17 ANTI-MJ/NXP-2 ANTIBODIES ARE THE MOST COMMON SPECIFCITY IN A COHORT OF ADULT CAUCASIAN PATIENTS WITH DERMATOMYOSITIS

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Background/objective Specific autoantibodies in patients with polymyositis/dermatomyositis (PM/DM) are associated with unique subsets, and they are useful in monitoring clinical course and predicting outcome. Anti-MJ antibodies, which recognise the nuclear protein NXP-2 in PML (promyelocytic leukemia) nuclear bodies, are a new specificity reported in 23–25% of juvenile DM, usually associated with severe muscle atrophy, functional impairment, and calcinosis. The aim of our study is to analyse the prevalence and clinical significance of anti-MJ antibodies in a cohort of adult Caucasian PM/DM patients.

Methods Autoantibodies in sera from 58 consecutive adult PM/DM patients (74% female, mean age 43+/–17, mean follow-up 55 months) were analysed by immunoprecipitation of 35S-labeled K562 cell extracts, ELISA (anti-MJ, Ro52, La, Jo-1), Western Blot and Indirect Immunofluorescence (IIF). Clinical association of anti-MJ was analysed using information from charts and database.

Results Anti-MJ antibodies are the most prevalent specificity (10/58; 17%) in our PM/DM cohort, followed by anti-Jo-1 (10%), -p155/140 (5%), -SRP (5%), -EJ (4%) and anti-Mi-2, -SMN complex, -OJ with one case each. Anti-MJ was found in 30% of DM and 8% of PM (p: 0.02). Among 10 cases of anti-MJ, 8 were DM and 2 were PM. When clinical features of 10 cases of anti-MJ (+) versus 48 cases anti-MJ (-) were compared, DM is more common (p: 0.03) and no overlap syndrome patients were found in anti-MJ group (0% vs 13%). Age of onset (25.5 vs 46.1 years) and age at initial visit (37.6 vs 54.6 years) were younger in anti-MJ group (p: 0.002), and 2 anti-MJ (+) were pediatric onset DM. In anti-MJ (+) patients, heliotrope rash (p:0.01) and calcinosis (p:0.057) were common, however, none of them had heart involvement (0% vs 27%, p:0.03), interstitial lung disease (0% vs 33%, p:0.048), or cancer (0% vs 8%). Myopathy in anti-MJ (+) patients showed a good response to steroid therapy. Only 6/10 anti-MJ (+) showed PML body nuclear dots staining in IIF, suggesting that IIF cannot be used for screening of anti-MJ antibodies.

Conclusion Anti-MJ antibodies are detected also in adult PM/DM, and they are the most frequent specificity in our cohort, found in 17% of PM/DM (30% in DM and 8% in PM). Anti-MJ (+) patients showed DM of young onset with a mild myopathy, severe calcinosis and without internal organ involvement. Anti-MJ will be a useful new addition of myositis-associated autoantibodies
to help clinical monitoring of patients with adult PM/DM.