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THE CLINICAL MANIFESTATIONS AT THE ONSET OF ANTISYNTHETASE SYNDROME: A CHAMELEON WITH MULTIPLE FACES

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BACKGROUND

Antisynthetase syndrome (ASS) is a rare systemic autoimmune myopathy characterized by fever, myositis, interstitial lung disease, joint involvement, “mechanic’s hands”, Raynaud’s phenomenon, and presence of antisynthetase autoantibodies. Although not scientifically proven, these clinical manifestations don’t occur simultaneously. Therefore, the aim of the study was to analyze chronologically the clinical manifestations at the onset of ASS.

MATERIALS AND METHODS

This is a retrospective, single-center cohort study, from 2000 to 2019, that included 54 patients with defined ASS (Connors et al., 2010). All patients had positivity to antisynthetase autoantibodies (anti-Jo-1, OJ, EJ, PL-7 or PL-12). The following clinical information were analyzed: lung manifestations (dyspnea, associated with changes in the pulmonary parenchyma observed in computed tomography and/or alteration in the pulmonary function test), joint (arthralgia or arthritis, non-erosive); muscle weakness associated with elevated creatine phosphokinase, electroneuromyography with myopathic pattern, muscle biopsy compatible with inflammatory myopathy and/or magnetic resonance with evidence of muscle edema), Raynaud’s phenomenon, “mechanic’s hands”, and fever (without an apparent cause).

RESULTS

The mean age of the patients was 42.4 years, with a predominance of females (77.8%) and white ethnicity (72.2%). The ASS started with fever (42.6%), joint symptoms (42.6%), muscle (38.9%), lung (37.0%), Raynaud’s phenomenon (18.5%) or “mechanic’s hands” (16.7%). The second clinical manifestation was the presence of muscle symptoms (37.0%), Raynaud’s phenomenon (33.3%), lung manifestation (31.5%), joint symptoms (27.8%), fever (25.9%), and “mechanic’s hands” (22.2%). The third clinical manifestation was the presence of “mechanic’s hands” (37.0%), Raynaud’s phenomenon (31.5%), lung manifestation (22.2%), joint symptoms (20.4%), fever (18.5%) or muscle affection (16.7%). Subsequent clinical symptoms emerged at varying times, enabling a variety of diagnoses and therapeutic schemes. In 2 out of 54 cases, joint, muscle and lung manifestations happened simultaneously. The median time between the symptom onset and the complete ASS clinical manifestation was 38.2 months, whereas the duration between the symptom onset and the suspicion or diagnosis of ASS was 48.7 months. Due to incomplete and variety of clinical manifestations, the initial diagnosis confounding diseases were polymyositis (29.6%), dermatomyositis (20.4%), rheumatoid arthritis (14.8%), non-specific interstitial pneumopathy (13.0%) or idiopathic pulmonary interstitial fibrosis (7.4%).

CONCLUSION

Clinical features at the onset of ASS are highly variable. Consequently, confounding factors are marked delays for the final and definitive diagnosis of ASS. Therefore, ASS should be considered as a differential diagnosis in patients with initial symptoms of joint, lung and/or muscle involvements, in addition to fever, “mechanic’s hands” and/or Raynaud’s phenomenon manifestations.