The Other Psoriasis: Strategies *for* Recognition *and* Diagnosis *of* Pustular Psoriasis

Provided by AAPA American Academy of PAs and The France Foundation

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Disclosures

Activity Staff Disclosures

- The planners, reviewers, editors, staff, CME committee, or other members at the AAPA and TFF who control content have no relevant financial relationships to disclose

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- Leigh Ann Pansch, MSN, FNP-BC, DCNP- Consultant-Novartis, Pfizer; Speaker's Bureau-Abbvie, Eli Lilly, and Sanofi-Regeneron

When you are done with this module….

- You should be able to compare and contrast the different pathophysiologic mechanisms of generalized pustular psoriasis (GPP) and palmoplantar pustulosis (PPP) versus plaque psoriasis

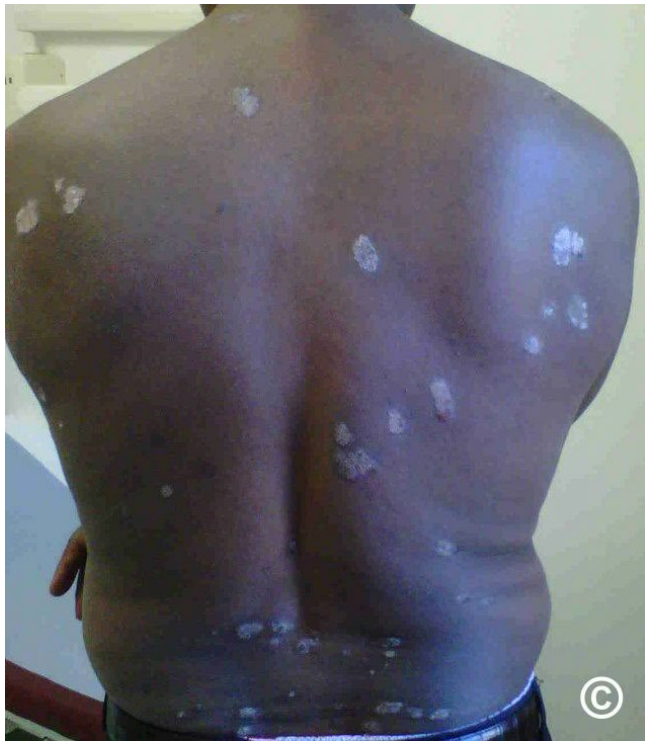
- You should be able to distinguish among the clinical presentations of GPP, PPP, and plaque psoriasis for diagnosis

- You should be able to formulate a plan describing the steps to urgently refer patients with GPP and PPP to a specialist

Question 1: Which of the following is thought to be a driver of pustular psoriasis but less so in plaque psoriasis?

A. IL-17
B. TNF-α
C. T-cells and neutrophils
D. IL-36

Question 2: Which of the following is more likely to be pustular psoriasis?



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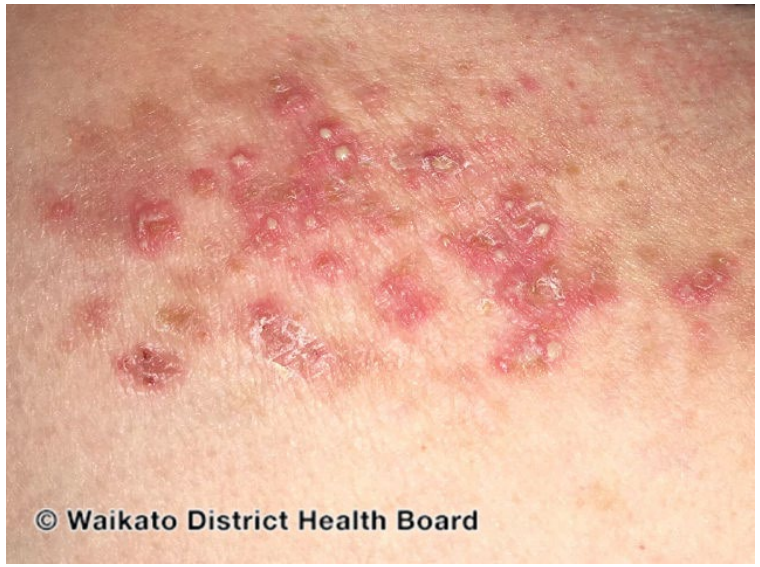

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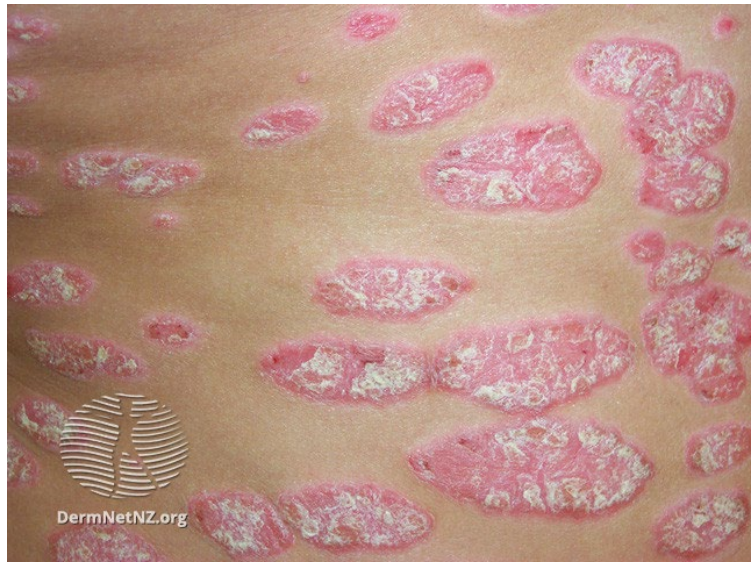

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Question 3

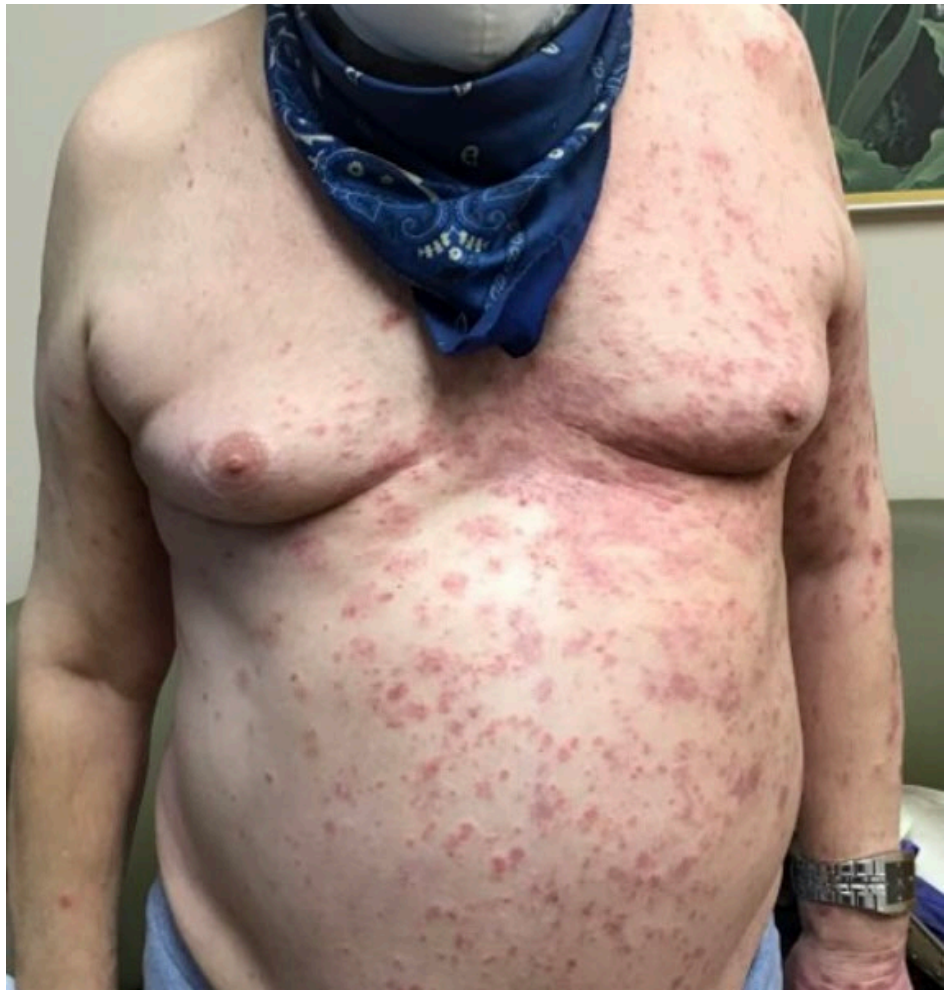
Tammy is a 43-year-old woman who reports to your primary practice presenting with a rash on her trunk with pustules on her palms and arches of her feet. Four weeks ago she was diagnosed with *Influenza A* and has a long history of plaque psoriasis. Her dermatologist has her on systemic adalimumab for control of her psoriasis.

