

# Dermatologic Manifestations of Rheumatologic Diseases

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## CONFLICTS OF INTEREST & DISCLOSURES

- > I have no conflicts of interest
- > I have no pertinent disclosures

## OVERVIEW

- > Dermatomyositis
- > Psoriasis
- > Rheumatoid Arthritis
- > Vasculitis
- > Drug Complications

# DERMATO- MYOSITIS

## DERMATOMYOSITIS: DIAGNOSTIC CRITERIA

- > Classic DM is simplistically:
  - > Cutaneous findings
  - > Muscle findings



Image source: VisualDerm.com

## DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Facial erythema
- > Periorbital Edema
- > Heliotrope sign
- > Gottron's sign
- > Gottron's papules
- > Mechanics hands
- > Ragged cuticles
- > Nailfold capillary dilation and hemorrhage
- > V-neck erythema
- > Shawl sign
- > Low back erythema
- > Holster sign
- > Scalp involvement
- > Poikiloderma
- > Calcinosis
- > Ulcerations

### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Malar erythema
  - > May extend over nasal bridge
  - > Often involves the nasolabial folds
  - > More extensive involvement can be seen in other areas including the forehead, lateral face, and ears.



Mervi et al. Clinical presentation and evaluation of dermatomyositis. *Indian J Dermatol*. 2012; 57(5): 375-381.  
Dogan E, A Haber, F Miller, L Rider for the International Myositis Assessment and Clinical Studies Group (IMACS). Photorecay of the cutaneous manifestations of the idiopathic inflammatory myopathies. *DOI: 2009 / JCO / qph*

### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Periorbital Edema



Collins and Herberman. Dermatomyositis. *Clinics in Dermatology*. 2006; 24: 363-373.

### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Heliotrope rash
  - > Violaceous eyelid rash
  - > Often looks dry
  - > Frequently asymptomatic



Image source: VisualDx/visualdx.com

### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Gottron's sign
- > erythematous macules and patches overlying the elbows and/or knees

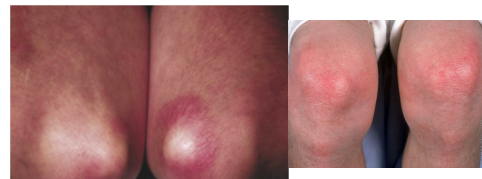


Image source: VisualDx/visualdx.com

### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Gottron's papules
  - > Scaly erythematous to violaceous papules and plaques over the extensor surfaces of the metacarpophalangeal and interphalangeal joints
  - > Darker skin types can be more challenging



### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Mechanic's Hands
  - > Rough, cracked, hyperkeratotic hands with predilection for the lateral and palmar areas of the fingers



### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Ragged cuticles, Nailfold capillary dilation/hemorrhage, Periungual erythema



Colten and Wortmann. Dermatomyositis. Clinics in Dermatology. 2006; 24: 363-373.

### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > V neck erythema, Shawl sign, Low back erythema
- > Confluent macular erythema



Image source: VisualDx/visualdx.com

### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Holster sign

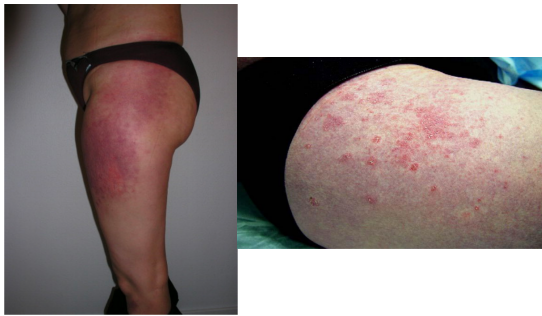


Image source: VisualDx/visualdx.com

### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Scalp involvement
- > Scale, erythema
- > May cause alopecia



Image source: VisualDx/visualdx.com

### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Poikiloderma
- > Hyperpigmentation + hypopigmentation + telangiectasias

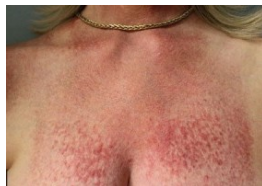
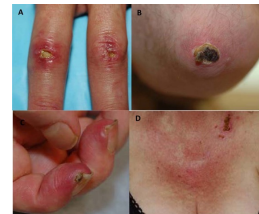


Image source: VisualDx/visualdx.com

### DERMATOMYOSITIS: CUTANEOUS MANIFESTATIONS

- > Calcinosis
- > Ulcerations
- > Common locations: extensor surfaces overlying joints (particularly fingers, elbows, and knees), lateral nailfolds or digital pulp, and sun-exposed areas (e.g. chest, ear helix)



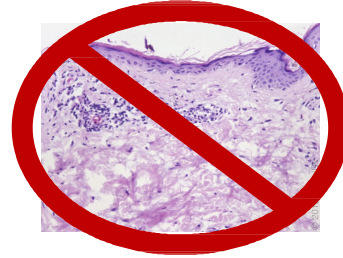
Narong et al. Cutaneous Ulcerations in Dermatomyositis: Association With Anti-Melanoma Differentiation-Associated Gene 2 Antibodies and Interstitial Lung Disease. Arthritis Care & Research. 2013;67(3): 467-472.

