Bust reputedly of "Psyche," an athlete in the employ of the Duke of Northumberland
The Blackamoor
Bust of a Black Man
Date: ca. 1758
Medium: Black limestone on a yellow marble socle
Dimensions: Overall: 28 × 20 × 10 1/2 inches (71.1 × 50.8 × 26.7 cm) and Base or socle: 8 1/2 inches (21.6 cm)
Credit Line: Yale Center for British Art, Paul Mellon Collection
FUTURE MEETINGS OF THE NEW ENGLAND DERMATOLOGICAL SOCIETY

December 4, 2021 – Online Clinical Meeting
Hosted by:
UMass Medical School
Worcester, MA

February 5 or 12, 2022 – Didactic Meeting*
Hosted by:
Boston Children’s Hospital
Waltham, MA

*meeting format still tbc
October 2, 2021

Dear Attendee:

I would like to take this opportunity to invite you to become a member of the New England Dermatological Society (NEDS). A key benefit to membership in the Society is complete and full access to the NEDS website. Such access allows members to search for all uploaded cases presented at past clinical meetings. These cases provide a valuable database of unusual dermatological disorders and their treatment.

The New England Dermatological Society offers a 50% discount on the first year’s membership dues for any person who applies within one year of completing their residency or fellowship training. The Society also offers a $50 rebate on membership dues to any member who refers a new member to the Society.

Initiate your application by applying online and then forward your CV and letter of recommendation to our administrator, Gayle Sommer at NEDS@mms.org. You’ll find membership application information and materials in the ‘Membership’ section of www.nederm.org.

If you have any further questions, please contact Gayle at 781-434-7731 or NEDS@mms.org. Your application will be reviewed at the next scheduled Council meeting once all required application information is received.

With best regards,

Avery LaChance, MD, MPH
Secretary, New England Dermatological Society
www.nederm.org
Case of the Year, Century Scholar and Book Awards

Case of the Year
The Case of the Year Award is given to the best clinical case presented during the academic year. Each program submits their “best” case at the conclusion of each academic year so that the Council can vote and decide on the award recipient. If the winner then presents the case at the American Academy of Dermatology (AAD) in the gross and microscopic session he/she will be awarded $500 to help with travel expenses. $500 will also be presented to the recipient’s residency program’s educational fund.

Century Scholar Award
The Century Scholar Award is given to the dermatology resident or trainee in their dermatology rotation who wrote up the winning case. He or she is awarded a $500 Amazon gift card towards the purchase of books.

If the case is accepted for presentation at a conference and the trainee travels to present it, $500 is awarded to the trainee to help with travel expenses. Their write up must specify that the case was presented at a New England Dermatological Society Conference and the case must be accepted within two years to receive the travel funding.

Book Award
The New England Dermatological Society will award $500 towards the purchase of a medical textbook to any dermatology resident at a member institution who satisfies the following requirements:

1. The resident is a first author of a report based on a case presented at a meeting of the New England Dermatological Society.

2. The report is accepted for publication by a peer-reviewed journal within one year of the meeting and contains language indicating that the “Case was presented at a meeting of the New England Dermatological Society at (institution) on (date).”

3. The resident supplies the Society’s Secretary with a copy of the final journal acceptance letter and a receipt for the purchased medical textbook. The Society will then award the recipient with a gift certificate in the amount of $500 towards the purchase of the medical textbook.
IMPORTANT

Conference Evaluation and CME Information

Registered attendees will receive an email within 7-10 business days of this event from nejmcust@mms.org with a link to the online evaluation for this activity.

Once you complete the online evaluation you will receive instructions on how to claim your CME credit and receive your certificate.

