AUTOIMMUNE CONNECTIVE TISSUE DISEASES

AMYOPATHIC DERMATOMYOSITIS WITH ANTI-MDA5 ANTIBODY: TWO CASES AND REVIEW OF THE LITERATURE

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Background: Anti-MDA5 dermatomyositis is an unusual form of dermatomyositis identified by associating cutaneous involvement, aggressive pulmonary involvement and minimal or no muscle involvement. Its particularly affects the Asian and Caucasian population. The treatment remains uncodified until now. We describe two new cases on Moroccan population.

Observation 1: A 52-years-old woman was hospitalized for arthralgia and skin rash. The clinical examination revealed an erythema of the lower eyelids, faces of extensions of the knees and elbows, Gottron's papules on hands and feet, a manicure sign, a minimal muscular weakness and a breathlessness. The electromyogram found a myogenic syndrome, muscle enzymes were low, immunological assessment and autoantibodies of the myositis specific syndrome were negative. She received prednisolone and cyclophosphamide pulse without any clinical or radiological improvements. Two months later, hyperkeratotic papules, necrotic ulcerations with livedoid appearance next to the soles of the feet and painful ulcers within the Gottron's papules appears, in addition to a dyspnea. The chest CT-scan revealed a bilateral pulmonary fibrosis. The anti-MDA5 was positive. The patient received Mycophenolate Mofetil with a good improvement.
Observation 2: A 38-years-old woman followed for amyopathic's dermatomyositis with erythema of the eyelids, Gottron's papules, panniculitis lesions in the arms, without muscle weakness. The muscle enzymes and the muscle biopsy were normal and immunological tests were negative. This cutaneous involvement was associated to a diffuse interstitial pneumonitis. A corticosteroids and hydroxychloroquine were initiated. Six months later, painful ulcerations on palmar folds and Gottron's papules associated to a dyspnea appeared. The anti-MDA5 antibody was positive. The patient was treated by Mycophenolate Mofetil with clinical improvement.

Key message: This entity has been described in Maghreb population including one case in Tunisia and Algeria, but it is also found in the Moroccan population. Mycophenolate mofetil as an immunosuppressant appears to be adapted in this particular dermatomyositis phenotype.