Distinct clinicopathologic and radiological manifestations of skin, lung, and muscle diseases in dermatomyositis patients positive for anti-aminoacyl tRNA synthetase antibodies

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[Background]

- Patients with antisynthetase syndrome (ASS), which is diagnosed by the presence of an anti-aminoacyl tRNA synthetase (ARS) antibody, may constitute a unique subtype of dermatomyositis patients.
- So far, eight anti-ARS antibodies (Jo-1, PL-7, PL-12, EJ, OJ, KS, Zo, and Ha) have been found.

[Aim of the present study]

- To examine the autoantibody profiles of patients with dermatomyositis, and compare the results with clinical manifestations between the ASS and non-ASS group.

[Patients]

- Fifty-nine patients with DM who visited our clinics from 2008 to 2016.
- Fifty-two patients met the criteria for the classification tree model by the International Myositis Classification Criteria Project (IMCCP)*.
- The remaining 7 patients include:
 - 4 “possible” cases of Bohan and Peter’s criteria,
 - 1 “probable” case,
 - 2 cases of amyopathic DM with ILD.

[Methods]

- ELISA kit: MESACUP anti-ARS test (Jo-1, PL-7, PL-12, EJ, and KS)
- All sera were examined for antibodies to TIF1- γ , MDA5 and Mi-2.
- Evaluation of cutaneous and systemic symptoms, including radiological images of interstitial lung disease (ILD) and myositis, in a blind fashion.
- The results were analyzed in relation to serum levels of KL-6, CK, ALD, and ALD/CK ratios between the ASS and non-ASS groups.

[Result 1] Antibody profile of the ASS and non-ASS group

[Result 2] Cutaneous and systemic symptoms

Clinical symptoms and signs	ASS	non-ASS	p value
Age of onset, y	51.7 $\pm$ 14.7	59.7 $\pm$ 14.5	0.043
Sex ratio, M : F	7:13	13:26	1.00
Heliotrope rash	5/20 (25%)	20/39 (51%)	0.094
Gotttron's papule	12/20 (60%)	33/39 (85%)	0.053
Erythema on the extensor surface of extremities	15/20 (75%)	27/39 (69%)	0.77
Mechanic's hand	14/20 (70%)	13/39 (33%)	0.012
Shawl-sign/V-sign	3/20 (15%)	20/39 (51%)	0.010
Raynaud phenomenon	3/20 (15%)	4/39 (10%)	0.68
Periungal erythema	11/20 (55%)	28/39 (72%)	0.25
Poikiloderma	0/20 (0%)	7/39 (18%)	0.08
Scratch dermatitis	3/20 (15%)	11/39 (28%)	0.34
Proximal muscle weakness	10/20 (50%)	26/39 (67%)	0.26
Muscle grasping and spontaneous pain	10/20 (50%)	23/39 (59%)	0.59
Elevated serum ALD level	12/12 (100%)	18/26 (69%)	0.0022
Arthralgia/arthritis	13/20 (65%)	9/39 (23%)	0.0037
Elevated serum CRP level	16/20 (80%)	15/39 (38%)	0.0029
Fever of over 38°C	8/16 (50%)	6/39 (14%)	0.019
Incidence of interstitial pneumonia	19/20 (95%)	17/39 (44%)	0.0001
Incidence of malignancy	4/20 (20%)	13/39 (33%)	0.37
History of collagen disease in relatives	6/20 (30%)	0/39 (0%)	0.0009

[Result 3] Incidence of ILD and CT image

[Result 4] Myositis pattern and serum ALD levels

group (cases with significant MRI findings/all MRI performed cases)	myositis or myositis dominance	fasciitis or fasciitis dominance	comparable
ASS (n=12/14)	5	6	1
non-ASS (n=23/31)	20	2	1

[Conclusion]

Patients with ASS have a higher incidence of mechanic's hands, shawl sign, systemic inflammation, ILD, and inflammatory myopathy associated with fasciitis, and elevated serum levels of ALD.

[References]

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