What would be your next step?

A. Begin treatment with corticosteroids
B. Change the systemic biologic she is on for treatment of her plaque psoriasis
C. Immediate referral to her dermatologist
D. Watch and wait for pustule clearance
E. Prescribe broad-spectrum antibiotics

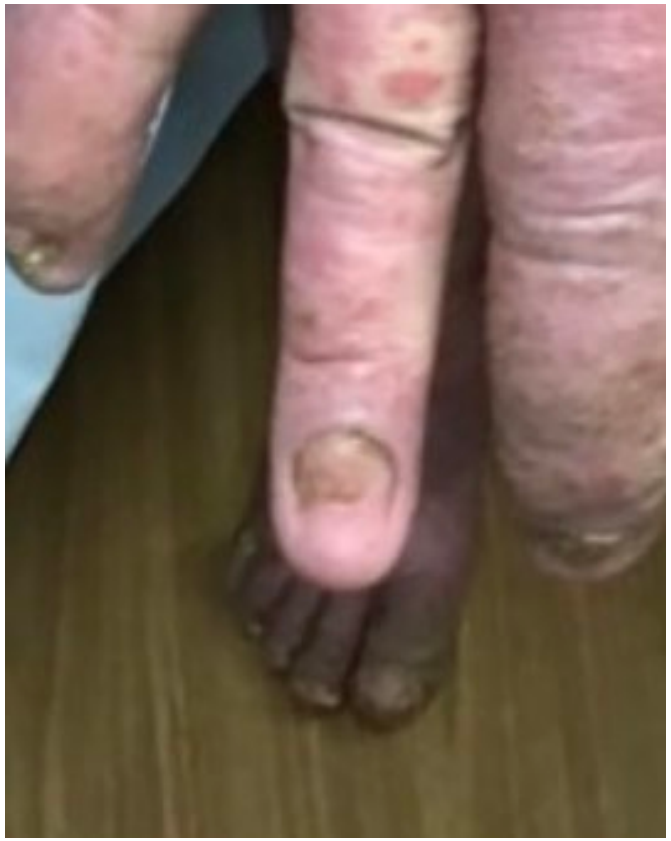
Meet Mike

- 56-year old male patient
- Worsening rash with pustules on his palms, soles, and new areas of rash
- Afebrile
- Cough (dry and persistent)
- Malaise
- Joint pain, severe sensitivity of fingertips and toe tips
- Onycholysis and anonychia

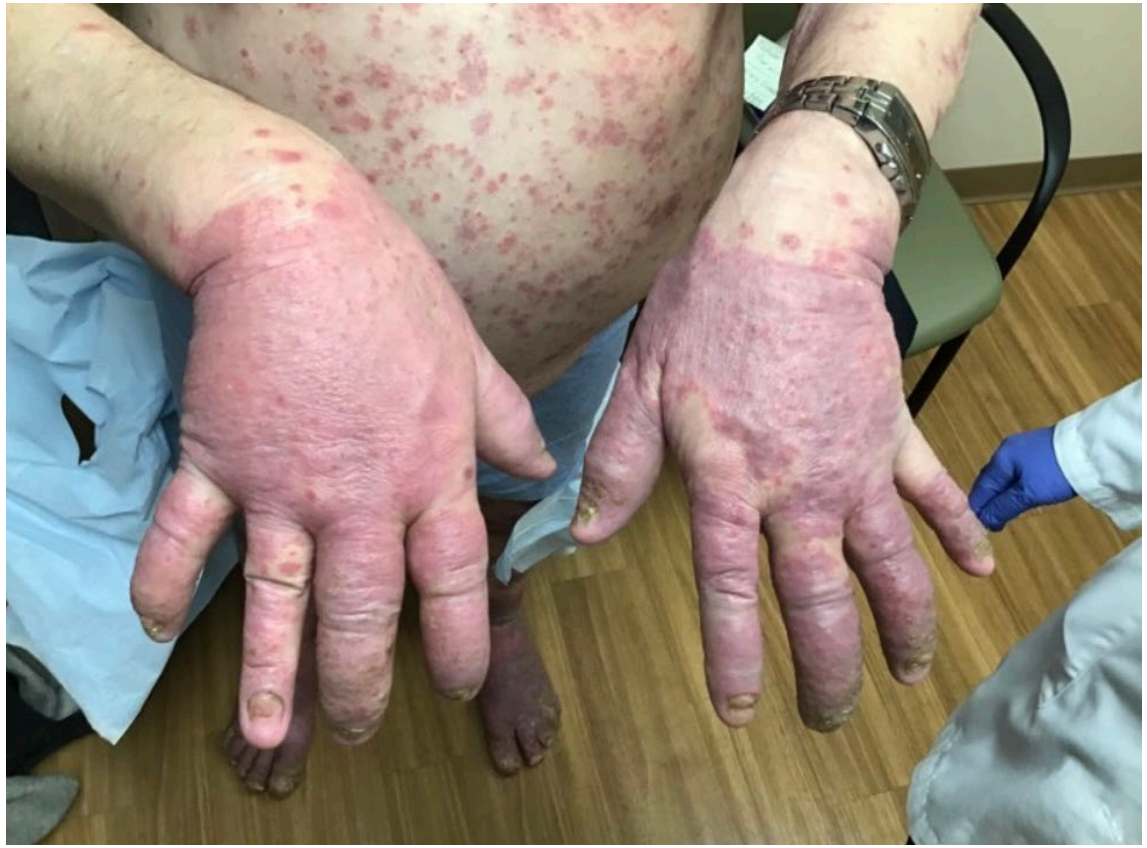


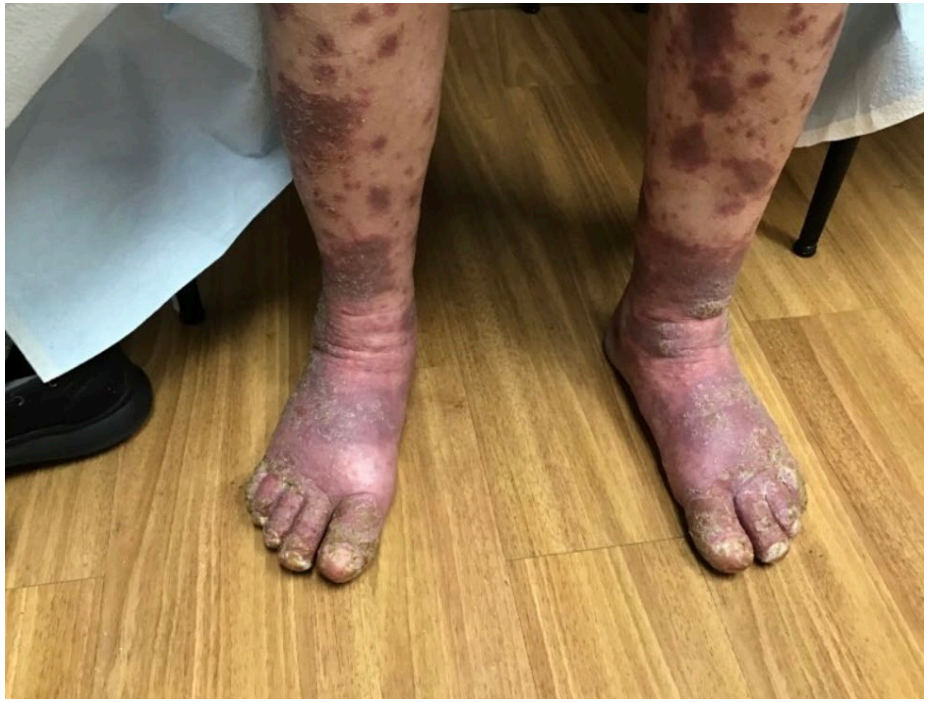
Meet Mike

- History:
  - Long time smoker, 1-2 pack(s)/day
  - History of plaque psoriasis
- Recent medical history:
  - Urgent care: diagnosed with bronchitis
  - Given oral and IM steroids
  - Developed rash, fever, joint pain, and returned to urgent care
  - Given second round of oral and IM steroids
  - Referred to dermatology