For questions on the evaluation or certificates, please contact Customer Service at 800-843-6356 or nejmcust@mms.org.
Clinical meeting hosted by Yale Medical School Dermatology Department

October 2, 2021

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<td>Verrica</td>
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<tr>
<td>7:30 – 8:30 am</td>
<td>Online Patient Case Viewing/Whole Slide Scanned Images Viewing</td>
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<td>8:30 – 8:40 am</td>
<td>Welcome and Introduction</td>
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<td>Christine Ko, MD</td>
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<tr>
<td>8:40 – 9:00 am</td>
<td>Follow-up Cases:</td>
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<tr>
<td>9:00 – 9:15 am</td>
<td>Deliberate Practice Session: Visual Recognition of Skin Disease</td>
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<td>9:45 – 10:15 am</td>
<td>In Depth Breakout Session Part 1 (choice of topics 1-11)</td>
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<tr>
<td>10:15 – 10:30 am</td>
<td>NEDS Business Meeting*</td>
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<td>10:30 – 11:00 am</td>
<td>Coffee Break / Optional Visit to Exhibit Portal*</td>
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<td>Optional Review of Patient Casebook/Whole Scanned Images</td>
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<tr>
<td>11:00 – 11:30 am</td>
<td>Presentation: “Moleculary Targeted Therapy for Sarcoïdosis and Granuloma Annulare: From Bedside to Benchtop and Back”</td>
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<td>William Damsky, MD, PhD</td>
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<tr>
<td>11:30 am – 12:00 pm</td>
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<td>12:00 pm - 12:30 pm</td>
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<tr>
<td>12:30 – 12:45 pm</td>
<td>Follow-up Deliberate Practice Session: Visual Recognition of Skin Disease</td>
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<td>Moderator: Christine Ko, MD</td>
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<td>12:45 – 1:00 pm</td>
<td>Concluding Remarks</td>
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<td>Christine Ko, MD</td>
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ACKNOWLEDGMENTS

Welcome to the New England Dermatological Society Meeting at Yale-New Haven Hospital on October 2, 2021. We hope that you find the virtual cases presented today to be interesting and of educational value. We would like to take this opportunity to thank the following individuals who have been instrumental in the planning and execution of this meeting.

This meeting would not have been possible without the support of all the members of the Yale University Department of Dermatology. We would especially like to thank the faculty advisors and dermatopathologists whose contributions to the case presentations were indispensable.

We would like to extend a special thank you to our leader and moderator for this meeting Christine Ko, MD; without her unbridled dedication and guidance this meeting would not be possible. We also thank our speakers for helping to make this NEDS experience as educational as possible: Alicia Little, MD, PhD; Amanda Zubek, MD, PhD; Angela Galan, MD; Brett King MD; Britt Craiglow, MD; Bruce Strober, MD, PhD; Caroline Nelson, MD; Chris Bunick, MD, PhD; Earl Glusac, MD; Fotios Koumpouras, MD; Gauri Panse, MD; Ian Odell, MD, PhD; Irwin Braverman, MD; Jean Bologna, MD; Jeffrey Cohen, MD; Jonathan Leventhal, MD; Kalman Watsky, MD; Kathleen Suozzi, MD; Marc Grossman, MD; Mary Tomayko, MD, PhD; Matthew Vesely, MD, PhD; Monique Hinchcliff, MD; Paul Schneiderman, MD; Peter Heald, MD; Richard Antaya, MD; Sara Perkins, MD; Sarika Ramachandran, MD; Sean Christensen, MD, PhD; Shawn Cowper, MD; and William Damksy, MD, PhD.

Assistance for the meeting and expert administrative skills were generously provided by Lisa Rao. We also thank Gayle Sommer for providing guidance regarding the novel framework of the meeting.

Lastly, a special thanks to the wonderful residents of the Yale Department of Dermatology, the authors of this book, for their case presentations and hard work preparing for this meeting: Alexander Fogel, MD, MBA; Michael Gowen, MD, MBA; Mary Laird, MD; Emma Weiss, MD, Goran Micevic, MD, PhD; Michal Kidacki MD, PhD; Sarah Kim MD; Noel Turner MD; Theodore Zaki MD; Margaret Johnston, MD; Catherine Baker, MD; Andrew Johnston MD, PhD; Elizabeth Tkachenko MD; William Shipman III MD, PhD; and Joseph Sarhan, MD, PhD. We would additionally like to thank the Yale medical students whose assistance in our preparations was invaluable.

