Video abstract transcript

Long-Term Prognosis of Antimelanoma Differentiation–Associated Gene 5–Positive Dermatomyositis With Interstitial Lung Disease

doi.org/10.3899/jrheum.2023-0371

Slide 1:

Hello, I'm Tsuneo Sasai, from Kyoto University. It' s honor to present our study at this video abstract. I would like to share with you the key highlights from our paper about long term prognosis of antimelanoma differentiation associated gene 5 positive dermatomyositis with interstitial lung disease.

Slide 2:

Interstitial lung disease (ILD) accompanied by anti-MDA5-positive dermatomyositis often progresses rapidly and takes poor prognostic outcomes. We previously reported the efficacy of combination therapy with high-dose glucocorticoid (GC), calcineurin inhibitor (CNI) and intravenous cyclophosphamide (IVCY) in the multicenter clinical trial. As you can see this figure, survival rate of short time had been improved by this treatment regimen. However, there is no evidence of their management during remission maintenance phase. Therefore, we evaluated their long-term outcomes and the effect of induction therapy on remission-maintenance phase.

Slide 3:

This study includes two researches. First, we examined the long-term prognosis of patients enrolled in a multicenter prospective study which we previously examined the efficacy of

induction therapy. Specifically, we analyzed the relapse-free rate of ILD and the withdrawal rates from steroids and other drugs after achieving remission. The remission criteria was defined as survival for more than 6 months without recurrence of ILD after initial treatment. Second research is a comparison with conventional therapy. After adding patients achieving remission at our institute, we divided these patients into two groups by their content of induction therapy. First group is triple therapy group: patients treated glucocorticoid, calcineurin inhibitor and cyclophosphamide. Second group is conventional therapy group: patients treated either only glucocorticoid or glucocorticoid and calcineurin inhibitor. Between both groups, we compared the relapse-free rate of ILD, and the drug withdrawal rates of immunosuppressants.

Slide 4:

Let me show the results. First, I will show you the long-term prognostic data of the multicenter study.

Slide 5:

The recurrence-free rate of ILD at 5 years from the initial treatment was 100%. The withdrawal rates of CNI and GC were 70% and 53%, respectively. The withdrawal rate of all immunosuppressants, I mean both GC and CNI, was 38%.

Slide 6:

These are the changes of clinical parameters from the initial treatment to the maintenance phase. The ferritin and KL-6 levels, which were considered poor prognostic factors, were significantly lower in the maintenance period than at the beginning, and the median values remained within the standard range. You can see this figure; the median value has decreased to the standard value almost one year after the initial treatment.

Anti-MDA5 titers also remained within standard range in the remission phase. The median antibody titer had decreased to within the standard value in one year.

Pulmonary function test significantly improved in the remission phase compared to the induction phase.

Slide 7:

Next, I will show you the comparative data on long-term prognosis by categorizing the content of initial induction therapy. I call the triple therapy group as Group T, and the conventional therapy group as Group C.

Slide 8:

This is the patient background. There was no differences between both groups in terms of age, gender, and serum data such as CRP and ferritin. The predicted diffusing capacity for carbon monoxide was significantly lower in Group C. This is because some patients in group C were diagnosed older time and the concept of ILD with anti-MDA5 were not familiar. As a result, some patients in group C might had been delayed diagnosis and worsened respiratory function. With regard to treatment, the initial dose of prednisolone was significantly higher in Group T.

Slide 9:

This is the results of the prognosis. The recurrence-free rate of ILD in group T was 90% and significantly higher than that in group C. The withdrawal rates of CNI and GC in group T were also significantly higher than those in group C, which were 79% and 43%, respectively. About the withdrawal rate of both CNI and GC, there was no

significant difference between both groups. However, some patients in only group T achieved complete withdrawing from both CNI and GC.

Slide 10:

This is the change of prednisolone-dose. As mentioned earlier in the patient background, prednisolone dose at the start of treatment was significantly higher in Group T. However, at 3 years, the median prednisolone dose in Group T was 3 mg, which was significantly lower than that in group C.

Slide 11:

Based on these results, we turn to the conclusions. Triple combination therapy at induction phase brought not only improvement of survival rate during the induction phase, but also retention of high recurrence-free rate during the maintenance phase. Moreover, some cases withdrew immunosuppressants completely. Compared with conventional therapy, triple combination therapy brought higher recurrence-free rate, higher withdrawal rate of immunosuppressants, and lower maintenance dose of prednisolone. Induction therapy with triple combination therapy is seemed to be beneficial during not only induction phase, but also in the long term. This is the end of my presentation. Thank you for your listening.

Sasai et al. J Rheumatol 2023. doi.org/10.3899/jrheum.2023-0371