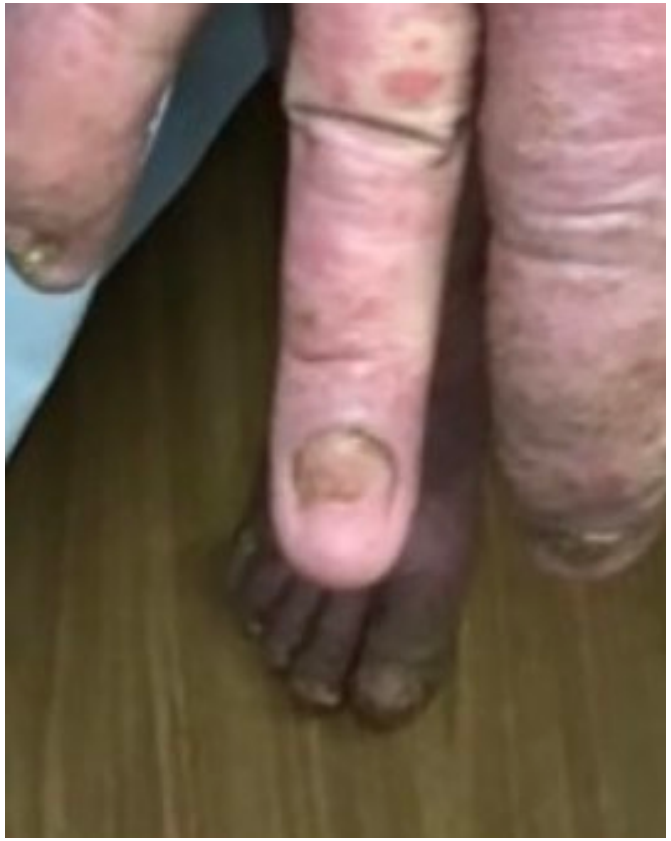
Erythema and Dactylitis





Differential Diagnoses

Differential Diagnoses	Rationale
Pemphigus foliaceus	Less likely, no blistering
Dermatitis herpetiformis	Less likely, no history of celiac, no abdominal
Impetigo	Possibility (wound culture)
Septicemia	Less likely, afebrile
Superinfected atopic dermatitis	Less likely, no history of atopy,
Generalized pustular psoriasis	Possibility (plaque psoriasis history, recent systemic steroids, anonychia)



Diagnostic Work-up

Immediate referral to a dermatologist who treats psoriasis is a great idea at any point!

Test	GPP Notes	Results
Wound culture	GPP pustules are sterile	Normal flora
Punch biopsy	IMPORTANT: specimen needs to go to a dermatopathologist for correct analysis	PAS stain negative for fungi Histopathology + pustular psoriasis
CBC with differential CMP UA		All within normal limits
QuantiFERON-TB-Gold Hepatitis HIV	To determine eligibility for large molecule protein biologic	Negative
Chest X-ray (postanterior and lateral)		Unremarkable

Treatment

CAUTION: The use of systemic corticosteroids in patients with a history of psoriasis can increase the risk of GPP. If necessary, use tapered doses for weaning.

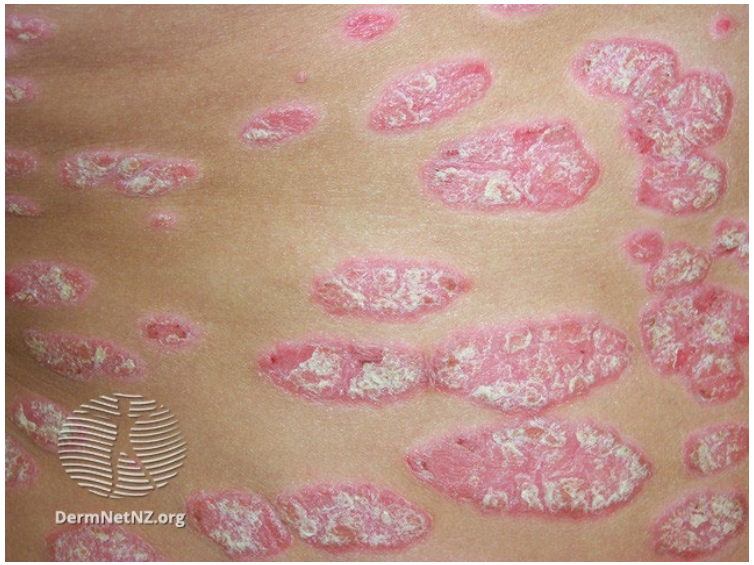
Treatments	Notes
Systemic oral corticosteroids	Two week tapered course
Clobetasol ointment	For pain at fingertips
Occlusive wet wrap	For pain at fingertips

What lead us to the diagnosis of GPP?

- ✓History of plaque psoriasis
- ✓Recent illness
- ✓Recent systemic steroids
- ✓Sterile pustules
- ✓Systemic rash
- ✓Palmar and plantar erythroderma
- ✓Anonychia
- ✓Skin biopsy: parakeratosis, acanthosis, hyperkeratosis, elongation of the rete ridges, and diminished stratum granulosum

Plaque Psoriasis vs. Pustular Psoriasis

A chronic, relapsing, auto-immune disease commonly presenting as **silvery hyperkeratotic plaque** on an erythematous base



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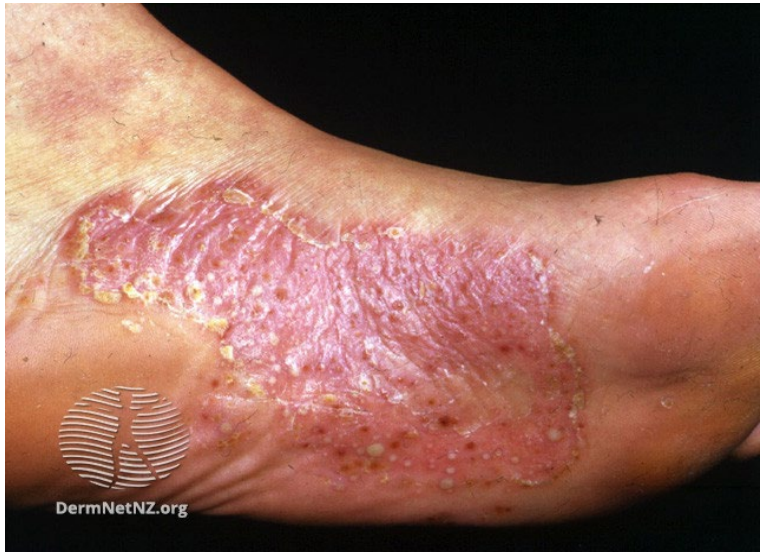
A rare, auto-immune systemic disorder commonly presenting as sterile pustules on an erythematous base



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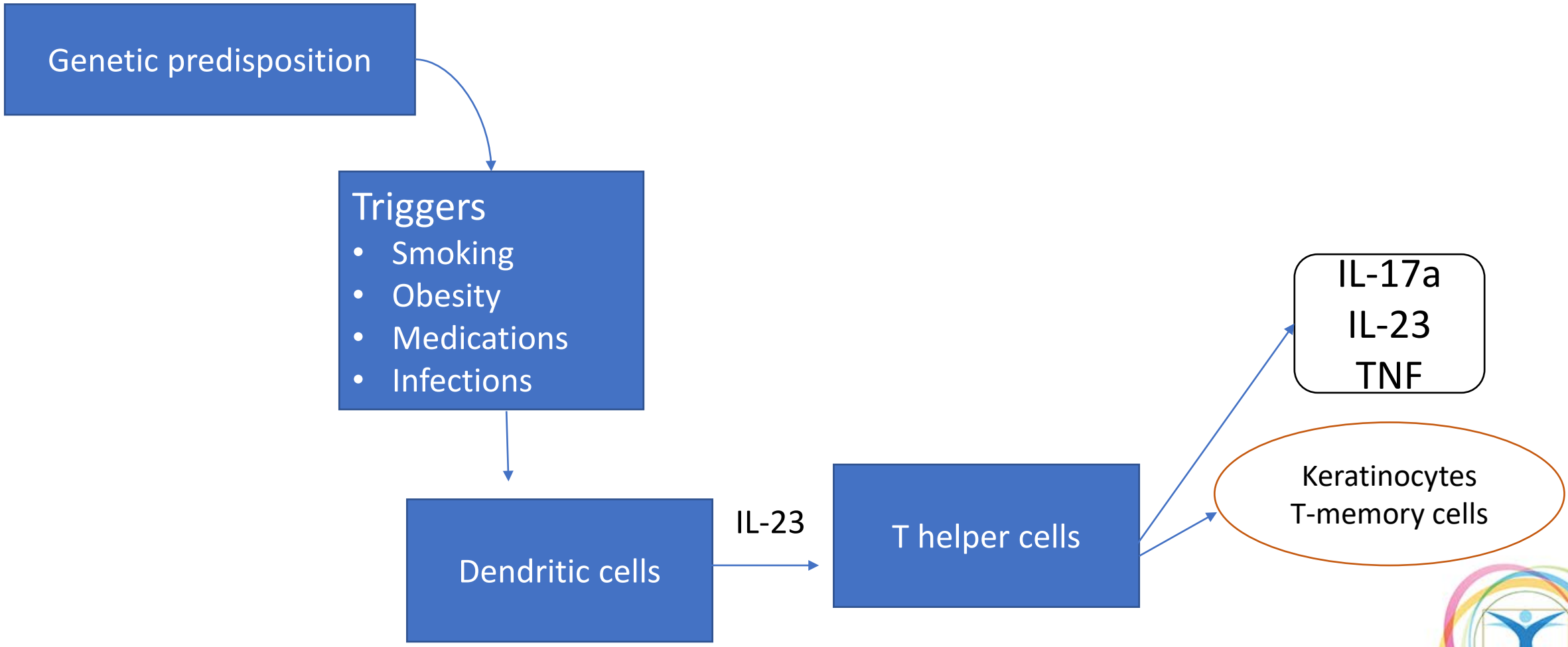
Plaque Psoriasis Pathophysiology



Pathophysiology Pustular Psoriasis

- Involvement of similar cytokines and cells found in plaque psoriasis
- Incomplete understanding
  - Sweating abnormality
- IL-36 plays an important role
  - IL-36, while active in plaque psoriasis, is hyperactive in pustular psoriasis
  - Mutations in *IL36RN,* an IL-36 receptor antagonist, predisposes for pustular psoriasis
- Other mutations associated with pustular psoriasis
  - CARD14, AO1S3, and myeloperoxidase

Griffiths CEM, et al. *Lancet.* 2021 Apr 3;397(10281):1301-1315.

