EDITORIAL

Treatment and Clinical Outcome of Juvenile Dermatomyositis

Autoimmune diseases in pediatric patients are heterogeneous, including systemic lupus erythematosus, juvenile rheumatoid arthritis, and juvenile dermatomyositis (JDM). JDM is a rare autoimmune disease involving multiple body systems and accounts for 85% of idiopathic inflammatory myopathies in children. JDM may lead to death or long-term disability; therefore, investigations into the important prognostic factors for guiding the treatment of JDM are crucial. However, to date, there is no standard guideline for the treatment of JDM. In this issue, Sun et al report a 20-year retrospective analysis of treatment and clinical outcomes to investigate outcomes of JDM in Taiwan and to identify predictors or factors associated with outcomes. The study gives us new insight into JDM.

JDM is an immune-mediated inflammatory disease involving the microvasculature of skin and muscle. The most common initial presentations are Gottron’s papules and muscle weakness. The clinical features are associated with systemic vasculopathy and are critical to the diagnosis. However, recent studies have suggested that magnetic resonance imaging is also useful in diagnosing JDM and that it may be used as a disease assessment tool. A recent clinical study showed that peripheral blood regulator T cells (Tregs) of active JDM patients were less capable of suppressing effector T-cell activation in vitro compared with Tregs of JDM in clinical remission. Therefore, the functional impairment of Tregs plays an important role in JDM inflammation in a proportion of patients with active disease. JDM is a heterogeneous disease and autoantibodies may be potentially useful biomarkers to classify patients into homogeneous subgroups and inform on disease prognosis. Autoantibody status and age at disease onset have also been shown to influence the clinical phenotype and overall prognosis in JDM. In JDM, the development of calcinosis has been reported to be associated with delayed diagnosis, a chronic disease course, and inadequately treated disease. Sun et al reported important prognostic factors of JDM in children for predicting clinical outcomes of such patients. Their findings suggest that calcinosis, skin ulcerations, and muscle weakness may develop several years later during follow-up, because aggressive treatment is suggested by some clinicians. The prognosis of amyopathic DM, unlike that in adult groups with the increased risks of interstitial lung disease and malignancy, has a generally good prognosis among pediatric patients.

There is no standard treatment protocol for JDM to date. Since the introduction of corticosteroids to treat JDM, significant improvements in clinical and functional prognosis have been achieved, and therefore, they remain the mainstay of treatment. However, systemic corticosteroids are associated with significant side effects after long-term use. Either immunosuppressive agents or intravenous immunoglobulin is a supplemental therapy for JDM patients with poor treatment responses. Biologic drugs, which are synthesized within a biologic system, are designed to target specific molecules involved in cytokine signaling or cell–cell interactions. The major targets of these biologic drugs are cytokines, immune cells, and some costimulation molecules. The three classic inflammatory cytokines, namely, tumor necrosis factor-α (TNF-α), interleukin-6 (IL-6), and IL-1, are the major targets of biologic therapies. The major immune cells targeted are B cells and T cells. In the past 2 decades, biologic drugs have seen a revolution in the range of effective treatments for pediatric rheumatic diseases, particularly juvenile idiopathic arthritis. As a result, minimal long-term disease-associated damage is increasingly becoming achievable. In the study by Sun et al, four patients received etanercept, an anti-TNF agent. Two of these four patients achieved complete clinical response after 15 months and 18 months of treatment, respectively (including etanercept and corticosteroid); however, one of these patients later developed disease flares during follow-up and required corticosteroid control. The other two patients showed poor response to anti-TNF agents and remain under multiple medications. Treatment with anti-TNF agents may be effective for adults with refractory DM or polymyositis, but these were not promising in the current study of JDM.

This article by Sun et al covers many important aspects, including prognostic factors and current treatment options for JDM, which will surely provide a better understanding about the role of various predictive factors in JDM. However, their
case numbers are still low. Large population and long-term studies on JDM aimed at determining the prognostic factors and treatment options, especially in severe cases, are still needed.

Conflicts of interest

The author declares no conflicts of interest.

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