RhAPP

RHEUMATOLOGY ADVANCED PRACTICE PROVIDERS

RHAPP NATIONAL CONFERENCE

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Rashes and Rheum

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SKIN & CANCER CENTER OF SCOTTSDALE

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Faculty Disclosure

- Advisor, Speaker: Abbvie, Eli Lilly, UCB, Incyte, Amgen, Arcutis, Sun

- Advisor, Speaker, Consultant: BMS

Objectives

- Look at physical exam findings of rashes commonly seen in rheumatology
- Identify “imitator” dermatoses that can present similarly to these rashes
- Compare them to one another and improve ability to differentiate them
  - GOLDEN RULE #1: Though these dermatoses have distinguishing features, it is often HISTORY that will best separate them from one another
  - GOLDEN RULE #2: Never say never, and never say always. (i.e., seldom do these dermatoses follow a specific pattern, 100% of the time)

Cutaneous Lupus

- Three main subtypes:
  1. Acute cutaneous lupus (ACLE)
  2. Subacute cutaneous lupus (SCLE)
  3. Chronic cutaneous lupus (CCLE/Discoid)

*Skin disease is seen in 35-60% of pt's with SLE

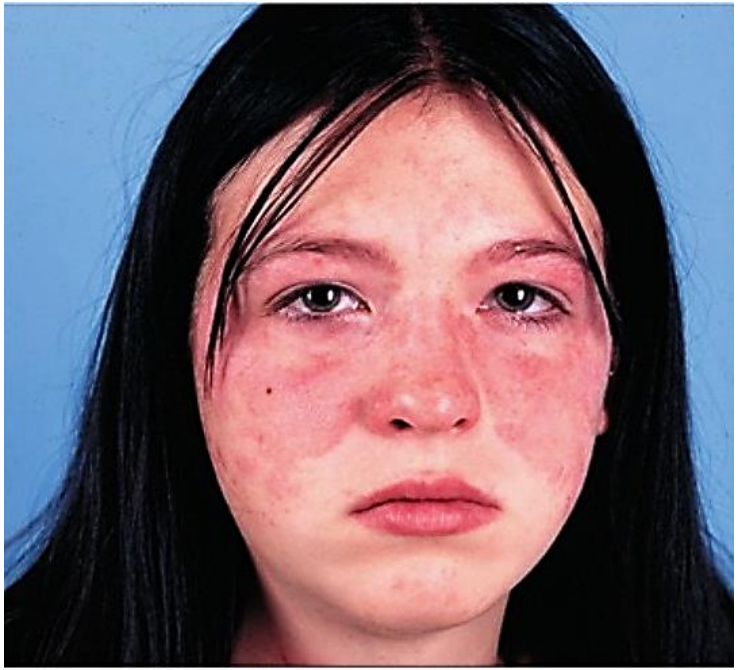
ACLE Most Common Skin Findings:

- Malar rash that is erythematous, and typically not indurated (firm or hard)
- Generalized rash that is papulosquamous

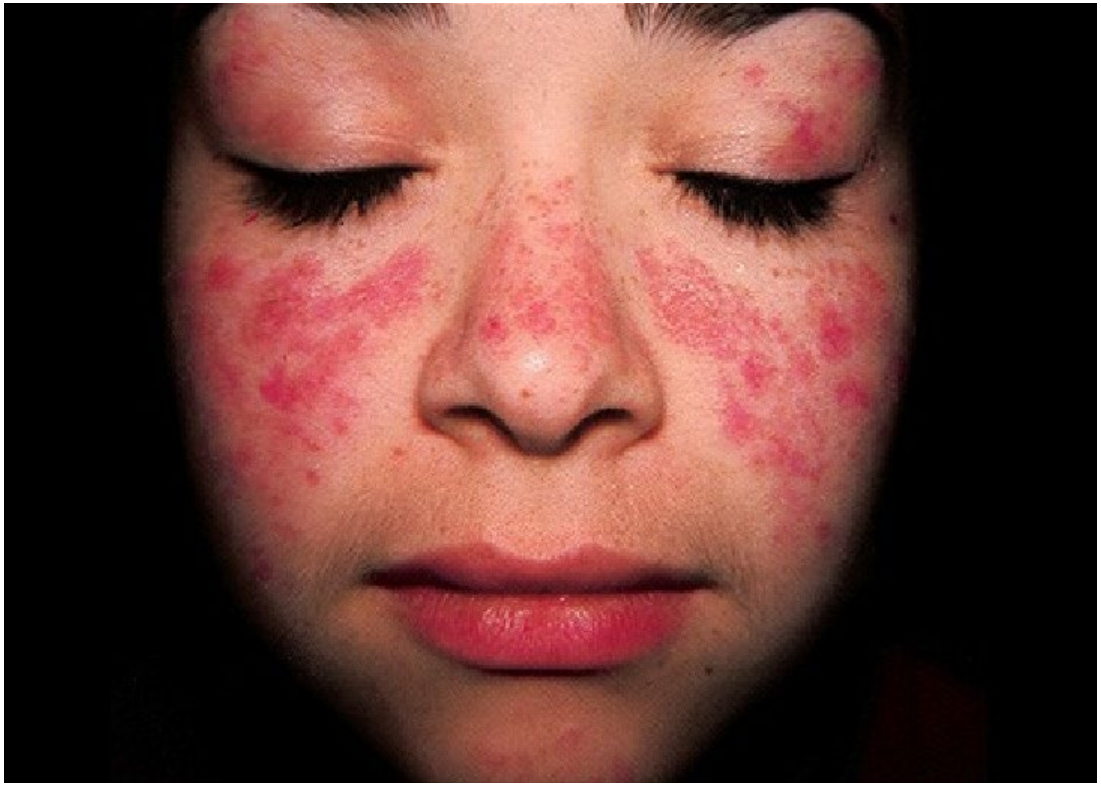
Malar Rash

- Up to 50% of those with SLE will present with this at on time or another
- Typically precipitated or exacerbated by UV light
- May last hours, days, weeks or rarely more prolonged
- ACLE occurs in the presence of flaring SLE and will wax and wane with underlying SLE disease (including nephritis)

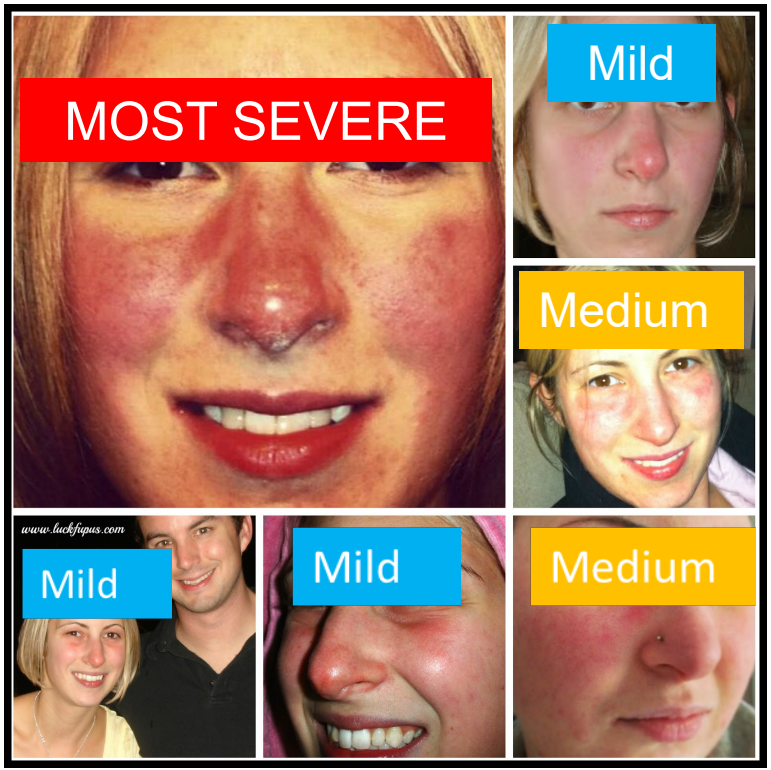
Classic Malar Rash



- Butterfly morphology
- Can be scaly (or not!)
- Red in appearance
- SPARES THE NASOLABIAL FOLD
- May or may not involve the eyelids



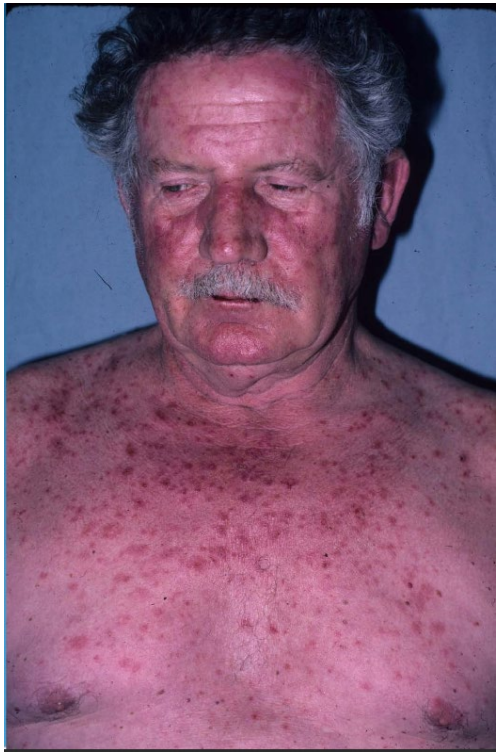
Cunliffe, Dr Tim, et al. (21st April 2015). *Lupus erythematosus*. Retrieved from http://www.pcds.org.uk.



- Severity of systemic disease can be reflected in severity of rash

MarlaJan et al. (September 5, 2013). *Cover up with makeup in the mirror…* Retrieved from http://www.luckfupus.com.

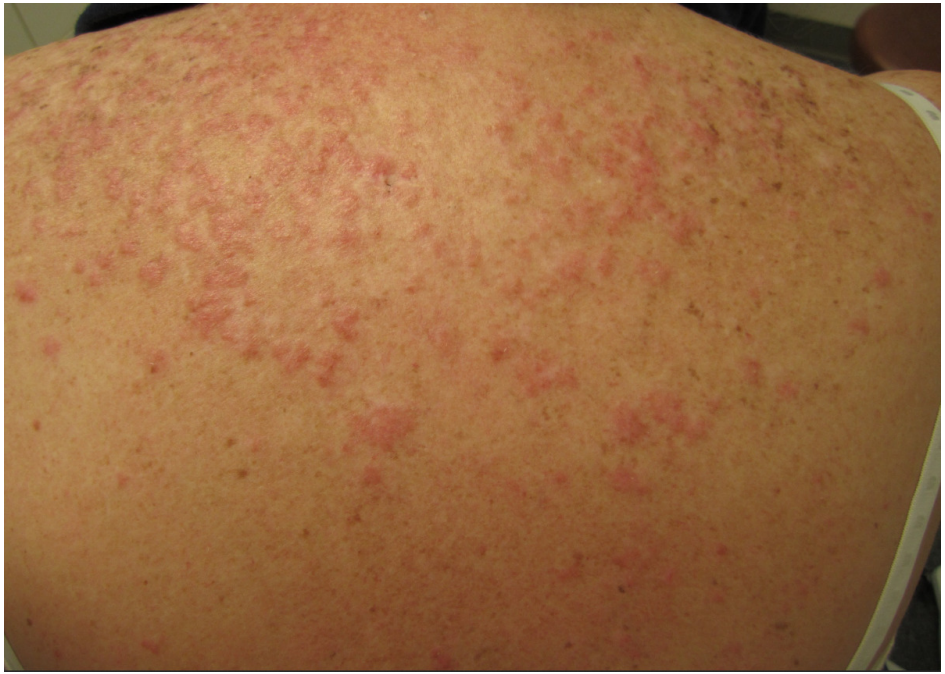
Generalized ACLE



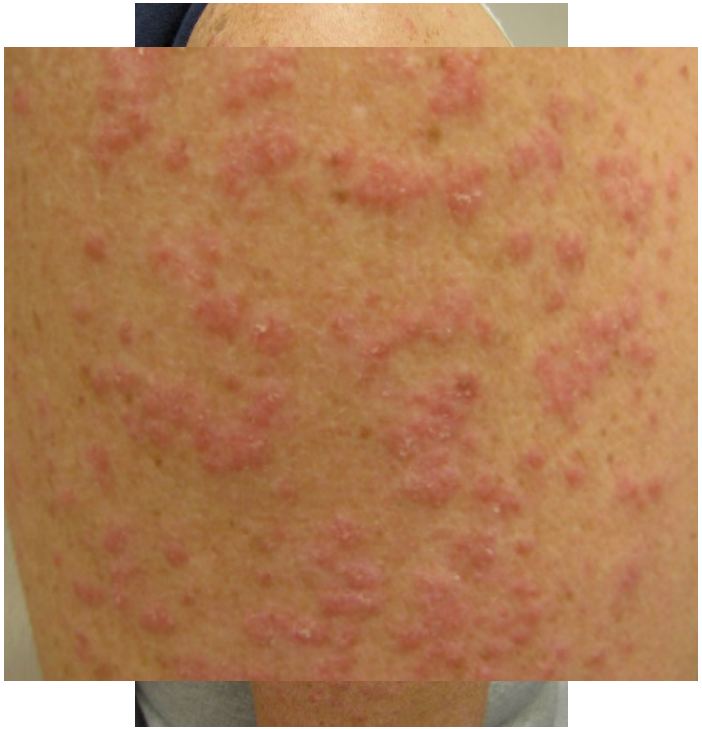
- Morbilliform or exanthematous eruption focuses over the extensor aspects of the arms and hands, sparing the knuckles
- Palms, soles, elbows, knees, or buttocks may be persistently erythematous or purplish, sometimes with overlying scale

Retrieved from http://www.dermpathmd.com/Clinical%20Dermatology/Cutaneous_Lupus_Erythematosus.pdf.