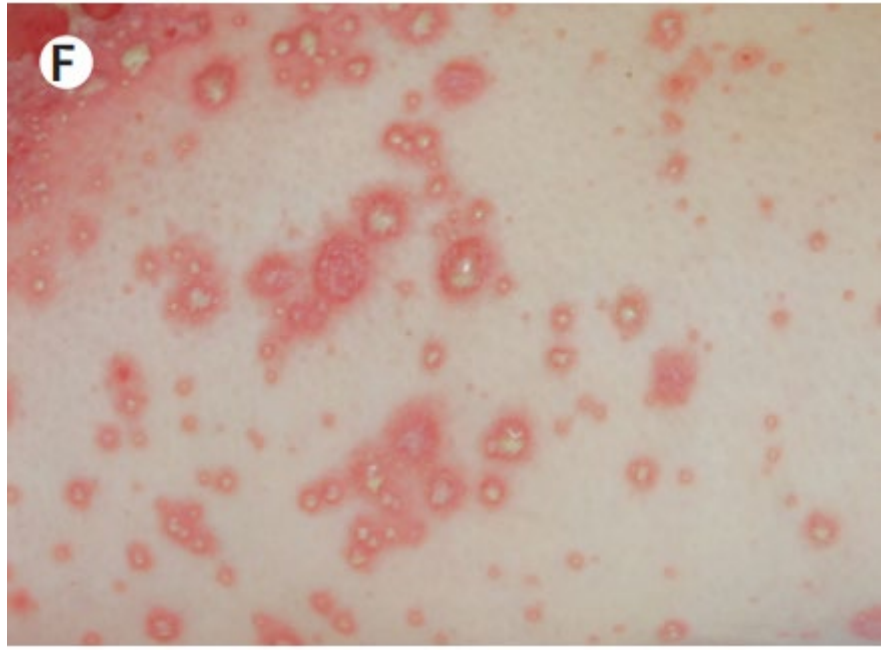
Triggers for Pustular Psoriasis

- **Smoking**
  - Need for *behavioral* change
- Rapid tapering of oral/intramuscular/topical corticosteroids
- Female predominant
- Metal allergies
- Pregnancy (impetigo herpetiformis)
- Hypocalcemia
- Stress
- Infections
- Medications (TNF-α inhibitors)
- Other diseases

Pustular Psoriasis

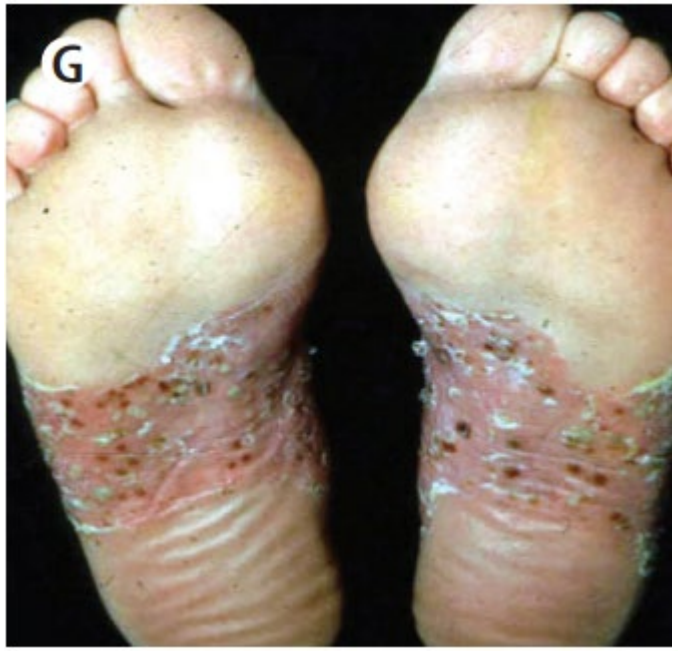
Generalized Pustular Psoriasis (GPP) or Von Zumbusch

(Diffuse generalized pustular eruption with Systemic symptoms including fever/joint pain)



Palmoplantar Pustulosis (PPP)

(Focalized on Palms/Soles)



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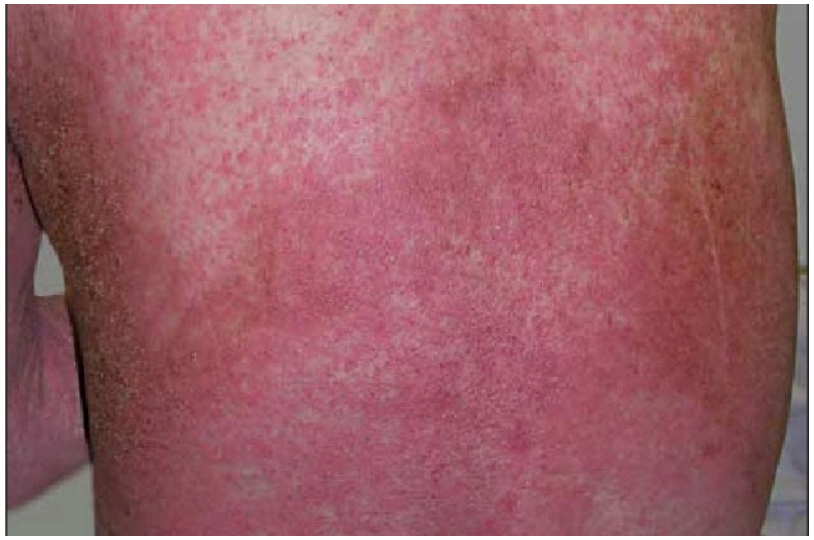
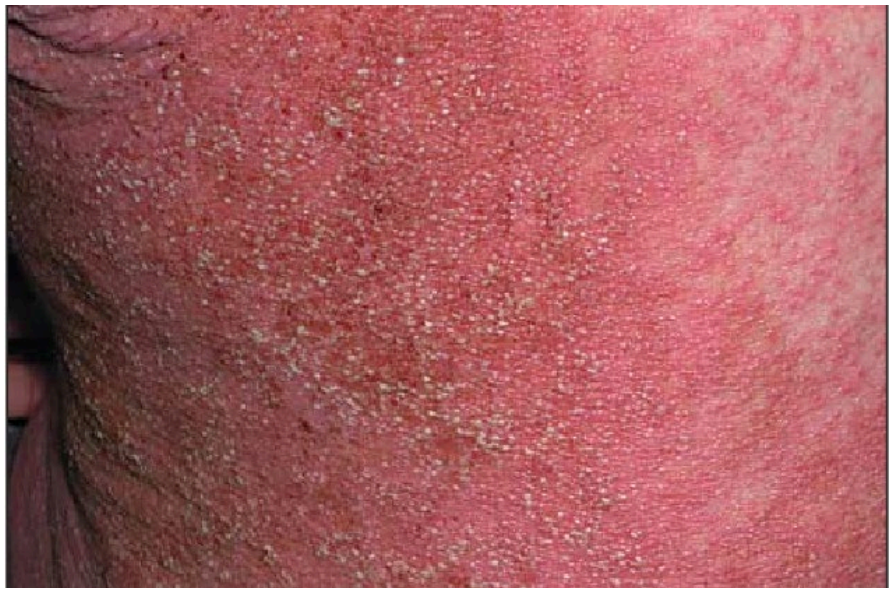
Generalized Pustular Psoriasis

Acute

- Abrupt
- Generalized, wide-spread
- Erythematous patches with pinhead-sized sterile pustules
- Can progress to erythroderma (> 90% of skin)
- Fever, malaise, joint pain
- Lower extremity edema
- Jaundice
- Eye concerns
- CAN BE LIFE THREATENING–REFER IMMEDIATELY TO ED!

