Extramuscular manifestations of idiopathic inflammatory myopathies

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Myositis is a heterogeneous disease

**Diagnosis**
- Polymyositis
- Dermatomyositis
- Juvenile DM (JPM)
- Paraneoplastic
- Myositis in overlap
- IBM
- Necrotizing myopathy
- Antisynthetase syndrome
- Amyopathic DM
- DM without dermatitis
- Nonspecific myositis

**Organ involvement**
- Muscle
- Skin
- Lung
- Heart
- Oesophagus
- Joints
- Calcinosis

Hallmark inflammatory skin lesions of DM

- Göttron’s papules.
- Göttron’s sign of DM.
- Peri orbital (heliotrope) violaceous erythema.
- Periungual telangiectasia.
- Confluent, macular, violaceous erythema over the dorsal aspect of the hands and fingers.
- Mechanic’s hand lesion.
- Poikiloderma atrophicans vasculare (poikilodermatomyositis)
- Calcinosis cutis.

Göttron’s papules

- Papules with a violaceous hue overlying the dorsal-lateral aspect of the interphalangeal and/or metacarpophalangeal joints. When fully formed, these papules become slightly depressed at the centre, which can assume a white, atrophic appearance. Associated telangiectasia can be present.

Göttron’s signs of DM

Symmetrical, confluent, macular, violaceous erythema with or without oedema overlying the dorsal aspect of the interphalangeal/metacarpophalangeal joints, olecranon processes, patellae and medial malleoli.

Peri orbital (heliotrope) violaceous erythema

- Confluent, macular, violaceous erythema of the eyelids and periorbital tissue with or without associated oedema.
Periungual telangiectasia

- Grossly visible with or without accompanying dystrophic cuticles and cuticular haemorrhage

Confluent, macular, violaceous erythema over the dorsal aspect of the hands and fingers

- This change often tracks along the extensor tendons. Also involves the extensor aspects of the arms and forearms, deltoids, posterior shoulders and neck (shawl sign), V-area of the anterior neck and upper chest (V-sign), central aspect of the face and forehead, and scalp

Mechanic's hand lesion

- Bilaterally symmetrical, confluent hyperkeratosis having the appearance of that produced by manual labour distributed along the ulnar aspect of the thumb and radial aspect of the fingers; also prominent on the index and middle fingers with occasional extension to the palmar surfaces

Poikiloderma atrophicans vasculare (poikilodermatomyositis)

- Circumscribed violaceous erythema with associated telangiectasia, hypopigmentation, hyperpigmentation and superficial atrophy most commonly found over the posterior shoulders, back, buttocks and V-area of the anterior neck and chest. Often asymmetrical.

Holster sign

Ulcration
Flagellar erythema

Alopecia

Palmar papules and plaques

Calcinosi

- **Superficial plaques and nodules**
  - Superficial dermal or subcutaneous papules, plaques and nodules that affect areas of potential trauma and inflammation, such as the elbows, knees, and posterior thigh.

- **Tumorous deposits**
  - Deep dermal and subcutaneous deposition, of calcium in sites of inflammation and subclinical trauma

- **Fascial plane deposition**
  - Calcinosi deposited along the intermuscular fascial plane. This often presents as a palpable indurated, well-demarcated linear deposit along the fascial planes of the extremities.

- **Exoskeleton**
  - External calcinosi that extensively encases soft tissues

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Antisynthetase syndrome

- Myositis
- Interstitial lung disease (89%)
- Arthritis (94%)
- Raynaud’s phenomenon (67%)
- Fears (87%)
- Mechanic’s hands (71%)
- Anti- Jo-1 – similar pathology
  - Periarticular fragmentation
  - Macrophage predominance
  - Perifascicular changes (atrophy, regeneration, some necrosis)
  - Normal capillary density

Arthritis

- Most frequent in ASS
- Also in other forms
- 33% of patients with IIMs
- Considered less severe and less destructive than RA
- Nonerosive arthritis
- Erosive and destructive
- Subluxing arthropathy

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Arthritis at Any Time</th>
<th>Arthritis at Disease Onset**</th>
<th>Current Arthritis† (≥ 1 swollen joint)</th>
</tr>
</thead>
<tbody>
<tr>
<td>PM (46)</td>
<td>25 (59)</td>
<td>19 (41)</td>
<td>17 (40)</td>
</tr>
<tr>
<td>DM (40)</td>
<td>22 (55)</td>
<td>15 (38)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>CAM (8)</td>
<td>2 (25)</td>
<td>1 (13)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>IMNM (11)</td>
<td>4 (36)</td>
<td>2 (18)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>IRM (1)</td>
<td>1 (100)</td>
<td>1 (100)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Total (106)</td>
<td>56 (53)</td>
<td>39 (37)</td>
<td>31 (29)</td>
</tr>
</tbody>
</table>

When at disease onset - then in 50% before muscle weakness

Table 2. Arthritis in myositis subtypes. Data are n (%).