Morbilliform = “Looks Like Measles”

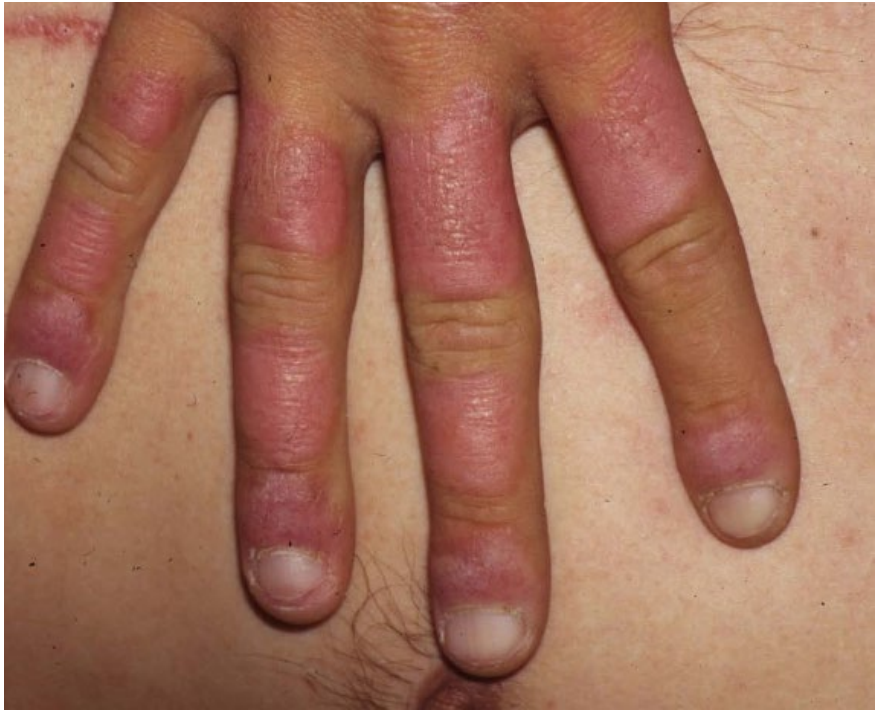


Galen Foulke, MD et al. 2013. *Cutaneous SLICC Classification Criteria for SLE and How to Treat Them* [PowerPoint slides]. Retrieved from http://ssms.weblinkconnect.com/CWT/EXTERNAL/WCPAGES_PRS/PDF/MEETINGS/2017_ANNUAL_MEETING/PDF/FOULKE.PDF

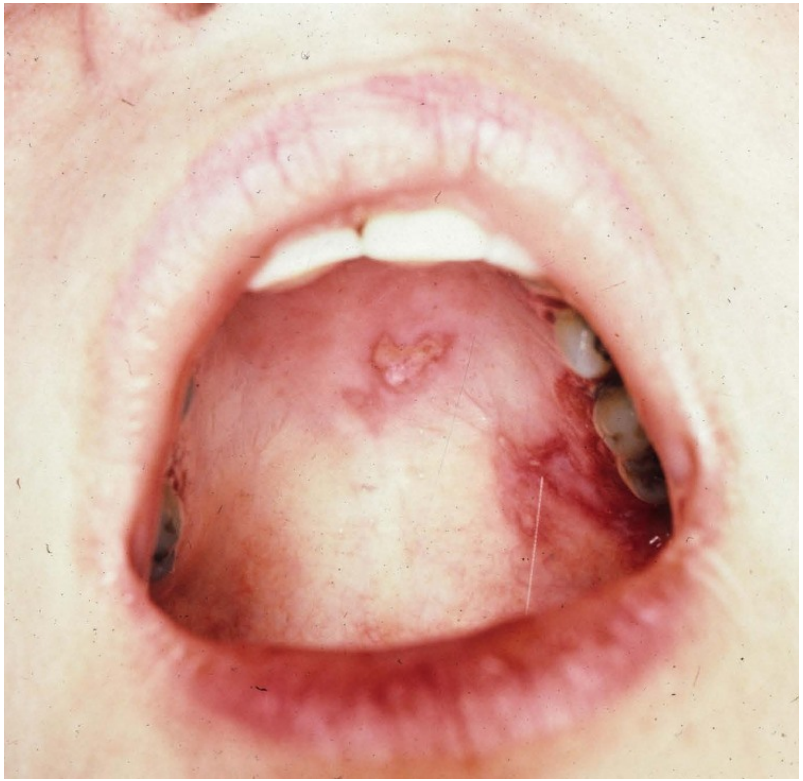


- Can be in a photosensitized pattern
- Papulosquamous = “raised and scaly”
- The scale is typically finer in nature

Galen Foulke, MD et al. 2013. *Cutaneous SLICC Classification Criteria for SLE and How to Treat Them* [PowerPoint slides]. Retrieved from http://ssms.weblinkconnect.com/CWT/EXTERNAL/WCPAGES_PRS/PDF/MEETINGS/2017_ANNUAL_MEETING/PDF/FOULKE.PDF



- Tends to involve extensor surfaces
- SPARES THE KNUCKLES



- Oral ulcerations can be seen in up to 40% of those with SLE (literature widely varies)
- Literature suggests the lesions are typically painless, but mehhhhhhh….
- Can sometimes present as the only skin manifestation
- Can present with DLE as well, but is much more common with ACLE

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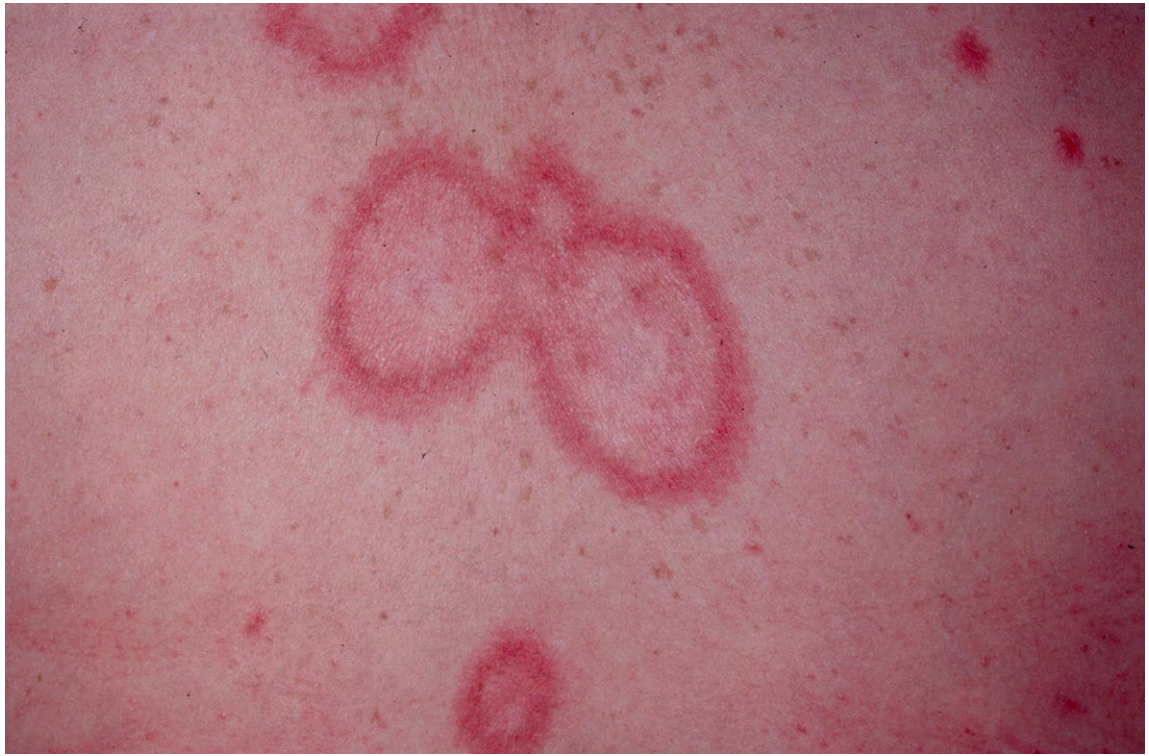
Subacute Cutaneous LE

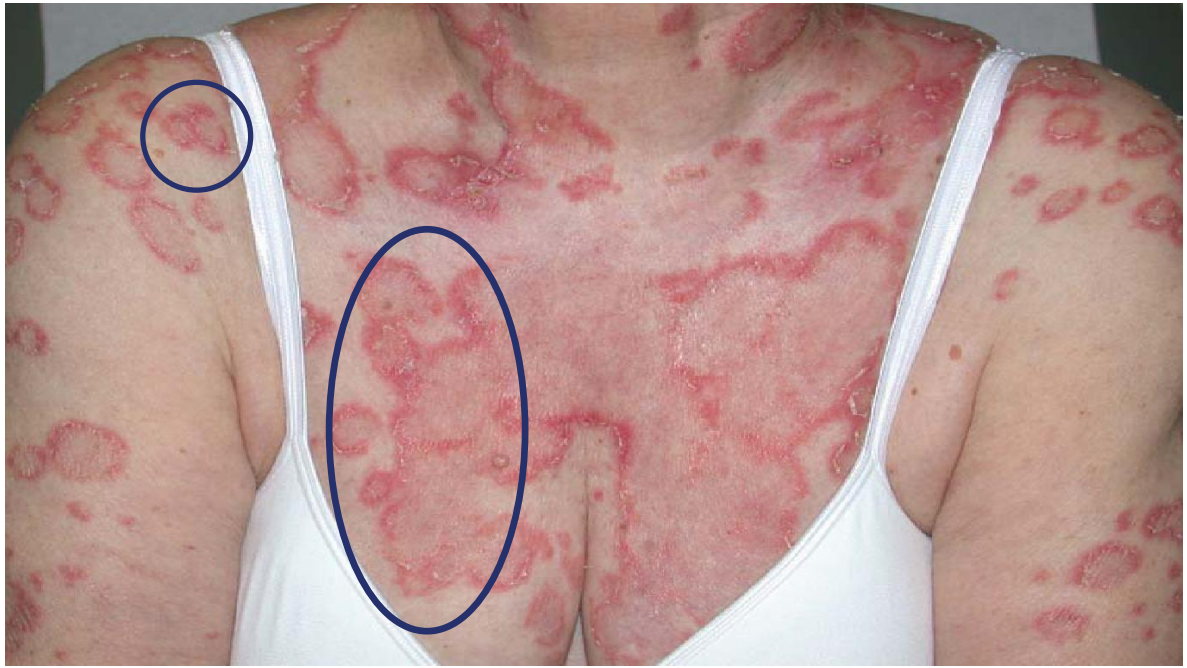
- THERE ARE 2 SUBTYPES: Annular AND psoriasiform (papulosquamous)
- Lesions begin as scaly erythematous macules and/or papules
- Most have antibodies to Ro/SSA antigen (70-90%), but not necessary to make a diagnosis of SCLE
- 30-50% of SCLE patients are (+) for anti-La/SSB. Most are positive for HLA-DR3. 80% have a (+) ANA test (usually in a particulate pattern)
- 50% of patients meet criteria for SLE
- 75% of patients have arthralgia or arthritis, 20% have leukopenia

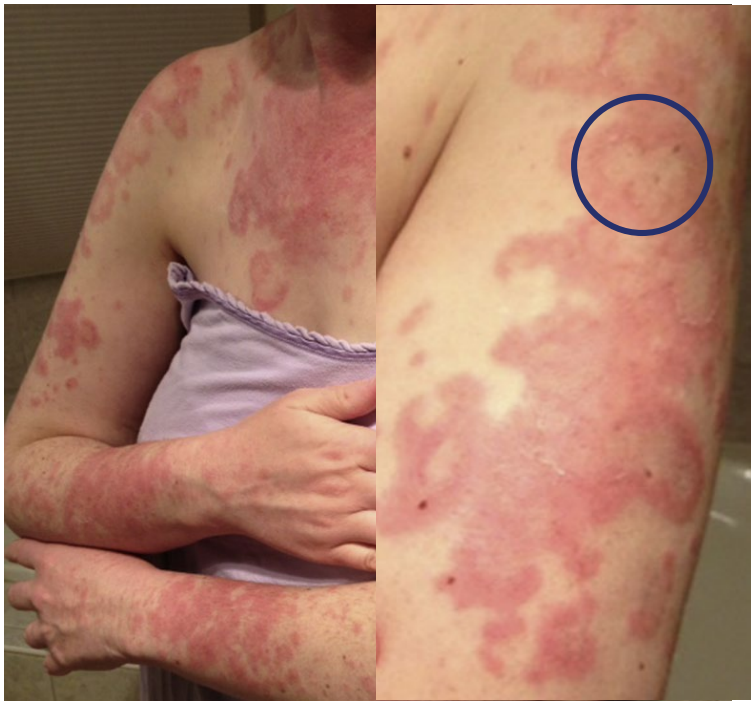
Annular



Lin, Janice et al. (2018). *Subacute cutaneous lupus erythmeatosus* (SCLE). Retrieved from https://emedicine.medscape.com.

Polycyclic = Two or More Connected Rings

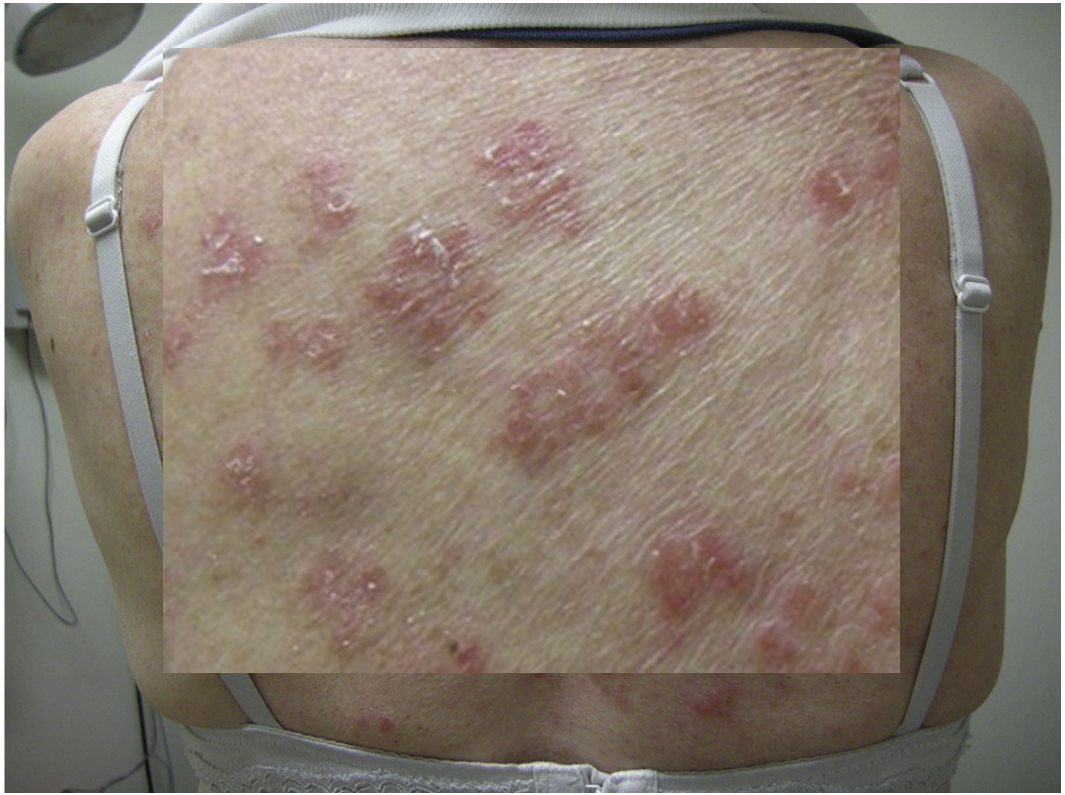




- Sun exposed surfaces and face and neck, the v- portion of chest and back, and sun exposed areas of arms
- Polycyclic lesions
- Follicles not involved

L.G. Okon, V.P. et al. *Werth / Best Practice & Research Clinical Rheumatology*. 27 (2013) 391–404.

Psoriasiform SCLE



L.G. Okon, V.P. et al. *Werth / Best Practice & Research Clinical Rheumatology*. 27 (2013) 391–404.

