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FRI0338 ANTI-MDA5 IDIOPATHIC INFLAMMATORY MYOSITIS (IIM) CONFERS POOR PROGNOSIS BUT NEGATIVE MYOSITIS SPECIFIC ANTIBODY (MSA) IS NOT BENIGN EITHER

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Background: MSA test is useful to diagnose IIM and subcategorize patients by disease phenotypes.

Objectives: The study aims to evaluate the survival of IIM patients of different MSA patterns.

Methods: An IIM registry had been set up in a tertiary referral centre since 2014 by recruiting prevalent and incident cases. Patients were followed-up prospectively. This study included patients fulfilling the 2017 EULAR/ACR classification criteria for IIM¹ and excluded those aged < 18 at disease onset. Immunoblot EUROLINE autoimmune inflammatory myopathies 16 antigens strip (EUROIMMUNE AG, Lubeck, Germany) was used. Information including baseline demographic data, disease manifestations, MSA results, co-existing malignancy, duration of survival and causes of death were collected. IIM patients were divided into seven groups, which included 1) anti-aminoacyl tRNA synthetase (ARS) 2) anti-MDA5, 3) anti-TIF1γ/anti-NXP2, 4) double positive MSA, 5) other MSAs, 6) negative MSA/MAA (myositis associated antibodies) and 7) positive MAA only. Survival probabilities were compared among different MSA groups by using the Kaplan-Meier method and log-rank test. A two-tailed probability value (p) < 0.05 was considered significant. The study was approved by Kowloon Central Cluster Ethic Committee (ref.: KC/KE-17-0177/ER-3)

Results: Among 112 IIM patients, 79 (70.5%) were female, and the median age of onset was 55 (18-90) years old; 63.4% were dermatomyositis (DM), 17.9% polymyositis (PM) and 18.8% clinically amyotrophic DM (CADM). Co-existing interstitial lung disease (ILD) was common and found in 65 (58%) patients; 16 (14.3%) had rapidly progressive interstitial lung disease (RPILD), and 16 (14.3%) died within the observed period. Overall, the commonest cause of death was RPILD, followed by infection and malignancy. While anti-MDA5 was strongly associated with RPILD (odds ratio = 33.0 [95% CI: 7.2-151.8], p<0.001), anti-MDA5 group had the worst survival, with 1-year and 5-year survival both at 43%, compared to above 80% in all other groups (log-rank test p<0.001) (Table 1). There were nine patients with double positive MSA and 28 had negative MSA/MAA. Analysis between MSA sub-groups found that the double positive MSA, MAA only and other MSAs group had no mortality during study period while the negative MSA/MAA group had the second highest mortality following the anti-MDA5 group (Figure 1). Infection and malignancy were the two major causes of death in the MSA/MAA negative group.

Conclusion: Anti-MDA5 associated RPILD was the leading cause of mortality in IIM. However, those tested negative for both MSA and MAA by current immunoblot technique also had guarded prognosis related to the risk of infection and malignancy.

Figure 1. Kaplan-Meier survival of IIM

Figure 1. Kaplan-Meier survival of IIM

Table 1. Survival rates in different MSA groups

Survival	1 year	5 years
Anti-ARS	100%	92%
Anti-MDA5	43%	43%
Double positive	100%	100%
Negative MSA/MAA	89%	82%
Only positive MAA	100%	100%
Anti-TIF1γ/anti-NXP2	100%	88%
Other MSAs	100%	100%

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FRI0339 CLINICAL CHARACTERISTICS OF ANTI-RO52A AND ANTI-RO52B ANTIBODIES IN DERMATOMYOSITIS/ POLYMYOSITIS

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Background: The anti-Ro52 antibody, found in numerous systemic autoimmune conditions, is one of the most common autoantibodies in inflammatory myositis. It is known to be associated with interstitial lung disease (ILD) and to coexist with anti-Jo-1¹. There are two spliced forms: Ro52α and Ro52β. Ro52β was originally reported in 1995 as a spliced form of Ro52α in the human heart². Ro52β antigen cDNA has a defective exon 4, which encodes part of an autoepitope hotspot².

Objectives: We investigated the clinical and laboratory characteristics of anti-Ro52α and anti-Ro52β. We also analyzed the characteristics of anti-Ro52α/β within each coexisting inflammatory myositis specific autoantibody (MSA) group to reduce the influences of coexisting MSA characteristics.

Methods: Among 229 dermatomyositis (DM) and polymyositis (PM) patients, 167 patients (DM 152, PM 10, juvenile DM 5) fulfilled the criteria of Bohan and Peter, and 62 clinical amyopathic DM cases fulfilled the criteria of Sontheimer. Anti-Ro52α and anti-Ro52β antibodies were detected by ELISA.

Results: 46 of the 229 patients were anti-Ro52α-positive (20%). ILD was a frequent complication in the anti-Ro52α-positive patients (35/42: 76%) (P=0.0016), and anti-Ro52α was highly positive in the anti-aminoacyl tRNA synthetase (ARS) antibody-positive group (19/32: 60%) (P<0.0001) (Table 1). However, the ILD frequencies of anti-Ro52α-positive and anti-Ro52α-negative patients within each group of coexisting MSA (anti-ARS, anti-TIF, anti-MDA5, etc.) were almost same (P=1). The anti-Ro52α-positive rates were similar to the positive rates of anti-Jo1 (61%) and other anti-ARSS (52%) (P=0.7).

Of the 46 anti-Ro52α-positive patients, 26 patients were anti-Ro52β-positive. No patient was only anti-Ro52β positive. The anti-Ro52α-positive and anti-Ro52β-positive groups were older than the anti-Ro52α-only-positive group (P=0.009), and the average of maximum creatine kinase was higher in both of the positive groups (P=0.065). The 6 patients without coexisting MSA were all both anti-Ro52α-positive and anti-Ro52β-positive (P=0.03) (Table 2).

Conclusion: Anti-Ro52 is highly positive when anti-ARS antibodies are present, but it is not anti-Jo-1-specific. Anti-Ro52 positivity is not associated with elevated risk of ILD in any of the MSA-positive groups. The anti-Ro52β antibody might be an indicator of myositis.

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