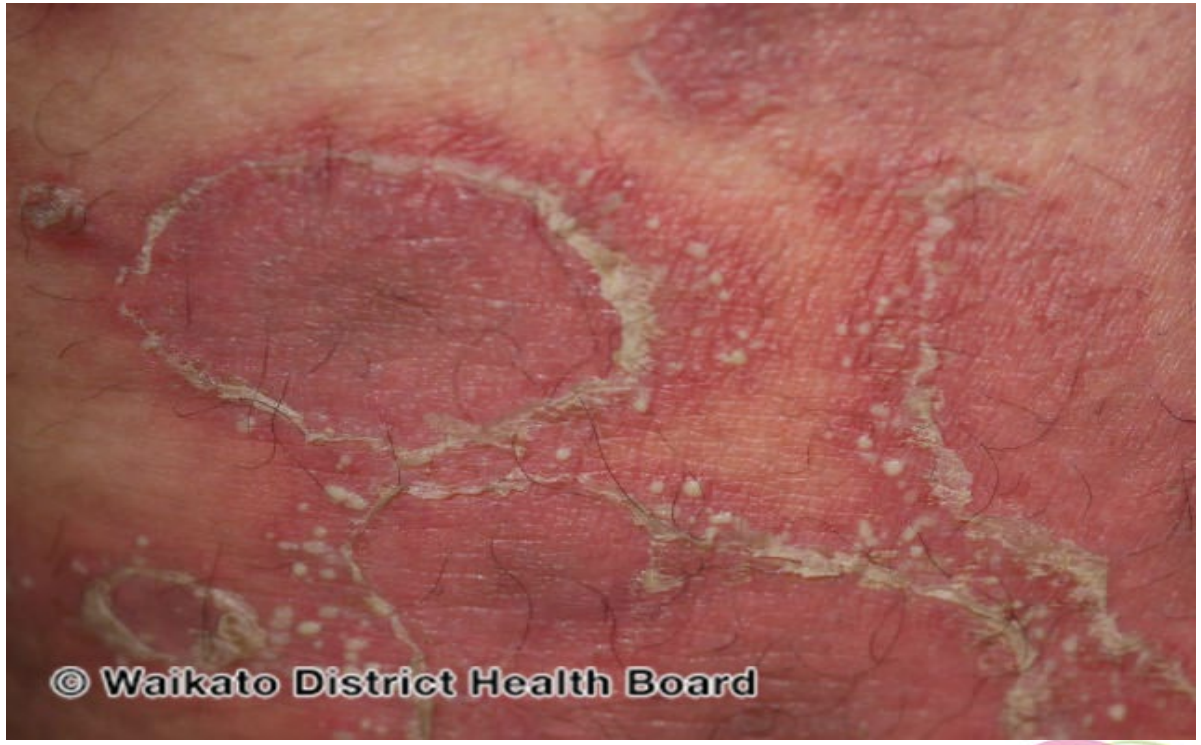




Generalized Pustular Psoriasis

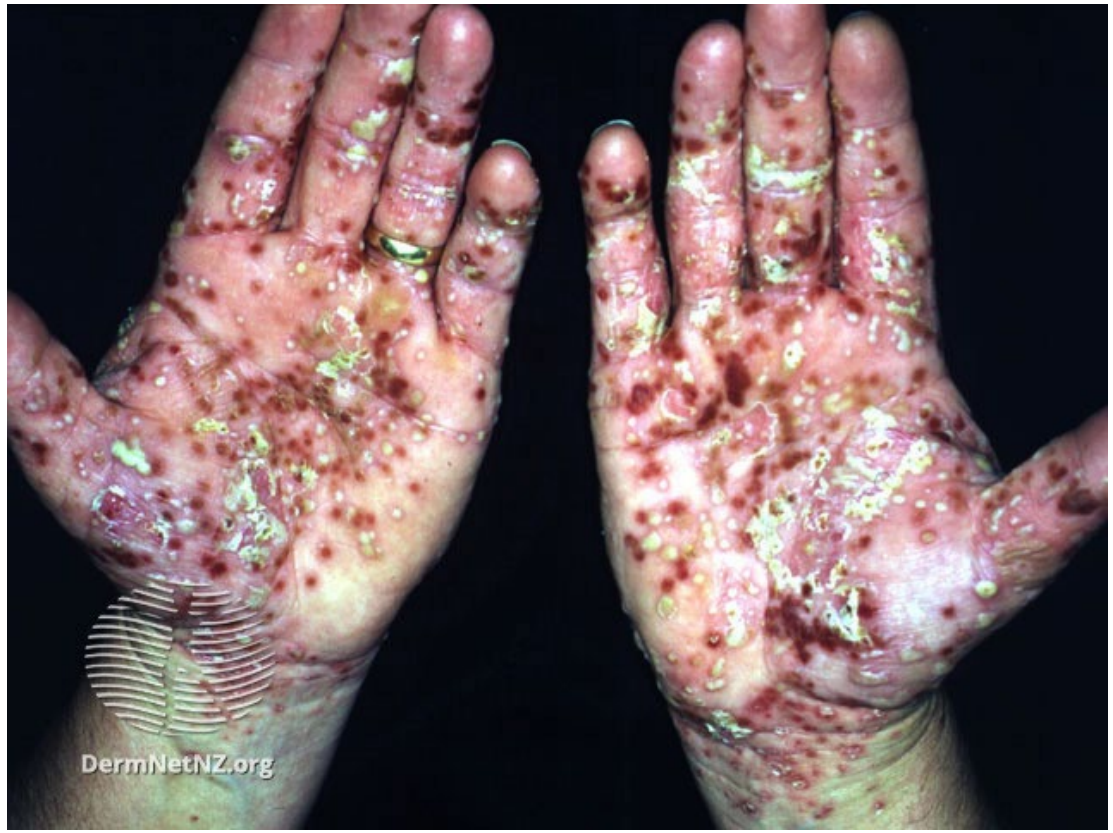
Annular

- Sub-acute
- Re-occurring
- Pustules on edge of plaques



Palmoplantar Pustular Psoriasis (PPP)

- Palms and soles
- Sterile yellow pustules resolving to red/brown macules
- Pustules 1-10 mm
- Often reoccurring, chronic
- Concomitant plaque psoriasis is common



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Refer Pustular Psoriasis to Dermatology

- Any time a patient is suspected to have pustular psoriasis they should be referred to dermatologist that treats psoriasis
- They will likely need systemic therapy

RED FLAG: If a patient has suspected pustular psoriasis and has constitutional symptoms (fever, body aches, signs of dehydration, etc.) *immediately refer to Emergency Department-this can be life threatening!*

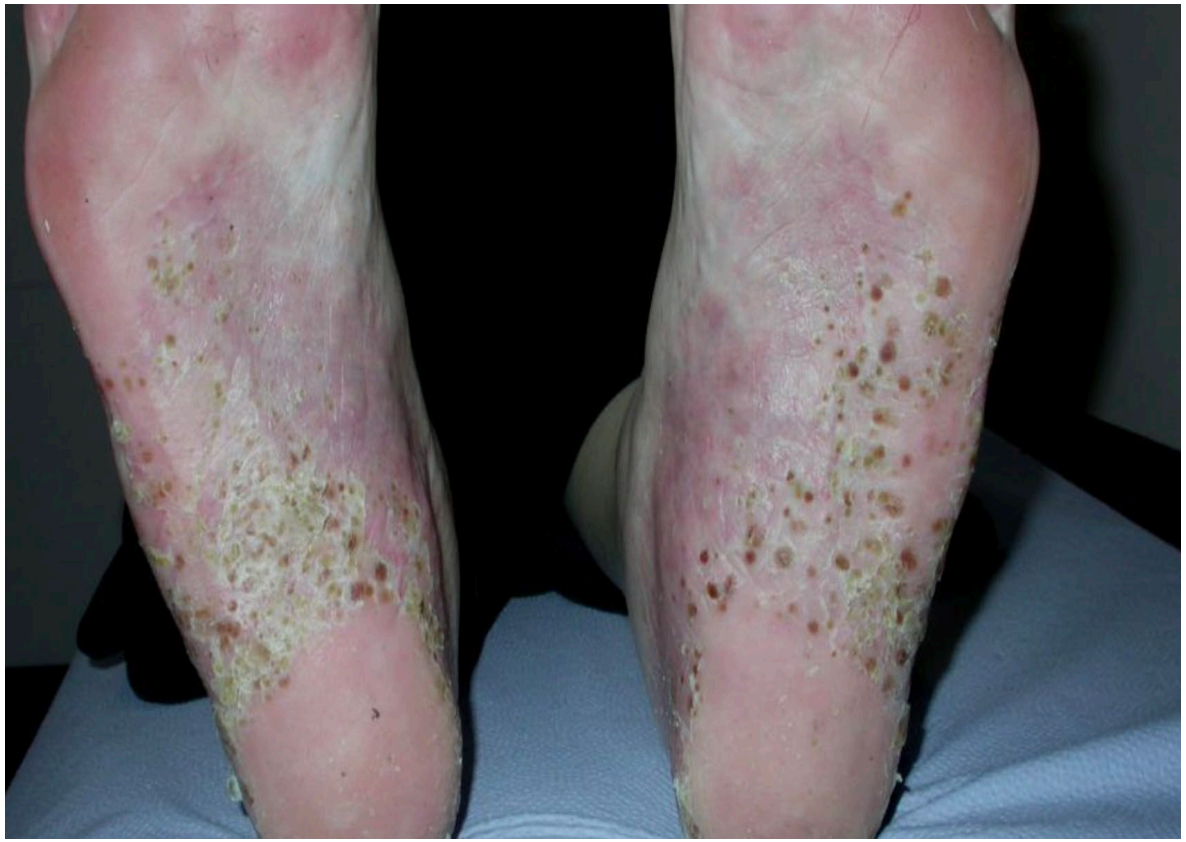
Building a Referral Network

- Society of Dermatology Physician Assistants (SDPA)–www.dermpa.org
- State PA/NP annual conferences
- National conferences
- Local dinner programs
- Colleagues from school

