Case Challenge: Mimics and Masqueraders

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I have no relevant relationships with industry.

Mimics and Masquerades

55 yo hospitalized patient s/p MVA complicated by multiple abdominal surgeries

11/13/13
Chest biopsy: Suppurative folliculitis with pityrospora

Treated with fluconazole, doxycycline, and ketoconazole wash

12/2/13
Zn 7 mcg/dL nl (60-130)
- Was receiving 1.3 mg/d of zinc in his TPN
- 3 mg/d is required in patients without GI losses, and up to 12 mg/d in patients with diarrhea or fistula losses
- Improved rapidly following additional zinc supplementation

12/11/13
Zinc Deficiency

- Acrodermatitis enteropathica
  - AR altered zinc metabolism
- Acquired AE
  - Infants on formula
  - Alcoholics
  - Malabsorption syndromes
  - Limited supply of injectable zinc in the US resulting in several cases of zinc deficiency in those receiving TPN

Acrodermatitis Enteropathica

- Acral and periorificial pustulobullous dermatitis
- Pustular paronychia
- Angular stomatitis
- Diarrhea
- Alopecia

s/p small bowel resection due to SMA occlusion

Take Two
**Nutritional Deficiencies**

- Pallor of the superficial epidermis with or without necrosis
- Confluent parakeratosis and psoriasiform epidermal hyperplasia

**Nutritional Deficiency**

- Niacin deficiency
- 3 D’s: Diarrhea, Dementia and a collar-like Dermatitis (Casal's necklace)
Nutritional Deficiency
Necrolytic Migratory Erythema

- Glucagon-secreting tumor of the pancreas
- Periorificial, flexural, intertriginous, and acral
- Circinate erythematosus patches to plaques with necrosis, erosion, and crusting
- Red shiny erosive fingertips
- Weight loss, beefy-red tongue, angular cheilitis

Take Home

- Acrodermatitis enteropathica
  - Zinc deficiency
  - Acral and periorificial pustulobullous dermatitis
  - Pustular paronychia
  - Early can present with pustular folliculitis

38 year old female

- History of melanoma and dysplastic nevi
- First presented to our clinic in 2006 with a several year history of a pruritic rash involving the arms, legs, chest, back and neck
- Flared several times a year without full resolution
- Clinical suspicion of lichenoid dermatitis
BX# 2 in 2006

Diagnosis?
BX# 6 in 2008

- Clinical: Lichenoid-like eruption, not improving, not confirmed by biopsy
- DDX: Lichenoid process.
BX# 6

- **Skin, left arm:** Impetiginized crust with areas of superficial epidermal acantholysis and sparse dyskeratosis.
- **Comment:** The histologic differential diagnosis includes bullous impetigo or impetiginized Grover's disease. The presence of sparse dyskeratotic cells militates against an acantholytic process such as pemphigus. There is no evidence of a lichenoid infiltrate.

**Diagnosis?**
Final Pathologic Diagnosis:
Skin, right posterior neck, shave biopsy: lentigo-like epithelial pattern and some additional broader buds of epithelium.

Diagnosis?
Derm Group Conf 2015

- Skin, left arm: Impetiginized crust with areas of superficial epidermal acantholysis and sparse dyskeratosis.

- No family history and normal nails but... Darier's???
Diagnosis?
Galli-Galli disease is an acantholytic variant of Dowling-Degos disease: Additional genetic evidence in a German family.

Galli-Galli disease is an acantholytic variant of Dowling-Degos disease: Additional genetic evidence in a German family

Galli-Galli Disease

Her 29 year old son may have similar eruption

Reticulate Hyperpigmentary Disorders

- Rare genetic pigmentary abnormalities which include:
  - Reticulate acropigmentation of Kitamura
  - Dowling-Degos disease
  - Galli-Galli disease
Reticulate Hyperpigmentary Disorders

- Reticulate acropigmentation of Kitamura
- Dowling-Degos disease
  - Galli-Galli disease
- Dyschromatosis universalis hereditaria
- Dyschromatosis symmetrica hereditaria

Take Home

- Flash back
- Clinical Images
- Galli-Galli Disease
  - Darier’s like eruption and hyperpigmented macules
  - Adenoid proliferation of the rete with basal hyperpigmentation and acantholysis


11 year old male

- 3 week history of draining lesions on the left lower leg
- No history of trauma (does have a dirt bike)
- Treated with I&D, clindamycin, and sulfamethoxazole-trimethoprim by PCP

2 Weeks Later

- Image of the healed lesions
Special stains for organisms and subsequent suggested tissue culture were negative

Diagnosis?
Pyoderma Gangrenosum?

- No bowel symptoms and endoscopy/colonoscopy negative
- Developed firm subcutaneous nodule on the left chest

Special Stains Negative
Diagnosis?

4 months from onset
- Admitted for IV solumedrol
- Work-up for autoimmune and systemic granulomatous disease
Diagnosis?

