ABSTRACT NUMBER: 2314

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

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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.

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