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REFRACTORY CUTANEOUS LUPUS: USE OF THALIDOMIDE OR LENALIDOMIDE FOR REFRACTORY LUPUS SKIN DISEASE

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Cutaneous lupus erythematosus (CLE) may cause extensive skin damage leading to aesthetic prejudice and poor quality of life. Antimalarial agents are recommended as first-line systemic therapy for CLE patients with moderate-to-severe skin lesions. In case of failure of antimalarial agents, a European expert consensus for CLE treatment and EULAR recommendation for systemic lupus erythematosus (SLE) recommend adding quina-craine, methotrexate, retinoids, dapsone or mycophenolate mofetil.¹ However, the overall efficacy of these second-line treatments is approximately 50% depending on CLE subtypes and may be associated with potential toxicity. Thalidomide (a-N-phthalimidoglutarimide) is currently recommended as a ‘rescue’ therapy in patients with severe and refractory CLE treatment. However, in France, a ‘Temporary Recommendations for Use’ has been granted for thalidomide in CLE after failure of antimalarials as second-line agent.^{1–2} Moreover, in a systematic literature review including 21 observational studies and 548 patients the pooled rate of response to thalidomide 50–100mg/day was 90% (95% CI, 85–94), with similar response rates between CLE subtypes.³ The clinical benefits need to be balanced against potential adverse events with a pooled rate of thalidomide withdrawal related to adverse events of 24% (95% CI 14–35) including high teratogenicity, peripheral neuropathy 16% (95% CI 9–25), thromboembolic events in 2% (95% CI 1–3). Moreover, the efficacy of thalidomide is only suppressive with a rate of relapse of 71% (95% CI 65–77) after thalidomide withdrawal which supports the use of a minimal maintenance dose.³ In case of failure of thalidomide, lenalidomide 5 mg/day a 4-amino-glutamyl analogue of thalidomide has shown promising results with partial response of 88% in a retrospective study of 40 patients.⁴ Importantly, no cases of new or worsening peripheral thalidomide-induced neuropathy was reported with lenalidomide.⁴ A case of progression to SLE with renal involvement was reported in a patient with isolated CLE after starting lenalidomide.⁵ Recent data did not confirm this finding but suggested that lenalidomide has little or no effect on global SLE activity.

Learning Objectives

- Describe the therapeutic strategy in CLE
- Explain when to prescribe thalidomide and lenalidomide
- Discuss the safety profile of thalidomide and lenalidomide

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OSTEONECROSIS: PREVENTION AND TREATMENT

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Osteonecrosis (ON) is a well-known component of damage accrual in patients with systemic lupus erythematosus (SLE), it frequently causes disability and considerably affects patients’ quality of life.

The increased incidence of ON in SLE patients as compared to the general population, patients with other autoimmune diseases, or diseases requiring high doses of corticosteroids (CS) suggest that the disease itself and/or the underlying genetic background of the patients might dictate the susceptibility in its occurrence.^{1–5} Several modifiable risk factors have been associated with an increased risk of ON, such as alcohol intake, cigarette smoking, high triglyceride levels, and heavy physical work. The use of CS has been recognized as a major risk factor for the development of ON. The average daily dose of CS, its highest dose, and the total cumulative CS dose, as well as the use of pulse therapy, and the presence of Cushingoid appearance have all been associated with ON. The prevalence of ON in patients with SLE ranges between 5–15% but it may be as high as 40% when patients with silent patterns are included. The clinical presentation of ON is variable, being related to the size and location of the affected bone(s). The hip and knee are the most frequently affected joints followed by ankle and shoulder. Multifocal ON has been reported in up to half of patients with SLE.

The early diagnosis of ON is challenging because it frequently occurs silently; there is often a time lag between the development of ON and the onset of symptoms. Preventing ON involves controlling modifiable risk factors. Corticosteroids, if needed, should be prescribed at the lowest possible dose and for the shortest period of time, regardless of patient’s disease manifestations. Anticoagulant therapy, and statins/lipid-lowering drugs, may reduce the incidence of CS-induced ON.

The therapeutic approach depends largely on the joint involved and the extent of the injury. Immobilization has a role for small lesions that will spontaneously heal. Various medications have been used anecdotally with some benefit, including lipid-lowering drugs, anticoagulants, vasodilators, and bisphosphonates. Stem cell treatment of femoral head ON has

been reported as useful therapy; however, this therapeutic approach has not been standardized and will need to be studied further. The type of surgical therapy is based on the severity of joint damage. For early ON, core decompression and percutaneous debridement and drilling is recommended. For ON lesions prior to bone collapse, bone grafting and osteotomies are also a possibility. Once subchondral fracture collapse is evident, bone grafting, hemi-resurfacing and total hip arthroplasty are the treatment options.

Learning Objectives

- Describe the epidemiology and clinical presentations of ON in patients with SLE
- Explain the modifiable and non-modifiable risk factors for ON in patients with SLE
- Discuss the preventive measures for ON in patients with SLE
- Describe how patients with SLE and ON should be treated

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ALVEOLAR HEMORRHAGE

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Diffuse alveolar hemorrhage (DAH) has been described in a number of systemic autoimmune diseases, including systemic lupus erythematosus (SLE), antiphospholipid syndrome (APS), Behçet's disease, microscopic polyarteritis, cryoglobulinaemic

vasculitis, Henoch-Schönlein purpura, Goodpasture's syndrome, granulomatous vasculitis and others.¹

The potential clinical importance of this complication needs to be stressed. It is likely that the true frequency is significantly higher than the very low prevalence suggested by the paucity of reported cases. Diffuse alveolar hemorrhage syndromes are notoriously difficult to diagnose. In the majority of cases, there is little or no haemoptysis, even with large volume intra-alveolar bleeding. The radiological signs are sometimes florid but often highly non-specific, consisting of amorphous ground-glass attenuation on chest radiography or high-resolution computed scans of the thorax. Invasive or semi-invasive evaluation is generally required, specifically bronchoscopy, with or without a surgical lung biopsy.

In general, in treated autoimmune diseases, infiltrative lung disorders can, for practical therapeutic purposes, be divided broadly into opportunistic infection, which demands specific antimicrobial therapy and a reduction in immunosuppression, and a wide range of immunologically mediated processes, which demand the opposite approach: intensification of immunosuppressive therapy. Refinement of the differential diagnosis in the latter group is important, but less important than the exclusion of infection. Bronchoalveolar lavage (BAL) to exclude infection has been the pivotal investigation that has allowed an empirical immunosuppressive approach. A pragmatic approach of standard immunosuppressive therapy after the exclusion of infection is not, in itself, sufficient. Active steps must be taken to diagnose DAH.²

Learning Objectives

- Explain the main challenges in the differential diagnosis of alveolar hemorrhage in SLE
- Describe the options for the treatment of alveolar hemorrhage in SLE
- Discuss new trends in research on new markers for alveolar hemorrhage in SLE

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