Autoimmune and Systemic Granulomatous Disease Work-up

- Microscopic hematuria
- Proteinuria
- Pauci-inflammatory crescentic glomerulonephritis
- Anti-proteinase 3 ab 148.7
- C-ANCA pattern

**Diagnosis?**

**Granulomatosis with Polyangitis**
- Aka Wegener’s granulomatous
- C-ANCA positive in 81% with cutaneous disease
- Most in men in their 4th decade
- Upper and lower airway
- Renal involvement
- Skin involvement in up to 45%
  - May be the first manifestation 15%
Consider Wegener’s granulomatous especially with PG-like ulcers of unusual sites like face/neck/ear.
Granulomatous septal panniculitis

PNGD

9 months later
Wound care, mycophenolate and prednisone

Take Home Points
- Pyoderma gangrenosum in kids isn’t just IBD
- Granulomatosis with polyangiitis
  - PG-like lesions in unusual sites
  - Not just granulomatous vasculitis
  - Suppurative/acneiform
  - PNGD
  - Churg-Strauss necrotizing extravascular granuloma
13 yo febrile male with syncope and progressive weakness
Painful purpuric lesions of palms and soles
Facial rash for 1 month prior
Denied sexual activity and drug use
No N/V/abdominal pain, joint pain or headache
Work-up

- Negative blood cultures
- Anemia
- Transaminitis
- Proteinuria
- Increased ESR
- Hypocomplementemia

- Normal platelets
- Normal anti-myeloperoxidase and anti-proteinase antibodies

Work-up

- Positive Antibodies
  - DsDNA
  - SSA
  - Sm
  - RNP
  - Cardiolipin
**Libman-Sacks Endocarditis**
- Manifestation of SLE
- Verrucous, non-bacterial thrombotic endocarditis
- Usually silent but can result in embolic phenomena
- Associated with antiphospholipid antibody

**Take Home**
- Acral purpuric lesions due to pauci-inflammatory fibrin thrombi
- Coagulopathy
- Embolic (infectious endocarditis, cholesterol, atrial myxoma, aseptic endocarditis ...)

**44 year-old male**

**Clinical:** Erythematous scaling papules symmetrically on the forearms, abdomen and lower back

**DDX:** Allergic contact dermatitis vs drug rash vs irritant dermatitis vs scabies vs pigmented purpuric dermatitis
Follicular Hyperkeratosis

- Discoid lupus
- Pityriasis rubra pilaris
- Lichen sclerosus
- Keratosis pilaris
Follicular Hyperkeratosis

- Discoid lupus
- Pityriasis rubra pilaris
- Lichen sclerosus
- Keratosis pilaris

Vacuolar Interface

Skin, left volar forearm:
Interface dermatitis with follicular plugging

**Comment:** The microscopic differential diagnosis includes drug or viral eruption, connective tissue disease (including Wong type dermatomyositis), pigmented purpuric dermatoses, and pityriasis lichenoides.
Additional History

- ROS positive for weakness and dysphagia
- 2 weeks prior PCP noted enlarge lymph nodes in the neck
- Metastatic tonsillar squamous cell carcinoma

Wong-type dermatomyositis

- Variant of DM with erythematous hyperkeratotic follicular papules similar to PRP
- Reported in children and adults
- Myositis occurs at similar rate of classic DM
- In Wong’s original report 52% developed malignancy
  - Subsequent reports have shown no increased rate of malignancy
- Palmoplantar keratoderma is sometimes present

Matsumoto A, Wang R, Carlson J.A.

Take Home

PRP-like + Vacuolar Interface = Wong Type DM
32 year-old female

**DDx:** ? Scarring alopecia

![Sad Face Emoji]
**32 year-old female**

**Clinical:** Erythematous scalp with hair loss, scaling. Fungal culture was negative. Has Crohn’s. Patient on Remicaide.

**DDx:** ? Scarring alopecia.

- 5 months of scaling and hair loss
- Failed dandruff shampoo, econazole, and betamethasone
TNF-Inhibitor Induced Alopecia

- 3/2016: started remicade
- 10/2017: c/o scaling and hair loss

Psoriatic Alopecia/Alopecia Areata-Like Reactions Secondary to Anti-Tumor Necrosis Factor α Therapy: A Novel Cause of Noncicatricial Alopecia


Psoriasis + Alopecia Areata + Mixed Infiltrate


TABLE 1. Clinical Criteria for the Diagnosis of Anti-TNF-related Alopecia

1. Recent initiation of TNF inhibitor therapy
2. No prior history of psoriasis
3. Flare of psoriasis after starting the anti-TNF treatment
4. Alopecic patches on the scalp
5. Other, erythematous scaly patches and/or purplish lesions on the scalp and elsewhere on the body

Take Home

- **TNF Inhibitor delayed reactions**
  - Psoriasiform dermatitis
    - Often with interface, spongiosis and mixed infiltrate
  - Alopecia
    - AA histology with psoriasis, mixed infiltrate and atrophic sebaceous glands
30 year old male

- Diffuse erythematous papular and vesicular rash
Disseminated zoster?