Above all, we would like to thank our patients and their families for agreeing to be a part of this clinical meeting today; they are truly our greatest source of learning.

We hope you enjoy the day.

On behalf of the Yale Dermatology Residents,
Mike Gowen, MD
Case 1: An ~40–year-old woman with alopecia

The patient presented with 2 months of alopecia, 1 month of malar rash, pleurisy, oral ulcers, sicca, arthralgias, myalgias, fatigue, organic brain syndrome and lymphadenopathy. Anti-dsDNA, anti-SM, anti-RNP, anti-chromatin, anti-SSA, anti-SSB were elevated. C3 and C4 were decreased. Proteinuria with normal creatinine was also present. CBC was normal; antiphospholipid antibodies were not detected.

Case courtesy Michal Kidacki, MD PhD, Shawn Cowper, MD, and Brett King, MD PhD
Nonscarring alopecia of lupus erythematosus

**Course and Treatment**
The patient has systemic lupus erythematosus (SLE). She met 2019 ACR/EULAR criteria with a score of 26. Her alopecia was diagnosed as non-scarring alopecia of LE; the patient was treated with azathioprine, hydroxychloroquine, minoxidil, spironolactone, topical clobetasol ointment and intralesional triamcinolone. Patient initially improved on the therapy but shortly after experienced a lupus flare requiring hospitalization.

**Teaching Points**
- Inflammation in darker skin can range from pink-red to dark brown-black
- Non-scarring alopecia is more frequently seen than scarring alopecia in SLE
- Non-scarring alopecia is less likely to show dyspigmentation on exam
- Non-scarring alopecia of LE may be difficult to distinguish from alopecia areata but will show inflammation involving the upper follicle and in a perivascular and perieccrine distribution

<table>
<thead>
<tr>
<th>Telogen Effluvium</th>
<th>Non-cicatricial alopecia of LE</th>
<th>Alopecia Areata</th>
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<td><img src="image1.png" alt="Image of Telogen Effluvium" /></td>
<td><img src="image2.png" alt="Image of Non-cicatricial Alopecia of LE" /></td>
<td><img src="image3.png" alt="Image of Alopecia Areata" /></td>
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<tr>
<td>~15-20% of hairs in telogen</td>
<td>Inflammation at all levels of the follicle and in perivascular and periaxillary foci</td>
<td>&gt;90% of hairs in telogen</td>
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</table>
Case 2: A young baby with a facial rash

Neonate with dozens of depressed pink macules in a predominantly periocular and perioral distribution and over cheeks.
Neonatal lupus erythematosus (NLE)

Course
No family history of lupus; mother reported arthritis and Raynaud’s phenomenon. The mother was found to have a positive ANA (1:2560) and positive anti-Smith antibody, awaiting rheumatology consultation. Patient with normal cardiac evaluation and pending laboratory studies; no clinical concern to date with normal development.

Teaching Points
• Serologies for both infant and mother are critical, as NLE may be the presenting sign of autoimmune disease in mother
• Infants with NLE require cardiac workup and EKG to evaluate for heart block; hematologic and liver disease should also be ruled out
• In the congenital presentation the lesions may scar compared to the neonatal presentation, in which there is no scarring
• The patient is of Dominican & Arabic descent and developed significant post-inflammatory hyperpigmentation

Patient at 6 months

Another patient with NLE

Courtesy Richard Antaya, MD

Courtesy Michael Gowen, MD
Case 3: An ~40-year-old woman with pink-brown plaques in ears and on scalp

Slightly atrophic pink and brown plaques in the bilateral conchal bowls, with previous involvement of the scalp, present for several months. ANA negative. No systemic symptoms.