### DERMATOMYOSITIS: DIFFERENTIAL DIAGNOSIS

Acute Cutaneous Lupus Erythematosus	Dermatomyositis
Malar rash without nasolabial fold involvement	Facial rash with nasolabial fold involvement
Skin manifestations do not localize to joints	Skin manifestations local to joints
Minimal pruritus	Very pruritic
+/- DLE	Heliotrope
Mucosal ulcerations	Poikiloderma
Systemic involvement - Arthritis, serositis, nephritis, seizures/psychosis, cytopenias	Holster sign
Autoantibodies - ANA, anti-dna, anti-sm, SSA, SSB	Confluent scalp involvement
	Systemic involvement - myositis, I.D, malignancy

### DERMATOMYOSITIS: DIFFERENTIAL DIAGNOSIS

- > CLE vs DM: Histopathology does NOT distinguish between the two



### DERMATOMYOSITIS: DIFFERENTIAL DIAGNOSIS

- > Psoriasis vs DM

Nail changes: Pits, Oil spots, onycholysis
Cuticles: normal capillaries
Involvement of gluteal cleft
Occipital scalp and EAC
Koebnerizes
Doesn't usually localize to dorsal hand joints
Sharply demarcated
Facial involvement less common

Image source: VisualDx/visualdx.com

### DERMATOMYOSITIS: DIFFERENTIAL DIAGNOSIS

- > Psoriasis vs DM
  - > Histopathology IS helpful

### DERMATOMYOSITIS: DIFFERENTIAL DIAGNOSIS

- > Multicentric Reticulohistiocytosis vs DM
  - > Rare, systemic non-Langerhans histiocytosis
  - > Symmetric erosive polyarthritis and mucocutaneous nodules
    - > Can also infiltrate other tissues
  - > Approximately 30% associated with internal malignancy
    - > Breast and gastric most common



Bhorat et al. Multicentric Reticulohistiocytosis: A Multicenter Case Series and Review of Literature. J Clin Invest. 2011; 121(1):45-49. Tipton et al. Multicentric reticulohistiocytosis. Clinics Dermatol. 2006; 24:486-492.

### DERMATOMYOSITIS: DIFFERENTIAL DIAGNOSIS

- > MRH
  - > Coral beading on nail folds
  - > Facial papules
  - > X-rays can show punched out erosions/reabsorption of juxta-articular space
    - > Can mimic RA or PsA

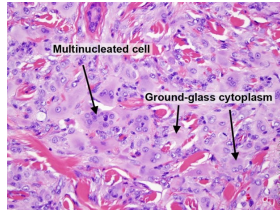


Image source: VisualDx/visualdx.com



## DERMATOMYOSITIS: DIFFERENTIAL DIAGNOSIS

- > MRH vs DM:
  - > Histopathology IS distinctive



- > Clinical DM, but path looks like MRH = MRH with DM-like clinical findings

## DERMATOMYOSITIS: DIFFERENTIAL DIAGNOSIS

### MRH presenting as Dermatomyositis

Case	Author	Sex/Age	Cutaneous features	Joint involvement	Other manifestations
1	Hsiung, JAAD 2003	F/37	Photodistributed erythematous rash	Severe polyarthritis	Raynaud's Periungual telangiectasia
2	Hsiung, JAAD 2003	F/56	Photodistributed erythematous rash	No	Photosensitivity PBC
3	Hsiung, JAAD 2003	F/37	Photodistributed erythematous rash	Arthralgias and arm stiffness	Myalgia
4	McIlwain, J Rheum 2005	F/?	Photodistributed erythematous rash	Symmetric polyarthritis	Proximal muscle weakness
5	Tait, Br J Rheum 1994	M/49	Rash over knuckles, elbows, neck	Erosive polyarthritis	Periungual telangiectasia
6	Munoz-Santos, Derm 2007	F/66	Photodistributed erythematous rash Photosensitivity	Polyarthralgias	Raynaud's Myalgia
7	Fett, Liu, 2011	F/50	Photodistributed erythematous rash Photosensitivity	Polyarthralgias	Periungual telangiectasia Photosensitivity

Munoz-Santos et al. Multicentric Reticulohistiocytosis-Mimicking Dermatomyositis. Dermatology. 2007; 214:268-271.