Diagnosis?

vZV
Additional History

- 11/5 ER for coughing and shortness of breath along with drooping of right eyelid and double vision
- CT head: negative
- 11/6 Requested to return to ER for elevated D-dimer
- CT chest/abdomen: abdominal mass and liver lesions
- Lost 20 pounds in 5 weeks
- 11/10 Rash started 5 days ago on abdomen, back and groin so PCP started valtrex

Additional History 11/21

- Worsening SOB and rash
- Admitted to outlying hospital
- Inconclusive liver biopsy
- Transferred to GMC based on telederm consult
**Additional History**

- Lymph node and liver biopsy suspicious for malignancy possibly of histiocytic/dendritic cell origin
- BM negative

**Paraneoplastic Pemphigus**

- Most with NHL but also CLL, Castleman disease, thymoma, Waldenstrom
- Usually presents with severe painful oral ulcers
- Can involve conjunctiva with symblepharon and lungs with obstructive disease

**Paraneoplastic Pemphigus**

- Autoantibodies to plakin family: envoplakin (210-kd), periplakin (190-kd), bullous pemphigoid antigen I (230-kd), desmoplakin I (250-kd), desmoplakin II (210-kd), plectin (500-kd), and alpha2-macroglobulin-like-1 (170-kd)
- Can also exhibit antibodies to antigens associated with pemphigus vulgaris (desmoglein 3, 130-kd) and pemphigus foliaceus (desmoglein 1, 160-kd)
Take Home

- PNP can be herpetiform
- Oral involvement is not always first or the most severe
- Pemphigus can show a punctate intercellular pattern on DIF

Mimics and Masquerades
Pearls from Wound Care
Ashley Group, MD
Associate Professor of Dermatology
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Disclosures

I have no conflicts of interest to disclose.

TIPS ON HEALING SKIN LESIONS
If it’s Wet... Dry It.
(Apply Dry Dressing)
If it’s Dry... Wet It.
Pearl #1
Debridement can help wounds heal!
Know how to do it and what your options are.

Types of Debridement
• Sharp
• Enzymatic
• Autolytic
• Maggot therapy

77 year old male – traumatic ulcer on shin present 3-4 months. Occurred after falling off chair and hitting his leg.
Sharp Debridement

- Apply topical anesthetic
- Cover with a gauze or film dressing or saran wrap
- Let sit for 20-30 minutes
- Use instrument of choice

50 year old male – 4 days ago he accidentally spilled boiling water on his foot
Hypergranulation Tissue Treatment

5 weeks later
- s/p 3 silver nitrate treatments

Sharp Debridement
- What if the fibrinous debris is too adherent, or too deep to gently remove with topical lidocaine jelly?

and/or
Sharp Debridement

Pre-scissor debridement

Post-scissor/curette debridement

Debridement Coding

- 97597 – Sharp selective debridement of necrotic tissue of an open wound with scissors, scalpel, and forceps (i.e. removal of fibres, devitalized epidermis, and/or dermis, exudate, debris, biofilm)
- Debridement codes have zero global surgical days, but they should only be reported once a week.
**Documentation Must Have's**

- Anatomical site treated
- Measurement of surface area
- Was anesthesia required?
- Surgical method performed
- Type and depth of tissue removed
- Depth of lesion
- Wound area
- Wound edges
- Wound type

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72 year old CF who slammed the toilet seat down on her finger 1 month ago. Painful, but she thinks it is healing slowly.

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**Enzymatic Debridement**

Collagenase – FDA approved

- Selectively removes necrotic tissue
- Apply 2 mm thickness to wound
- Keep wound moist
- Change dressing daily (ideally)
- Silver or iodine containing dressings may make it less effective
Enzymatic Debridement

**Tip**
- If there is a thick, dry eschar, you can lightly score the eschar with a #15 blade
- This helps collagenase to penetrate

**Disclaimer**
- Collagenase ointment can be very expensive!
- It is brand name only and isn’t covered by all insurances

Enzymatic Debridement

This is also helpful when needing to debride deeper wounds or wounds post-sharp debridement.

33 year old male with 5 week history of a wound on the leg, thought to be due to spider bite.

3 weeks later
Autolytic Debridement

What is this?
The patient’s own phagocytic cells and endogenous enzymes help debride the wound.

What kind of dressings may help?

Hydrocolloids

Hydrogels

Hydrogels

• Composed of up to 96% water → maintain a moist environment
• Donate moisture to rehydrate dry wounds
• Soothing & cooling
• Promotes autolytic debridement

Hydrogels

• You can cut to size
• Needs a secondary dressing
Hydrogel Border

- Border dressings should not be cut
- No need for secondary dressing

Autolytic Debridement

- MEDIHONEY Dressings
  - Debride acute wounds, chronic wounds, and burns
  - MOA: high osmolality
  - Promotes outflow of wound fluid that helps to debride non-viable tissue

MEDIHONEY Case

79 year old male – traumatic ulcer developed after falling down stairs. Present for several months. Healing skilled so referred.
MEDIHONEY Case + Sharp Debridement

79 year old CF with severe arterial insufficiency had a traumatic wound to the leg after hitting her leg on a bucket 1 week later

The best part…

it’s very affordable!

