

# Antisynthetase Syndrome Without Muscle Involvement

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Antisynthetase syndrome 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.

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### 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.

**Palabras clave:** Síndrome antisintetasa. Enfermedad pulmonar intersticial. Anticuerpos antisintetasa. Miopatía inflamatoria y manos de mecánico.

### Introduction

A case of antisynthetase syndrome without myopathic affection treated with leflunomide and oral corticosteroid therapy is presented.

### Case Report

A 68-year-old male with a personal history of dyslipidemia and who had undergone surgery due to a prostatic adenoma, came to the emergency room due to malaise, fever, and dyspnea which had lasted 2 months. Lung examination showed decreased breath sounds and rales on both lung bases, but predominant on the left. The chest x-ray showed an infiltrate in both pulmonary bases that obscured the diaphragm and the cardiac profile. An arterial blood gas test revealed respiratory insufficiency (PO<sub>2</sub>, 50.7 mm Hg; PCO<sub>2</sub>, 41 mm Hg, and SatO<sub>2</sub>, 87.6%), and the hemogram showed hypereosinophilia (7.4%), elevated LDH (633 U/L), and CPK (140 U/L), the rest being normal.

The patient was hospitalized (neumology department) and started on antibiotics. A lung computer tomography showed bilateral infiltrates predominant in the periphery of the lower with a bronchogram, septum widening, and zones with an "unpolished glass" aspect in the lower right lobe. The bacilloscopy, sputum cultures, and serologic tests for atypical microorganisms were negative. Mantoux testing was negative. A bronchoscopy was interrupted before a bronchoalveolar lavage or a transbronchial biopsy could be performed. Other test performed were ESR (33 mm/h) and CRP (36 mg/dL), with aldolase, CPK, GOT, and GPT, and their results were normal. The immunologic study was normal. Because of the suspicion of an interstitial lung disease, treatment with steroids was started (1 mg/kg/day of methylprednisolone, orally, with a gradual decrease in dose). Clinical improvement was noticeable at 2 weeks. The chest x-ray and the respiratory functional tests were normalized over the course of the following months, and a spirometry done after discharge showed a restrictive pattern of disease (FEF<sub>25%-75%</sub>, 85%; FEV<sub>1</sub>, 70%; FVC, 65%, with FEV<sub>1</sub>/FVC of 76%, and a decrease of DL<sub>co</sub> of 77%). Twelve months after, coinciding with a decrease in the steroid dose to 10 mg/day of prednisone,

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**Figure.** Detail of the “mechanics’ hands” (presence of creases and finger hyperpigmentation).

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.<sup>1,2</sup> Interstitial lung disease is the characteristic clinical affection and is manifested as a dry cough and dyspnea associated to physical activity.<sup>3</sup> 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.<sup>4,5</sup> Other

common manifestations are constitutional symptoms, fever, polyarthritis, mechanics’ hands, and Raynaud’s. The presence of antisynthetase antibodies (specific antibodies of myositis) is characteristic.<sup>6</sup> 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.<sup>7</sup> 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.<sup>8</sup> Treatment of antisynthetase syndrome consists of steroids in association to immunosuppressants (azathyoprine and methotrexate), especially if there is lung and muscle affection. Good outcomes have been reported with the use of rituximab.<sup>9</sup> 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.<sup>10</sup>

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