Case Practice

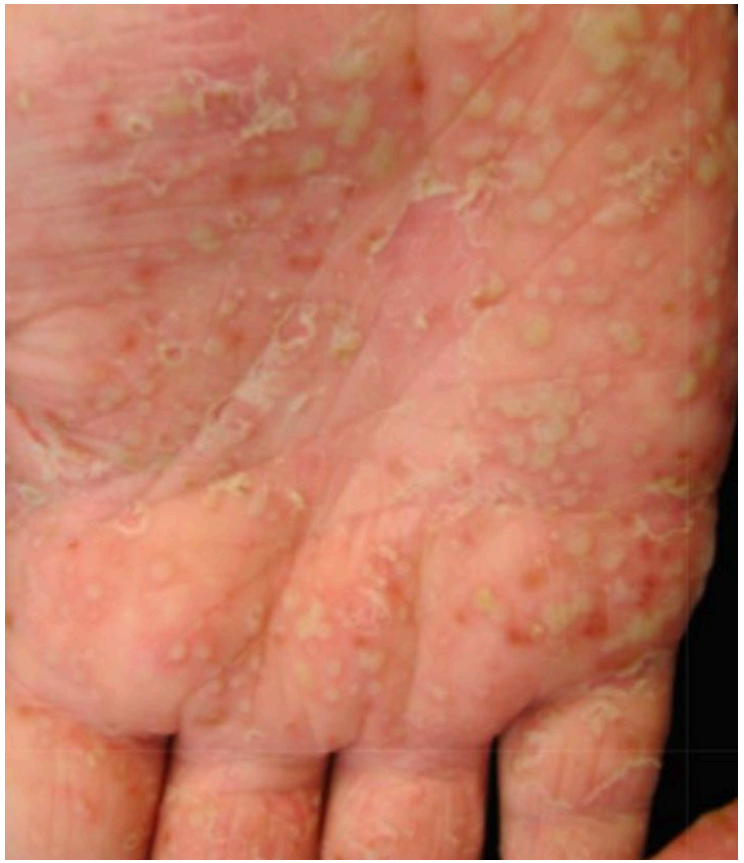
Jesse

- 55-year-old male
- Current complaint is painful, itchy blisters on his palms and soles for the past month
- Patient reports difficulty performing his job as a construction foreman due to pain, fissures, and bleeding



Jesse

- Past medical history:
  - Hypertension
  - Hypothyroidism
  - Recent “sinus surgery”
  - “Pre-diabetic”
  - Recurrent “athlete’s foot.” Reports he’s had “athlete’s foot for years, comes and goes–never goes away forever.”
- Current Medications
  - Losartan
  - Levothyroxine
  - Daily low-dose aspirin
  - OTC tolnaftate (Tinactin) 1% spray
- Socio/behavioral: Smoker
- Physical: No constitutional symptoms



https://www.dermcoll.edu.au/atoz/palmoplantar-pustulosis/

Differential Diagnoses

- Tinea pedis/manuum
- Dyshidrotic eczema
- Palmoplantar pustulosis (PPP)
- Contact dermatitis

What do you think are the next best steps for Jesse?

A. Comprehensive skin exam to evaluate for rash
B. Wound culture
C. CBC differential, CMP, and UA
D. Refer to dermatology

Testing and Results

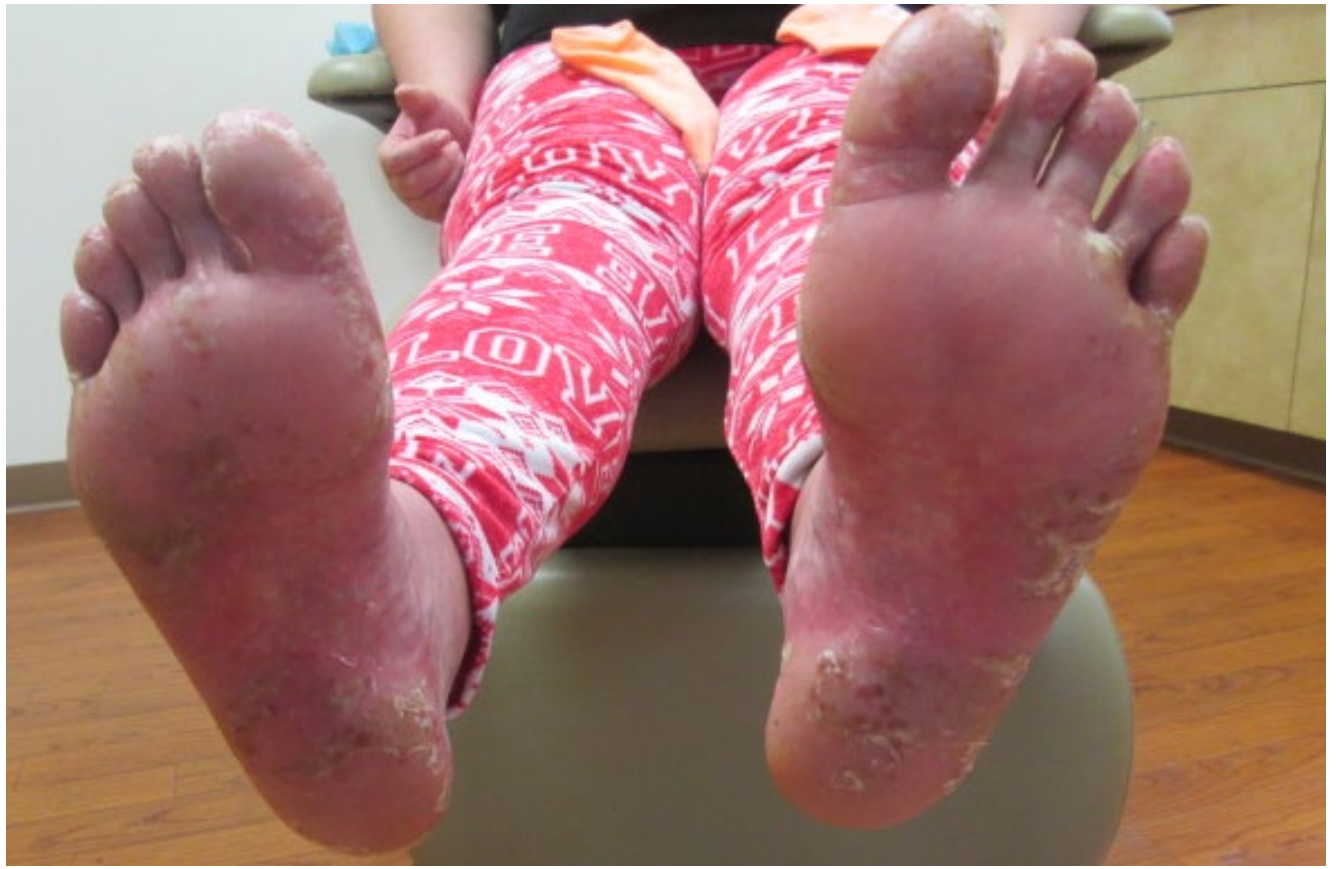
Tests	Results
Wound culture	Normal flora
CBC differential	Unremarkable
CMP	BG elevated at 146 and otherwise unremarkable
UA	Positive for 2+ ketones

Possible Next Steps

- Punch biopsy for confirmation of diagnosis
- Referral to a dermatology provider who treats psoriasis
- Trial of high-potency TCS (possibly under occlusion)
- Systemic corticosteroid treatment

Trena

- 50-year-old female
- Current complaint: Painful, itchy “blisters” on palms and soles
- Thickened nails