Case:

- 72 year old CM with DM2 and venous insufficiency
- Non-healing wounds and recurrent cellulitis of left foot x 4 months
- Chronic Gram (–) toe web infection
  - Tissue culture grew Pseudomonas
- Management:
  - Oral antibiotics (levofloxacin)
  - Sharp debridement
  - Enzymatic debridement – collagenase
  - Compression
Minimal improvement after 4 months

Maggot Debridement Therapy

Indication:
For debridement of non-healing necrotic skin and soft tissue wounds, including pressure ulcers, venous stasis ulcers, neuropathic foot ulcers, and non-healing traumatic or post-surgical wounds

- Sterile, lab grown larvae
- Bagged vs free range

Maggot Debridement Therapy

- One bag of medical maggots was applied x 4 days
- Zinc oxide applied to peri-wound skin
- Covered larvae with saline moistened gauze, kerlix, and coban (changed dressing daily)

*Important not to get the dressing wet!

*Also, important not to occlude with bag while showering.
Maggot Debridement Therapy

After 1 day  
After 2 days

Maggot Debridement Therapy

After 3 days  
After 4 days - removal

MDT – How do they do it?

20 days after removal

1. Selectively remove necrotic tissue
   • Secret proteolytic digestive enzymes
   • Liquefy necrotic tissue
   • Mechanical – mouth hooks + body spines
   • Loosen necrotic tissue

2. Reduce bacterial burden
   • Antimicrobial substances inhibit growth of G+ and G- organisms
   • Including MRSA and Pseudomonas aeruginosa
   • Breakdown & prevent biofilms

3. Anti-inflammatory
   • Excretions/secretions reduce complement

4. Promote angiogenesis via salivary secretions
   • Increase granulation tissue

Pearl #2
Use compression! (when appropriate)

Before Applying Compression
• Check an ABI to exclude arterial disease
  – ABI < 0.8 - indicates arterial disease - use caution when applying compression
  – ABI < 0.5 - compression is contraindicated

• Ask about CHF

Types of compression
• Multi-layer compression wraps
• Tubular dressing
• Stockings
  • Single layer
  • Dual layer
• Pneumatic compression devices
**Multi-layer Compression Wraps**

- Unna boot
  - Short stretch (inelastic)
  - Best for mobile patients - encourage exercise!

**What do you need for an Unna Boot?**

- Unna boot – calamine and zinc oxide impregnated gauze
- Kerlix gauze
- Coban

**3 layer wrap**

**How to Apply an Unna Boot**

1. Dress wound
2. Wrap Unna boot
3. Wrap Kerlix
4. Wrap Coban

- Wrap from the distal foot all the way up to 2 cm below the knee. Include the heel.
- Initially change twice weekly, then as edema & drainage improve can change weekly (unless heavy drainage).
- Keep it dry
- Apply compression at 30-40 mmHg
Prescribed Exercise With Compression vs Compression Alone in Treating Patients With Venous Leg Ulcers
A Systematic Review and Meta-analysis

• Found exercise to be associated with increased VLE healing at 12 weeks
• Progressive resistance exercise + physical activity is most effective
  • Calf muscle strengthening – i.e. heel raises
  • Walking – i.e. .30 minutes 3x/week

• SO...recommend exercise!

Multi-layer Compression Wraps

• Profore
  • 4 layer wrap
  • Long stretch (elastic) – this will work for mobile or non-mobile patients
  • Lite version (3 layers)

• Coban 2 layer compression systems
  • Short stretch design
  • Provides compression at 35-40 mmHg
  • Indicated for patients with ABI >= 0.8
  • Lite version
    • 25% decreased resting pressure – 25-30 mmHg
    • Good for patients with poor tolerance or ABI >= 0.5

Tubular Dressings

Tubigrip
  • Elastic tubular dressing
  • Gives light compression
  • When applied in a double layer to an appropriate size limb, exerts 10-15 mmHg
  • Multiple sizes, A-G
Stockings

- Apply when they get up in the morning, take off at bedtime
- Replace every 6 months
- Medicare will cover if:
  - Patient has an open venous leg ulcer
  - 30-40 mmHg or higher

“I can’t get my compression stockings on”

Jobst Stocking Donner

Emy-6 compression stocking applicator

- Comes in S, M, L

Compression

- They also make “dual layer” stockings
  - Easier to apply
  - 1st sock – 10-20 mmHg
  - 2nd sock – 20-30 mmHg
  - Combined effect ➔ greater compression

- Velcro wraps
  - Adjustable level of compression
  - I.e.
    - Circuid Justadite lower leg
    - Farrow wraps
Compression

- Pneumatic compression devices
- Indicated for lymphedema

How can you diagnose lymphedema?

Pearl #3

Help patients with their wound care at home
Order dressings or home health
Who might need wound care/dressings at home?

- Hidradenitis suppurativa
- Bullous pemphigoid (and other bullous disorders)
- Epidermolysis bullosa
- Pressure ulcers
- Leg ulcers
- Neurotic excoriations

Can they do wound care by themselves?

Home Health

- Do they already have home health?
  - Just send a new order to their company

- If not, have they had home health before? Do they have a preference on company?

- Several options – look for one with “skilled nursing services”
Hyperbaric oxygen can help treat some (specific) surgical complications

Approved Indications for HBO₂

- Air or gas embolism
- Carbon monoxide poisoning, +/- cyanide poisoning
- Clostridial myositis and myonecrosis (gas gangrene)
- Crush injury, compartment syndromes, & other acute ischemias
- Enhancement of selected problem wounds:
  - Arterial insufficiencies
  - Wagner grade III or higher diabetic wound (deep ulcer w/osteomyelitis or abscess)
- Exceptional blood loss anemia
- Intracranial abscess
- Necrotizing soft tissue infections
- Chronic refractory osteomyelitis
- Delayed radiation injury
- Thermal burns (not approved by Medicare)
- Decompression sickness
- Compromised skin grafts and flaps
What is hyperbaric oxygen?