Case courtesy Shaman Bhullar MS4, Joseph Sarhan, MD PhD, Anjela Galan, MD, and Jonathan Leventhal, MD
Discoid lupus erythematosus

Course and Treatment
The patient fulfilled the following classical clinical criteria for discoid lupus erythematosus (DLE) including: atrophic scarring, location on head and neck, dyspigmentation, and follicular hyperkeratosis/plugging. She was treated with hydroxychloroquine 200 mg BID and topical clobetasol solution followed by tacrolimus ointment maintenance with improvement. There was not evidence of systemic involvement.

Teaching Points
• Black individuals have a 4-5 times increased incidence of DLE compared with white individuals
• Black individuals develop the disease at an earlier age and have a higher mortality rate.
• Risk of developing SLE is significant in patients with DLE (~15-20%), but it is unclear if patients with skin of color have a higher risk of progression.
Chronic cutaneous lupus erythematosus

Dyspigmentation, atrophy, and alopecia with biopsy findings including absent hair follicles, deep lymphocytic inflammation, and clusters of CD123-positive cells. *Clinical image courtesy Jonathan Leventhal, MD.*

Dyspigmentation in a lesion with little inflammation apparent in tissue sections. There is hyperkeratosis, loss of hair follicles, melanin in the basal layer, and pigment incontinence. *Clinical image courtesy, Michal Kidacki, MD PhD*
Chronic cutaneous lupus erythematosus
Toxic epidermal necrolysis-like lupus erythematosus

Images courtesy, Jeff Gehlhausen, MD PhD, and the Yale Dermatology Residents’ Collection
Toxic epidermal necrolysis

At presentation

Day 6

Images courtesy, Yale Dermatology Residents’ Collection
Case 4: An ~30-year-old woman with papules on the forearms and an inner arm plaque

Case courtesy Dana Correale, MD, Jennifer McNiff, MD, and Robert Patrignelli, MD

Patient with Fitzpatrick skin type VI, with “new” papules on the forearms and plaque on the inner arm. Per records, patient has had a similar rash documented by dermatology over the past 6 years. She also is followed by rheumatology with a diagnosis of systemic lupus erythematosus; she fits criteria with alopecia, photosensitivity, documented lesions of discoid lupus erythematosus, intermittent arthralgias and arthritis, ANA of 1:1280 (coarse speckled), and positive anti-dsDNA. Her complete blood count was normal in July, 2021.
Reactive granulomatous dermatitis

**Course**
The patient fulfills criteria for systemic lupus erythematosus (SLE). She is intermittently treated by rheumatology with azathioprine (currently 150 mg qd), hydroxychloroquine (200 mg bid), and prednisone 20 mg po qd. The skin lesions partially respond to intralesional triamcinolone or topical steroids but recur.

**Teaching Points**
- Reactive granulomatous dermatitis (RGD) has been proposed as a term to encompass palisaded neutrophilic and granulomatous dermatitis (PNGD), interstitial granulomatous dermatitis (IGD), and interstitial granulomatous drug reaction
- RGD is a useful umbrella term in patients like this with multiple morphologies; the patient’s symmetric forearm papules and the upper arm annular erythematous to hyperpigmented plaque fit well with PNGD and IGD, respectively
- Diagnosis of reactive granulomatous dermatitis allows consideration of association with autoimmune diseases (especially lupus erythematosus and rheumatoid arthritis), an inciting medication (e.g. calcium-channel blockers, beta-blockers, ACE inhibitors, lipid-lowering agents), or internal malignancy (especially hematologic)

*July 2021, right axilla and L5 finger*
Case 5: An ~80–year-old woman with rheumatoid arthritis on JAKi

An ~80-year-old woman with a long history of rheumatoid arthritis presented with unilateral, crusted papules on the left hand, forearm and elbow with an initial biopsy of a left elbow lesion showing a dense neutrophilic infiltrate (left column, 3rd image). A second biopsy 1 month later from the finger showed palisading granulomas with sparse neutrophils (not shown). Three months later, the eruption became symmetric (left column, 2nd image); she concurrently developed palpable purpura on the lower legs with biopsy showing leukocytoclastic vasculitis (right column).
Skin manifestations of rheumatoid arthritis

Course
• The patient was diagnosed with palisaded neutrophilic and granulomatous dermatitis (PNGD) of the upper extremities and small vessel vasculitis of the lower extremities. Her skin lesions have partially responded to treatment of her refractory rheumatoid arthritis (RA) with an IL-6 receptor antagonist; due to incomplete relief of joint pain, treatment will be switched to rituximab.

