Antisynthetase syndrome without muscle involvement is a well-defined syndrome characterized by the presence of interstitial lung disease in association with arthritis, myositis, mechanic’s hands, and Raynaud’s phenomenon in the presence of antisynthetase antibodies, especially Ac anti-Jo1. We described the case of a 68-year-old man with this syndrome in the absence of inflammatory muscle disease.

Key words: Antisynthetase syndrome. Interstitial lung disease. Antisynthetase antibodies. Inflammatory myopathy and mechanic’s hands.

Síndrome antisintetasa sin afectación muscular

El síndrome antisintetasa se caracteriza por la asociación de enfermedad pulmonar intersticial, clínica inflamatoria sistémica y artritis, miositis, manos de mecánico y fenómeno de Raynaud, todo ello en presencia de anticuerpos antisintetasa, fundamentalmente anti-Jo1. La ausencia de afectación muscular es infrecuente. Presentamos un caso de un paciente varón de 68 años con síndrome antisintetasa en ausencia de miopatía inflamatoria.

the patients presents pain and swelling of the hands, Raynaud’s phenomenon, and finger coarsening with hyperpigmentation, which results in a consultation with rheumatology.

Upon physical examination he had arthritis of the wrists, PIP and DIP joints and mechanics’ hands (Figure) without any muscle weakness. He persisted with elevated ESR and CRP in the tests but CPK and aldolase were normal. Immunologic testing showed high positive titres of anti-Jo1 and anti-RO/SSA antibodies. Treatment was started with steroids and leflunomide with a remission of the joint (polyarthritis) and systemic (normal ESR and CRP) symptoms after 4 months.

Discussion

Antisynthetase syndrome is an infrequent disease classically associated to idiopathic inflammatory myopathies (IIM), polymyositis, and dermatomyositis.1,2 Interstitial lung disease is the characteristic clinical affection and is manifested as a dry cough and dyspnea associated to physical activity.3 In a great sense it determines the prognosis of this process. Practically all of the patients have muscle affection. This can occasionally happen years after the onset of lung disease. Its absence, though documented in the literature, is exceptional.4,5 Other common manifestations are constitutional symptoms, fever, polyarthritis, mechanics’ hands, and Raynaud’s. The presence of antisynthetase antibodies (specific antibodies of myositis) is characteristic.6 These antibodies intervene in the binding of different aminoacids to RNA (during the formation of tRNA) by acting against its respective synthase enzyme. Anti-Jo1 antibodies (vs histidine) is the most frequently encountered in these patients and seems to be related to the development and severity of lung disease.7 Our patient also presented anti-RO/SSA antibodies. Their presence has been associated in IIM (myopathy-associated antibodies) to a worse prognosis of lung disease.8 Treatment of antisynthetase syndrome consists of steroids in association to immunosuppresants (azathyprine and methotrexate), especially if there is lung and muscle affection. Good outcomes have been reported with the use of rituximab.9 In our case, the choice of leflunomide was determined by the predominance of joint clinical symptoms and signs and it has been employed, with good results, in different systemic diseases, including antisynthetase syndrome.10

References