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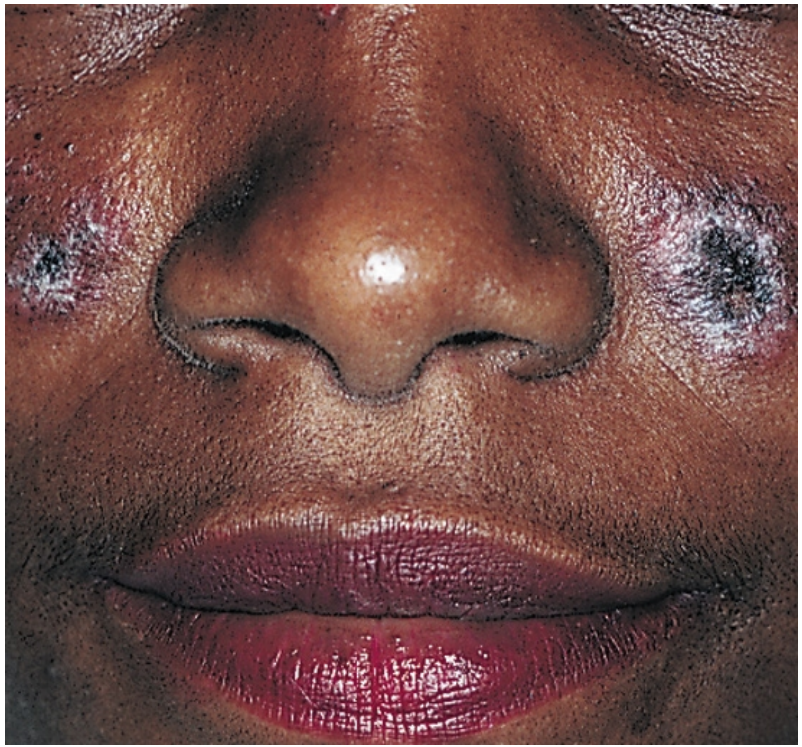
CCLE

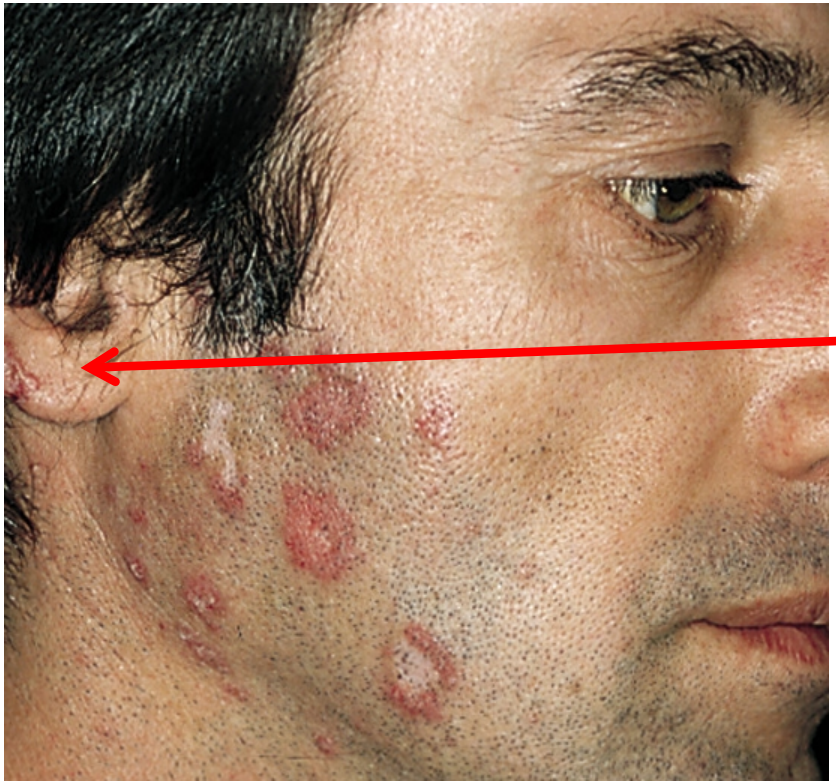
- Most common presentation: Discoid Lupus Erythematosus (DLE)
- Often presents in young adults
- Men more likely to have DLE
- Likely more common in African Americans
- Age of onset usually between 20-40 years of age
- Often occurs in the absence of SLE
- ACLE in general is 7x more common than DLE
- Present in 15-30% of SLE population

Discoid Lupus Erythematosus

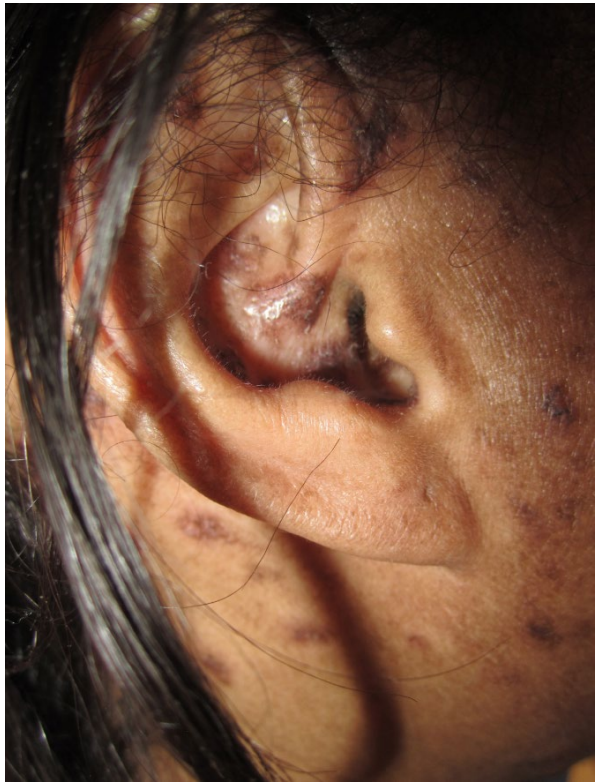
- Majority are Localized to head, above neck
- There is a generalized form; by definition can include lesions both above and below the neck
- it is potentiated by sunlight, but not as much as with ACLE or SCLE



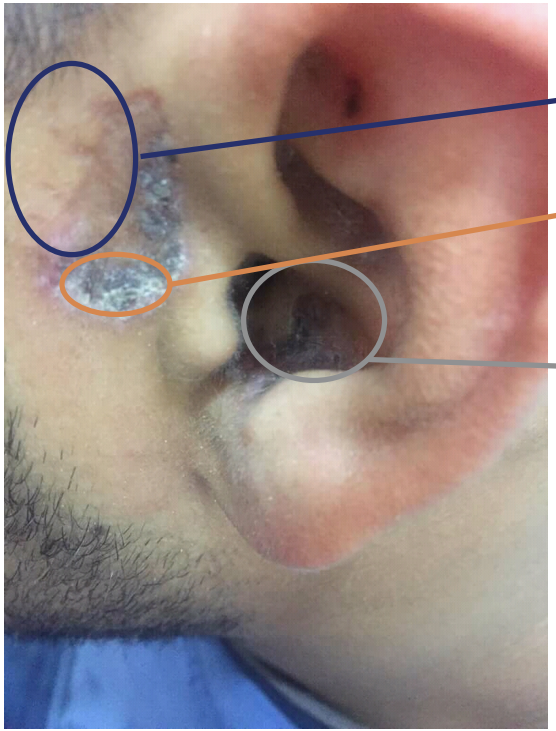
- Discoid = shaped like a disc
- Tend to be pigmented
- Over time, become scarred and atrophic and then actually become hypopigmented



- Notice the atrophy and pigment loss in the beard line lesions
- And wait, what is that on his ear

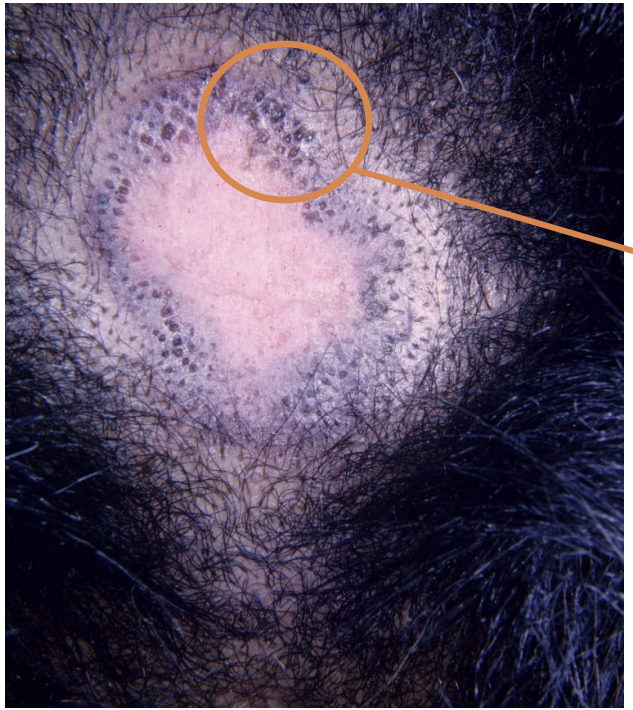


- The ears are often involved. (if you see any lupus like lesion with ear involvement, think DLE)
- If it involves the conchae, it is known as “Shuster’s sign.”



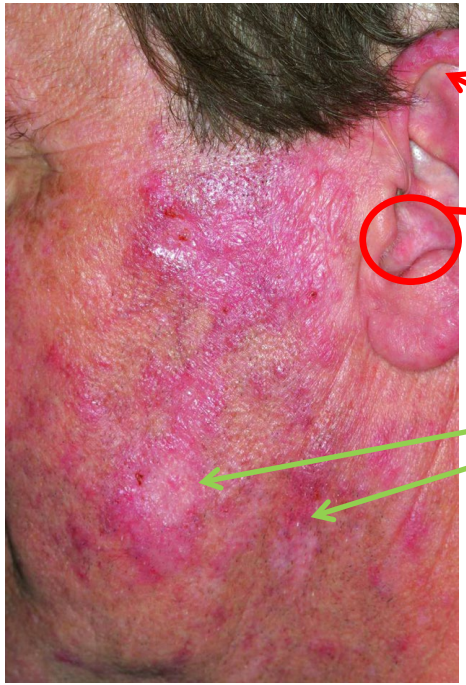
- Atrophic changes
- Hyperpigmented, scaly plaques
- Shuster sign (involves the conchae)

Mohammad et al. 2018. (2018). Retrieved from www.wikimedia.org.



- Scalp lesions are common (if you see any lupus like lesion with scalp involvement, think DLE)
- Hair follicles are involved and become indurated, and have a “plugged” appearance (happens in scalp and skin lesions)
- Can lead to scarring alopecia and permanent hair loss

http://www.dermpathmd.com/Clinical%20Dermatology/Cutaneous_Lupus_Erythematosus.pdf.



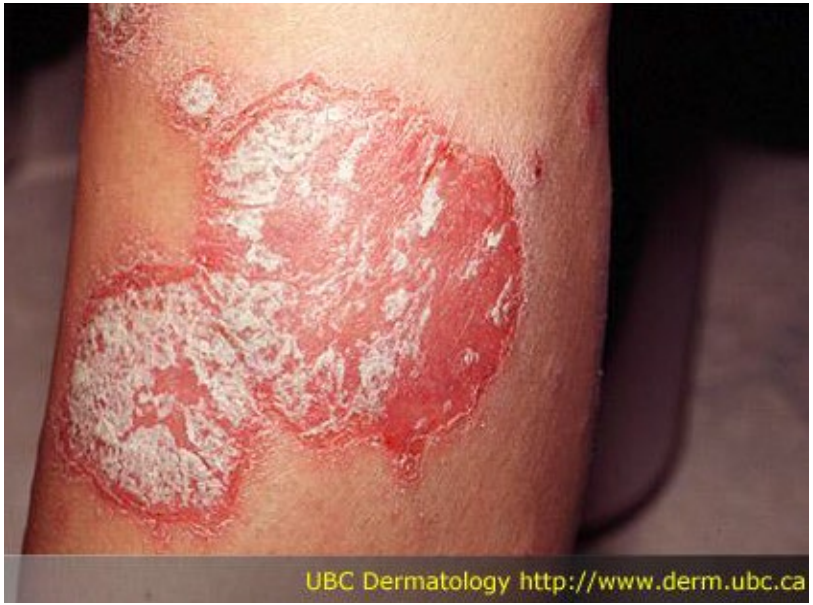
- Though this is less “disc” like in appearance, notice:
  1. The distinct involvement of the ear and conchae
  2. The atrophic changes in the distal aspect of the cheek and lateral neck
  3. The lateral face is most likely to be affected.

Alana M. Nevares, MD (2018). Systemic Lupus Erythematosus (SLE)(Disseminated Lupus Erythematosus). Retrieved from www.Merckmanuals.com.

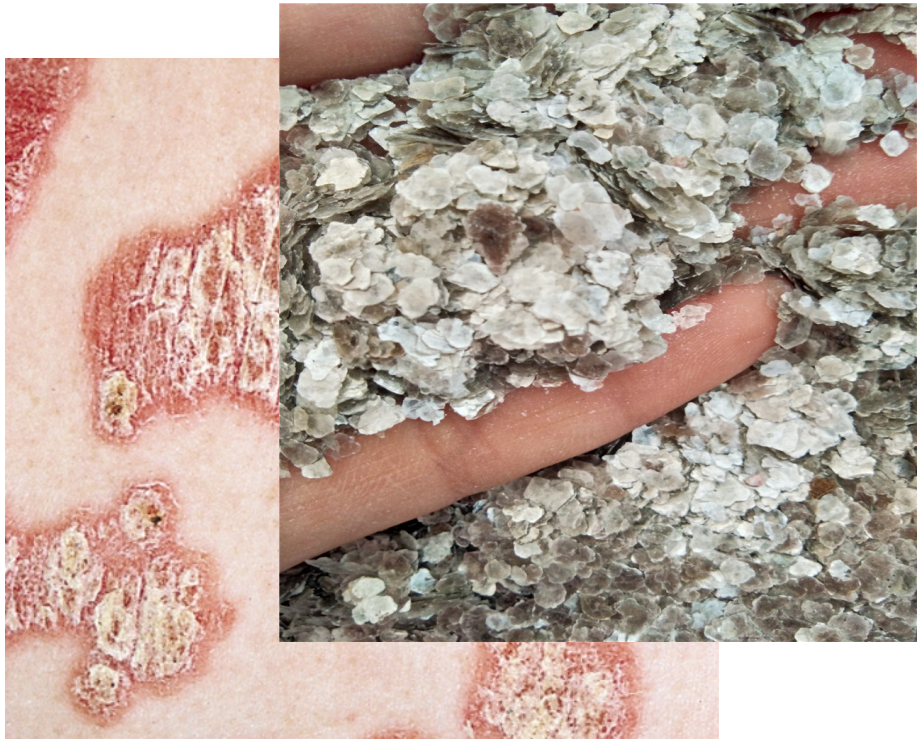
Differentiating SCLE/ACLE/CCLE (DLE)