## DERMATOMYOSITIS: DIFFERENTIAL DIAGNOSIS

- > MRH vs DM
- > Important to distinguish between the two for screening and therapeutic purposes
  - > malignancy screening
  - > MRH
    - > severely deforming erosive arthritis, infiltration of internal organs, muscles, and salivary glands
  - > Dermatomyositis
    - > myositis, ILD, UV minimization

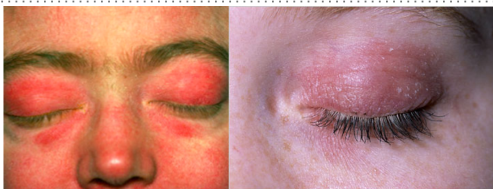
## SORT



Mechanic's Hands	Hand Eczema
Not usually pruritic	Pruritic
Symmetric	Often worse on dominant hand
Ulnar aspect of thumb + radial aspect of fingers (predominance for index/middle)	Lateral aspects of fingers, often palmar involvement. Sometimes dorsal hands involved.
No seasonal variation	Often seasonal variation
No vesicles	Can have vesicles

Schäfers et al. Mechanic's hands revisited: is this sign still useful for diagnosis in patients with lung involvement of collagen vascular diseases? BMC Res Notes. 2014; 7:303.

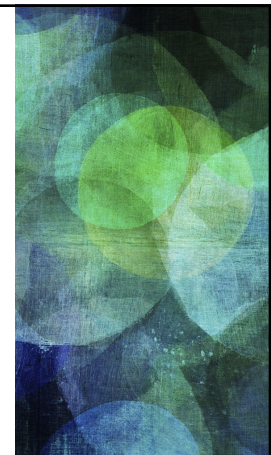
## HELIOTROPE VS EYELID DERMATITIS



Heliotrope rash	Eyelid Dermatitis
Asymptomatic	Pruritic
Violaceous	Erythematous
+/- scale	Usually has scale
	DDx: eczema, allergic/irritant contact dermatitis, seborrheic dermatitis, blepharitis

Image source: VisualDerm.com

## PSORIASIS



### PSORIASIS: PLAQUE PSORIASIS

- > Erythematous, well-demarcated plaques with silvery scale
- > +/- Pruritus
- > Nail pits/oil spots



Image source: VisualDx/visualdx.com

### PSORIASIS: INVERSE

- > Symmetric, shiny, erythematous, thin, sharply demarcated plaques
- > Intertriginous areas: axillae, groin, genitals, inframammary area
- > Due to moisture, the scale is usually not appreciated
- > +/- Pruritus
- > Check nails and all other intertriginous areas as well as typical plaque psoriasis areas (scalp, extensor surfaces, etc.)
- > Present in ~30% of patients who have plaque psoriasis
- > Inguinal folds most common area
- > External genitalia is involved in up to 80% of individuals with inverse psoriasis



Image source: VisualDx/visualdx.com

### PSORIASIS: INVERSE



Image source: VisualDx/visualdx.com

### DDX

- > Acanthosis nigricans
- > Tinea
- > Candidal intertrigo
- > Seborrheic dermatitis

Image source: VisualDx/visualdx.com

### PSORIASIS: PUSTULAR

- > Pustules are sterile
- > Widespread or limited
- > Annular
  - > Pustules at the periphery of erythematous, annular lesions, located most commonly on the trunk
  - > Expand peripherally, with healing occurring in the center
- > Palmar/Plantar
  - > Often on the medial foot and can look like tinea pedis or eczema/dyshidrosis
- > Acrodermatitis continua of Hallopeau
  - > Pustules on the fingertips and nail bed, often with subsequent nail dystrophy and loss



### PSORIASIS: PUSTULAR

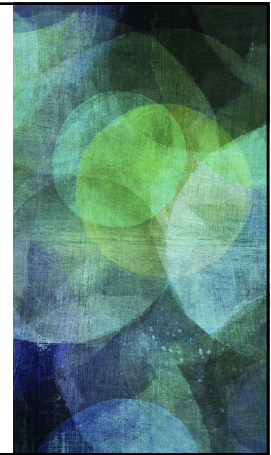


Image source: VisualDx/visualdx.com



Image source: VisualDx/visualdx.com

# RHEUMATOID ARTHRITIS



## RHEUMATOID ARTHRITIS

- > Chronic inflammatory joint and systemic disease affecting 1% of the population
- > Polyarticular arthritis
- > Rheumatoid nodules are the most common cutaneous manifestation
  - > Occur in 20% of patients
  - > Extensor surfaces and pressure areas
- > Rheumatoid vasculitis is rare