• This is a treatment in which a patient breathes 100% oxygen intermittently while inside a pressurized (>1.4 ATA) hyperbaric chamber.

Delayed Radiation Injury

• Develops after a latent period of 6 months or more after radiation exposure
  • Can be seen many years later
• Previously irradiated tissue is:
  • Hypoxic
  • Hypovascular
  • Hypocellular

• Often precipitated by additional tissue insult → surgery or trauma within radiation field
  • Surgical sites may not heal
  • Wounds may have late dehiscence
  • Prepare for complications when cutting into previously irradiated tissue!

• Intermittent O2 tension during HBO, can:
  • Stimulate collagen synthesis & fibroblast proliferation
  • Induce angiogenesis
  • Enhance cellular proliferation

Photo courtesy of Dr. Sorabh Kandelwaahl
Failing Flaps and Autologous Grafts

**HBO₂ Improves Flap/Graft Survival by:**

- Provides immediate hyperoxygenation of blood plasma → increases perfusion of ischemic tissue
- Stimulates new blood vessel growth – angiogenesis + vasculogenesis
- Upregulates fibroblast activity → collagen deposition
- Alters flap circulatory dynamics:
  - blood flow, edema → decreased congestion

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**HBO₂ therapy is time dependent!!**
- (with respect to salvage)
- Refer urgently at the first sign of flap/graft failure
  - Grafts - dusky appearance, epidermolysis, necrosis
  - Flaps: arterial insufficiency - pale, delayed capillary refill, decreased turgor, cool temp
    - venous congestion - bluish-purple, brisk capillary refill, increased turgor, warm temp
    - complete arterial/venous occlusion

- BID treatments
- Once flap/graft is viable/stable, decrease to daily treatments
- Usually 10-30 treatments

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**Same day – s/p 1 day of BID HBO₂**

Photos courtesy of Dr. Jeannie Le

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Compromised Flap Salvaged with HBO₂

- Urgent start
- BID treatments x 6 days, then daily
- Total of 18 HBO₂ treatments

- Key point
  - Don’t wait for outright dehiscence
  - The earlier the start, the better the salvage

Failing Skin Graft (from dog bite)

Before HBO₂  After HBO₂

Photos courtesy of Dr. Jeannie Le

Threatened Flap (from abdominoplasty)

Before HBO₂  After HBO₂

Photos courtesy of the University of Pennsylvania, per Dr. Jeannie Le
Pearl #5
Dietary Tips

Horse Chestnut Seed Extract

- Escin → active component
  - May overall prevent vascular leakage
    - Inhibits elastase & hyaluronidase
    - May prevent leukocyte activation
  - Improves signs & symptoms of CVI
    - Decreases leg pain
    - Decreases leg volume/swelling
  - Safe and well tolerated
    - Adverse events were mild and infrequent
      - Pruritus, GI issues, headache, nausea, dizziness in 0.7% of patients

Protein and Wound Healing

- Protein needs increase when wounds are present
- Optimal wound healing requires adequate nutrition
- Increased protein intake can accelerate healing of chronic wounds

$15.98 on Amazon
How do you get the right amount of protein?

Common Foods with at least 25 g – 2 servings per day

- 4 oz salmon/flounder
- 8 shrimp
- ½ cup canned tuna
- 4 oz chicken/turkey breast
- 3 oz beef
- 3 oz nuts
- 4 oz cheese
- 1 cup cottage cheese
- 4 eggs
- 3 cups mild
- 2 cups pinto beans
- 6 tbsp peanut butter
- 2 cups Greek yogurt

Other ways to get increased protein

Available at Walmart, Costco, Sam's, Amazon

- 12 pack for $22
- 12 pack for $20
- 12 pack for $17
- 12 pack for $24

Ready-made meal ideas
Summary

1. Debridement can help wounds heal
2. Use compression, when appropriate
3. Help patients with their wound care at home – order dressings or home health
4. Consider hyperbaric oxygen for failing flaps and grafts, as well as wound healing problems in areas of previously irradiated skin.
5. Recommend increased protein intake to speed wound healing
Survival Guide to Inpatient Consults

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DISCLOSURE OF RELEVANT RELATIONSHIPS WITH INDUSTRY:
I have no relevant relationships with industry to disclose

Consultative Dermatology—Objectives

I. Discuss the impact of dermatology consultation on the care of medically complex hospitalized patients

II. Discuss ways in which the dermatology consultant can be an effective contributor to patient management

III. Use case examples to review high-yield inpatient dermatology topics, with diagnosis and management pearls
Background

- Dermatology is an outpatient specialty: less than half of dermatologists in the US see inpatient consults, and most spend less than 1 hr/week doing so

- This means the care of some of the most complex dermatology cases has shifted to non-dermatologists in the hospital setting

Background

- Is this a problem?

