EJR European Journal of Rheumatology

DOI: 10.5152/eurjrheum.2020.20045

Letter to the Editor

Successful low-dose cyclosporine A treatment of a case of juvenile dermatomyositis with interstitial lung disease

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Dear Editor,

The optimal treatment for dermatomyositis (DM) with interstitial lung diseases (ILDs) and antimelanoma differentiation-associated gene 5 (MDA5) antibody positivity is not known (1). An article published in this journal described the efficacy of high-dose intravenous immunoglobulin therapy for such cases (1). For juvenile DM (JDM), ILD with serum anti-MDA5 antibody positivity and high serum Krebs von den Lungen (KL)-6 titer are high risk factors for poor outcome (2). Thus, effective and less cytotoxic management strategies for such cases are desirable (3). Ueki et al. (4) reported that 4 of 5 patients with JDM, ILD, and anti-MDA5 antibody positivity who underwent intensive immunosuppressive therapy showed favorable 4-year outcomes. Recently, we experienced a case of JDM associated with ILD and anti-MDA5 antibody positivity. After early initiation of cyclosporine A (CsA) with prednisolone (PDN), the patient showed a favorable 8-year outcome even with low dose of CsA.

The Japanese female patient was 22 years old. At the age of 14, she had skin erosion, arthralgia, slight fever, and muscle pain at. At presentation, malar rash, inverse Gottron's sign on the back dorsum, skin ulcer on the foot dorsa, and foot periungual erythema were observed, which suggested JDM. The laboratory results were as follows: erythrocyte sedimentation rate, 16 mm/h; leukocytes, 3,530/μL; hemoglobin, 12.8 g/dL; platelets, 154×10^3/μL; aspartate aminotransferase, 138 IU/L; alanine aminotransferase, 144 IU/L; lactate dehydrogenase, 417 U/L; creatine kinase, 72 U/L; aldolase, 12.3 IU/L (normal range, 2.1-6.1 U/L); C-reactive protein, 0.135 mg/dL; KL-6, 808 U/mL (normal range, <250 U/mL); and anti-MDA5 antibody positive (measured at the Department of Rheumatology, Tokai University School of Medicine, Isehara, Japan). Although the serum soluble interleukin-2 receptor level was significantly increased to 1,150 U/mL (normal range, 220-530 U/mL), other specific autoantibodies, such as anti-dsDNA, anti-SS-A, anti-U1RNP, anti-Scl-70, and anti-JO-1 antibodies, were negative. Buttock and thigh magnetic resonance imaging revealed specific findings of inflammatory myositis. Chest computed tomography (CT) revealed cryptogenic organizing pneumonia with random subpleural ground-glass opacity with consolidation patterns. Her forced expiratory volume percentage at 1 second was 73.1%. After the diagnosis of JDM with ILD and anti-MDA5 antibody positivity, immunosuppressive therapy consisting of PDN 30 mg/day (0.7 mg/kg) and CsA 75 mg/day (1.7 mg/kg, once a day, preprandial administration) was promptly initiated. The efficacy and safety of low-dose CsA administration given once daily were reported in some pediatric patients (5, 6). The blood CsA level 2 hours post-dosing was maintained at 400-600 ng/mL. Thereafter, clinical, muscular, and skin signs were dramatically resolved without any respiratory manifestations. The CT findings were gradually improved with decreased serum KL-6 titer. The anti-MDA5 antibody became negative 3 years after immunosuppressive therapy initiation. Thus, the PDN and CsA therapies could be discontinued 3 and 6 years after the first presentation, respectively. The patient is currently in drug-free remission without any sequelae.

Although she did not have any myositis-specific autoantibodies except for anti-MDA5 antibody, her IDL was evident. Thus, prompt initiation of immunosuppressive therapy with PDN and CsA was essential for this patient with risk factors for poor outcome (2-4). Interestingly, though given only once daily, low-dose administration of CsA protocol was effective for long-term complete remission in this patient. However, it has recently been reported that a pediatric male patient had a recurrence of JDM 8 years after remission (7). Thus, close long-term observation is needed for selected patients with JDM. Moreover, the efficacy of low-dose CsA treatment for patients with JDM remains to be determined in a larger number of patients. For patients with pediatric-onset rheumatic diseases, including JDM, more effective and less cytotoxic treatment strategies should be studied. In this context, our current experience may be useful for future reference.

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Cite this article as: Sato R, Tanaka H. Successful low-dose cyclosporine A treatment of a case of juvenile dermatomyositis with interstitial lung disease. Eur J Rheumatol 2020; 7(3): 138-9.

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Submitted: March 26, 2020
Accepted: May 23, 2020

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - H.T.; Design - H.T.; Supervision - H.T.; Materials - R.S.; Data Collection and/or Processing - R.S.; Writing Manuscript - R.S., H.T.

Conflict of Interest: The authors have no conflict of interest to declare.

Financial Disclosure: The authors declared that this study has received no financial support.

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