

Teaching Slide Collection of the Cutaneous Manifestations of the Myositis Disease Activity Assessment Tool

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Introduction

- The following is a slide collection of representative examples of some of the cutaneous lesions associated with juvenile and adult dermatomyositis (DM) and other idiopathic inflammatory myopathies (IIM) which are included in the Myositis Intention to Treat Activity Index (MITAX).
- Guidance on scoring these lesions using the MITAX and the Myositis Disease Activity Assessment Visual Analogue Scales (MYOACT) is also provided.

Introduction - II

- This work is part of a larger photo-essay of the cutaneous manifestations of the IIM, being prepared by the International Myositis Outcome Assessment Collaborative Study Group (IMOACSG). Please contact the authors for permission to reproduce in any way.

MITAX Scoring, General Principles I

- **Category A:** Presence of any of the following lesions with score of 2 (same), 3 (worse), or 4 (new):
 - Cutaneous ulceration
 - Erythroderma
 - Erythema accompanied by vesiculobullous or erosive changes or necrosis
 - Panniculitis involving $\geq 10\%$ body surface area.
- These lesions become **category B** (score = 1, improving) when they are improving such that therapy can be reduced below level in category A (moderate doses of steroids or immunosuppressives).
- These lesions become **category C** when they are stable or improving such that therapy can be reduced below category B (ie., low doses of corticosteroid or other immunosuppressive agents, antimalarials, topical steroids).

MITAX Scoring, General Principles II

- The following are **category B** lesions when scored 2 (same), 3 (worse) or 4 (new):
 - Erythema without secondary changes
 - Panniculitis involving < 10% body surface area
 - Heliotrope rash
 - Gottron's papules or sign
 - Periungual capillary changes
 - Alopecia: severe, diffuse
 - Mechanic's hands
- These lesions become **category C** when scored 1 (improving), such that therapy can be reduced below category B to symptomatic therapy (low doses steroids and other agents, topical steroids, antimalarials).

MITAX Scoring, General Principles III

- **Category C:**

- Any Category A or B lesion improving, as noted above
- Alopecia: mild, focal
- Vascular changes: livedo reticularis or telangiectasias

VAS Scoring – General Principles

- The VAS score integrates the severity of activity based upon all of the cutaneous manifestations present (not all of which may be part of the MITAX).
- Maximum value (10 cm) guidelines for cutaneous VAS score:
 - Ulceration to muscle, tendon, or bone
 - Erythroderma involving $\geq 80\%$ body
- See specific examples as follows for further guidance in scoring. The proposed scoring in the following examples pre-supposes each lesion is the only cutaneous manifestation.

Cutaneous Ulceration

Definition:

Primary injury of the skin due to vascular insufficiency, with necrosis of the epidermis, dermis or deeper.

Cutaneous Ulceration- Examples



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- Close up of isolated deep ulceration on posterior thigh which extends to the biceps femoris and semitendinosus muscles from a girl with JDM.
- **Proposed Scoring:**
 - MITAX score: 4 (new), category A
 - VAS scale: 10 cm

Ulceration - Examples



- Well circumscribed, relatively shallow healing ulcer involving the forearm, with secondary crust and surrounding mild erythema in a boy with juvenile DM.
- **Proposed Scoring:**
 - MITAX: 1 (improving), category B
 - VAS Scale (if this is the only lesion): 5 cm

