



Images in clinical rheumatology

Muscle weakness and dyspnea

Debilidad muscular y disnea

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Case report

We present the case of a 71-year-old male, ex-smoker of 20 packets/year. He did not report visits to foreign countries, contact with animals nor any other background of interest. The signs and symptoms began with muscular weakness in the pelvic and scapular girdles. Once the diagnosis of polymyalgia rheumatica had been established, treatment with 0.3 mg/kg/day of methylprednisolone was initiated. The patient returned after 6 weeks with fever, dyspnoea, non-productive cough and pleuritic ribcage pain. These symptoms were accompanied by symmetric arthralgias in wrists and ankles. Examination revealed general bad health and a fever of 38.5 °C, tachycardia of 110 beats per minute, tachypnoea of 30 breaths per minute and bilateral dry, crepitant rale. He presented minimal inflammation of both wrists and Raynaud's phenomenon on both palms (Figure 1). Analytical analyses showed a leukocyte count of $11,280 \times 10^3/\mu\text{l}$, a neutrophil count of $8,800 \times 10^3/\mu\text{l}$, lactate dehydrogenase at 1,160 IU/l, aspartate-aminotransferase at 93 IU/l, alanine-aminotransferase at 110 IU/l, alkaline phosphatase at 100 IU/l, gamma-glutamyltransferase at 135 IU/l and a globular sedimentation velocity of 79 mm/1st h. Creatine kinase and creatine kinase MB isoenzyme, troponin I, myoglobin and aldolase enzymes were all within normal levels, as was haemostasis. The gas analysis was compatible with a partial respiratory insufficiency. The thoracic X-ray revealed a diffuse bilateral interstitial pattern, predominantly in the right lung (Figure 2). The patient was admitted with an initial diagnosis of interstitial pneumopathy, of a possible infectious origin. He was treated with ceftriaxone and doxycycline, with a negative initial evolution, which led to a bronchoscopy and high-resolution computerized axial tomography (HRCT) being requested. Repeated blood and sputum cultures were tested for bacteria, mycobacteria and fungi with negative results. The same result was obtained from the serology

for atypical bacteria. Bronchoalveolar lavage with bronchial aspiration showed a neutrophilic inflammatory component.

The HRCT showed a predominantly peripheral diffuse interstitial disease in relation with possible cryptogenic organizing pneumonia



Figure 1. Biphasic Raynaud's phenomenon in both hands.

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Figure 2. Posteroanterior chest radiograph. A more marked bilateral interstitial pattern can be observed in the right side of the thorax.

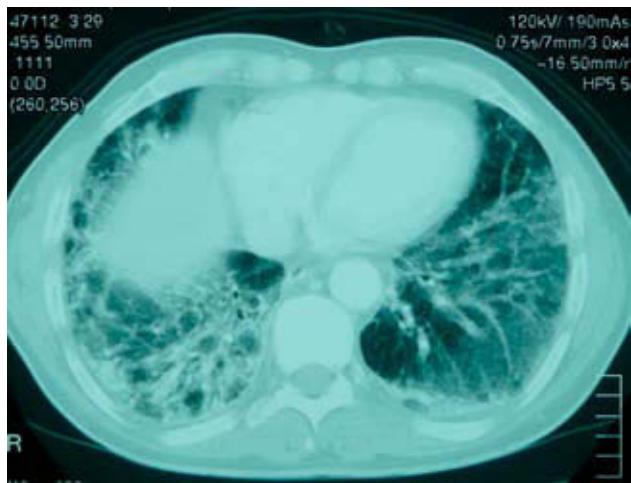


Figure 3. High-resolution computed tomography of the thorax showing opaque glass pattern.

(COP) (Figure 3). The autoantibodies were received and the diagnosis became clear: anti-Jo-1 antibodies, 376 IU/ml [No.: 0-80]; antinuclear antibodies, 1/320; rheumatoid factor, 41.8 IU/ml. Treatment with methylprednisolone (1 mg/kg/day) was initiated. The electromyographic study was normal and the spirometry showed a moderate restrictive pattern with decreased carbon monoxide diffusion capacity.

Diagnosis

Anti-synthetase syndrome (ASS) with cryptogenic organizing pneumonia.

Evolution

The response to glucocorticoids was good, with an improvement in respiratory, muscular and defervescence symptoms within 72 hours. The patient was discharged with 1 mg/kg/day of methylprednisolone and 0.75 mg/kg/day of azathioprine. In subsequent outpatient controls, the patient was asymptomatic, with radiographic resolution.

Discussion

Anti-synthetase syndrome is a condition with an approximate incidence of 1.25-2.50 cases per million inhabitants.¹ It is included within idiopathic inflammatory myopathies with pulmonary involvement. Among the possible hypotheses for its pathogenesis, the most notable are the viral theory (based on immune stimulation produced by the picornavirus family) and the immunological explanation (based on CD8+ T lymphocytes and macrophages).¹

The clinical semiology is fairly homogeneous. The most common manifestation is diffuse interstitial lung disease, which is present in 70% of patients² with different patterns: non-specific interstitial pneumonia, usual interstitial pneumonia and COP, which has a better prognosis.³ Other manifestations of ASS include fever, myositis, polyarthritis, Raynaud's phenomenon and "mechanic's hands", which is a characteristic cutaneous affection of this disease.⁴ In spite of the evident muscular manifestations, the patient did not present an elevation of enzymes specific to muscle damage (described in 5% of cases) and the electromyogram was normal (11% of patients).¹

Antisynthetase antibodies, with a high specificity, are necessary for the diagnosis of this disease. For this reason, when faced with the onset of an interstitial lung disease, or during its evolution, their determination is recommended if a definitive diagnosis cannot be established. This is also the case in patients suspected of unconfirmed connective tissue disease.⁵

The optimal treatment has not been well established. The therapeutic base is treatment with glucocorticoids along with immunosuppressants (azathioprine, methotrexate, cyclophosphamide, etc.). Patients with ASS generally have a better prognosis than those with other interstitial pulmonary diseases, pulmonary involvement being the most important clinical factor.⁶

As a conclusion, we point out the need for a comprehensive view of patients with interstitial pulmonary involvement, since the key for a correct diagnosis may occasionally lie in the extrapulmonary manifestations.

References

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