Teaching Points
• Skin manifestations of RA include rheumatoid nodules, small and medium vessel vasculitis, neutrophilic dermatoses, PNGD, and interstitial granulomatous dermatitis (IGD). Reactive granulomatous dermatitis (RGD) is a proposed term that unifies PNGD and IGD
• RGD can be a predominantly neutrophilic process +/- vasculitis as in the patient’s initial biopsy; adding qualifiers to the histopathologic diagnosis [i.e. “PNGD or RGD, eosinophil-rich” or “PNGD or RGD, histiocyte rich”] would underscore the range of inflammatory cells seen in this entity

Medium vessel vasculitis in another patient with rheumatoid arthritis
Reactive granulomatous dermatitis, neutrophil-rich

Teaching Points (Continued)
• Early lesions of reactive granulomatous dermatitis may have prominent neutrophils, with the histopathologic differential diagnosis including infection

Reference
Teaching Points (Continued)

- Well-developed lesions of reactive granulomatous dermatitis display necrobiosis with palisades of histiocytes and variable numbers of neutrophils.

Reference
Case 6: An ~60-year-old man with facial papulonodules, telangiectasia, and dactylitis

The patient had a twenty-year history of “severe breakouts” of the face, arms, and feet. Skin findings included bilateral medial lower eyelid plaques, extending to the medial lower cheek on the left with prominent telangiectasia, and a nodule at the site of a scar on the forehead; other scalp and extremity lesions are not shown. Several toes were purplish, swollen, and displayed onychorhexis. He also had amputation of the R3 toe in the early 2000s due to severe pain. No cervical, axillary, or inguinal lymphadenopathy was noted. Biopsy of a plaque showed non-necrotizing granulomas.
Angiolupoid sarcoidosis with sarcoid dactylitis

Course and Treatment:
The patient had a known history of ocular and nasopulmonary sarcoidosis, hyperlipidemia, hypertension, and osteopenia; he was treated with prednisone and methotrexate for his internal involvement with minimal improvement of cutaneous lesions. He was enrolled in a clinical trial of tofacitinib, with gradual improvement. He had partial remission of facial lesions, improvement of subcutaneous nodules on the hands, and improvement of sarcoid dactylitis. Treatment also led to improvement of sinus symptoms. His response on tofacitinib allowed discontinuation of prednisone and methotrexate.

Teaching Points
• Angiolupoid sarcoidosis is a unique presentation of cutaneous sarcoidosis with papulonodular lesions and telangiectasias of the central face
• Sarcoidosis can present with dactylitis with or without underlying bony changes
• Nail dystrophy (onychorrhexus in this case) can be secondary to sarcoidosis

References
Another example of sarcoidosis of the digit with sarcoidal granulomas below the nail matrix epithelium. Resolution of sarcoidal nail dystrophy with residual postinflammatory hyperpigmentation, status post treatment and maintenance with tofacitinib 5 mg po bid. 
*Courtesy William Damsky, MD PhD.*
Teaching Points (Continued)

- Cutaneous sarcoidosis in darker skin may be minimally erythematous and may appear hypopigmented, hyperpigmented, or infiltrative
- Sarcoidosis can be associated with significant scarring and pigmentary alteration
Case 7: An ex-24 week, now 4-week-old baby girl with a brown plaque

Female infant, Fitzpatrick skin type VI, with brown plaque on right 2\textsuperscript{nd} toe. According to parents, right 2\textsuperscript{nd} toe has been “reddish” over the last few weeks and in last few weeks has become more edematous and darker in color with no known trauma.
Infantile hemangioma