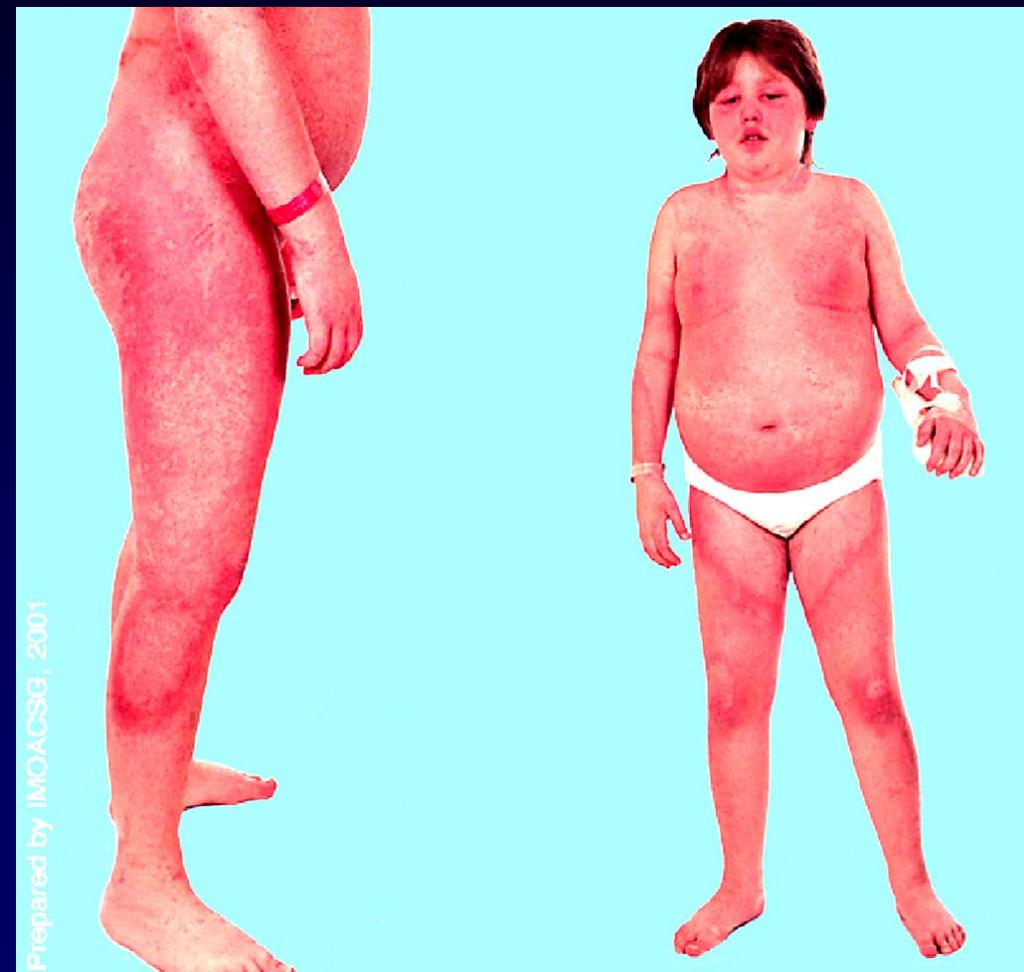
Erythroderma

Definition:

Generalized confluent erythema, with or without scale, involving both sun-exposed and non sun-exposed skin; can involve the entire cutaneous surface.

Erythroderma - Example

- Composite view showing confluent erythema involving both sun-exposed and non sun-exposed skin in a boy with severe juvenile DM. Note the generalized edema.
- **Proposed Scoring:**
 - MITAX: 3 (worse), category A
 - VAS scale: 8 cm



Erythematous Rashes

Definition:

Includes sun-exposed and non sun-exposed erythema, such as malar, facial, linear extensor erythema, V- and shawl-sign, erythema in periungual distribution, and non sun-exposed erythema.

Accompanied by Secondary Changes – Vesiculobullous Changes, Erosions, Necrosis

Definitions:

Vesiculobullous: Circumscribed, elevated fluid-containing lesions; vesicles measure ≤ 0.5 cm diameter, bullae are greater in size.

Erosions: Moist, slightly depressed lesions in which part or all of the epidermis has been denuded.

Necrosis: Cell death or devitalized tissue, which can manifest in the skin as a black eschar, ulceration, as well as dry or wet gangrene.

Erythema Accompanied by Vesiculobullous Changes- Example



- Partially-intact bullae with superficial desquamation (peeling) on a background of macular erythema on the neck and trunk of a patient with DM. When the blisters are broken down, superficial erosions develop with secondary hemorrhagic serous exudates forming crusts.
- **Proposed Scoring:**
 - MITAX: 3 (worse), category A
 - VAS scale: 8 cm

Erythema Accompanied by Erosions- Examples



- Confluent, macular erythema involving non sun-exposed skin with secondary linear excoriations in a girl with juvenile DM.
- **Proposed Scoring:**
 - MITAX: 3 (worse), category A
 - VAS scale: 6 cm

Erythema Accompanied by Necrosis- Example

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- Ulceration with central necrosis and surrounding erythema at the site of a Gottron's papule involving the elbow in a boy with severe juvenile DM.
- **Proposed Scoring:**
 - MITAX: 3 (worse), category A
 - VAS scale: 9 cm

Erythematous Rashes without Secondary Changes - Examples



- *Malar/facial erythema:*
Acute, confluent macular erythema in a periorbital and malar distribution with extension to the chin; note sparing of perioral and nasolabial skin in this girl with JDM.
- **Proposed Scoring:**
 - MITAX: 4 (new), category B
 - VAS scale: 6 cm

