9. Systemic sclerosis, myositis and related syndromes - etiology, pathogenesis and animal models.

AB0149

PERIPHERAL T HELPER SUBSET PROFILING DIFFERS IN VARIOUS SUBSETS OF IDIOPATHIC INFLAMMATORY MYOSITIS

A. Anuja¹, H. Singh¹, D. Misra¹, V. Agarwal¹, L. Gupta¹, ¹Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India

Background: There is dearth of biomarkers in Idiopathic Inflammatory Myositis(IIM) to identify ongoing inflammation in the muscle and distinguish it from inactivity or damage.

Objectives: Since myositis is autoantibody mediated and tertiary lymphoid organogenesis (TLO) reported in the diseased muscles, we investigated peripheral blood T helper subset profiling as a reflection of ongoing muscle inflammation.

Methods: Twenty-six patients of IIM (ACR EULAR criteria) were compared with 15 healthy controls (HC) and 21 patients with sarcoidosis (Table 1). Peripheral blood mononuclear cells were stained with combinations of antibodies to identify Th1, Th17, Th17.1 and Treg cells after stimulation assays (BD Biosciences). Myositis Specific and Associated autoantibodies were tested by the line immunoassay (Euroimmune, Germany).

Table 1. Baseline characteristics of patients with inflammatory myositis

Characteristics	Demographic details (n, % or median, IQR)	Healthy Control (median, IQR)
Age	37±25.25	26.0±32
Gender(M:F)	5 vs. 21	12 vs. 3
Diagnosis		
PM	3	
DM	15	
OM	4	
ASS	4	
Disease course		
Monocyclic	5	
Polycyclic	7	
Chronic continuous	1	
Undefined	13	
Clinical Profile		
Myositis	4 (15.3%)	
ILD	5 (19.23%)	
Rash	3 (11.53%)	
Arthritis	6 (23.07%)	
Other	16 (23.69%)	
Disease duration (years)	1.3 ± 6.91	
Disease activity		
Active	12 [PM(1), OM(1), ASS(4), DM(5)]	
Inactive	14 [PM(2), OM(2), ASS(0), DM(8)]	
Antinuclear Antibodies		
Positive		
Nuclear		
Speckled	9 (34.61%)	
Homogenous	4 (15.38%)	
Nucleolar	1 (3.8%)	
Other	5 (19.23%)	
Cytoplasmic	3 (11.53%)	
Negative	4 (15.38%)	
Myositis Specific Antibodies		
Positive		
ARS	2(7.6%)	
Mi-2	3(11.53%)	
SAE-1	2(7.6%)	
NXP2	2(7.6%)	
MDA5	0	
MAA		
Ku	1(3.8%)	
dsDNA	0	
U1RNP	0	
Ro52	4(15.38%)	
Negative	12(46.15%)	

Results: All T helper subsets were higher in myositis as compared with healthy controls (figure 1A a-d). Between various IIM subsets, polymyositis had higher Th1 and Treg cells (Figure 1B b, c) while Th17 and Th17.1 cells(c) were higher in Overlap Myositis (Figure 1B a, d) as compared with healthy controls. Patients with sarcoidosis had similar subset profiling as myositis. (Figure 5a-f)

Figure 1 A. Representative plot depicting all T helper subsets quantified were higher in myositis as compared with healthy controls1B: Representative plot comparing %T cell subsets in various subsets of myositis with healthy controls showing that % Th1 cells (a) and Tregs (d) are highest in Polymyositis than controls while % Th17 (b) and % Th17.1 cells (c) are higher in Overlap Myositis

Patients who were had either arthritis or were positive for myositis specific autoantibodies had higher Th17.1 cells (Figure 3 a(iii) & b(iii)) than those negative for MSA. There was no difference in T cell profile between the various autoantibody subsets (Figure 6a-d).

There was no difference in subsets between active and inactive disease although active disease had lower Th1/Treg, Th17/Treg and Th17.1/Treg ratios.

Conclusion: T Helper cell subsets are distinct from HC but similar to sarcoidosis patients. However, they differ in various subsets of myositis, suggesting different pathogenic mechanisms are operative. Autoantibody positivity is associated with elevated Th17.1 population suggesting plasticity in TLO which needs to be explored further. However, T cell profiling cannot distinguish active from inactive disease limited predictive potential as a biomarker.

Figure 2 A. Comparisons between various phenotypic subsets suggest patients positive for MSA had higher Th17.1 cells (Figure 2A a(iii)) than those negative for MSA. Similarly, patients with arthritis had higher Th17.1 cells(Figure 2A b (iii)). 2B Representative dot plot of T cell subsets ratio (Th1, Th17 & Th17.1) with Treg subsets (a) Th17/Treg ratio observed higher in lower cells in active as compared with inactive disease. 2C Representative dot plot T cell subsets in Sarcoidosis and myositis2D Representative dot plot comparing percentage of T cell subsets in various antibody subsets of myositis

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ADAPTOR PROTEINS ACTIVATE FIBROBLASTS IN SYSTEMIC SCLEROSIS

Y. Ariza^{1,2}, S. Weber¹, N. Neise¹, A. Kreuter³, G. Schett¹, J. Distler¹, A. Ramming¹, ¹Friedrich-Alexander-University (FAU) Erlangen-Nürnberg and Universitätsklinikum Erlangen, Department of Internal Medicine 3