- ACLE lesions more transient usually, and heal with less pigmentary change (vs. DLE which readily scars)
- ACLE usually malar. SCLE usually involves neck/ shoulders, dorsal UE and trunk, or lateral face.
- DLE more severe than either SCLE or ACLE, in terms of long term skin sequelae (greater hyper or hypo pigmentation, scarring, follicular plugging and scale with alopecia)
- DLE is typically indurated, ACLE and SCLE are typically not
- SCLE patients are typically “sick” vs. the ACLE and DLE

Psoriasis



- Silvery scaly
- Erythematous
- Sharply demarcated
- Classically involves scalp elbows, knees, low back

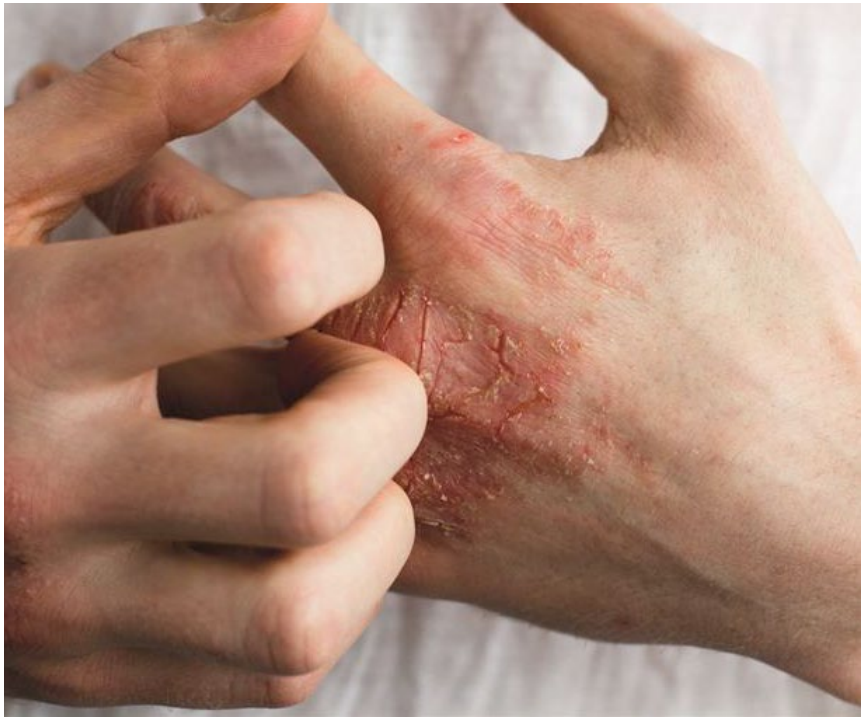


- Micaceous scale = shiny, rock-like scale.

Philip E. LeBoit, MD, Brian Hinds, MD, et al. *Lupus Erythematosus*. 2018. Retrieved from https://www.derm101.com/clinical-atlas/lupus-erythematosus.

Atopic Eczema

- Literal translation “thrown out by heat.”
- In dermatology, it refers to a collection of rashes that present with scale.
- For this talk, we will focus on atopic eczema

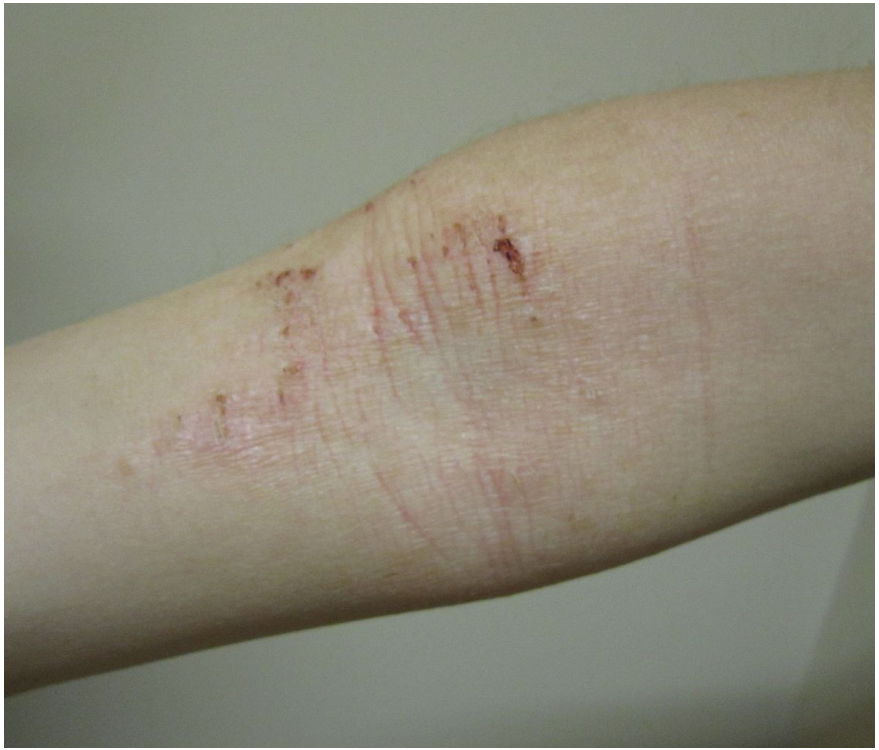


- Dry
- Scaly
- Erythematous
- Fissures
- Excoriations
- Hyperlinearity of skin lines
- Itch/ scratch cycle
- Atopic triad: Asthma, allergies (hay fever), eczema
- Strongly familial

Sparks, Dana et al. *Home Remedies: The itchy irritation of eczema*. 2018. Retrieved fromhttps://newsnetwork.mayoclinic.org.

Hyperlinearity of Lesional AND Non-Lesional Skin

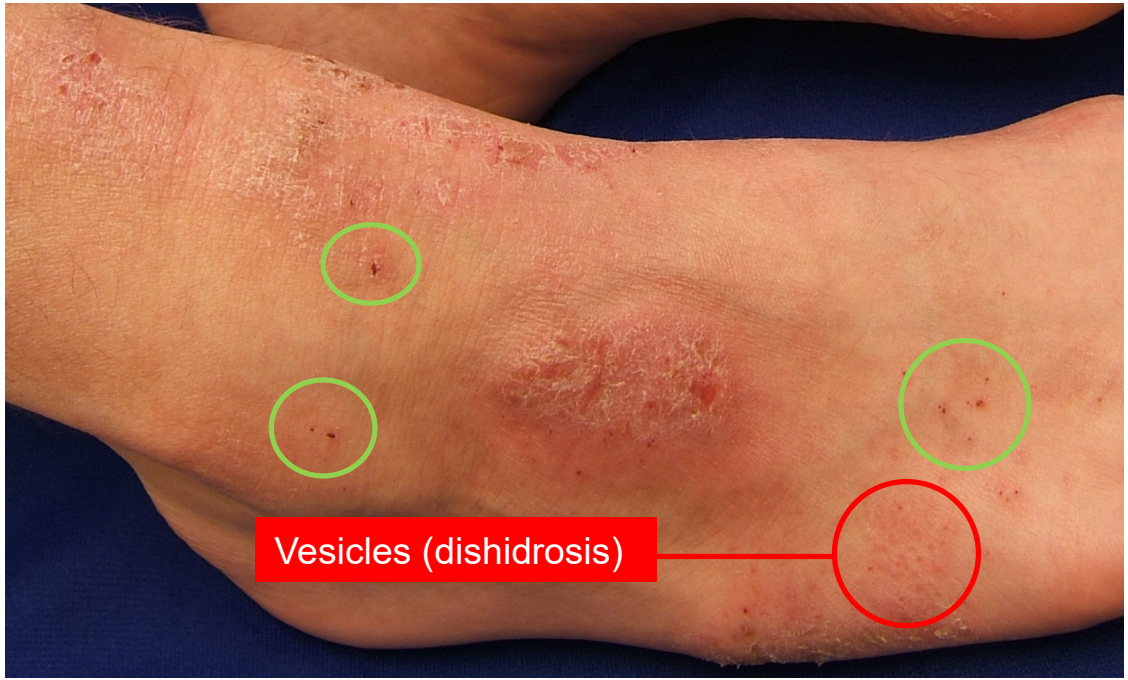
Sasha Fox Walters et al. (2015). Retrieved from www.istockphoto.com.



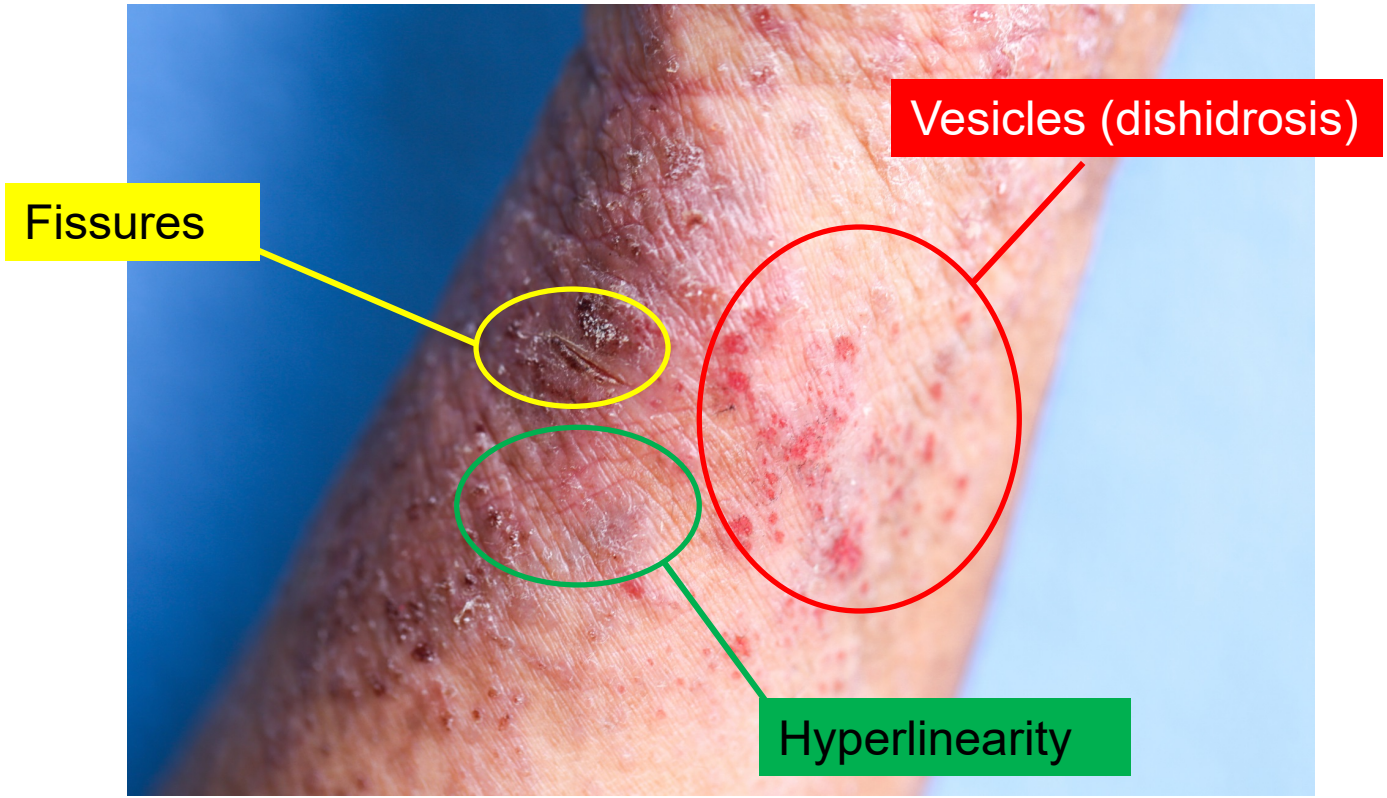
- Patterns change with time, (in the flexures for children to early teens) but overall the lesions have similar characteristics
- Dry
- Scaly
- Erythematous
- Excoriations
- Hyper-linear

Atopic Dermatitis. (2005). Retrieved from https://en.wikipedia.org/w/index.php?title=Atopic_dermatitis.

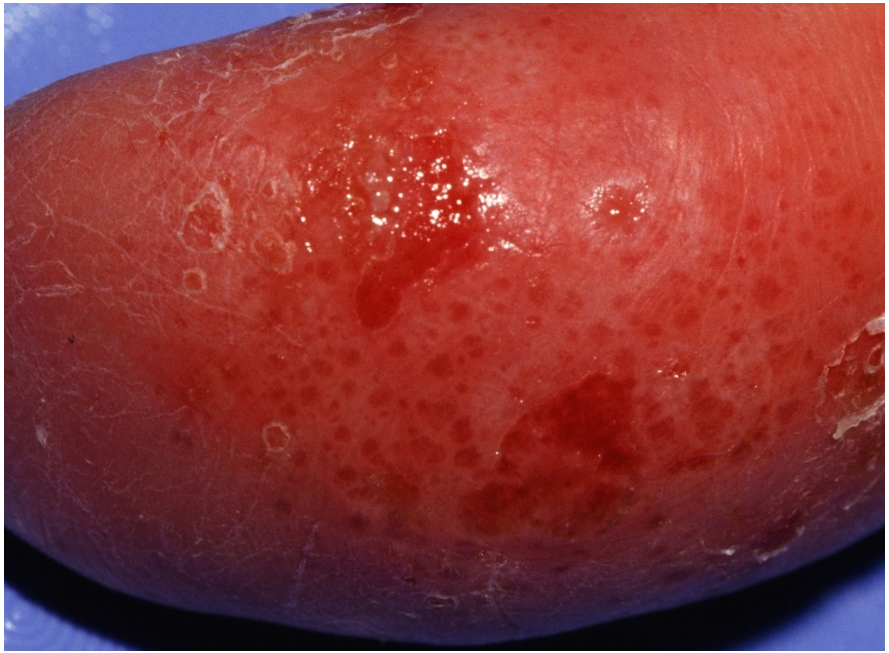
Excoriations Are Common



What is Eczema? Retrieved from https://nationaleczema.org/eczema.