Course and Treatment

- Ultrasonic Doppler flow detector demonstrated increased flow on the dorsal, thicker aspect.
- Initially, given location and size of lesion, and overall health of the patient, no treatment was given.
- At the one month inpatient follow-up, the lesion was growing - topical timolol 0.5% qd for 2-3 days then bid thereafter (with orders to monitor heart rate, blood pressure, and respiratory status during first few applications).
- Three days later, there was concern that the lesion was beginning to ulcerate. PO propranolol 0.5mg/kg/day divided q8 hours was started.
- Four days later, PO propranolol was increased to 1.0 mg/kg/day divided q8 hours; however, NICU course was complicated by bronchopulmonary dysplasia, bilateral retinopathy of prematurity stage 2, anemia of prematurity, and osteopenia of prematurity, and necrotizing enterocolitis stage II with volvulus necessitating resection of distal ileum to proximal transverse colon and ileostomy placement - requiring no PO meds, so propranolol was held. Hemangioma remained stable.
- Propranolol was restarted 1 month later at 0.5 mg/kg/day then increased to 1.0 mg/kg/day 24 hours later.
- 3 months later, hemangioma continued to significantly improve and PO propranolol was continued at 1.0 mg/kg/day

Ultrasonic Doppler flow detector

5 months after treatment
Infantile hemangioma, other examples

Teaching Points

- In lighter skin types superficial infantile hemangiomas (IH) usually appear bright red, while deeper IH appear skin colored or bluish; however, in darker skin types superficial and deeper IH may appear bluish, violaceous, or even brown.
- For deep IH, and for both superficial and deep IH in skin of color, an ultrasonic Doppler flow detector may help confirm the diagnosis of infantile hemangioma.

Images courtesy Richard Antaya, MD
Verrucous venulocapillary malformation

Courtesy Yale Dermatology Residents’ Collection
Case 8: An ~40–year-old man with alopecia and pink-brown pruritic plaques

A 37-year-old man presented with a 10-year-history of a pruritic rash unresponsive to topical therapy and phototherapy.
Psoriasis

Course and Treatment
Despite treatment with methotrexate 17.5 mg weekly, his plaque psoriasis progressed to involve 30% of his body surface area. Switching methotrexate to adalimumab, and subsequently risankizumab, resulted in 100% clearance of his active psoriasis lesions. His psoriasis was replaced by hyperpigmented patches (see below).

Teaching points
• Psoriasis in patients with heavily pigmented skin may present as dark gray to violaceous to bluish to pink-red plaques with minimal gray to white scale
• There is an increased tendency toward post-inflammatory pigmentary alteration

Psoriasis and post-inflammatory hyperpigmentation in another patient with Fitzpatrick V skin. Courtesy Jeffrey Cohen MD.
Psoriasis, inverse

- Pink to violaceous plaque with white-silver scale
- Confluent parakeratosis
- Hypogranulosis
- Acanthosis
- Increased small vessels in the papillary dermis
- Background melanin pigment
Psoriasis, pustular

Pustular psoriasis in a Hispanic patient with Fitzpatrick skin type V. *Clinical image courtesy Mary Tomayko, MD PhD.*

**References**


Case 9: An ~40–year-old woman with a brown patch

Past medical history of limited systemic sclerosis (SSc) including mild sclerodactyly, GERD, and Raynaud phenomenon treated with mycophenolate mofetil; presented with new hyperpigmentation of her chest and abdomen along with worsening subjective skin tightness. Her modified Rodnan skin score (mRSS) was 10, and a biopsy of the hyperpigmented patch was obtained. Biopsy findings included thickened collagen bundles associated with a perivascular infiltrate composed of lymphocytes and plasma cells.
**Course and Treatment**
In this case, proximal fibrosis changed her diagnosis from limited to diffuse cutaneous SSc. Antibody tests were positive for Scl-70 >8 (ref neg <1) and ANA at 1:1280, homogenous and 1:40, nucleolar patterns. With worsening skin-limited disease, extracorporeal photopheresis was added to her treatment regimen. Despite this addition, over the next 1-2 months she developed rapid progression of her skin fibrosis to a mRSS of 30 along with interstitial lung disease. Therefore, treatment by autologous stem cell transplant was performed. One year later, her mRSS improved to 18 yet she continued to have diffuse hyperpigmentation of the trunk and extremities.