- Non-dermatologists are often uncomfortable recognizing and treating cutaneous diseases
  - May not perform a comprehensive skin exam
  - Lack training to know what to look for
  - May not appreciate significance of specific skin findings

Background

- The primary team may not identify 77% of skin findings

- Dermatology consultation appears to change the final diagnosis ~60-70% of the time
Impact of Dermatology Consultation

- 204 of 902 (23%) of new consults from hematology / bone marrow transplant service
- 52% neutropenic; 30% bone marrow transplant recipients → Complexity of cases
- In 121 (59%) of the cases, the final dermatologic diagnosis was not included in the Dx of the primary team

Impact of Dermatology Consultation

- Among presumed cases of cellulitis, ~30% can be re-classified as pseudocellulitis, ~90% of these can stop unnecessary antibiotics, and half can be discharged from inpatient or observation units
- Dermatology consultation may reduce length of stay and readmission rate among those with inflammatory skin conditions

Background

- This is an indication of the value of a dermatology consult and, for dermatologists and other sub-specialists, the importance of serving in this capacity (as a consultant)
- ...as well as the importance of improving dermatology training for medical students, residents, and colleagues in other specialties
Keys to successful inpatient consultation...

I. Know when a situation demands your urgent attention; relish opportunities to make a real difference for your patients

II. Don't be afraid to advocate for your patients

III. Don't sell yourself short; never assume the primary team "knows better"

IV. Put your nickel down, but state out loud what you might be missing

→ Put yourself in a position to end up right even when you are wrong

Case 1

A 56 yo man with refractory AML on salvage chemotherapy develops a violaceous plaque on the anterior neck that expands rapidly over the course of one day

Febrile and neutropenic with negative cultures

On empiric meropenem, vancomycin, voriconazole
What should be on the differential diagnosis here?

In this profoundly immunosuppressed patient, what can you not afford to miss?

What would you do next?
- A. Draw repeat blood cultures
- B. Order a CT scan of the head and neck
- C. Perform a skin biopsy for frozen section processing
- D. Perform a skin biopsy for routine permanent section processing

Case 1
Angioinvasive mold, likely mucormycosis Localized, or disseminated?
Rapid diagnosis crucial to guide antibiotic coverage, poss. debridement
Zygomycetes are resistant to voriconazole (may be a risk factor)

"Bull's-eye infarct" of zygomycosis
Trichosporonosis

- Growing cause of opportunistic infection
- Closely related to Cryptococcus
- Serum cryptococcal antigen is often positive

Photo courtesy of Dr. Evan Piette

*Bull’s-eye infarct* of zygomycosis
Trichosporonosis
• KOH examination
• Septate hyphae and spores

Photo courtesy of Dr. Evan Piette

Aspergillus
• Soil saprophyte
• Lungs, sinuses
• Skin lesions disseminated or primary cutaneous
• Risk factors: Immunosuppression (neutropenia, steroids)

Aspergillus (touch prep) → rapid diagnosis informing management decisions in real-time
Case 2

23yo man with refractory ALL admitted for salvage chemotherapy

Neutropenic and febrile

Developed tender swelling of the lower leg; faint purpura but otherwise unremarkable

Stenotrophomonas
Stenotrophomonas:
• Most common in neutropenic patients on broad-spectrum Abx
• Highly resistant
• High-dose TMP-SMX is first-line therapy

In this case, tissue culture identified the organism 48hrs before the blood cultures were positive.

**Keys to successful inpatient consultation...**

I. Know when a situation demands your urgent attention; relish opportunities to make a real difference for your patients (and colleagues)
   ➔ Looking for guidance; “ruling out” sometimes as important as “ruling in”

II. Don’t be afraid to advocate for your patients

III. Don’t sell yourself short; never assume the primary medical team “knows better”

IV. Put your nickel down, but state out loud what you might be missing
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**Case 3**

52yo man with no significant past medical history presents to an outside hospital with a “bug bite” on the right hand
Case 3

- He is treated with broad-spectrum antibiotics for possible cellulitis
- The site becomes necrotic, and he goes for surgical debridement
- The surgical wound breaks down and becomes necrotic again
- He undergoes repeated debridements and then amputations
- Blood and tissue cultures repeatedly negative
- Ultimately transferred to Penn with a request for hyperbaric oxygen therapy
When something does not respond to a particular treatment as you expect it to…

...re-evaluate and re-assess; do you have the right diagnosis?

Case 3

• Presentation at the time of transfer notable for:
  – HR 123, BP 90/50s, RR 30, O2 93% on 4L nasal cannula
  – Confusion / altered mental status
  – WBC count 41 (89% neutrophils); diffuse pulmonary infiltrates on chest x-ray

Meeting Sepsis / SIRS criteria
Is this patient infected?
A) Yes  
B) No

Case 3—Pyoderma Gangrenosum

Clues suggestive of PG:
- Clinical: gun-metal gray, violaceous, necrotic border, undermined ulcer
Clues suggestive of PG:
- **Clinical:** gun-metal gray, violaceous, necrotic border, undermined ulcer
- **History:** pathergy, a worsening or induction of new lesions due to trauma (only 1/3 of PG patients, but very characteristic)

Case 3—Pyoderma Gangrenosum

**PG work-up:**
1. Perform biopsy and tissue culture of violaceous border to rule out other diagnoses
Case 3—Pyoderma Gangrenosum

PG work-up:

1. Perform biopsy and tissue culture of violaceous border to rule out other diagnoses

2. Screen for underlying medical conditions with physical exam and ROS (ask about GI, joint symptoms), ESR, CRP, CMP, CBC, blood smear, colonoscopy, ANCA

JAMA Dermatol. 2018 Apr;115(4):409-413
Patient with myelodysplastic syndrome (MDS) who developed PG at the site of port placement.