Erythematous Rashes without Secondary Changes - Examples



Prepared by IMOACSG, 2001

- *Linear extensor erythema:* Confluent macular erythema of the extensor aspects of the thighs and legs in a patient with DM.
- Proposed Scoring:
 - MITAX: 3 (worse), category B
 - VAS: 6 cm

Erythematous Rashes without Secondary Changes - Examples



Prepared by IMOACSG, 2001

- *Non sun-exposed erythema:*
A well- circumscribed, intensely erythematous patch involving the medial thigh of a girl with juvenile DM.
- Proposed Scoring:
 - MITAX: 4 (new), category B
 - VAS: 3 - 4 cm

Panniculitis

Definition:

Tender erythematous or violaceous nodules or depressions due to inflammation in the subcutaneous fat. For the MITAX, lesions $\geq 10\%$ body surface area are scored as Category A, and those $< 10\%$ BSA are scored Category B, when either are new, worse or the same (see standard burn charts or *Harriet Lane Handbook*).

Panniculitis- Example



- Panniculitis involving the upper arm, which presented as swelling, redness and pain in a woman with DM. A biopsy showed superficial and deep perivascular and peri-appendageal mononuclear cell infiltrates extending into the subcutaneous fat septae.
- **Proposed Scoring:**
 - MITAX: 3 (worse), category B
 - VAS: 7 cm

Heliotrope Rash

Definition:

Discrete or confluent erythematous to violaceous macules usually involving the upper eyelids; can extend periorbitally. Can be associated with periorbital edema and telangiectasias of the eyelids.

Heliotrope Rash - Examples



- Confluent macular erythema confined to the upper eyelid, with associated periorbital edema.
- Proposed Scoring:
 - MITAX: 2 (same), category B
 - VAS: 3 cm

Heliotrope Rash - Examples



- Heliotrope, resolving as telangiectasia. Heliotrope was previously confined to the upper eyelid. Only telangiectasias and atrophy are apparent at present.
- **Proposed Scoring:**
 - MITAX: 1 (improving), category C
 - VAS: 2 cm

Gottron's Papules/Gottron's Sign

Definition:

Erythematous to violaceous papules and plaques (Gottron's papules), or macules (Gottron's sign), over extensor surfaces of joints, generally in a symmetric distribution.

Gottron's Papules - Example



- Symmetric erythematous to violaceous papules overlying the metacarpal and interphalangeal joints in a juvenile DM patient.
- Proposed Scoring:
 - MITAX: 2 (same), category B
 - VAS: 5 cm

Periungual Capillary Changes

Definition:

Dilatation of the nailfold capillaries which may be accompanied by vessel dropout and which is visible with the naked eye or by using additional magnification with otoscopy.

Periungual Capillary Changes - Example



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- Telangiectasia with prominent tortuosity and vessel dropout, demonstrated under an oil droplet in a boy with juvenile DM.
- Proposed Scoring:
 - MITAX: 2 (same), Category B
 - VAS: 5 cm

Alopecia

Definition:

Hair loss. May be mild or severe, depending on whether it is focal or diffuse, scarring or non-scarring.

Alopecia: Severe, Diffuse - Example



- Moderately severe, diffuse alopecia in a young woman with juvenile DM.
- Proposed Scoring:
 - MITAX: 2 (same), Category B
 - VAS: 5 cm

Alopecia: Mild, Focal - Example



- Focal alopecia in juvenile DM with associated scale.
- Proposed Scoring:
 - MITAX: 2 (same), Category C
 - VAS: 2 cm

Mechanic's Hands

Definition:

Hyperkeratosis and scaling with frequent fissuring and cracking along the lateral and palmar aspects of the fingers.

Mechanic's Hands - Example

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- Scale, cracking and fissuring with bleeding along the lateral and palmar aspects of the fingers in a PM patient.
- Proposed Scoring:
 - MITAX: 3 (worse), Category B
 - VAS: 4 cm

Vascular Changes

Definitions:

Livedo reticularis: Fixed peripheral vascular condition characterized by bluish-red netlike mottling of the skin on the trunk or extremities, which persists after the skin has warmed.

Telangiectasias: Fixed dilatation of superficial capillaries conferring a blanchable erythema to the involved skin.

Livedo Reticularis - Example



- Fixed erythema in reticular or netlike pattern involving the thigh in a patient with DM.
- Proposed Scoring:
 - MITAX: 3 (worse), Category C
 - VAS: 3 cm

Telangiectasia - Example



- Prominent telangiectasia on a background of blanchable erythema involving the upper chest (V-sign) in a DM patient.
- Proposed Scoring:
 - MITAX: 2 (same), Category C
 - VAS: 4 cm

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