Table 3. Examples of maximal score

| Severe arthritis with extreme loss of function (bedridden, inability for self care) |
|-------------------------------|---------------------------------------------|
| 10                           | 9                            |
| 8                             | 7                            |
| 6                             | 5                            |
| 4                             | 3                            |
| 3                             | 2                            |
| 2                             | 1                            |

Skeletal disease activity

<table>
<thead>
<tr>
<th>Skeletal Disease Activity</th>
<th>Absent</th>
<th>(Maximum)</th>
<th>Examples of maximal score</th>
</tr>
</thead>
<tbody>
<tr>
<td>13. Arthritis</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>a) Severe active inflammatory polyarthritis</td>
<td>0 1 2 3 4</td>
<td>N/A</td>
<td></td>
</tr>
<tr>
<td>b) Moderately active inflammatory arthritis</td>
<td>0 1 2 3 4</td>
<td>N/A</td>
<td></td>
</tr>
<tr>
<td>c) Mfl arthritis</td>
<td>0 1 2 3 4</td>
<td>N/A</td>
<td></td>
</tr>
<tr>
<td>14. Arthritis</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>a) Severe: arthritis of two or more joints with clinically significant loss of the functional range of movement and requiring assistance with activities of daily living</td>
<td></td>
<td></td>
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<tr>
<td>b) Moderate: arthritis of one or more joints with some loss of functional range of movement, but not requiring assistance with activities of daily living</td>
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<tr>
<td>c) Mild: arthritis of one or more joints with neither loss of range of motion nor impaired activities of daily living</td>
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</table>

Arthritis: joint pain with or without stiffness but due to an inflammatory process in two or more joints.
Interstitial lung disease (ILD)

- Nonspecific interstitial pneumonia (NSIP)
- Usual interstitial pneumonia (UIP)
- Organizing pneumonia (OP)
- Lymphocytic interstitial pneumonia (LIP)
- Acute interstitial pneumonia (AIP)/diffuse alveolar damage (DAD)

Dysphagia

- More severe disease and worse prognosis, fibrosis to cricopharyngeal muscle
- Aspiration - pneumonia or lung abscess.
- IBM, PM, DM, IMNM
  - Avoid barium esophagography
  - Alternatives: manometry or RT MRI
  - Upper endoscopy to rule out cancer

Pulmonary involvement

- 40% (wide range)
- Interstitial lung disease (ILD)
- Aspiration pneumonia
- Drug-induced lung disease
- Ventilatory muscle weakness
  - Respiratory failure
  - Significant dyspnea
- Pulmonary hypertension

Interstitial lung disease in myositis (46%)

<table>
<thead>
<tr>
<th>ILD in 107 PM/DM patients</th>
<th>6M deterioration (m=47)</th>
<th>Absence of ILD deterioration (m=39)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dyspnoea</td>
<td>95</td>
<td>63</td>
</tr>
<tr>
<td>Cough</td>
<td>94</td>
<td>63</td>
</tr>
<tr>
<td>Asympt.</td>
<td>6</td>
<td>36</td>
</tr>
<tr>
<td>FVC</td>
<td>66</td>
<td>71</td>
</tr>
<tr>
<td>VC</td>
<td>70</td>
<td>75</td>
</tr>
<tr>
<td>DLCO</td>
<td>36</td>
<td>54</td>
</tr>
<tr>
<td>UIP HRCT</td>
<td>55</td>
<td>24</td>
</tr>
<tr>
<td>NSIP</td>
<td>22</td>
<td>42</td>
</tr>
<tr>
<td>Biopsy</td>
<td>67</td>
<td>32</td>
</tr>
<tr>
<td>Mortality</td>
<td>47</td>
<td>3</td>
</tr>
</tbody>
</table>

**HRCT patterns of ILD in myositis**

- **Reticular opacity**
- **Groundglass attenuation**
- **Consolidation-dominant pattern in patients with DM**
- **Typical presentation of subpleural band**

Honeycombing, in a pattern consistent with usual interstitial pneumonia.

**Cardiovascular involvement in myositis**

**Increased risk in the first years**
- Myocarditis
- Myocardial infarction
- Venous thromboembolism - DVT and PE

**Increased risk in long-term IIM**
- Subclinical cardiac involvement effecting both heart function and rhythm, and conduction abnormalities, which in part might be because of myocarditis
Relative risk of incident myocardial infarction according to polymyositis/dermatomyositis status

<table>
<thead>
<tr>
<th>Myocardial infarction</th>
<th>Polymyositis (n=431)</th>
<th>Non-polymyositis (n=4496)</th>
<th>Dermatomyositis (n=352)</th>
<th>Non-dermatomyositis (n=3528)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Events, n</td>
<td>29</td>
<td>198</td>
<td>13</td>
<td>74</td>
</tr>
<tr>
<td>Total follow-up time</td>
<td>1287</td>
<td>19 650</td>
<td>1035</td>
<td>15 764</td>
</tr>
<tr>
<td>Incidence rate per 1000 person-years</td>
<td>22.52</td>
<td>5.50</td>
<td>12.56</td>
<td>4.69</td>
</tr>
<tr>
<td>Age-, sex-, and entry time–matched cox HR (95% CI)</td>
<td>5.20 (3.31, 8.17)</td>
<td>1.0</td>
<td>3.51 (1.88, 6.54)</td>
<td>1.0</td>
</tr>
<tr>
<td>Fully-adjusted age-, sex-, and entry time–matched cox HR (95% CI)</td>
<td>3.89 (2.28, 6.65)</td>
<td>1.0</td>
<td>2.92 (1.48, 5.78)</td>
<td>1.0</td>
</tr>
</tbody>
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