The incidence rate of atherosclerotic CVE was 16.3/1000 patient-years (95% CI: 8.8-30.3), twice as high as expected by QRISK2. Analysis showed statistical difference with males (RR=6.49, $p=0.002$) and white patients (RR=5.04, $p=0.02$) although the statistical significance for white ethnicity was lost after adjusting for age and gender. Statistical analysis showed association between disease duration and higher incidence of atherosclerotic CVE, with a rate reduction of 0.7 per year from disease onset ($p=0.01$) after adjusting for age and gender.

Fig. 1. Atherosclerotic risk by disease duration

Fig. 2. Kaplan-Meier survival plot to atherosclerotic event

Conclusion: Our results suggest that atherosclerotic cardiovascular risk in inflammatory myopathies is underestimated, particularly early on the disease. Based on our results we would recommend a low threshold for primary prevention of atherosclerosis including the use of statins particularly for the first year after diagnosis.

Disclosure of Interests: None declared

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THU0336 IMMUNE CHECKPOINT INHIBITOR-RELATED MYOSITIS: A RETROSPECTIVE COHORT STUDY

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Background: Myositis is a rare immune checkpoint inhibitor (ICI)-related adverse event frequently associated with myasthenia gravis (MG) and myocarditis (MC) leading to mortality rates up to 52%.¹

Objectives: To characterize the presentation, course and outcomes of patients with ICI-related myositis alone or with overlap syndrome (myositis with MG or MC or both).

Methods: We retrospectively identified a cohort of patients treated with ICI at MD Anderson Cancer Center between 2016 and 2019. Suspected myositis was

identified using International Classification of Disease version 10 codes and confirmed by electronic medical record review of muscle enzymes, pathology, and other tests, when available. Patients with myositis alone or with overlap syndrome were compared using Fischer's exact tests and t tests.

Results: During the study period 8,636 patients received ICI, of which 31 (0.36%) were diagnosed with myositis: 14 (45%) with myositis alone and 17 (55%) with overlap (MG in 5, MC in 4, MG and MC in 8). Twenty patients received programmed death-1 (PD-1) or programmed death-ligand-1 (PDL-1) inhibitors, and 10 received combination PD-1/PDL-1 inhibitor with a cytotoxic T-lymphocyte associated antigen 4 (CTLA-4) inhibitor. One patient received single agent CTLA-4 inhibitor (excluded from pooled data). For the entire cohort the median age at diagnosis was 69 years (range: 40-95 years); the most common presenting symptoms were fatigue in 27 (90%) patients, weakness in 24 (80%), and myalgia in 23 (77%); median CK was 2,236 U/L (range: 23-19,794 U/L). For treatment, 22 of 30 (73%) patients received at least one therapy in addition to steroids: plasmapheresis in 15 (50%) patients, intravenous immune globulin (IVIG) in 12 (40%), biologics in 9 (30%) (rituximab in 6, infliximab in 5, tocilizumab in 3), tacrolimus in 6 (20%), and mycophenolate mofetil in 4 (13%). Median length of exposure to steroids was 47 days (range: 1-250 days). Five (17%) patients were rechallenged with ICI after myositis resolution (3 with myositis alone, 2 with overlap), of which 1 (20%) patient experienced a myositis flare. Twenty-five (83%) patients were not rechallenged on ICI and 3 (12%) of those patients had a flare. Differences between patients with myositis alone compared to those with overlap are shown in Table 1. Patients with overlap more often received a second therapy, specifically plasmapheresis and IVIG, had longer hospitalizations and greater symptom burden at discharge. Overall death between groups was similar; however death attributed to the adverse event occurred only in those with overlap.

Table 1. Myositis alone vs. Overlap

	Myositis alone (N=13)	Overlap (N=17)	P value
	N(%) / median days [range]		
Time to symptom onset	42 [10-161]	22 [9-149]	0.234
Initial steroid dose (mg/kg day)	1.7	1.8	0.187
Second therapy	7 (54)	15 (88)	0.049
Plasmapheresis	3 (23)	12 (71)	0.025
IVIG	1 (8)	11 (65)	0.002
Outcomes			
Hospitalization length	5 [2-50]	24 [7-92]	0.019
Respiratory failure	0 (0)	13 (76)	
Symptoms at discharge			0.047
Improved	8 (62)	6 (35)	
Resolved	3 (23)	1 (6)	
Death			
Overall	8 (62)	12 (71)	0.706
Adverse event	0 (0)	7 (41)	

Conclusion: Our results represent the largest cohort of ICI-related myositis to date. Patients with overlap syndrome are treated more aggressively and have worse outcomes than those with myositis alone. Prospective studies are warranted to determine risk factors for developing myositis or overlap syndrome and to determine optimal treatment.

References:

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THU0337 THE EFFECTIVENESS OF PLASMA EXCHANGE THERAPY FOR ANTI-MDA5 ANTIBODY-POSITIVE REFRACTORY INTERSTITIAL LUNG DISEASE

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Background: This is an extended report of our study [1]. Anti-melanoma differentiation-associated gene 5 (MDA5) antibodies, which are closely related to interstitial lung disease (ILD) with or without rapid progression, are a type of myositis-specific autoantibody. Since rapid progressive-ILD (RP-ILD) with anti-MDA5 antibodies is refractory and fatal, intensive immunosuppressive therapy with combination calcineurin inhibitor, and intravenous pulse cyclophosphamide was developed, and was shown to improve patient survival and prognosis [2]. However, 20–30% of cases were still fatal, and several additional