PG was bullous type PG (60% of cases due to malignancy).

Bone marrow biopsy revealed new finding of 20% blasts (AML).

Pyoderma gangrenosum heralding transformation of MDS into AML.

Always ask the “why here, why now?” question.

What we see on the skin may indicate an underlying disorder.

Case 3—Pyoderma Gangrenosum

Management:
- Two critical issues:
  1. Patient with SIRS/Sepsis (why?)
  2. Patient has lost R hand and now has limb-threatening L arm ulcer

Do you wait for biopsy and tissue culture to come back before treating?

A) Yes
B) No
Case 3—Pyoderma Gangrenosum

Management:

• SIRS/Sepsis:
  – Could he have a systemic infection?
  – Or, could he have a significant inflammatory response to PG itself?


Case 3—Pyoderma Gangrenosum

Management:

– PG can affect the lungs, liver, heart, bones, eyes, etc.
– It can cause a capillary leak phenomenon due to overwhelming inflammation
– Leukocytosis, confusion, tachypnea, pulm infiltrates, etc. can all be explained by PG
– PG itself can cause a patient to be systemically ill


Spectrum of Neutrophilic Dermatoses

Sweet syndrome:
– Acute febrile neutrophilic dermatosis
– Can also involve the joints, bones, liver, heart, spleen, kidneys
– Pulmonary manifestations can lead to respiratory failure
– Can develop SIRS and shock
– …and even be life-threatening
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Spectrum of Neutrophilic Dermatoses

Case 3—Pyoderma Gangrenosum

Management:
- In addition to causing systemic illness, this is also a limb-threatening process:
  - Rapid expansion of ulcer following biopsy (pathergy)
  - Deep, enlarging ulcer with major functional implications
Management:
• So, we know the cost of inaction

• What about the cost of action (i.e. high-dose steroids) when there is concern for infection?

Case 3—Pyoderma Gangrenosum

Management:
• Steroids can be used safely in infection
  – Especially when the patient is receiving treatment for a known or suspected infection
  – Steroids are given for pressor-refractory sepsis
  – Since PG responds rapidly to steroids, we usually know quickly whether we are right

• Ruling out other diagnoses is important, but quick action may be critical

• Have the courage to think through the possibilities, costs, and benefits, and take a stand (this usually means a discussion with the primary team)
After one day of 1.5mg/kg prednisone divided twice daily

Rapid improvement of the ulcer
Resolution of respiratory distress, confusion, and SIRS
Use the inflammatory, violaceous border to gauge response to therapy and guide steroid taper.

Wound breakdown following hernia repair; no response to antibiotics and debridement.

Five days after starting high-dose prednisone, the ulcer "looks great."
Back to our patient…
Complete healing after ~5 weeks
Even the largest of ulcers can heal; successful treatment of PG can be extremely rewarding.
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SJS/TEN

What key advocacy roles can the dermatologist play in SJS/TEN?

- Make the correct diagnosis
  - In one study, 1/3 of those biopsied received an alternate diagnosis
- Identify and stop the culprit drug
  - Earlier withdrawal = better
  - OR of death = 0.69 for each day sooner
  - OR of death = 4.9 for drugs with long half lives
- Coordinate management (ICU or burn unit)
  - In one study, mortality = 32%
  - Mortality of those transferred after > 1 week = 51%
**SJS/TEN**

The level of care is a surrogate for supportive care:

- Fluids
- Electrolytes
- Temperature
- Nutrition
- Pain control
- Airway maintenance
- Wound care
- Infection surveillance
- Ophthalmology
- Gynecology
- Urology
- Hematology

*Role of dermatology in 1) diagnosis, 2) identification of culprit drug, and as 3) team “quarterback”

*Overall critical role in patient advocacy*

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**Case 4**

Palpable purpura in 86yo patient with GI bleed, new lung cancer

Biopsy showed small vessel vasculitis

DIF showed perivascular IgA

* IgA Vasculitis (aka Henoch-Schonlein purpura)
When a patient presents with lesions suspicious for vasculitis, initial work-up should try to answer three basic questions:

1) Are the lesions due to vasculitis?
2) Are there findings which help establish a particular diagnosis?
3) Are other organ systems involved?

