PP14. COMPARISON OF THE UTILITY AND VALIDITY OF THREE SCORING TOOLS TO DETECT SKIN DISEASE IN PATIENTS WITH JUVENILE DERMATOMYOSITIS

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Background: In JDM the involvement and assessment of skin is a vital aspect. The abbreviated Cutaneous Assessment Tool (CAT) encompassing active skin disease and skin damage, the DAS and the Myositis Intention to Treat Activity Index (MITAX), both with skin components, have been suggested for the measurement of skin disease in JDM patients; however, the optimal tool is unknown.

Aims: To compare the three tools and correlate them with the physician’s 10 cm visual analogue scale (skin VAS) to define which tool best assesses skin disease.

Methods: Patients recruited to the UK JDM Cohort & Biomarker Study who fulfilled Bohan–Peter criteria were included. Each patient was assessed using the CAT, DAS, MITAX and a skin VAS. Markers of muscle disease [Childhood Myositis Assessment Scale (CMAS), MMT8, creatine kinase U/l), inflammatory markers (CRP mg/l and ESR mm/h) and physician’s global score were recorded. Spearman’s correlation (r) was used and a relationship >0.75 was considered strong. A P-value <0.05 was considered significant.

Results: 67 JDM patients were assessed; 59.7% were female. The mean (±s.d.) age was 8.96 (3.37) years, with mean age at diagnosis 6.59 (3.42) years and mean disease duration 3.26 (3.08) years. The DAS skin had the strongest correlation with the skin VAS (Table 1). The MITAX skin and CAT activity scores were significantly correlated with the skin VAS. They were all correlated with CMAS and MMT8 scores; no significant correlations were noted with the CK.

Conclusion: These data demonstrate the potential application of using a skin assessment tool to evaluate and monitor skin involvement in JDM patients. It demonstrates that the DAS skin appears to be the best of the tools using the skin VAS as the gold standard. The DAS skin was concise, quick to use and easy to score.

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Table 1. Spearman’s correlation between items shown as r and corresponding P value

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<tbody>
<tr>
<td>Skin VAS</td>
<td>P = 0.831 r = 0.823</td>
<td>P = 0.954 r = 0.940</td>
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<td>CMAS</td>
<td>P = 0.501 r = 0.501</td>
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<td>MMT8</td>
<td>P = 0.933 r = 0.933</td>
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<td>CK</td>
<td>P = 0.921 r = 0.921</td>
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<td>ESR</td>
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CMAS: Childhood Myositis Assessment Scale; VAS: visual analogue scale.

PP15. JUVENILE-ONSET SYSTEMIC LUPUS ERYSITEMATOSUS WITH OVERLAP FEATURES OF ANCA-ASSOCIATED VASCULITIS: A CASE REPORT

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Background: Vasculitis in SLE is well documented. However, ANCA positivity has been shown to correlate poorly with vasculitis activity in SLE patients [1]. A small case series has shown juvenile-onset SLE (JSLE) and ANCA-associated vasculitis (AAV) as an overlapping syndrome with good response to cyclophosphamide [2].

Aims: To discuss a case of an adolescent girl presenting with JSLE, ANCA positivity and features of possible vasculitis.

Methods: A 14-year-old girl of Arabic ethnicity had been diagnosed 3 years previously in Eastern Asia with JSLE. Disease course had been complicated by vasculo-inflammatory disease with gangrene of a digit due to possible APS or a vasculitic process. She presented in the UK with features of lupus-like illness including low complement levels and erythematous maculopapular rash. Features of possible vasculitis included nasal crusting, epistaxis, chronic cough and haemoptysis. She also had episcleritis, intermittent dactylitis and pericardial effusion, which are described in both lupus and AAV. She had active disease despite previous rituximab in Kuwait and ongoing treatment with HCQ, MMF and long-standing high dose prednisolone (1 mg/kg/day).

Results: Investigations revealed a positive pANCA, targeted against myeloperoxidase (MPO) at 23 (-9). Interestingly, anti-dsDNA antibodies were negative and complement deficiency was unlikely, with normal CH50 and AP100. High resolution CT revealed interstitial lung disease and bronchoscopy showed increased secretions with a brown tinge and bronchoalveolar lavage cytology revealed macrophages laden with haemosiderin. Skin biopsy supported SLE with neutrophilic dermatosis and no evidence of granulomata. The risk of an open lung biopsy was thought to outweigh the benefits. She was therefore commenced on cyclophosphamide with the aim of achieving remission and as a steroid-sparing agent.

Conclusion: Our case highlights a possible overlapping syndrome, where an adolescent patient presented with features of JSLE and later, possible AAV. Long-term immunosuppressant therapies can mask overlapping features, making it difficult to obtain proven biopsy changes suggestive of AAV. However, awareness of this association can be used to guide further investigations and treatment tailored to specific disease manifestations.

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References

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