**Interstitial lung disease**
NSIP pattern outlined by dashes

NSIP = nonspecific interstitial pneumonia; pattern of involvement predominantly affecting the periphery of the lower lobes, +/- ground glass opacities, with subpleural sparing

**Diffuse hyperpigmentation**

Courtesy Ami Rubinowitz, MD
Systemic sclerosis (SSc), pigmentary alteration

**Teaching Points**
- SSc-associated skin fibrosis can be accompanied by changes in pigmentation
- Hyperpigmentation may be diffuse (as in this case) and may only be appreciated relative to the patient’s baseline; splattered hypopigmentation is possible
- Dyspigmentation can have a salt and pepper appearance due to retained follicular pigment

Hyperpigmentation in two patients with systemic sclerosis (left column; courtesy Ian Odell, MD PhD). Salt-and-pepper dyspigmentation (right column) in two other patients with systemic sclerosis.
modified Rodnan skin score

Based on skin thickness of 17 body sites
Maximum score = 51

Bilateral:
Upper arms
Forearms
Hands
Fingers
Thighs
Lower legs
Feet

Unilateral:
Face
Chest
Abdomen

Minimal clinically important difference = Δ3 to 5

Score 0 to 3
0 = No thickening = fine wrinkles present, no thickness with pinching
1 = Mild thickening = fine wrinkles present, easy to pinch a thickened skin fold between 2 fingers
2 = Moderate thickening = no fine wrinkles, difficulty in making a skin fold between 2 fingers
3 = Severe thickening = inability to make a skin fold between 2 fingers

References
Teaching Points (Continued)

- In addition to pigmentary changes, nailfold erythema and mat-like telangiectasia may be less pronounced or not appreciable in darker skin, but ragged cuticles can still be apparent.
- Recognition of skin color and textural changes is essential to assess for worsening fibrotic disease; textural changes are measured by the modified Rodnan Skin Score (mRSS).

Images courtesy Ian Odell, MD PhD
Systemic sclerosis, calcification and ulceration

Teaching Points (Continued)

- Dystrophic calcification and digital ulcers can be highly problematic irrespective of background skin color.

Images courtesy Ian Odell, MD PhD
Case 10: An ~10–year-old boy with hypopigmented macules and patches

The patient presented with a one-year history of asymptomatic hypopigmented macules and patches over >50% of his body surface area. No history of atopy, personal or in family members, except for a personal history of seasonal allergies. No lymphadenopathy.
Course and Treatment
The patient was presented at Yale Grand Rounds, and the consensus diagnosis was hypopigmented mycosis fungoides despite the paucicellular infiltrate. High throughput sequencing for T-cell receptor gene rearrangement did not detect clonality. The patient was treated with narrow-band UVB twice a week for 3 months and then once a week for 4 months with excellent response and resolution of lesions.

Teaching Points
• Hypopigmented mycosis fungoides can be a difficult diagnosis on histopathologic grounds alone, and clinical-pathologic correlation is useful
• High throughput sequencing of the T-cell receptor gene can (but not always) provide supportive data for the diagnosis of mycosis fungoides; in one study, sensitivity was 85%, using histopathology as the gold standard of diagnosis (clonality detected in 29 of 34 cases)

References
Case 11: An ~90–year-old man with a 10-year history of an expanding chest plaque

Prior treatments included topical 5-fluorouracil (discontinued due to erosions and pain), imiquimod, and 2 sessions of photodynamic therapy without improvement. Multiple biopsies all showed cornoid lamella, dyskeratotic keratinocytes and superficial lymphohistiocytic inflammation.
Giant porokeratosis

Course/treatment
Porokeratoses share a defect in the mevalonate pathway of cholesterol synthesis. This patient had a partial response in the clinical appearance and reduction of itch after two months of topical 2% cholesterol/2% lovastatin ointment. Previous reports have shown benefit in other subtypes of porokeratosis with this treatment.

