Pediatric Rheumatology

Oral presentation

8.4 Novel autoantibodies targeting a p140 protein are a major autoantigen system in juvenile dermatomyositis and a marker of calcinosis

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Background

To demonstrate that autoantibodies targeting a p140 protein are a major autoantigen in juvenile dermatomyositis (JDM) and describe the clinical associations in children recruited to the JDM Registry (JDRR).

Methods

156 children were studied. Serum was screened by immunofluorescence (IF) and radio-immunoprecipitation (IPP) [1]. Immunodepletion was used to establish whether p140 is different to p155/140 also recognised in JDM [1].

Results

21% of children were positive for anti-p140 on IPP, with a weak non-specific nuclear pattern or negative ANA on IF. No anti-p140 cases were positive for other autoantibody specificities. Immunodepletion confirmed that p140 and p155/140 are different autoantigens. Anti-p140 positives compared to negatives had a similar male:female ratio and age at diagnosis. No significant difference was observed in the type or distribution of rash when comparing anti-p140 positives vs. negatives except for more rash on the trunk in negative cases ($p = 0.017$). Calcinosis was significantly more frequent in anti-p140 positives (52%) compared to negatives (13%) ($p < 0.001$, OR 7.1 95% CI 3–16.8). When comparing anti-p140 and anti-p155/140 cases; cutaneous oedema ($p = 0.04$) and rash over the trunk ($p = 0.002$) and small joints ($p = 0.013$) was more frequent in anti-p155/140. Calcinosis in anti-p140 remained a significant feature compared to anti-p155/140 ($p = 0.005$, OR 6.4 95% CI 2–22).

Conclusion

Anti-p140 found in this cohort is likely to be the same as anti-MJ, described against nuclear matrix protein NXP-2 [2]; further confirmation is required. Anti-p140 is a major autoantibody subset in JDM. Further characterisation of this system will provide insights into the pathophysiology of calcinosis in JDM.

References

1. Gunawardena H, Wedderburn LR, North J, Betteridge ZE, Dunphy J, Chinoy H, Davidson JE, Cooper RG, McHugh NJ, for the Juvenile Dermatomyositis Research Group UK: **Clinical associations of autoantibodies to a p155/140 kDa doublet in juvenile dermatomyositis.** *Rheumatology* 2008, **47:**324-8.
2. Targoff IN, Trieu EP, Levy-Neto M, Fertig N, Oddis CV: **Sera with Autoantibodies to the MJ antigen react with NXP2.** *Arthritis Rheum* 2007, **56(Suppl 9):**S787.