

Conclusion: Anti-FHL1 autoantibodies were detected in 20.5% of IIM patients. In IBM and IMNM, the presence of anti-FHL1-autoantibodies was associated with a severe myopathy as suggested by presence of dysphagia and muscle atrophy.

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POS0884 THE ENHANCED LIVER FIBROSIS (ELF) SCORE AS A BIOMARKER OF SKIN FIBROSIS IN SYSTEMIC SCLEROSIS

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Background: Serum fibrotic markers for systemic sclerosis (SSc) remain limited. The Enhanced Liver Fibrosis (ELF) score, originally derived and validated in patients with chronic liver disease, is an algorithm combining 3 serum markers, known as procollagen type III amino terminal propeptide (PIIINP), tissue inhibitor of metalloproteinases 1 (TIMP-1), and hyaluronic acid (HA). The combined score was proved to be superior to the single components in reflecting the severity of liver fibrosis. However, the performance of ELF score and its components has not been fully validated in SSc.

Objectives: To investigate PIIINP, TIMP-1, HA, and the combined algorithm ELF score as fibrotic markers for SSc skin involvement.

Methods: Eighty SSc patients (44 dcSSc and 36 lcSSc), fulfilling the 2013 ACR/EULAR criteria with the absence of chronic liver diseases, were enrolled. Eighty age- and sex- matched healthy controls were also included. Serum PIIINP and HA levels were quantified by chemiluminescence immunoassay. Serum TIMP-1 levels were determined by enzyme-linked immunosorbent assay. The ELF score was calculated using the formula $ELF\ score = 2.494 + 0.846 * \ln(HA) + 0.735 * \ln(PIIINP) + 0.391 * \ln(TIMP-1)$. Results were correlated with clinical profiles including modified Rodnan skin score (mRSS) and interstitial lung disease (ILD).

Results: Compared with healthy controls, patients with SSc showed significantly elevated serum PIIINP (11.2 ± 4.8 vs. $5.73 \pm 1.4 \mu\text{g/L}$, $p < 0.001$), TIMP-1 (123.7 ± 78.6 vs. $67.8 \pm 26.5 \text{ ng/ml}$, $p < 0.001$), and ELF score (10.5 ± 0.9 vs. 9.7 ± 0.4 , $p < 0.001$). Even higher levels of PIIINP, TIMP-1, and ELF score were observed in dcSSc patients, compared with lcSSc patients ($p < 0.001$, $p = 0.024$, $p = 0.003$, respectively). No significant difference was found in the levels of serum HA between patients and controls. Strong correlations were observed between mRSS and ELF score ($r = 0.54$, $p < 0.001$), and between mRSS and PIIINP ($r = 0.62$, $p < 0.001$), whereas only weak correlations could be observed between mRSS and TIMP-1 ($r = 0.28$, $p = 0.02$), and between mRSS and HA ($r = 0.26$, $p = 0.03$). When stratified by ELF score, using cutoffs proposed for liver fibrosis and cirrhosis, SSc patients with $ELF < 9.8$ showed the lowest mRSS on average, while patients with $ELF > 11.3$ showed the highest ($p < 0.001$). When stratified by serum PIIINP levels, using the 25th and 75th percentiles, SSc patients with serum PIIINP levels $< 7.8 \mu\text{g/L}$ showed the lowest mRSS on average, while patients with $PIIINP > 14.0 \mu\text{g/L}$ showed the highest ($p < 0.001$). Neither the ELF score nor its components showed significant difference between patients with and without ILD.

Conclusion: The ELF score could be used for reflecting the severity of overall skin involvement in SSc, and serum PIIINP also increased in parallel with the increase of mRSS. Longitudinal prospective studies exploring ELF score or serum PIIINP as fibrotic markers and outcome measures of SSc are warranted.

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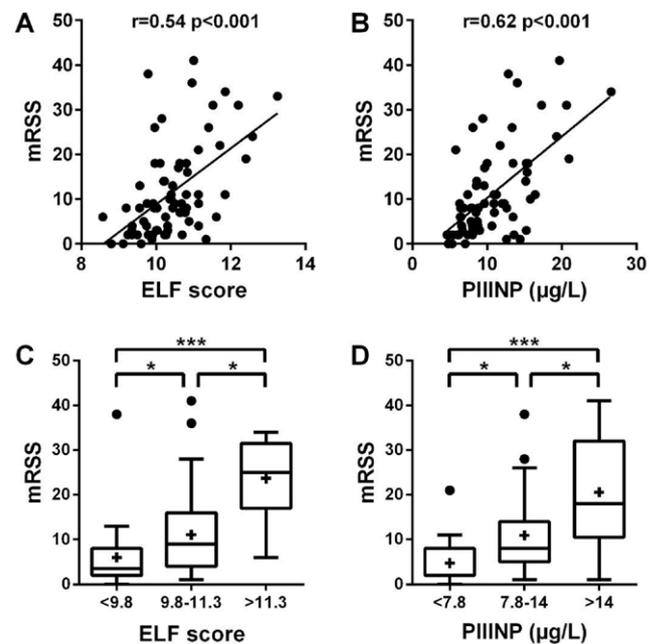


Figure 1. Correlations of mRSS with ELF score (A) and serum PIIINP (B) and distribution of mRSS among different ELF (C) and PIIINP (D) ranges.

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POS0885 HIGH INCIDENCE AND MORTALITY OF PNEUMOCYSTIS JIROVECI INFECTION IN ANTI-MDA5-ANTIBODY POSITIVE DERMATOMYOSITIS: EXPERIENCE FROM A SINGLE CENTER

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Background: Idiopathic inflammatory myopathies (IIM) was associated with a significantly higher risk of opportunistic infections that including Pneumocystis jiroveci pneumonia (PJP) which is potentially fatal opportunistic infection. However, no prior studies have evaluated the PJP infection in subtypes of IIM.

Objectives: To investigate the incidence rate and mortality rate of PJP infection in subgroups of IIM patients according to myopathy specific antibodies.

Methods: In the first part, we reviewed 463 consecutive patients with IIM retrospectively to analyze incidence of PJP infection. In the next part, we enrolled 30 consecutive PJP infection patients with any rheumatic disease was to identify the mortality rate and risk factors. Kaplan-Meier curve with log rank test was used to access differences in survival. Univariate and multivariate analyses were performed to identify prognostic factors using Cox regression.

Results: We found that 12 (7.5%) PJP cases occurred in 160 anti-MDA5-ab-positive DM patients, while only two (0.7%) PJP cases were found in 303 anti-MDA5-ab-negative DM/PM patients ($P < 0.05$). PJP infection typically happened in the first two months of the treatment for anti-MDA5-ab-positive DM patients who have a significant decrease in the CD4+ T cell counts and Lymphocyte counts ($P < 0.05$). Only two (16.7%) anti-MDA5-ab-positive DM patients recover from PJP, with lethally higher mortality than those PJP infection with other rheumatic diseases (83.3% vs. 38.9%, $P < 0.05$). We found no association between the time to anti-PJP treatment and treatment outcomes in anti-MDA5-ab-positive DM; yet we confirmed in PJP infection with other rheumatic diseases that anti-PJP treatment within 6 days crucially increased the survival ($P < 0.05$).

Conclusion: PJP infection has alarming high incidence and mortality in anti-MDA5-ab-positive DM patients. Unlike PJP infection with other rheumatic diseases, timely treatment for PJP doesn't improve the prognosis of this particular subtype. Therefore, the necessity of further study of PJP prophylaxis treatment in anti-MDA5-ab-positive DM patients is verified.