Image source: VisualDx/visualdx.com

## RHEUMATOID ARTHRITIS: DDX

- > Is it always a rheumatoid nodule?
- > Granuloma annulare



Image source: VisualDx/visualdx.com

## RHEUMATOID ARTHRITIS: DDX

- > Erythema Elevatum Diutinum (EED)



Image source: VisualDx/visualdx.com

## RHEUMATOID ARTHRITIS: DDX

Rheumatoid Nodules	Granuloma Annulare (subcutaneous)	EED
Usually asymptomatic	Asymptomatic or mildly pruritic	Arthralgias, pain, pruritus
Nodules may ulcerate	No ulceration	Rare ulceration
Extensor surfaces, pressure areas (sacrum), hands, knees, Achilles	Often on lower-upper extremities or acral surfaces	Predominantly over extensor surfaces
Size varies, may reach 5cm	Smaller in RA, larger in PM	Smaller in RA, larger in PM
No mucin on path, granuloma with degenerated CT center	+ Mucin, necrobiotic granulomas	LEV on histopathology
	May spontaneously resolve (~2yrs)	
	Look for other signs of RA (arthritis, smooth patches/plaques)	



Image source: VisualDx/visualdx.com

## RHEUMATOID ARTHRITIS: VASCULITIS

- > Affects 2-3% of patients
- > Small to medium sized vessels (rare cases of large vessel involvement)
- > Purpura, petechiae, ulcers, ischemic lesions
- > EED may be triggered by RA
- > May have associated systemic symptoms
- > Severe cases may have systemic involvement
  - > Heart, lungs, GI, nervous system



Marcucci et al. Extra-articular rheumatoid arthritis. *Rheumatism*. 2019. 79(4):212-224.  
Image source: <https://www.researchgate.net/publication/341111111>

## RHEUMATOID ARTHRITIS: VASCULITIS

- > Look for:
  - > Aeral petechiae or ulcerations, +/- nail fold infarcts
  - > Minute digital ulcerations
  - > Livedo reticularis
  - > Palpable purpura
  - > Vesicles/Bullae



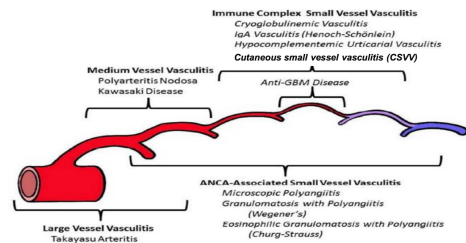
Image source: <https://www.researchgate.net/publication/341111111>

## VASCULITIS

*Work-Up Based on  
Physical Exam Findings*

Adapted from Nicole Fett, MD

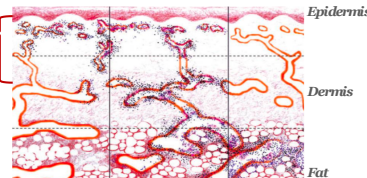
## VASCULITIS: CHAPEL HILL CONSENSUS CONFERENCE



Jennette et al. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis Rheum*. 2013; 65(1):1-11.

## VASCULITIS: CLINICAL PRESENTATION

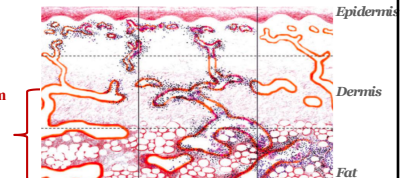
- Urticarial
- Palpable purpura
- Livedo reticularis/livedo racemosa/retiform purpura
- Nodules
- Ulcers
- Infarcts



JA Carlson. The histological assessment of cutaneous vasculitis. *Histopathol*. 2010. 56:3-23

## VASCULITIS: CLINICAL PRESENTATION

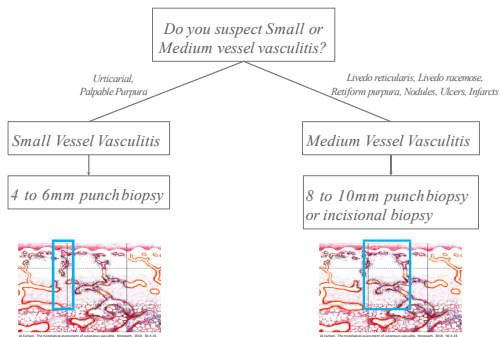
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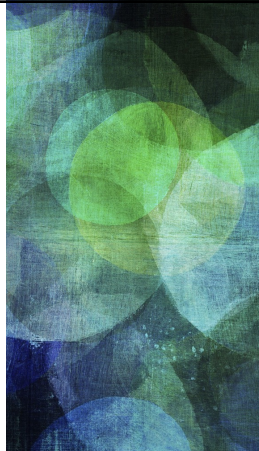
## VASCULITIS: ALGORITHM