Teaching Points
• Porokeratoses are caused by mutations in the mevalonate pathway
• Treatment with topical 2% cholesterol/2% lovastatin (from SkinMedicinals) can improve symptoms and appearance

References
Ugwu N et al, Two percent lovastatin ointment as a pathogenesis-directed monotherapy for porokeratosis. JAAD Case Rep 2020;6:1110-1112.
Teaching Points

- Pigmented porokeratosis can affect a range of skin types
- Facial involvement is more common in Asian individuals
- Pigment incontinence may be present in the dermis below the cornoid lamellae

Clinical image courtesy Kalman Watsky, MD

Cornoid lamella

Pigment incontinence

Dermoscopic image courtesy Nan Ring, MD
Porokeratosis, other variants

Linear porokeratosis. Courtesy Julie Cantatore-Francis, MD.

Porokeratosis ptychotropica. Courtesy Yale Dermatology Residents’ Collection.
Case 12: An ~70-year-old man with spiny yellow papules on the palms

Case courtesy Sa Rang Kim, MD, Gauri Panse, MD, and Christopher G Bunick, MD PhD

This patient with no prior medical diagnoses presented with numerous yellow keratotic papules on bilateral palms for 50 years. Patient had a history of exposure to Agent Orange a few years prior to the onset of the lesions. He has not had any prior treatments. No other family members affected.
Spiny keratoderma

Teaching Points

- Spiny keratoderma, also known as music box spine dermatosis, is a rare disease that presents with asymptomatic 1-2 mm keratotic papules over palms and soles.
- Keratotic papules may range from white to yellow in color.
- There are hereditary and acquired forms; the acquired form can be associated with chronic diseases (such as Darier disease, polycystic kidney disease) and malignancy.
- Clinical differentials include punctate porokeratosis and keratoderma, arsenical keratosis, and palmoplantar warts.
- Treatment is not necessary but includes keratolytics, oral retinoids and mechanical debridement.

References

Palmar keratotic lesions

Minute spines in spiny keratoderma

Thin spines in porokeratotic eccrine ostial and dermal duct nevus

Punctate keratoderma

Images courtesy Christopher G Bunick, MD PhD, Keith Choate, MD PhD, and Yale Dermatology Residents’ Collection.
CASE LIST

Case 1  Nonscarring alopecia of lupus erythematosus

Case 2  Neonatal lupus erythematosus

Case 3  Discoid lupus erythematosus
Related cases/other examples:
  Chronic cutaneous lupus erythematosus
  Toxic epidermal necrolysis-like lupus erythematosus
  Toxic epidermal necrolysis

Case 4  Reactive granulomatous dermatitis

Case 5  Rheumatoid arthritis
Histopathologic examples:
  Reactive granulomatous dermatitis, neutrophil-rich
  Reactive granulomatous dermatitis, histiocyte-rich

Case 6  Angiolupoid sarcoidosis with dactylitis
Related cases/other examples:
  Sarcoidosis of the digit
  Sarcoidosis, other examples

Case 7  Infantile hemangioma
Related cases:
  Other examples of infantile hemangioma
  Verrucous venulocapillary malformation

Case 8  Psoriasis
Related cases:
  Psoriasis, inverse
  Psoriasis, pustular

Case 9  Diffuse systemic sclerosis
Related cases/information:
  Systemic sclerosis, pigmentary alteration
  modified Rodnan skin score
  Systemic sclerosis, digital changes
  Systemic sclerosis, calcification and ulceration

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Case 11  Giant porokeratosis
Related cases:
  Porokeratosis, pigmented
  Porokeratosis, other variants

Case 12  Spiny keratoderma
Related cases:
  Palmar keratotic lesions