plans the probability of treatment adherence increases substantially. On average the SLE patient is more aware of their own symptoms, disease progression and medication than patients with less complicated or less heterogeneous diseases. They need to become experts in their disease because they are the ones living with the symptoms and can often 'feel' a flare coming on before the laboratory results show it. Health-related-quality of life (HRQoL) when living with a chronic disease like SLE very much depends on how you self-manage; living a healthy life, keeping active, prioritising everyday tasks according to energy-level etc. The engaged and informed patient can be both a help and a burden when it comes to the physican´s disease management. It helps if the patient has the right medical information and respects that the physician has the expertise to decide what is best for them.[1] A patient needs to know that not every symptom is because of lupus and that they might not be flaring, even though they feel like they are.

This 'complicated' care of an SLE patient often takes more than one health care professional. Apart from a multidisciplinary team of physicians each specialised in their own manifestations (like kidneys, lungs, heart etc.) it is often beneficial to involve other areas such as specialised nurses, physiotherapists, psychologists, occupational therapists etc. This multidisciplinary team of course requires a good coordination, which should not be put exclusively on the patient.

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Learning Objectives

- Explain the importance of involving patients in shared-decision-making
- Describe how a multidisciplinary team can be used in the SLE care
- Describe the role of the informed patient in their own care

23 THE NURSE'S PERSPECTIVE

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Lupus (Latin term for wolf) was used, since the Middle Ages to describe several types of diseases characterized by ulcerous lesions, mainly in the lower limbs. The true turning point in its history occurred at the beginning of the 19th century, with the distinction between lupus vulgaris and cutaneous lupus in its modern sense.[1]

Today, systemic lupus erythematosus (SLE/lupus) is described as a chronic systemic autoimmune disease of variable severity and course, distinguished by a tendency for flare. It is clinically and serologically, a diverse autoimmune disease that can affect any organ or system of the body and display a wide spectrum of manifestations.[2]

Evidently, due to the chronic relapsing-remitting nature of SLE,[3] a holistic approach, involving a multi-disciplinary team (Physician, Rheumatology/Lupus Nurse Specialist, Physiotherapist etc) is essential to providing high quality care to lupus patients.