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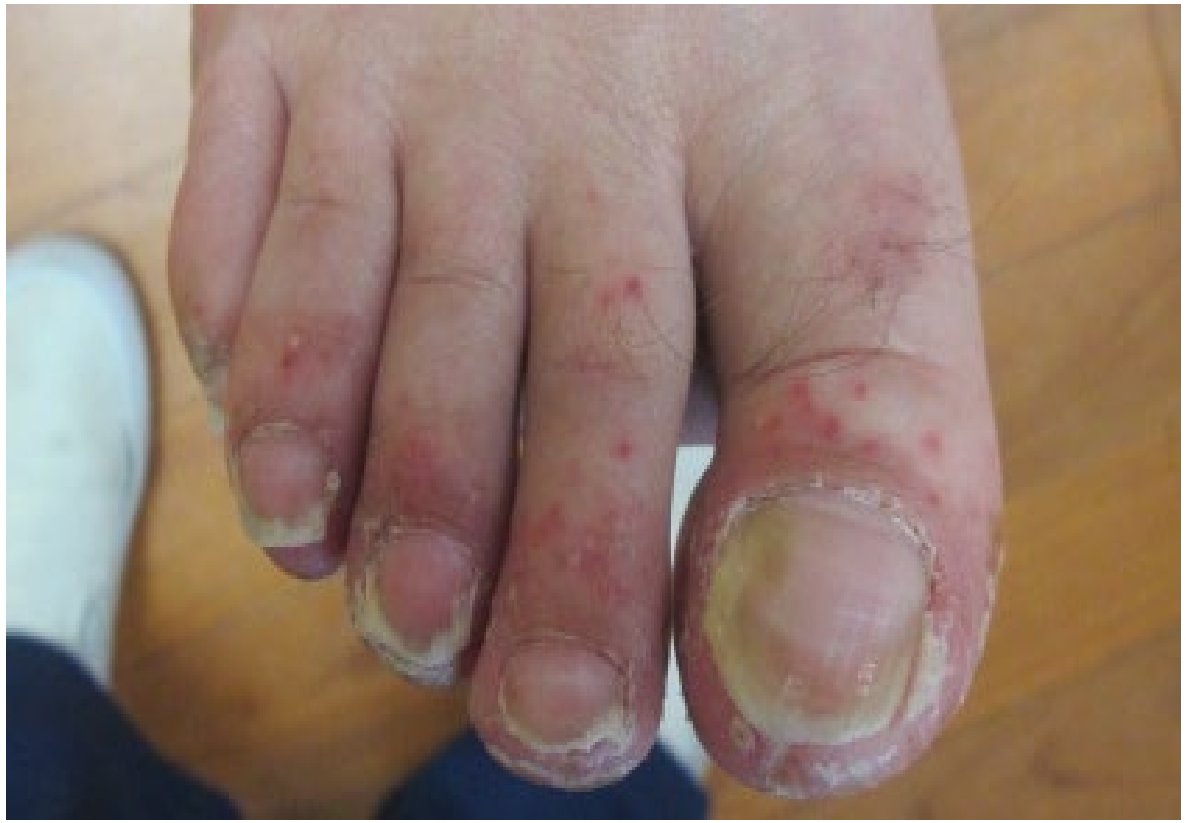


Past medical history:

- Bipolar disorder
- Thyroid cancer

Current medications:

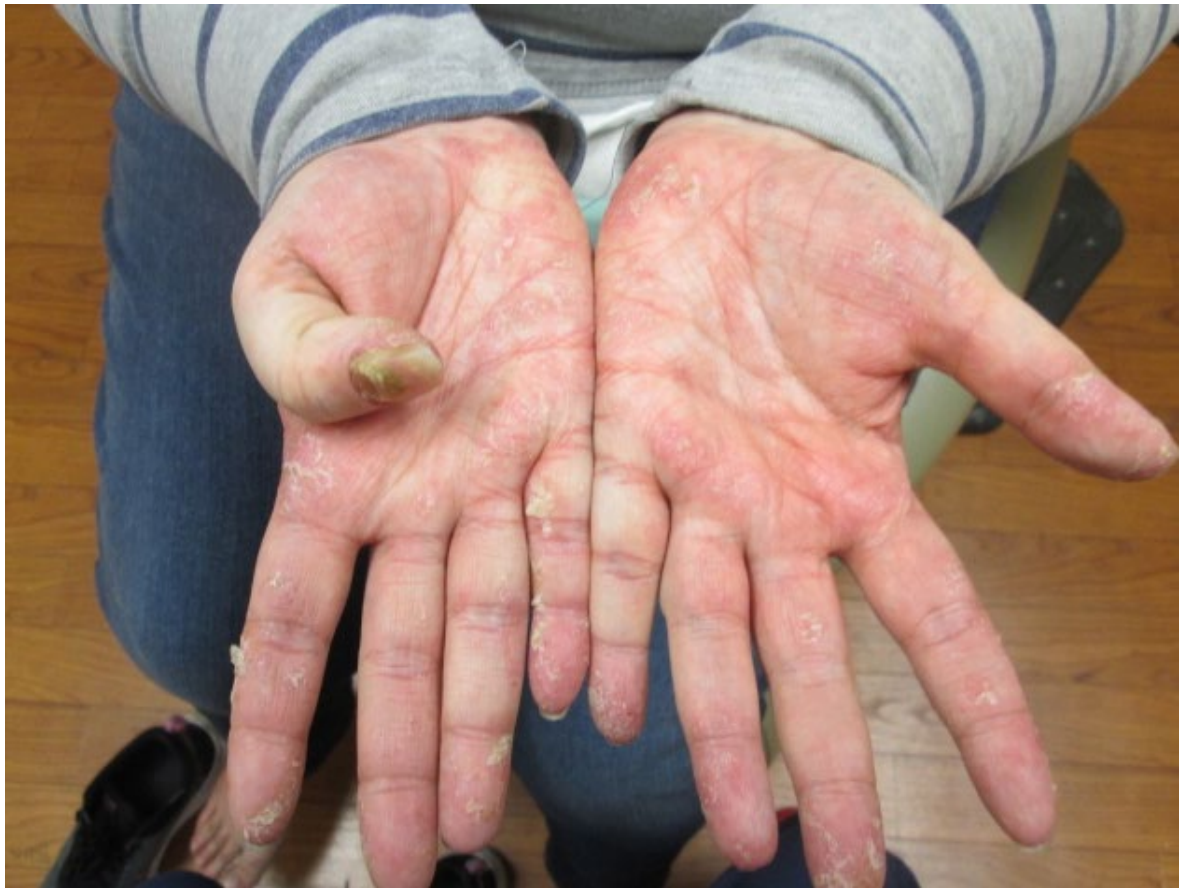
- Lithium
- Mirtazapine
- Levothyroxine
- Ondansetron
- Clonazepam
- Gabapentin
- Triamcinolone
- Efinaconazole

Trena

Socio/behavioral:

- Smoker, both cigarettes and marijuana (nausea)
- Single mom to 15-year-old son with cerebral palsy
- Works part time, nights as a cleaner

Physical: No constitutional symptoms



Differential Diagnoses

- Tinea pedis/manuum
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- Palmoplantar pustulosis (PPP)
- Contact dermatitis

What should you do next?

A. KOH
B. Bacterial culture
C. Nail culture
D. Punch biopsy
E. Refer to a dermatology provider that treats psoriasis

Tests/Results

Tests	Result
KOH	Negative
Bacterial culture	Sterile
Nail culture	No signs of onychomycosis; showed only a thickened nail plate
Clobetasol treatment	Poor, inadequate response

Referral to Dermatologist

At the Dermatology Office

Tests	Result
Complete skin exam	Pustules indicative of PPP; no sign of PsO
Punch biopsy (4 mm of pustule and surrounding erythema) send to **Dermatopathologist**	Pustules filled with neutrophils and eosinophils in the upper dermis with an accumulation of mast cells and eosinophils below the pustules.
Hepatitis panel	Negative
QuantiFERON Gold	Negative
HIV	Negative
CBC	Normal
CMP	Normal
Lipids	Normal

Summary

- Pustular psoriasis is a rare autoimmune disorder found in 0.05-0.12% of the population
- Pustular psoriasis can easily be mistaken for other common presentations
  - The key to diagnosis for primary care can be found in simple procedures which result in negative findings
- Pustular psoriasis can be debilitating and resistant to conventional therapies

Post-test 1: Which of the following is thought to be a driver of pustular psoriasis but less so in plaque psoriasis?