Vesicles (dishidrosis)
Fissures
Hyperlinearity

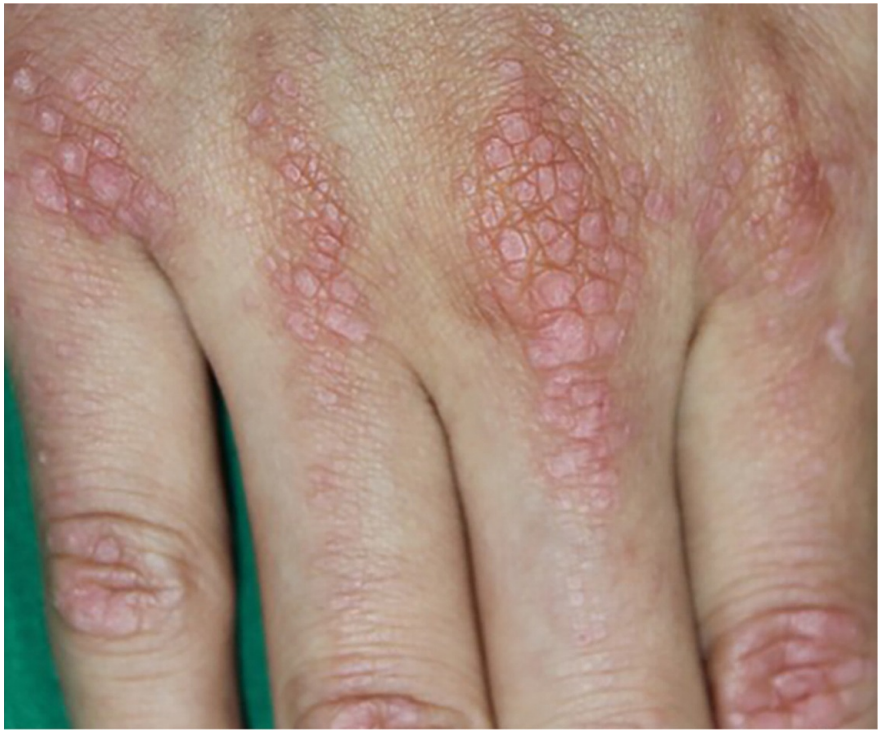
Pompholyx (Dishidrotic Eczema)



- If you see vesicles amongst a scaly rash, especially on the lateral fingers, lateral hands, and/or the lateral feet think eczema
- Often called “tapioca-like” in appearance

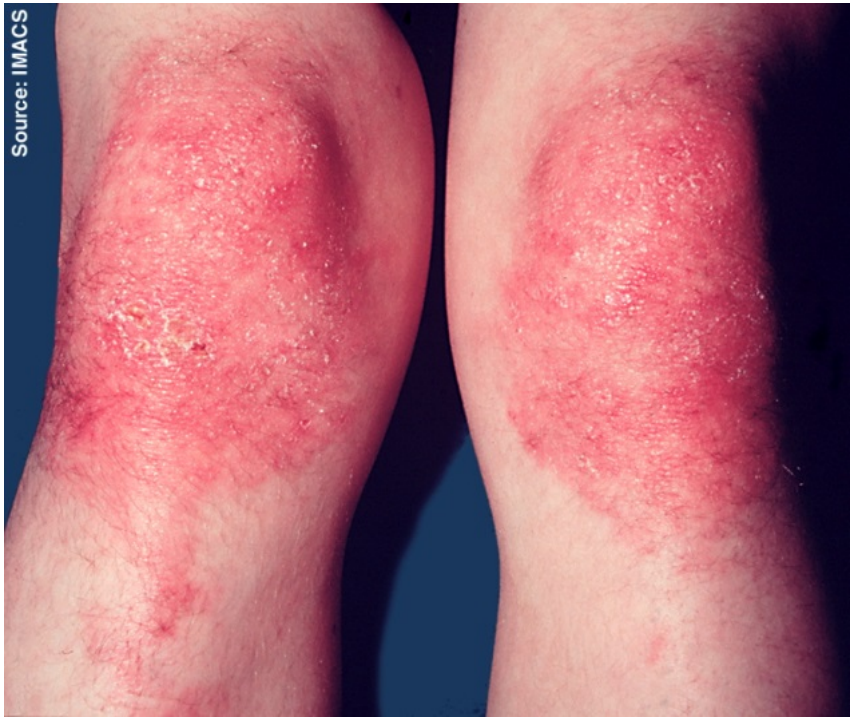
Dermatomyositis

- Rash usually precedes systemic symptoms.
- Reddish or bluish-purple patches mostly affect sun-exposed areas.
- A violaceous rash may also affect cheeks, nose, shoulders, upper chest and elbows.
- Up to 30% of those with cutaneous findings are without muscular symptoms
- Calcinosis cutis is more common in children than adults, and is rare in adults.



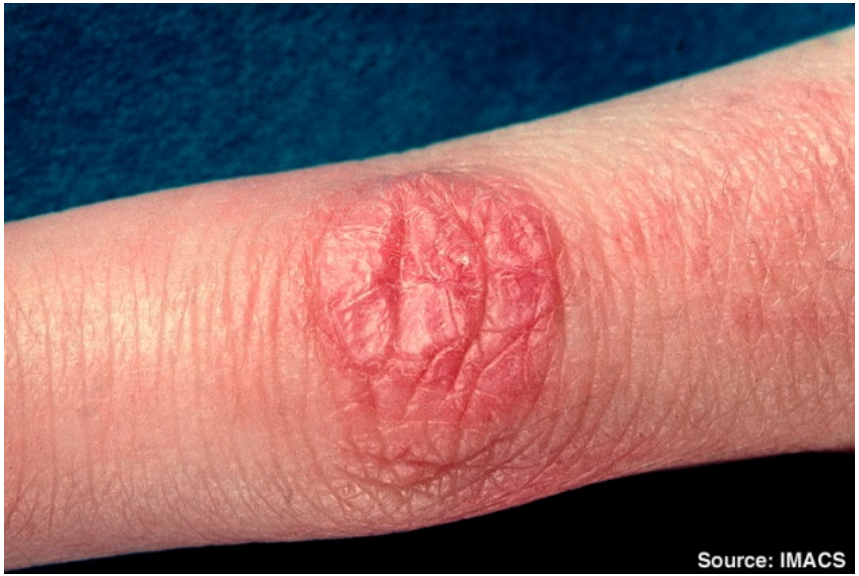
- GOTTRON’S PAPULES
- Flat topped erythematous papules

HEI SUNG KIM, MD, PhD; SO MIN KIM, MD; and JEONG DEUK LEE, MD, PhD, et al. “Erythematous Papules on Dorsum of Both Hands.” *Am Fam Physician*. 2017 Jun 15;95(12):803-804.



- Gottron’s sign: Confluent erythema overlying extensor joints
- (Versus Gottron’s papules)

Elizabeth M. Dugan, Adam M. Huber, Frederick W. Miller, Lisa G. Rider, et al. Retreived from http://dermatology.cdlib.org/1502/reviews/photoessay/12.jpg



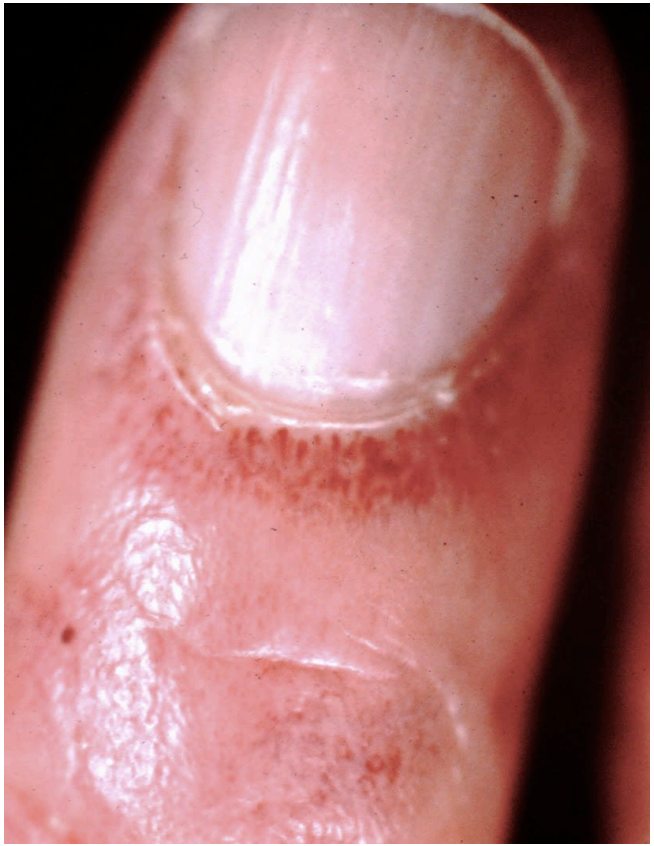
- Very little induration (tend to be flat and soft) in typical lesions, but they can become indurated with chronicity

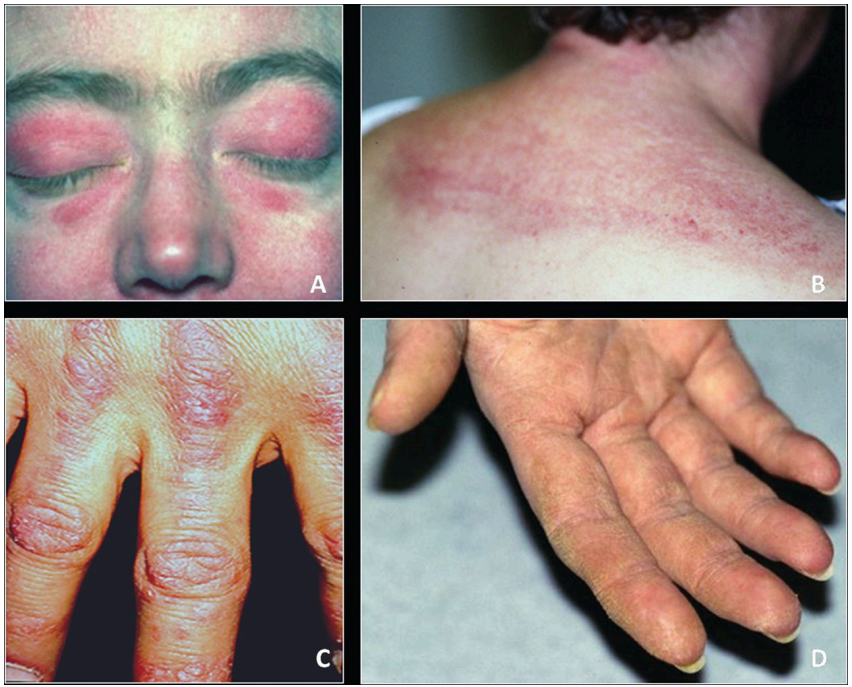
Heliotrope: A Violet Rash With or Without Edema of the Periorbits

By Photo (c)2006 Derek Ramsey (Ram-Man) - Self-photographed, CC BY-SA 2.5, https://commons.wikimedia.org/w/index.php?curid=1158053

Foulke G, Baccon J, Marks JG, Clarke JT, et al. Antimalarial Myopathy in Amyopathic Dermatomyositis. *Arch Dermatol*. 2012;148(9):1100–1101. doi:10.1001/archdermatol.2012.1711

Capillary Loops/Telangiectasia





A. Heliotrope
B. Shawl sign
C. Gottron’s
D. Mechanic’s hands

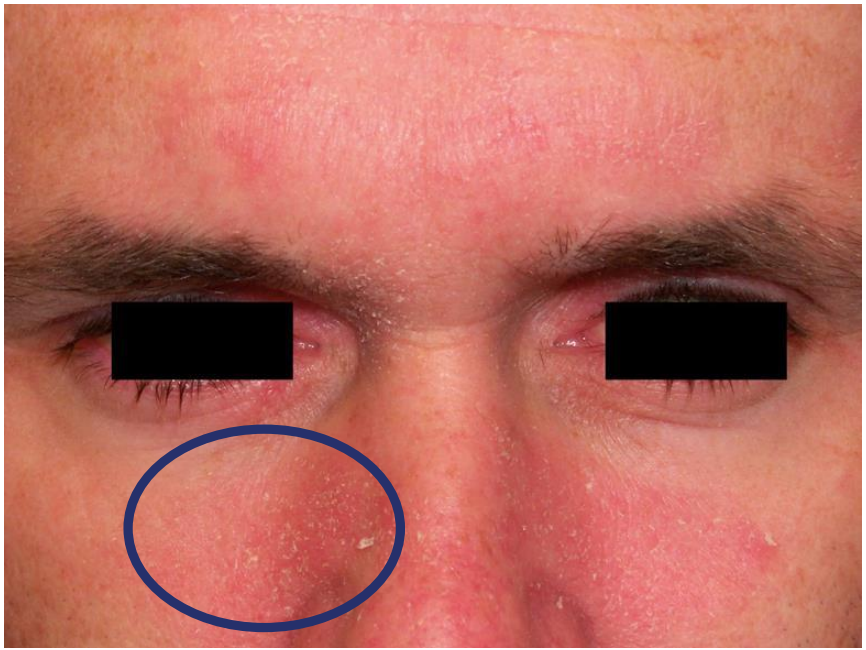
Ilias N. Lazarou and Pierre-André Guerne, et al. *The Journal of Rheumatology*. May 2013, 40 (5) 550-564;
DOI: https://doi.org/10.3899/jrheum.

Imitators

Seborrheic Dermatitis

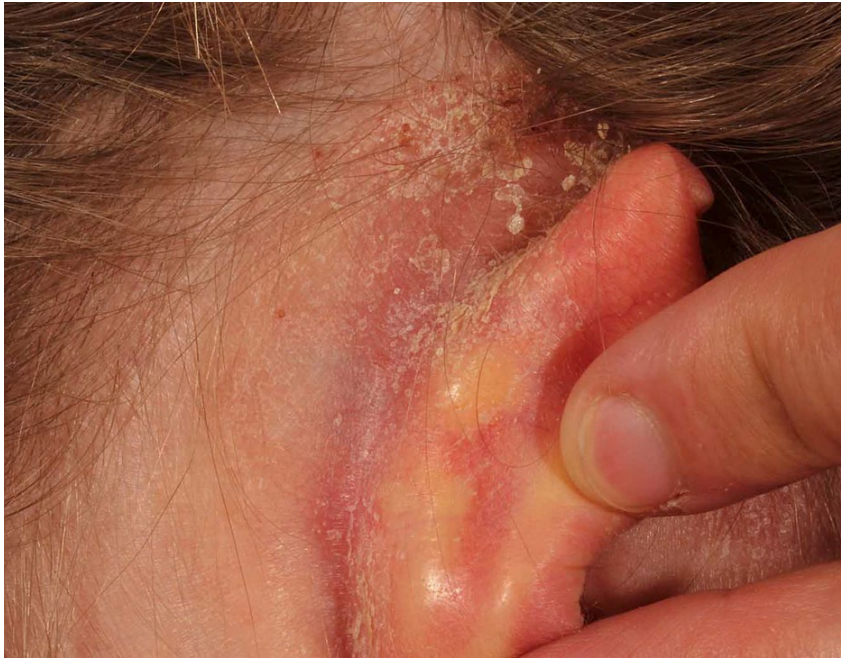
- Greasy, erythematous, loose scale (Due to the inherent nature for oil to build up, gather into flakey patches, and then cause a hypersensitivity reaction to all the buildup).
- Concentrates on the midface, and tends to be heaviest in folds and hair-bearing areas
- Often involves:
  - Midface
  - Eyebrows
  - Ears
  - Scalp
  - Beard/ mustache

FOR ALL INTENT AND PURPOSES, IT'S BASCIALLY DANDRUFF!



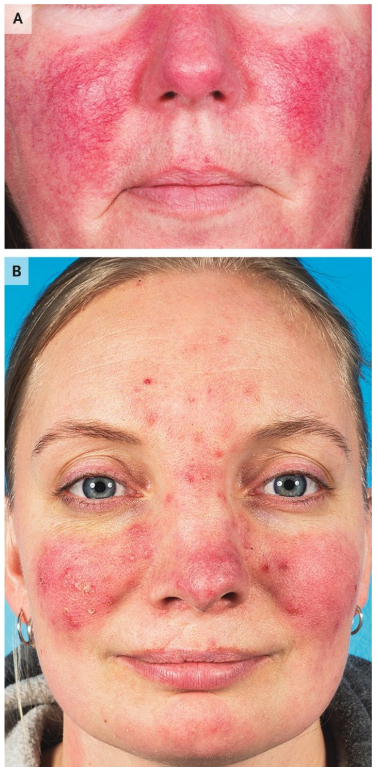
- Loose, greasy, scale
- Involves eyebrows and midface

Amanda Oakley et al. (1997). Retrieved form www.dermnetnz.com.



