Objectives: We evaluated the effect of plasma exchange (PE) on survival in patients with refractory RP-ILD who were positive for anti-MDA5 antibodies.

Methods: Among 167 patients newly diagnosed with PM/DM, clinically amyopathic DM, or cancer associated myositis from 2008 to 2019 at our hospital, 12 were diagnosed with refractory RP-ILD and were positive for anti-MDA5 antibodies. PE was used as an adjunct to standard therapy and consisted of fresh frozen plasma as replacement solution. The primary outcome was non-disease-specific mortality, and anti-MDA5 antibody titres were measured by ELISA using the MESCUP anti-MDA5 test in 155 patients whose serum was frozen and stored at the time of diagnosis.

Results: Anti-MDA5 antibodies were detected in 35 patients, of whom 26 were diagnosed with RP-ILD and 11 were refractory to intensive immunosuppressive therapy. Seven patients received PE (PE group) and four did not (non-PE group). The 1-year survival rate of the PE group was higher than that of the non-PE group (100% and 25%, respectively, P = 0.011). Regarding adverse events associated with PE, two patients had anaphylactic shock, one had high fever due to RP-ILD, and one had a catheter infection. All adverse events resolved with appropriate treatment.

Conclusion: We evaluated the association between 1-year survival rate and PE for refractory RP-ILD in patients positive for anti-MDA5 antibodies. Intensive immunosuppressive therapy improved the survival rate in RP-ILD patients with anti-MDA5 antibodies, but 20-30% of cases were still fatal. PE could be administered to patients with active infectious disease who were immunocompromised by intensive immunosuppressive therapy. PE may be considered in refractory RP-ILD patients positive for anti-MDA5 antibodies.

References:

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