ABSTRACT NUMBER: 2314

Anti-Mi2 Dermatomyositis Revisited: Pure DM Phenotype with Muscle Fiber Necrosis and High Risk of Malignancy

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Meeting: 2016 ACR/ARHP Annual Meeting

Date of first publication: September 28, 2016

Keywords: Cancer, dermatomyositis and myositis

SESSION INFORMATION

Date: Tuesday, November 15, 2016

Session Type: ACR Poster Session C

Session Title: Muscle Biology, Myositis and Myopathies - Poster II: Clinical

Session Time: 9:00AM-11:00AM

Background/Purpose: Anti-Mi2 autoantibodies (Aabs) have been proposed to be highly specific for dermatomyositis (DM) and to be associated with a DM classical phenotype consisting of typical skin rashes and low extra-muscular features. Cancer has been estimated in about 30% of all DM patients. Patients with anti-Mi2 DM are considered having a good prognosis, possibly related to a markedly lower risk of malignancy reported in this subset. Nonetheless, there has been only a few and small cohort descriptions of this DM subgroup. Our objective was therefore to describe the phenotype of anti-Mi-2 DM in a large French cohort.

Methods: A national multicenter retrospective cohort study was performed (15 medical centers) including all patients with a clinical phenotype suggestive of DM (cutaneous manifestations and/or muscle involvement) and a positive anti-Mi2 Aabs. Medical records were retrospectively reviewed. Muscle strength was assessed using the Medical Research Council (MRC) scale and

cancer-associated myositis (CAM) was defined as a cancer occurring ± 3 years of diagnosing myositis.

Results: A total of 65 patients were identified, 62% were female and mean age at diagnosis was 54 years old (yo) (±17 yo). DM skin rash was reported in 88% of patients, most frequently Gottron papules and/or sign (68%), periungueal erythema (51%) and heliotrope rash (40%). Peripheral muscle weakness was reported in 92% of patients and dysphagia was reported in 34% of patients. At diagnosis, patients displayed severe muscle weakness (MRC 3/5, ±1/5*)* with mean CK level of 5085 UI/L (±5535 UI/L). Systematic review of muscle biopsies (n=11) showed marked inflammatory infiltrates. Strikingly, necrosis and regeneration were identified in all patients (n=11/11). C5b-9 deposition was found in all patients mainly on non-necrotic fibers but only sparsely on capillaries and without prominent capillary loss. Arthritis, Raynaud phenomenon and interstitial lung disease were reported in less than 20 % of patients**.** CAM was identified in 20% of patients and detected within one year and a half of DM diagnosis in most patients (n=11/12). All CAM patients, but one (38 yo), were diagnosed over 50 yo. There was no predominant histological subtype of malignancy (gastro-intestinal, urological, gynecological and pulmonary) and cancer was metastatic in a third of patients. Survival rate was 83% after a mean follow-up of 4.9 years from cancer diagnosis. Ninety-eight percent of patients were initially treated with corticosteroids (CS), in combination with immunosuppressant (IS) in 60% of cases. Patients treated with CS monotherapy (n=14), needed second-line agents upon follow-up in 60% of cases. In all, 53% of patients relapsed upon CS and/or IS tapering.

Conclusion: In this large French cohort, patients with anti-Mi2 DM displayed a phenotype with 3 main characteristics (i) pure DM phenotype (low overlap features) (ii) necrotizing myositis (severe weakness, high CK level and muscle fiber necrosis) and (iii) higher than expected malignancy rate.

Disclosure: O. Landon-Cardinal, None; **G. Monseau**, None; **Y. Schoindre**, None; **A. Rigolet**, None; **N. Champtiaux**, None; **B. Hervier**, None; **A. Masseau**, None; **E. Hachulla**, None; **T. Papo**, None; **B. Terrier**, None; **A. Meyer**, None; **J. E. Kahn**, None; **F. Maurier**, None; **F. Gaches**, None; **E. Salort-Campana**, None; **T. Zenone**, None; **N. Costedoat-Chalumeau**, None; **F. Perez**, None; **M. Samson**, None; **A. M. Piette**, None; **G. Moulis**, None; **S. Audia**, None; **S. Genot**, None; **N. Schleinitz**, None; **G. Lefevre**, None; **L. Verneuil**, None; **O. Benveniste**, None; **Y. Allenbach**, None; **B. Bienvenu**, None.

To cite this abstract in AMA style:

Landon-Cardinal O, Monseau G, Schoindre Y, Rigolet A, Champtiaux N, Hervier B, Masseau A, Hachulla E, Papo T, Terrier B, Meyer A, Kahn JE, Maurier F, Gaches F, Salort-Campana E, Zenone T, Costedoat-Chalumeau N, Perez F, Samson M, Piette AM, Moulis G, Audia S, Genot S, Schleinitz N, Lefevre G, Verneuil L, Benveniste O, Allenbach Y, Bienvenu B. Anti-Mi2 Dermatomyositis Revisited: Pure DM Phenotype with Muscle Fiber Necrosis and High Risk of Malignancy [abstract]. *Arthritis Rheumatol.* 2016; 68 (suppl 10). http://acrabstracts.org/abstract/anti-mi2-dermatomyositis-revisited-pure-dm-phenotype-with-muscle-fiber-necrosis-and-high-risk-of-malignancy/. Accessed November 18, 2016.