- Loose, easily removed scale that feels greasy, and is heavy in the folds.

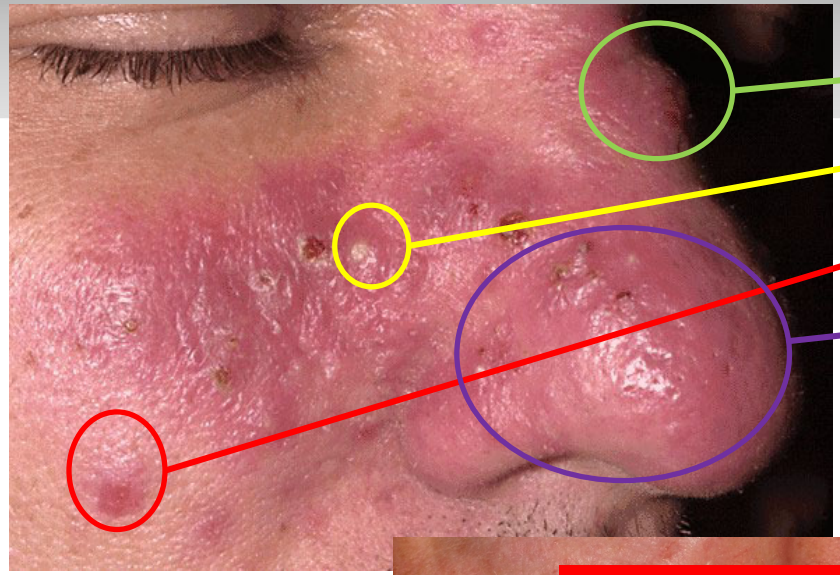
Rosacea



- Varying degrees of background erythema, most often of the midface
- TELANGIECTASIA

- Papules and/or pustules and/or nodules
- Adult women= most common demographic
- Exacerbated by anything that causes vasodilation (alcohol, heat, exercise, emotion, etc.)

Carla Rothaus et al. (2017). *Rosacea*. Retrieved from https://resident360.nejm.org

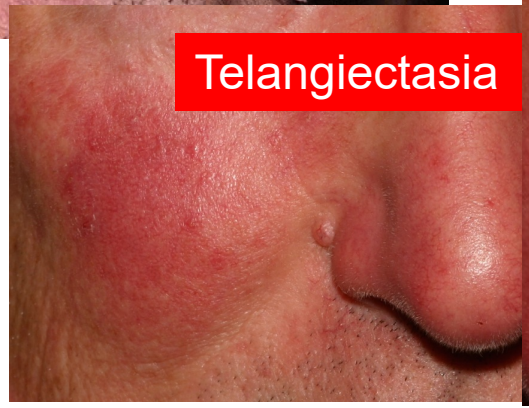


Nodules and/or

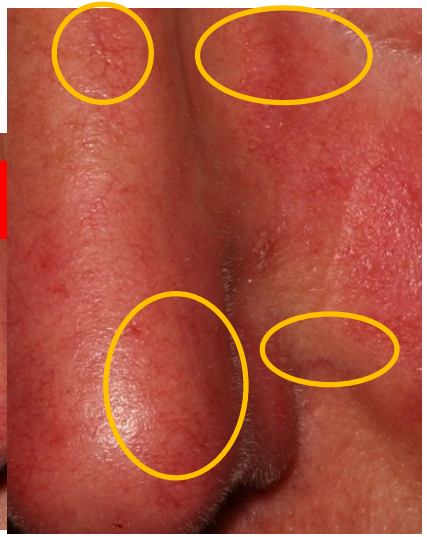
Pustules and/or

Papules

Rhinophyma
(bulbous nose)

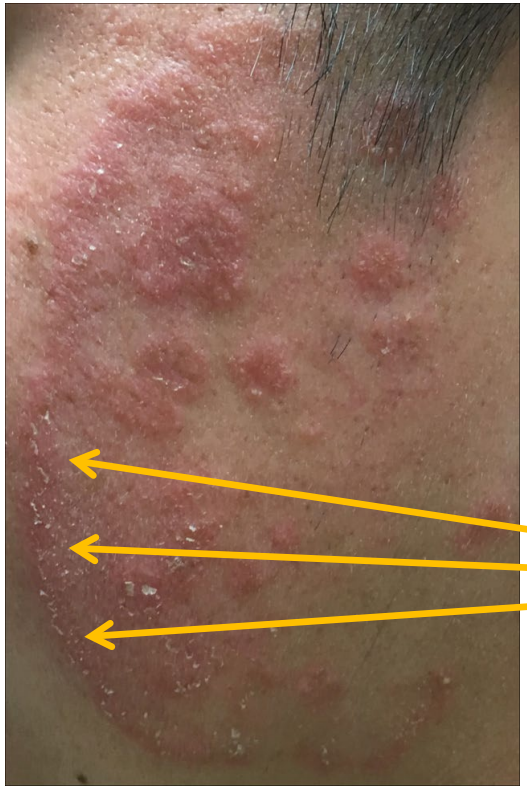


Telangiectasia

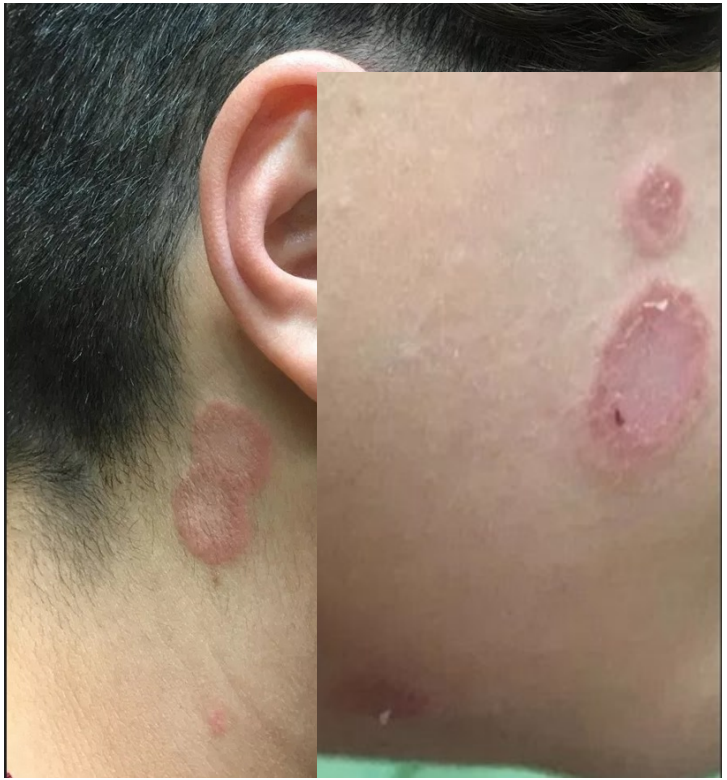


Health Jade team. *Rosacea*. Retrieved from https://healthjade.com/rosacea/.

Tinea



- Caused by a number of dermatophytes; exposures from pets (especially cats), contact with those already infected, fomites.
- Most distinguishing features are:
  - Arciform or nummular (coin-shaped) formation
  - Raised edges that can appear serpiginous (like a snake)
  - An edge of leading scale that appears to be “pushing forward”



- Classic central clearing with raised scaly border, and a leading edge of scale

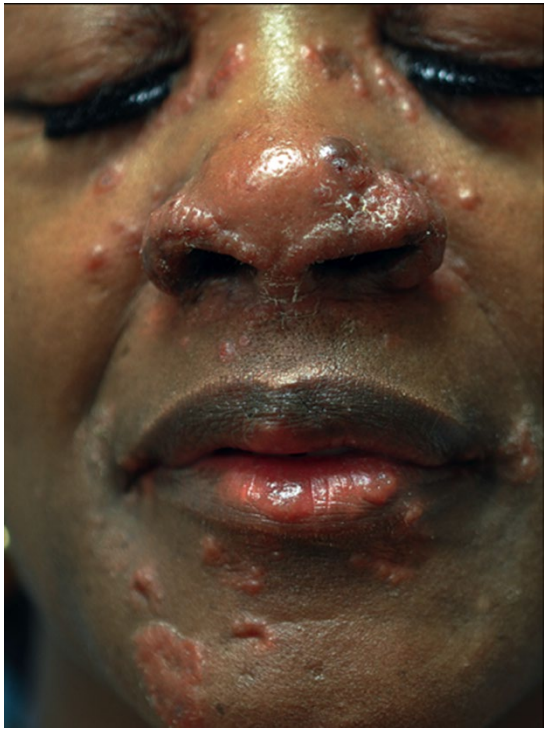
Retrieved from https://www.timeofcare.com/tinea-corporis/.

Sarcoid

"The only thing I know is, that I don't know."

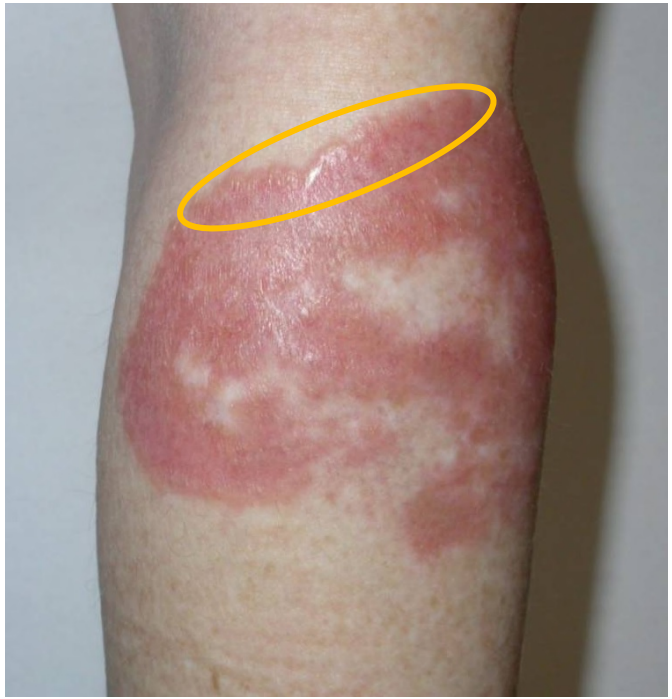
– Rob Casquejo

- In terms of morphology, anything goes!
  - Can be papular, nodular, targetoid, subcutaneous. The most common skin lesions are of the papular variety, on the face.
- Typical lesions are red to brown, tender, and often smooth in appearance
- This is a scenario in which history can be an invaluable tool.
  - Women > men
  - Predominantly those of African ethnicities
  - ROS: is crucial to weed out systemic disease- lung especially, constitutional, cardiac, musculoskeletal, etc.
    - But up to 1/3 of patients with skin disease will never manifest with systemic disease.

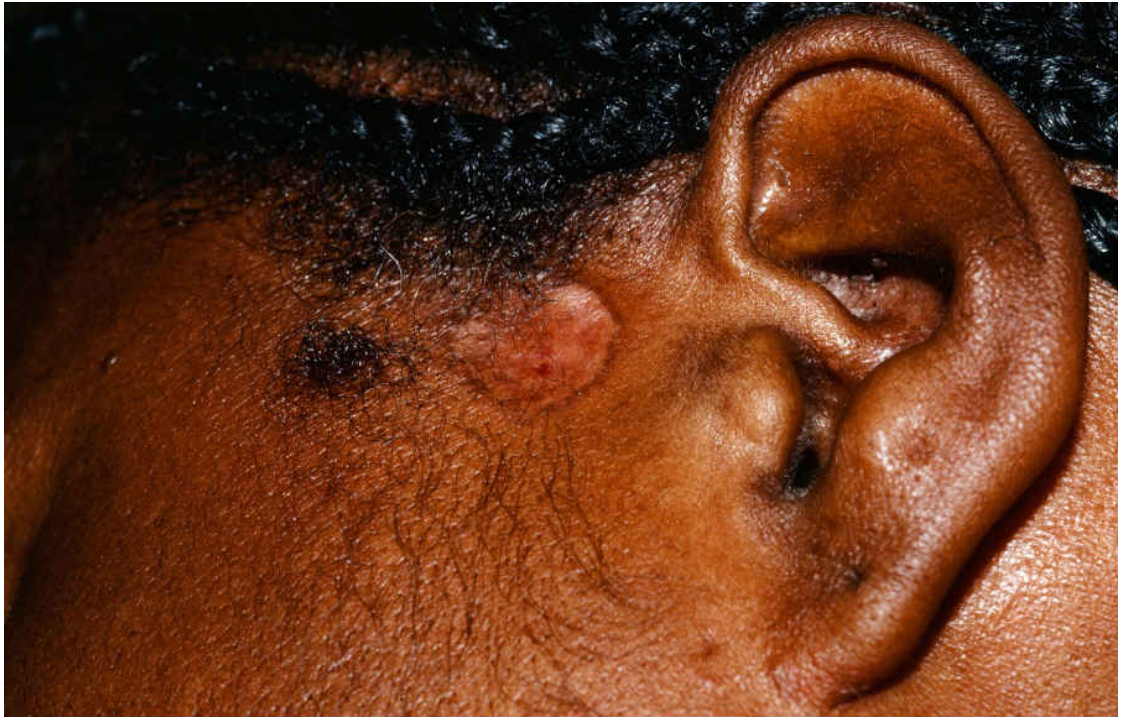


- African ethicnity
- Red to brown papules
- Facial lesions often involve cheeks, nose, peri-ocular and lips

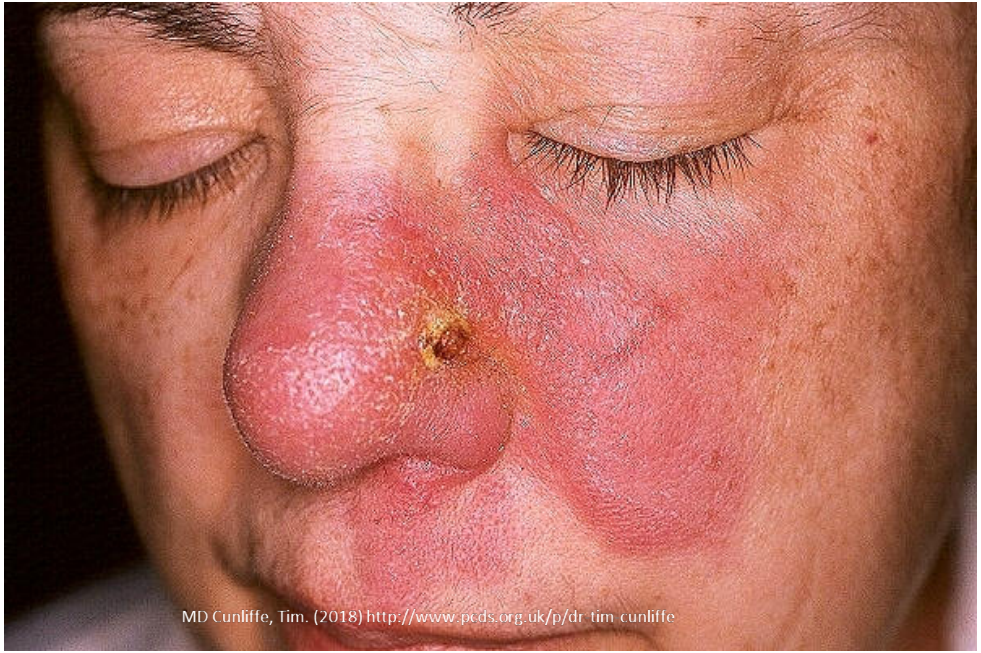
Wanat KA, Rosenbach M, et al. Case Series Demonstrating Improvement in Chronic Cutaneous Sarcoidosis Following Treatment With TNF Inhibitors. *Arch Dermatol*. 2012;148(9):1097–1100.