## VASCULITIS: BIOPSY BASICS

- > Timeframe
  - > Goal is to get a new lesion (24-48 hrs)
    - > Lesions < 48hrs old
      - > +fibrin in vessel wall
      - > PMNs in vessel wall
      - > Replaced by lymphs and macrophages >48hrs
- > DIF
  - > Lesional biopsy
  - > Early is best (<48 hrs)
    - > DIF can be negative at 72+ hours due to immunocomplex breakdown
- > DIF is useful in assessing presence or absence of immune complexes and predominant immune complex deposition
  - > Knowing which Ig's are involved impacts prognosis and helps narrow ddx
  - > No Ig's increases suspicion of ANCA assoc vasculitis

## DRUG COMPLICATIONS



## DRUG COMPLICATIONS: PLAQUENIL ASSOC HYPERPIGMENTATION

- > Plaquenil Induced Hyperpigmentation
  - > 13% of patients
  - > Typically bilateral and starts ~4 mo after treatment
  - > Improvement with discontinuation, but usually doesn't completely resolve



Tekgor et al. A case of exogenous ochronosis associated with hydroxychloroquine. *Eur J Rheumatol.* 2018; 5(3):206-208.  
Mir et al. Hydroxychloroquine-induced hyperpigmentation. *Dermatol Online J.* 2013; 19(12):20723.

## DRUG COMPLICATIONS: FIXED DRUG REACTION

- > Sharply demarcated round patch recurring at the same body site
- > Typically asymptomatic, but can be pruritic or painful
- > Can occur anywhere, but oral and anogenital most common
- > Common culprits:
  - > Antibiotics (sulfonamides, trimethoprim, fluoroquinolones, tetracyclines)
  - > NSAIDs
  - > OCPs
  - > Cetirizine, hydroxyzine



Image source: VisualDx.com/visuald.com

## DRUG COMPLICATIONS: FIXED DRUG REACTION



Image source: VisualDx.com/visuald.com

### DRUG COMPLICATIONS: TNF- $\alpha$ INDUCED PSORIASIS

- > most common presentation as psoriasis vulgaris, followed by palmoplantar pustular psoriasis and guttate psoriasis
- > Class effect
  - > Switching TNFi will not help
  - > Rechallenge with any TNFi will re-elicite response, often quickly and more severely



Wollina U, Hansel G, Koch A, et al. Tumor necrosis factor- $\alpha$  inhibitor induced psoriasis or psoriasiform exanthema: first 120 cases from the literature including a series of six new patients. *Am J Clin Dermatol*. 2008;9(1):1-14.

Shelling et al. A case of palmoplantar pustulosis induced by certolizumab pegol new anti-TNF- $\alpha$  demonstrates the same class effect. *J Clin Aesthet Dermatol*. 2012;5(5):40-41.

### DRUG COMPLICATIONS: AGEP

- > Acute Generalized Exanthematous Pustulosis (AGEP)
  - > Follows medication, mercury exposure or viral infection (enterovirus, adenovirus, CMV, EBV, HBV)
    - > Within 48 hours to 2 weeks of medication exposure
  - > Fever of  $\sim 39^{\circ}\text{C}$  (can persist for a week)
  - > Burning and pruritus
  - > Nonfollicular pustules become widespread within hours
    - > spares palms/soles
    - > Mucous membranes involved in 20%
  - > Stop drug, resolves in 2 weeks
  - > Watch for hypocalcemia



Image source: VisualDx (www.visualdx.com)

### DRUG COMPLICATIONS: AGEP



Image source: VisualDx (www.visualdx.com)

### REFERENCES

- > Callen and Wortmann. Dermatomyositis. *Clinics in Dermatology*. 2006. 24, 363-373.
- > Dugan E, A Huber, F Miller, L Rider for the International Myositis Assessment and Clinical Studies Group (IMACS). Photobias of the cutaneous manifestations of the idiopathic inflammatory myopathies. *DOJ*. 2009. 15(2): epub.
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- > Marvi et al. Clinical presentation and evaluation of dermatomyositis. *Indian J Dermatol*. 2012. 57(5):375-381.
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- > Sohara et al. Mechanic's hands revisited: is this sign still useful for diagnosis in patients with lung involvement of collagen vascular diseases? *BMC Res Notes*. 2014. 7:303.
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