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ANTI-JO1 ANTI-SYNTHETASE SYNDROME: PATIENTS' PROFILE IN A REFERENCE UNIT

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BACKGROUND

Anti-synthetase syndrome is a rare autoimmune disease of inflammatory myopathies. The triad of the syndrome includes myositis, diffuse pulmonary interstitial disease and anti-Aminoacyl tRNA antibodies, mainly anti-histidyl-tRNA (anti-Jo-1). Nowadays its known that the spectrum of the syndrome is larger. The aim of this study is to describe the profile of patients with anti-Jo-1 syndrome in a reference unit.

MATERIALS AND METHODS

In a clinic of diffuse connective tissue diseases (358 patients with inflammatory myopathies and systemic sclerosis) at a reference unit in Rio de Janeiro, all patients classified as anti-synthetase syndrome associated with anti-Jo- 1 were searched. The demographic picture and the pattern of systemic involvement of the included patients were retrospectively analyzed as age and time of diagnosis, gender, presence of myositis / creatine kinase level (CPK), Raynaud's phenomenon, interstitial lung disease, mechanic's hand cutaneous lesion, arthritis, fever, diffuse Respiratory Function Test pattern of Carbon Dioxide (PFR / DLCO) and computed tomography of the chest (CT Scan), neoplasia and use of immunosuppressants.

RESULTS

We found 14 patients with this syndrome; 13 women and one man; 3 black and 11 white; average age of diagnose 50.2 years. In relation to clinical: 12 had myositis, 7 with CPK increase; All of them had pulmonary interstitial disease confirmed for CT Scan and / or restrictive standard in PFR; all had mechanic's hands. Fever, arthritis and Raynaud occurred in a smaller number of patients. Only one patient have had concomitant neoplasia. All used immunosuppressants as base drug or maintenance (10 azathioprine and 4 others); 8 used cyclophosphamide therapy to treat diffuse interstitial pneumopathy; one patient made use of rituximab for the treatment of arthritis. One patient had died from infection.

CONCLUSION

Anti-synthetase syndrome presented as inflammatory myopathy associated with diffuse interstitial pneumopathy and mechanical hands. The severity of interstitial pneumopathy reinforces attention for screening and monitoring this complication.