IgA Vasculitis
Initial presentation often indistinguishable from other small vessel vasculitis
Abdominal pain, bleeding (65%), arthralgia/arthritis (63%)
IgA-associated glomerulonephritis (40%)
IgA Vasculitis

Initial presentation often indistinguishable from other small vessel vasculitis
Abdominal pain, bleeding (65%), arthralgias/arthritics (63%)
IgA-associated glomerulonephritis (40%)
UA w/ micro shows 50-100 RBCs
Not followed up / urine sediment not examined

IgA Vasculitis

Given 10mg daily prednisone from rheumatology for his skin
Presents after 1 month to dermatology clinic
Skin is clear, but repeat UA shows packed RBCs
UProt/Cr ratio = 10, Cr = 2
Arranged urgent evaluation by nephrology, diagnosed with glomerulonephritis

IgA Vasculitis (Henoch-Schönlein Purpura)

Most common in children
- Viral URI or Strep pharyngitis often precede onset by 1-2w
- Overall, 40% due to infection
- Drug trigger in around 20%
- Consider paraneoplasia in adults (90% male)

Medicine 2014;93(2):106-13
• Factors predicting renal involvement:
  • Age older than 6
  • Persistent purpura
  • Severe abdominal pain
  • Purpura above the umbilicus (controversial)
  • Renal symptoms at the time of diagnosis

IgA Vasculitis—Prognosis

• Prognosis of IgA vasculitis is generally favorable but depends on the severity of renal disease

• Close follow-up, with frequent repeat UA and BP monitoring while the rash is present, then monthly up to 6 months (renal involvement usually develops within 1 month)

• Usual therapy for IgA nephropathy is ACE inhibitor, steroids depending on degree of proteinuria, renal dysfunction; mycophenolate often used as steroid-sparing agent

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III. Don’t sell yourself short; never assume the primary team "knows better"
  → Don’t be afraid to help guide medical management when appropriate

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Case 5

- 54yo woman with GERD presents with purpuric macules and papules on the legs
- Seen multiple providers with multisystem complaints, including weakness, arthralgias, myalgias, weight loss, malaise, gingival bleeding
- Consulted for “vasculitis”

- Diagnostic key is purpura not quite typical for vasculitis—better described as perifollicular hemorrhage
- Corkscrew hairs
- Restricted diet due to reflux disease

Clinical diagnosis = Scurvy

Vitamin C level < 5umol/L (23-114 umol/L)
Case 5

- Within days, before results available, the patient developed seizures and was admitted to the ICU in status epilepticus
- Discussed with ICU team and able to argue successfully for IV vitamin C, after which the patient improved

Scurvy

- Vitamin C is an essential co-factor in collagen production
- Deficiency leads to corkscrew hairs, follicular hyperkeratosis, perifollicular hemorrhage
- Gingivitis, ecchymosis, impaired wound healing
- Fatigue, depression, bone and joint pain due to subperiosteal hemorrhage

Scurvy

- Can cause neuropsychiatric, cardiovascular, and pulmonary symptoms
- In late stages, may suffer cerebral hemorrhage and seizures (as in this case)
- Can be fatal if left untreated

Start vitamin C supplementation while awaiting laboratory confirmation.
Case 5

- Diagnostic momentum can be hard to slow
- Don’t be afraid to challenge assumptions or make treatment recommendations based on your concerns
- Be humble, but don’t doubt your insights as a specialist

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Case 6

A 63yo man with sarcoidosis s/p heart transplantation, on prednisone and tacrolimus, presents with headache and a pustular eruption on the shoulders

Recently diagnosed with prostatitis and discharged; returning with altered mental status
Case 6

What would you do next?

Is this folliculitis? Acne? Herpes?

Diagnosis not really clear clinically, but given the atypical appearance and immunosuppression, decided to:

- Biopsy for H&E and tissue culture
- Send HSV PCR, culture of pustule
- Perform Tzanck smear

Tzanck smear showing Cryptococcus
Case 6

- Cutaneous crypto is disseminated crypto until proven otherwise
- CSF studies, serum crypto antigen sent and also positive
- Amphotericin plus flucytosine → fluconazole

Disseminated crypto in an AIDS patient

Disseminated crypto in an AIDS patient
Case 7

45yo man with a history of heart transplantation in 2006 on tacrolimus, prednisone, and mycophenolate

Admitted with large erythematous patch on the lower leg, extending to the right medial thigh; WBC count of 14.6

Treated with cefepime and vancomycin for presumed cellulitis

Redness regressed but did not completely resolve, leaving behind two red-brown patches
Case 7

Patient clinically well-appearing

Provided a history that the cellulitis had improved markedly

Residual red-brown pigmentation attributed to hemorrhage into cellulitis, +/- dependency, perhaps some incidental trauma

Decided to continue current management but made a plan to re-evaluate and biopsy if not improving; reached out to ID to discuss
Case 7

- Lumbar puncture revealed elevated opening pressure and positive crypto antigen
- Started on 2 weeks of amphotericin, with improvement in cellulitis, and discharged on maintenance fluconazole

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4. Put your nickel down, but state out loud what you might be missing
   ➔ Put yourself in a position to end up right even when you are wrong
Don’t be beholden to yesterday’s (or someone else’s) Dx; avoid premature closure bias

> Gather sufficient information
> Develop a differential
> Identify and investigate “red flags” appropriately
> Consider the worst case scenario—what you don’t want to miss
> Consult with a colleague

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Consultative Dermatology—Summary

I. Dermatology, like other sub-specialties, encompasses a highly specialized body of knowledge

II. We have a tremendous ability to shape patient care but must be present and available to do so

III. Our reward is helping patients and guiding our colleagues like no one else can, and seeing amazing cases in the process
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The Dermatology Foundation has supported & advanced my career.

Thank you

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