



## IRA E-BULLETIN : ISSUE 2 | APRIL 2020

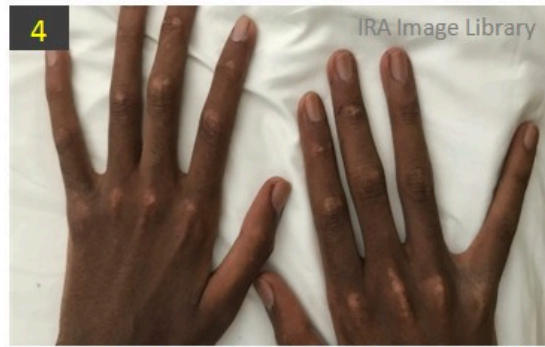
### THE HELIOTROPES RASH & THE GOTTRON OF ALL VARIETIES

		<ul style="list-style-type: none"><li>➤ Pink-violaceous erythema involving the periorbital skin</li><li>➤ Component of ACR/EULAR classification criteria having 3.1 points without muscle biopsy and 3.2 points with muscle biopsy</li><li>➤ An important differentiating finding from Lupus</li></ul>
<p>1a. &amp; 1b. -&gt; Heliotrope Rash</p>		
		<ul style="list-style-type: none"><li>➤ <b>Macular</b>, pink-violaceous erythema overlying other joints, such as the elbows or knees</li><li>➤ Component of ACR/EULAR classification criteria having 3.3 points without muscle biopsy and 3.7 points with muscle biopsy</li></ul>
<p>1a. &amp; 1b. -&gt; Gottron Sign at elbow &amp; Knee</p>		

### THE GOTTRON OF ALL VARIETIES



3. -> Gottron Papule



4. -> Healed Gottron (Poikiloderma)



5. ->Ulcerative Gottron



6. ->Inverse Gottron

**3. Gottron Papule**

- Pink-violaceous papules overlying interphalangeal and metacarpophalangeal joints
- Component of ACR/EULAR classification criteria having 2.1 points without muscle biopsy and 2.7 points with muscle biopsy

**4. Healed Gottron**

- Area of hypopigmentation with or without scarring
- Component of Myositis Damage Index

**5. & 6.**

**Ulcerative & Inverse Gottron**

- Inverse Gottron are palmar Gottron papules.
- Less commonly reported but sinister sign of Rapidly progressive Interstitial Lung Disease.
- More commonly observed with
  - Anti MDA-5 positivity
  - Amyopathic variant

## ANTI SYNTHETASE SYNDROME



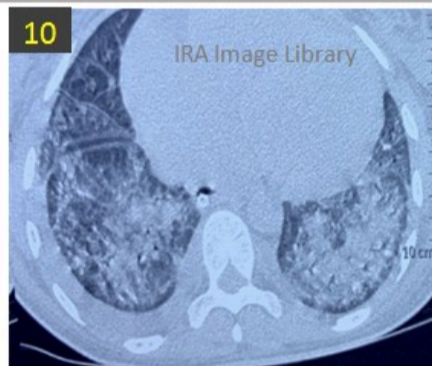
7. ->Mechanic's Hands



8. ->Mechanic's Hands & Hiker's Feet



9. ->Raynaud's Phenomenon



10. ->Interstitial Lung Disease

➤ **Characterized by**

- Autoantibodies against one of many aminoacyl transfer RNA (tRNA) synthetases with clinical features of myositis and
  - Interstitial lung disease (ILD),
  - Non-erosive arthritis,
  - Raynaud's phenomenon,
  - Unexplained fever and/or
  - mechanic's hands or Hiker's Feet

➤ **Mechanic's hands and hiker's feet**

- May serve as a disease activity marker.
- Improvement is often seen with successful treatment.

➤ **ANA Pattern**

- Cytoplasmic pattern as antigen targets are located in cytoplasm.

