CLINICAL IMAGE

Lymphocytic interstitial pneumonia as an extremely rare manifestation of systemic lupus erythematosus

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Systemic lupus erythematosus (SLE) is a chronic autoimmune disease, which may affect multiple organs, including the skin, kidneys, the central nervous system, joints, the hematopoietic system, and lungs. More than 50% of patients with SLE experience at least one pulmonary manifestation. Involvement of the respiratory system in SLE is associated with high mortality.[1] The most common pulmonary manifestations are pleurisy, chronic interstitial lung disease, diffuse alveolar hemorrhage, pulmonary hypertension, pulmonary embolism, and shrinking lung syndrome.[1] Lymphocytic interstitial pneumonia (LIP) is rarely associated with SLE and most commonly occurs in patients with Sjögren syndrome.[2,3]

A 30-year-old male smoker was admitted to the Department of Rheumatology due to fever up to 39 °C, loss of weight, arthritis, myalgia, nonscarring alopecia, photosensitivity, butterfly rash, and oral ulcers (FIGURE 1A). Additionally, he reported xerostomia and eye dryness. Laboratory tests revealed elevated inflammatory markers (C-reactive protein and erythrocyte sedimentation rate), hypergammaglobulinemia, and presence of rheumatoid factor and antinuclear antibodies at a titer of 1:10 240 with SSA, SSB, and ribosomal *P* antibodies. Diagnostic virologic testing excluded infection with hepatitis B virus, hepatitis C virus, and HIV. The patient was diagnosed with SLE based on the 2019 European League Against Rheumatism / American College of Rheumatology criteria. He also fulfilled Sjögren syndrome classification criteria. The patient was treated with glucocorticoids (GCs): initially with methylprednisolone at a dose of 80 mg intravenously for 4 days, followed by oral prednisone at a tapered dose. Treatment attempts with multiple disease-modifying antirheumatic drugs were made. Methotrexate was discontinued due to adverse effects (mouth ulcers), and azathioprine, cyclosporine, and hydroxychloroquine proved to be ineffective despite treatment with maximum doses. After 6 months, the patient was readmitted to the clinic due to severe erythematous-infiltrative skin lesions on the face, neck, chest, upper back, and shoulders. Skin lesions appeared after 30 minutes of exposure to sunlight and covered more than 70% of the surface of the skin (FIGURE 1B). A skin biopsy was performed and showed subacute cutaneous lupus erythematosus. Treatment with higher doses of GCs and intravenous immunoglobulins was used, and mycophenolate mofetil (MMF) 2 g/d was included as a disease-modifying antirheumatic drug resulting in significant improvement after one month (FIGURE 1C). However, despite sustained low disease activity for 6 months and no pulmonary symptoms, a routine chest X-ray revealed emphysematous bulla. Arterial blood gas measurement did not show any abnormalities. Spirometry showed moderate obstruction (forced expiratory volume in the first second of expiration, 67%) without reversibility in the bronchodilator test and diffusion capacity for carbon monoxide was mildly impaired (DLCO, 60%). We performed high-resolution computed tomography, which showed multiple cysts (maximum dimensions, 70 × 48 mm) and ground-glass opacities suggesting LIP (FIGURE 1D). After pulmonary consultation, treatment with MMF and GCs was maintained, and the patient is followed on an ambulatory basis.

LIP is a rare type of diffuse parenchymal disease, which is characterized by pulmonary infiltration of lymphocytes and plasma cells.[2,3] The first-line therapy of LIP is oral GCs.[2] Other reported treatment includes cyclophosphamide,

