Pediatric Rheumatology

Poster presentation

Myeloid cells which secrete S100 proteins in juvenile dermatomyositis may contribute to disease activity

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Background

Juvenile dermatomyositis (JDM) is thought to involve an autoimmune myositis, yet the elements of the immune response which damage muscle tissue in JDM remain unclear. Muscle tissue from early JDM shows infiltration by predominantly macrophage/myeloid cells. Traditional histopathology would suggest that these cells have a scavenger or 'repair' function; our data suggest otherwise. We have analysed production of the highly proinflammatory S100 proteins MRP8/14 in JDM patients.

Methods

40 children with JDM (32 female) were recruited through the UK JDM Registry and Repository. Muscle biopsy tissue (n = 33) serum (n = 39) and clinical data (physicians global assessment, CMAS, CK) were analysed. Serum MRP8/14 was measured by ELISA in JDM and 50 healthy age-matched children. Muscle biopsies were analysed by 2-colour immunofluorescence, stained with antibodies to human CD68, CD14, CD163, CD15, DC-LAMP, MRP14 and the heterodimer MRP8/14.

Results

MRP8/14 were significantly raised in serum from children with JDM (2428 ± 1717 ng/ml) compared to age matched controls (340 ± 40 ng/ml). Serum MRP8/14 levels correlated with CMAS (r = -0.525) PGA (r = 0.498) and CK (r = 0.688). Muscle biopsy analysis frequently showed early diffuse infiltrate of MRP14+ cells. Lineage analysis of infiltrating MRP14+ cells showed that the majority were CD68+ (76.6%) with a minority of MRP14+ cells being CD14+ (21.9%), CD163 (20.2%) or CD15+ (19.6%). Infiltrating DCs did not express MRP proteins.

Conclusion

MRP8/14 levels in serum of patients with JDM correlate with disease activity, and a subpopulation of pro-inflammatory macrophages in muscle tissue may be a source of these highly inflammatory proteins.