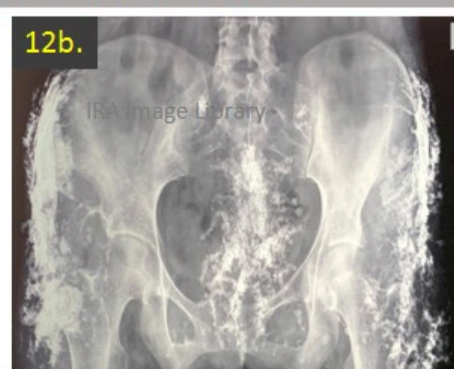
## CALCINOSIS CUTIS



11a. & 11b.-> Calcinosis Cutis, Elbow & Bilateral Knee



12a. & 12b.-> Radiographic Calcinosis Cutis



- Due to Dystrophic deposition of insoluble calcium salts in skin, fascia, muscle and subcutaneous tissue.

➤ **Association**

- More common with juvenile dermatomyositis
- Associated with NXP2 and TIF1 Gamma positivity

➤ **Calcinosis cutis in DM vs SSc**

- Usually bulky, nodular and mass like over trunk and extensor aspects of joints in DM
- Linear, superficial and predominantly involves hands in systemic sclerosis.

# SIGNS OF DERMATOMYOSITIS

<p>13a. -&gt; "V" Sign &amp; 13b. Shawl Sign</p>		<p>14. -&gt; Holster Sign</p>
		<p><b>13. "V" Sign &amp; Shawl Sign</b></p> <ul style="list-style-type: none"> <li>Classical signs of Photosensitivity seen in dermatomyositis</li> </ul> <p><b>14. Holster Sign</b></p> <ul style="list-style-type: none"> <li>Violaceous rash over the lateral hip</li> <li>Under reported sign of dermatomyositis which has 91% specificity and around 80% sensitivity (if examined !!).</li> </ul> <p><b>15 Nailfold Infarct</b></p> <ul style="list-style-type: none"> <li>Inflammatory vascular lesion, more common with juvenile dermatomyositis .</li> </ul> <p><b>16. Ragged Cuticle</b></p> <ul style="list-style-type: none"> <li>An important clinical finding mentioned in dermatomyositis and systemic sclerosis</li> </ul>
<p>15. -&gt; Nailfold Infarct</p>	<p>16. -&gt; Ragged Cuticle</p>	

## MRI THIGH: ACTIVITY & ATROPHY

<h3>MRI Thigh: Activity &amp; Atrophy</h3>		
		<p>The earliest change at MRI of inflamed muscles is muscle edema,</p> <ul style="list-style-type: none"> <li>Secondary to increased water content</li> <li>Usually diffuse in polymyositis , whereas in dermatomyositis it can be ill-defined and patchy. In addition, in dermatomyositis, edema can also spread to the subcutaneous tissue and myofascial structures</li> </ul> <p>Damage is detected by T1 weighted images</p> <ul style="list-style-type: none"> <li>characterized by muscle atrophy and/or fat replacement</li> </ul>
<p>18. -&gt; T2 with Fat Suppression (STIR) Sequence of MRI Thigh shows Increased uptake in vastus lateralis and intermedius bilaterally Suggestive of Active Myositis</p>	<p>19. -&gt; T1 Sequence of MRI Thigh shows Atrophic changes with fatty infiltration Suggestive of Damage due to Myositis</p>	

## A MIMIC TO DERMATITIS OF DERMATOMYOSITIS : VITAMIN A DEFICIENCY



- **Young girl with**
  - Periorbital edema,
  - Dry, flaky skin lesions over the shawl area, knees, legs
  - Muscle aches and mild muscle weakness.
- Clinically mimics **Inflammatory myositis**

- This patient also had stomatitis, chronic diarrhoea.
- Normal level of muscle enzymes, flaky dermatitis over large area, presence of Bitots spots, angular stomatitis, glossitis helped in diagnosis of Vitamin A deficiency and differentiating from the Inflammatory myositis.

## Image Contributors:

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Sangeetha KN	St. John's, Bengaluru	2a,16,19.
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