Cunliffe, Tim et al. (2018) Retrived from http://www.pcds.org.uk/clinical-guidance/sarcoid.

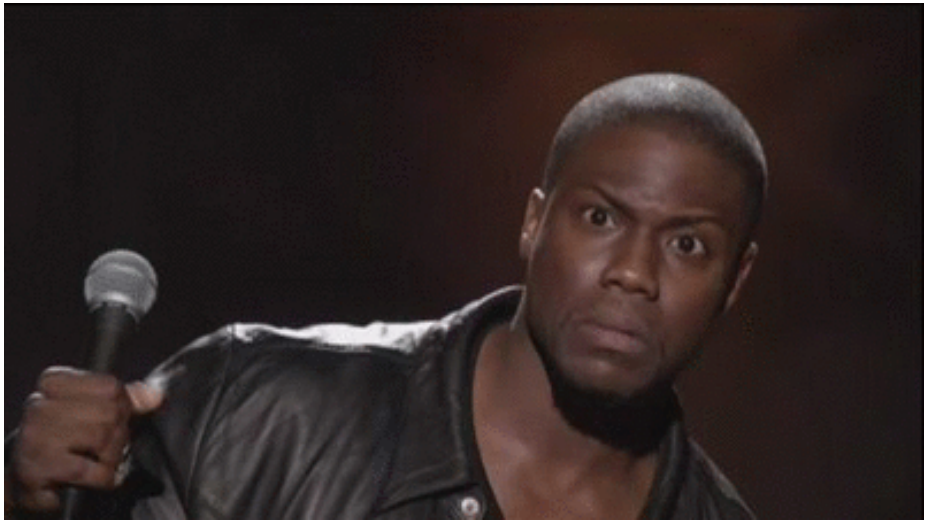


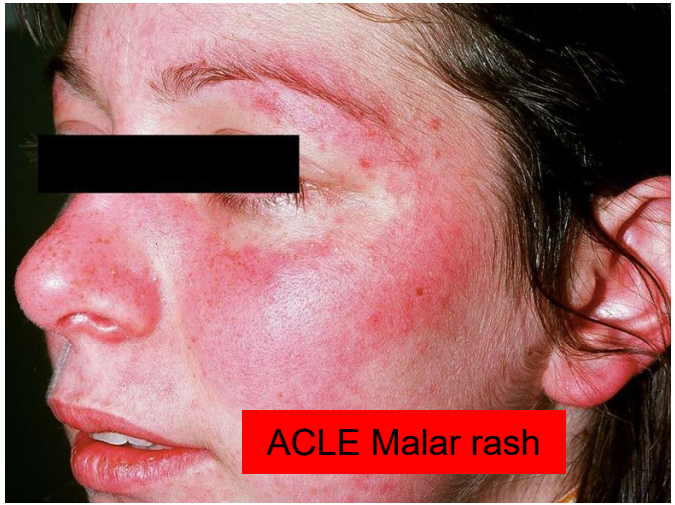
Cunliffe, Tim et al. (2018) Retrieved from http://www.pcds.org.uk/clinical-guidance/sarcoid.



MD Cunliffe, Tim et al. (2018) http://www.pcds.org.uk/p/dr-tim-cunliffe.

Let’s Test What We’ve Learned!





- Sparing of nasolabial folds
- Malar distribution

- Telangiectasia
- Crosses the NLF

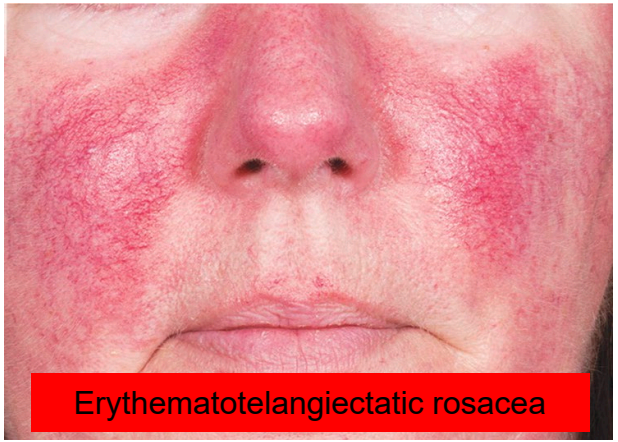
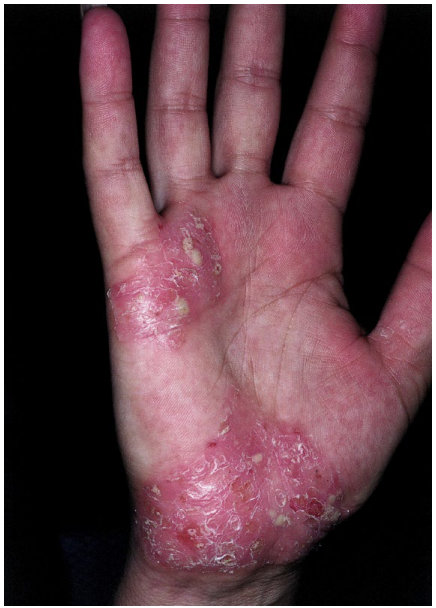


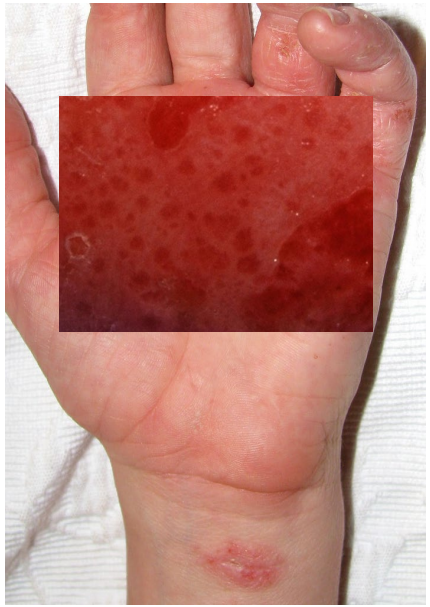
Photo credit: http://dermnetnz.org
Carla Rothaus et al. November 2017. *Rosacea*. Retrieved from https://resident360.nejm.org.

Pustular PSO



Painless pustules

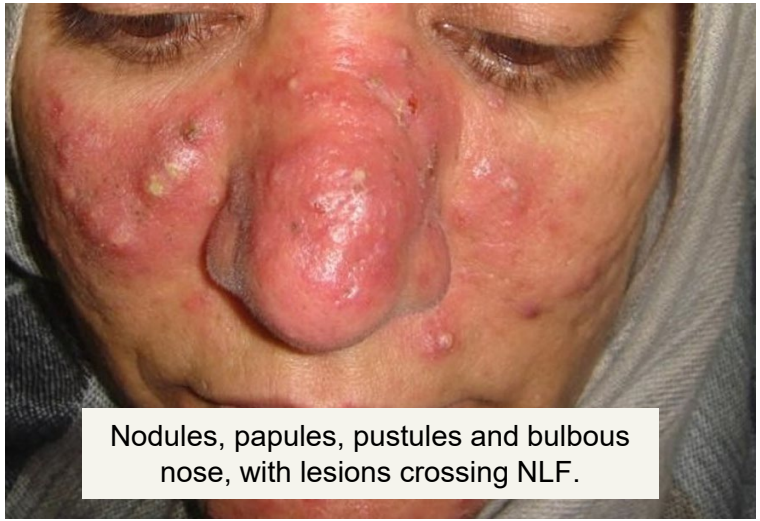
Atopic Eczema



Hyperlinear

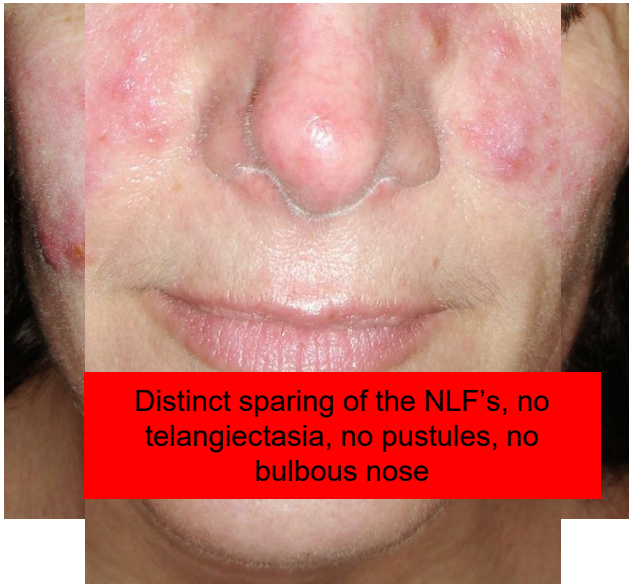
Care_SMC (2008). Retrieved from http://flickriver.com
Photo credit: Dr Allan Watson. Retrieved from https://www.dermcoll.edu.au/atoz/psoriasis/.

Rosacea



Nodules, papules, pustules and bulbous nose, with lesions crossing NLF.

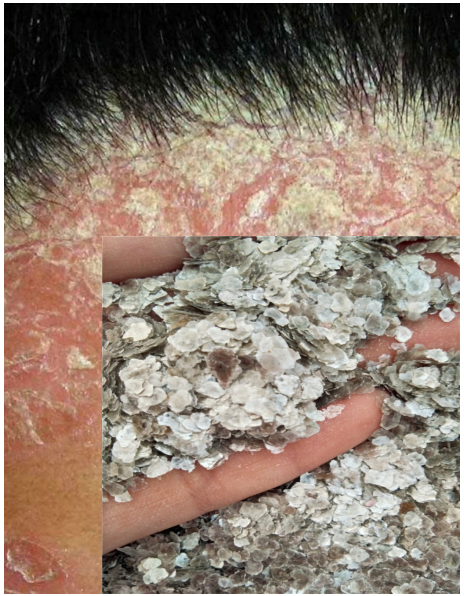
Acle



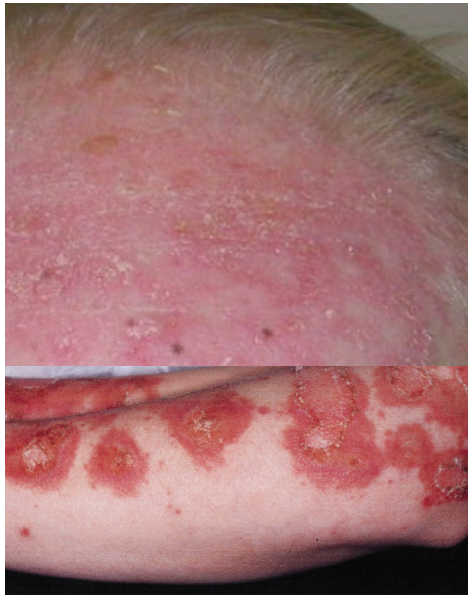
Distinct sparing of the NLF's, no telangiectasia, no pustules, no bulbous nose

Health Jade team. *Rosacea*. Retrieved from https://healthjade.com/rosacea/; felClinic. (2018). *Systemic Lupus Erythematosus* (Lupus or SLE). Retrieved from https://felclinic.com/systemic-lupus-erythematosus-sle-lupus/.

(Micaceous scale)
Psoriasis



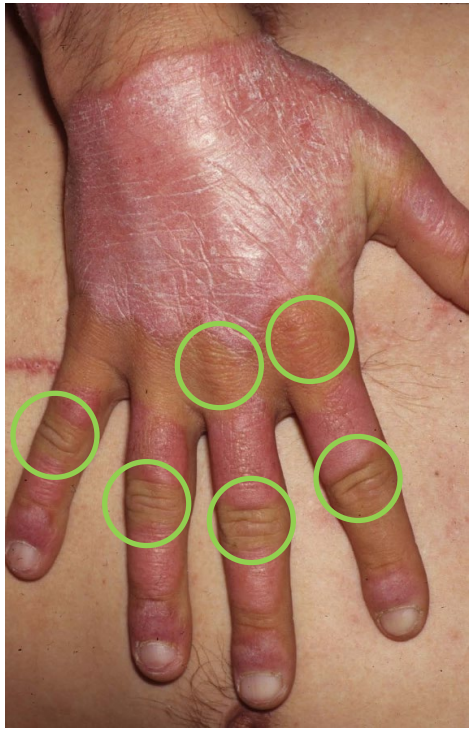
SCLE



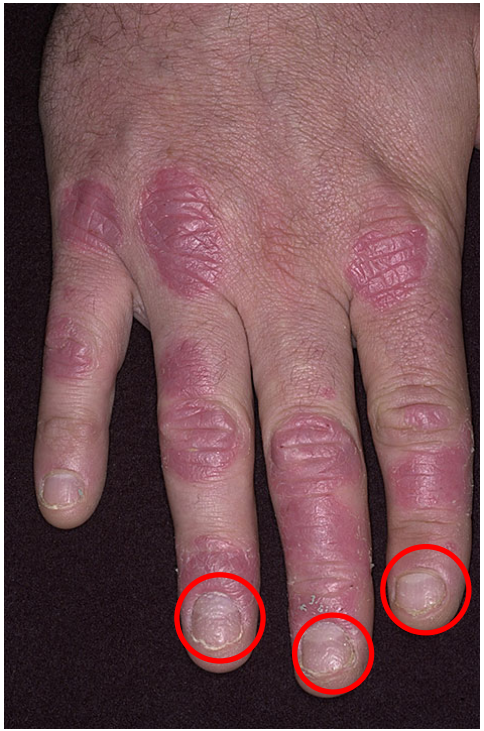
(Polycyclic lesions)

Galen Foulke, MD et al. 2013. *Cutaneous SLICC Classification Criteria for SLE and How to Treat Them* [PowerPoint slides]. Retrieved from http://ssms.weblinkconnect.com/CWT/EXTERNAL/WCPAGES_PRS/PDF/MEETINGS/2017_ANNUAL_MEETING/PDF/FOULKE.PDF.