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B. TNF-α
C. T-cells and neutrophils
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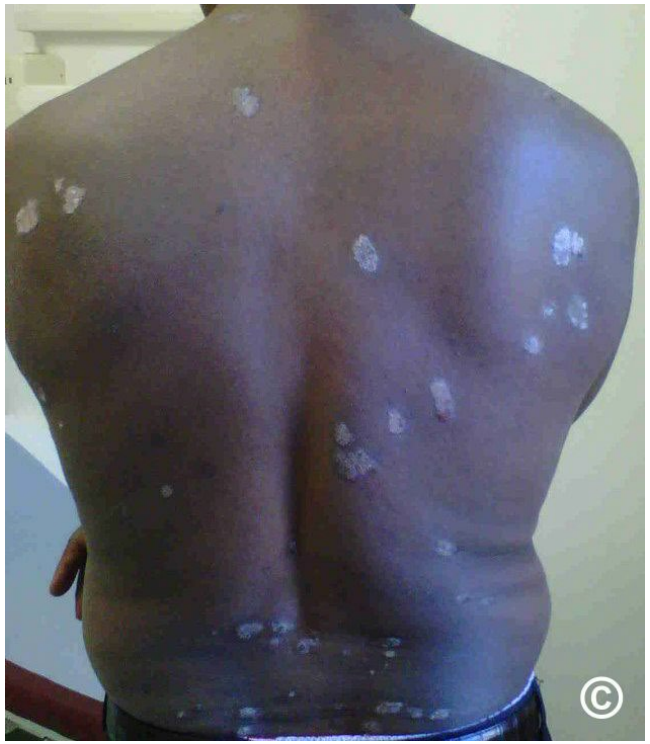

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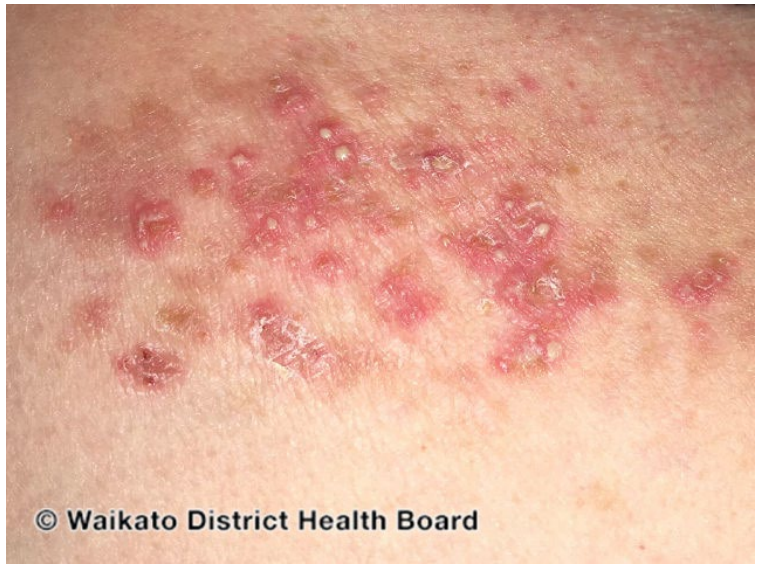

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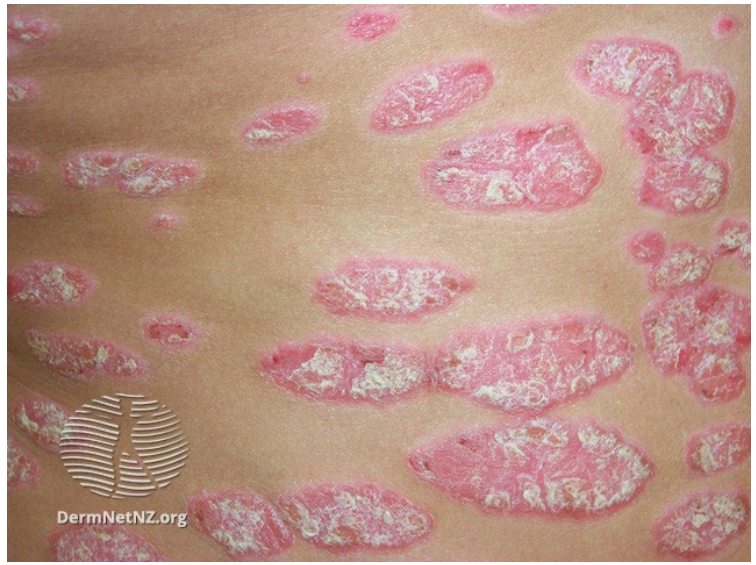

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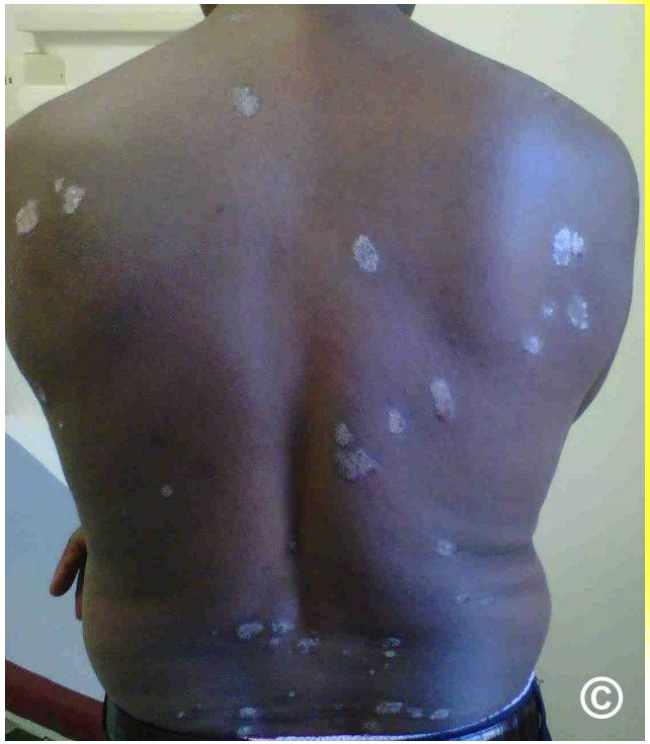

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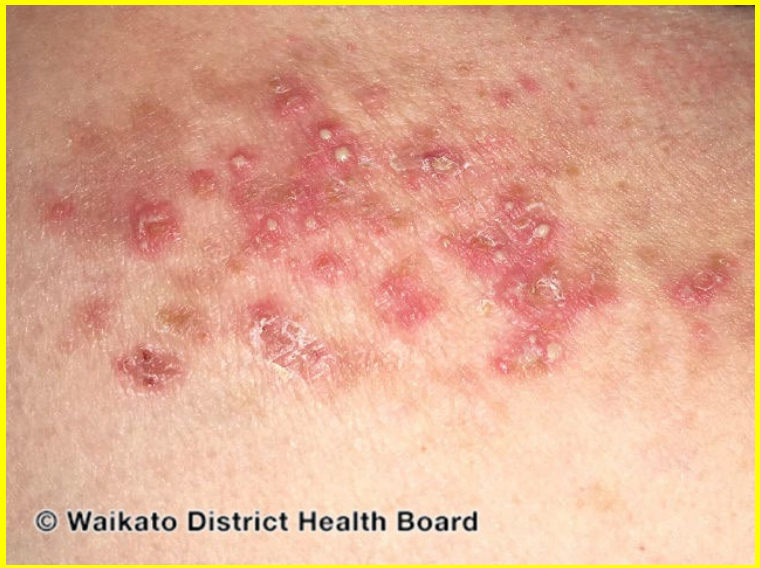

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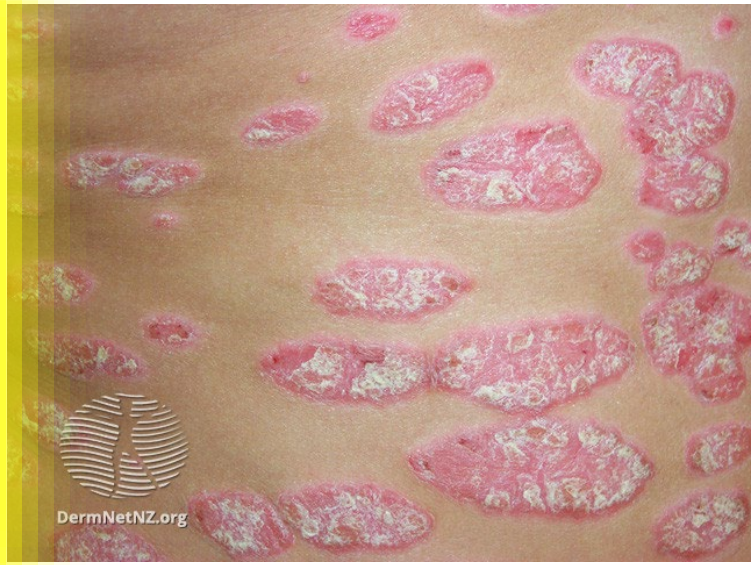

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Post-test 3:

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What would be your next step?

A. Begin treatment with corticosteroids
B. Change the systemic biologic she is on for treatment of her plaque psoriasis
C. Immediate referral to her dermatologist
D. Watch and wait for pustule clearance
E. Prescribe broad-spectrum antibiotics

What would be your next step?

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Questions?