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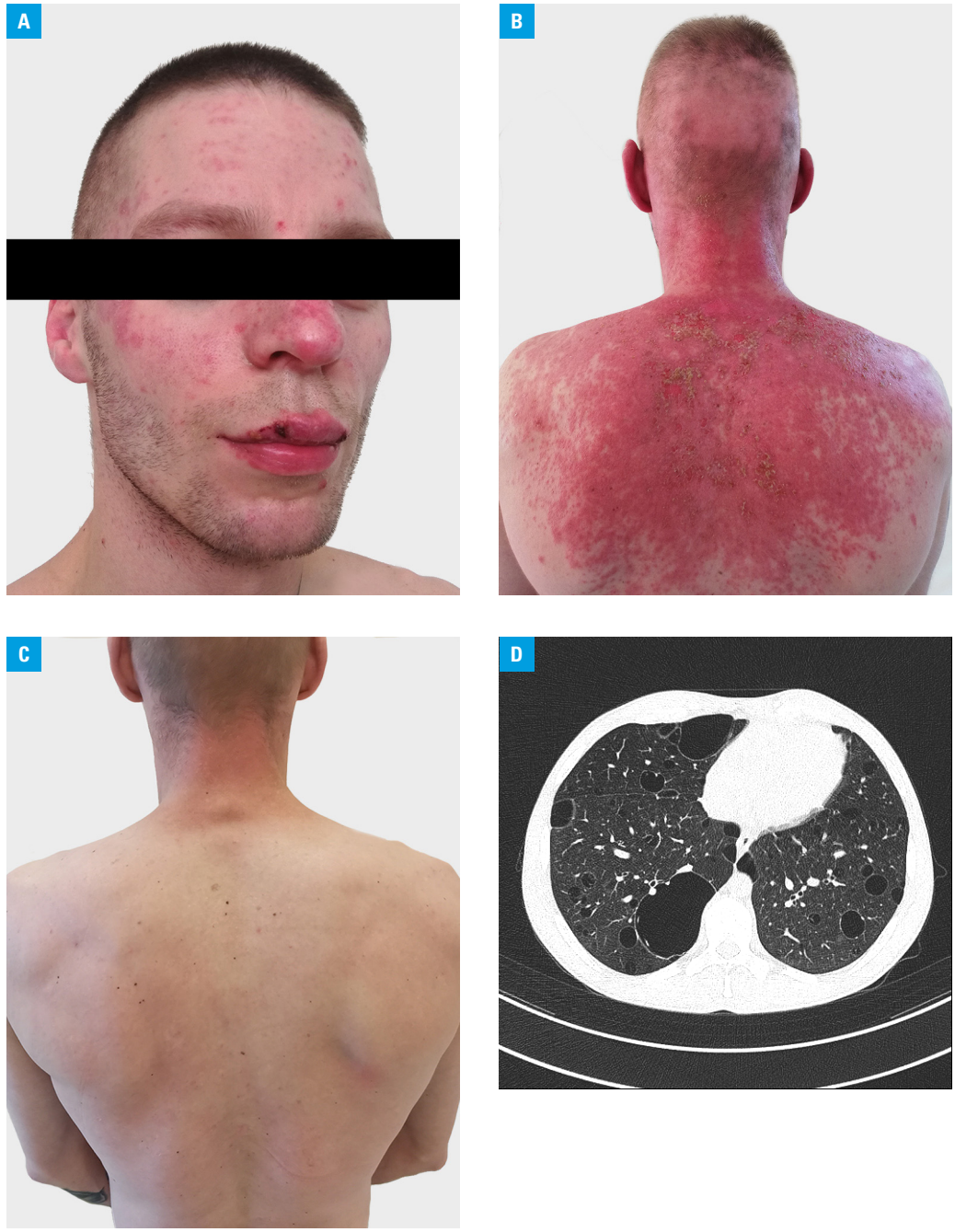

FIGURE 1 **A** – butterfly rash, oral ulcers, and mucocutaneous lesions on the face; **B** – severe erythematous-infiltrative skin lesions on the upper back and shoulders; nonscarring alopecia; **C** – resolution of skin lesions after mycophenolate mofetil therapy; **D** – high-resolution computed tomography showing ground-glass opacities and thin air cysts in both lungs, with the largest cyst (70 × 48 mm) located in the tenth segment of the right lung

rituximab, and MMF.[1,2,4] Even with immunomodulatory treatment, only 50% to 67% of patients survive more than 5 years after being diagnosed with LIP.[2] About 5% of cases of LIP also transform to lymphoma.[2] Our patient did not present any pulmonary symptoms, despite advanced abnormalities on imaging. For this reason, we suggest that all patients diagnosed with SLE (even asymptomatic) should be screened for pulmonary complications.

ARTICLE INFORMATION

PATIENT CONSENT The patient provided informed consent to publish the photographs.

CONFLICT OF INTEREST None declared.

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