Generalized ACLE

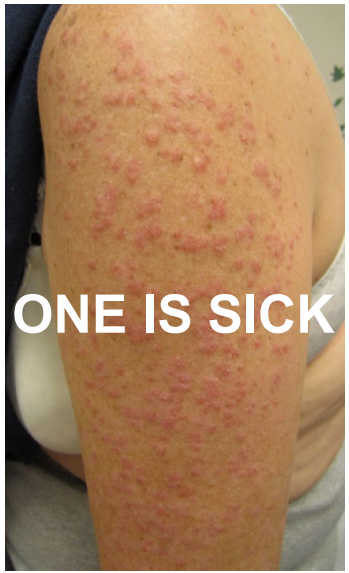


Psoriasis

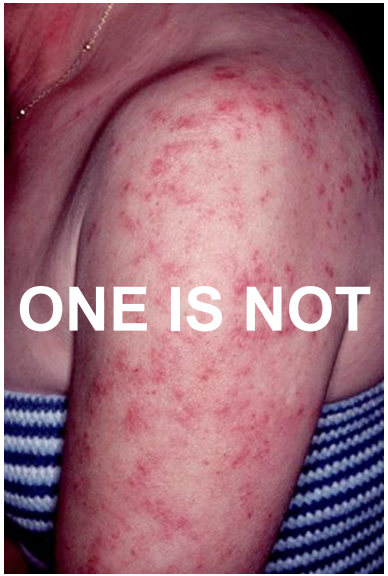


http://www.dermpathmd.com/Clinical%20Dermatology/Cutaneous_Lupus_Erythematosus.pdf.

Generalized ACLE, papulosquamous

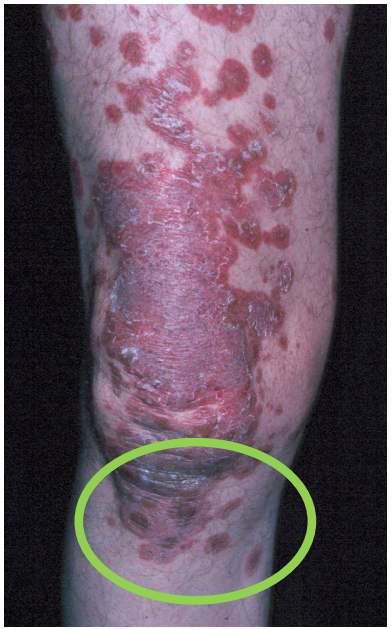


Early SCLE

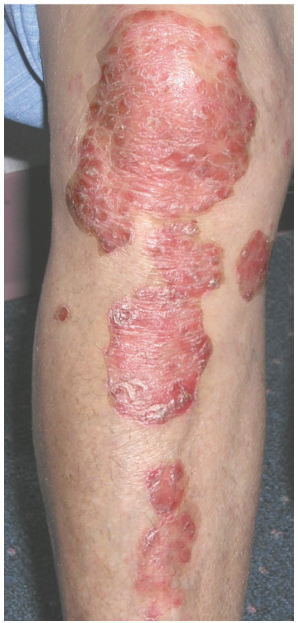


Galen Foulke, MD. 2013. Cutaneous SLICC Classification Criteria for SLE and How to Treat Them [PowerPoint slides]. Retrieved from http://ssms.weblinkconnect.com/CWT/EXTERNAL/WCPAGES_PRS/PDF/MEETINGS/2017_ANNUAL_MEETING/PDF/FOULKE.PDF; Janice Lin, MD, MPH et al. (2018). Subacute Cutaneous Lupus Erythematosus (SCLE). Retrieved from http://webmd.com.

SCLE

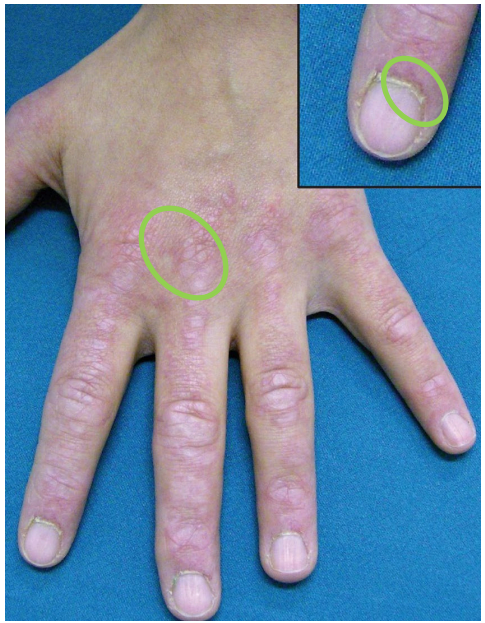


PSORIASIS

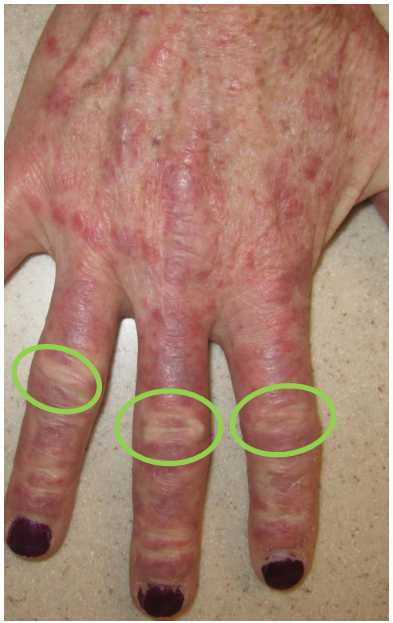


Janice Lin, MD, MPH. (2018). Subacute Cutaneous Lupus Erythematosus (SCLE). Retrieved from http://webmd.com;
Dr. Allan Watson. https://www.dermcoll.edu.au/atoz/psoriasis/.

Dermatomyositis



Generalized ACLE



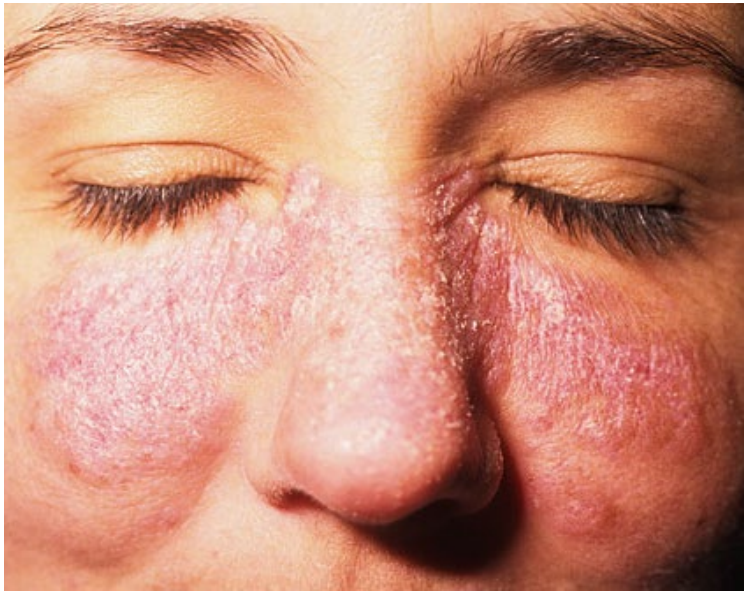
Federica Ricceri and Francesca Prignano et al. *Gottron papules*: a pathognomonic sign of dermatomyositis
CMAJ February 05, 2013 185 (2) 148; Galen Foulke, MD et al. 2013. *Cutaneous SLICC Classification Criteria for SLE and How to Treat Them* [PowerPoint slides]. Retrieved from http://ssms.weblinkconnect.com/CWT/EXTERNAL/WCPAGES_PRS/PDF/MEETINGS/2017_ANNUAL_MEETING/PDF/FOULKE.PDF.

Involves the conchae
Atrophic scarring w/ loss of pigment

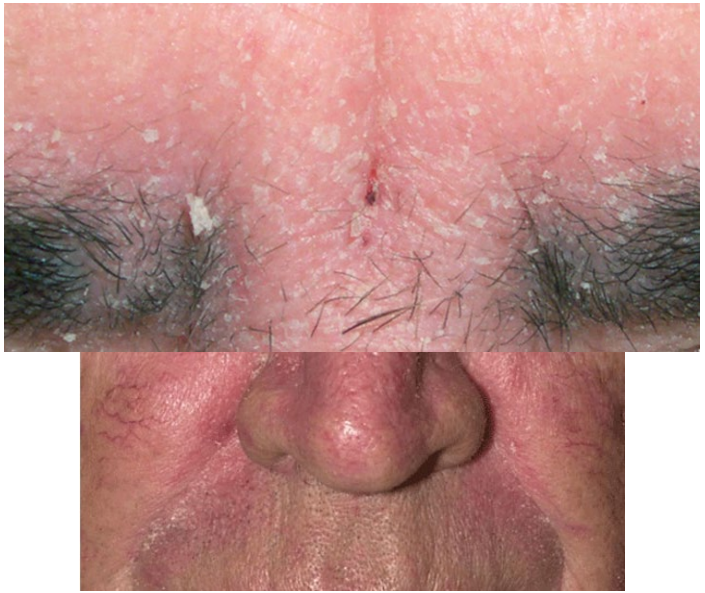
Less scarring
Involves the ear lobe NOT the conchae
Red to brown color

Philip E. LeBoit, MD, Brian Hinds, MD et al. (2018). *Lupus Erythematosus*. Retrieved from https://www.derm101.com/clinical-atlas/lupus-erythematosus; Philip E. LeBoit, MD, Brian Hinds, MD, et al. (2018). *Sarcoidosis*. Retrieved from https://www.derm101.com/clinical-atlas/lupus-erythematosus.

Malar rash of ACLE

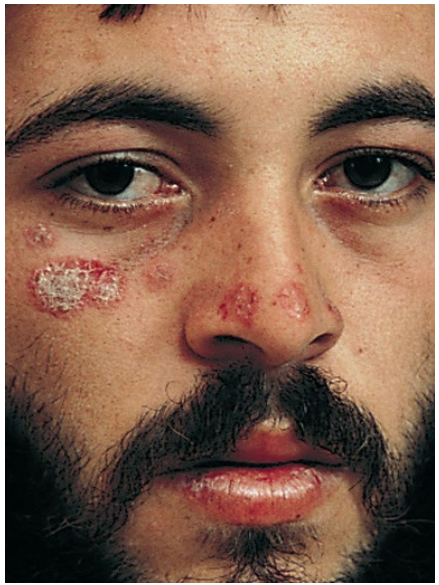


Seborrheic dermatitis

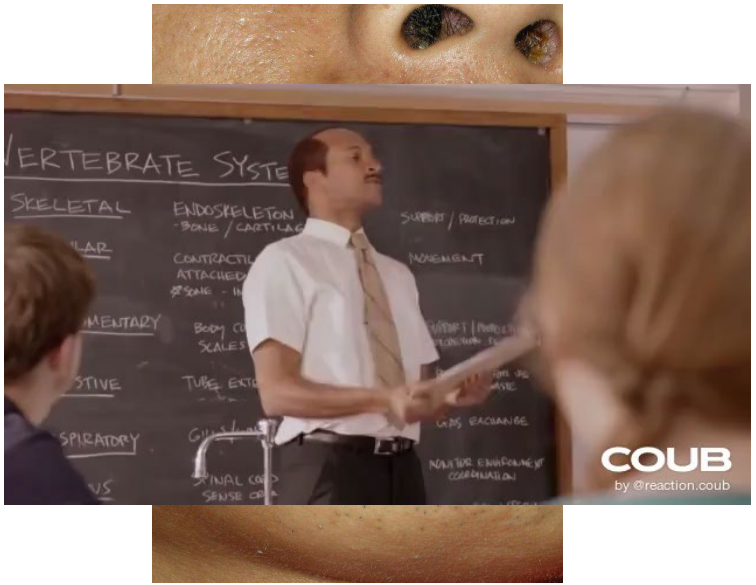


Jennifer Robinson, MD et al. (2018). *A Visual Guide to Understanding Lupus*. Retrieved from www.webmd.com; Dr. Thomas Habif. Retrieved from http://merckmanuals.com; Natalie Silver et al. (2018). *What Causes Eczema on the Scalp, and How Is It Treated?* Retrieved from https://www.healthline.com/health/skin-disorders/eczema-on-scalp.

DLE



SYPHILIS!



Philip E. LeBoit, MD, Brian Hinds, MD et al. (2018). Lupus Erythematosus. Retrieved from https://www.derm101.com/clinical-atlas/lupus-erythematosus; 1. Henry Foong FRCP Edin (2011) Secondary syphilis presenting as annular plaques on face and scalp. httpwww.vgrd.orgarchivecases2011sypsyp.html; 2. Key and Peele, Season 3 episode 1, “Substitute Teacher,” Keegan-Michael Key and Jordan Peele.

Conclusions

- Unfortunately, rashes do not consistently follow predictable patterns
- Inspect as much of their skin as possible, to find other signs of specific disease (eyes, scalp, nails, etc…)
- Just as we all learned in training, use history as part of your diagnostic conclusions (ie. Take note of their other presenting symptoms in other systems/ evolution of lesions, etc.)

Helen Vivienne Fletcher et al. (2011). Little Miss Autoimmune. Retrieved from http://littlemissautoimmune.blogspot.com /2011/12/why-couldnt-butterfly-rash-look-like.htmld.

1. Galen Foulke, MD et al. 2013. *Cutaneous SLICC Classification Criteria for SLE and How to Treat Them* [PowerPoint slides]. http://ssms.weblinkconnect.com/CWT/EXTERNAL/WCPAGES_PRS/PDF/MEETINGS/2017_ANNUAL_MEETING/PDF/FOULKE.PDF.
2. Jarukitsopa S, Hoganson DD, Crowson CS, et al. Epidemiology of systemic lupus erythematosus and cutaneous lupus erythematosus in a predominantly white population in the United States. *Arthritis Care Res* (Hoboken). 2015;67(6):817-28.
3. Kole AK, Ghosh A, et al. Cutaneous manifestations of systemic lupus erythematosus in a tertiary referral center. *Indian J Dermatol*. 2009;54(2):132-6.
4. Mohammed Hammoudeh, Ahmed Al-Momani, Husam Sarakbi, Prem Chandra, and Samer Hammoudeh, et al. “Oral Manifestations of Systemic Lupus Erythematosus Patients in Qatar: A Pilot Study,” *International Journal of Rheumatology*, vol. 2018, Article ID 6052326, 6 pages, 2018.
5. Presley BC, Bush JS, Watson SC, et al. Dermatomyositis with extensive calcification in an adult. *West J Emerg Med*. 2012;13(1):136-8.
6. Wysenbeek AJ, Guedj D, Amit M, Weinberger A, et al. Rash in systemic lupus erythematosus: prevalence and relation to cutaneous and non-cutaneous disease manifestations. *Ann Rheum Dis*. 1992;51(6):717-9.
7. https://www.hopkinslupus.org/lupus-info/lupus-affects-body/skin-lupus/.
8. http://www.dermpathmd.com/Clinical%20Dermatology/Cutaneous_Lupus_Erythematosus.pdf.
9. http://www.dermpathmd.com/Clinical%20Dermatology/Cutaneous_Lupus_Erythematosus.pdf.