

CASE REPORT

Prurigo Pigmentosa

An atypical case in a healthy young male without recognized risk factors

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Prurigo Pigmentosa: Background

Rare idiopathic inflammatory dermatosis - "Nagashima's disease," "keto rash"

Classically presents in young females of Japanese descent

KNOWN ASSOCIATIONS

- Ketosis (keto diet, DKA, anorexia)
- Diabetes mellitus
- H. pylori infection
- Sjögren's syndrome
- Adult-onset Still's disease

THREE CLINICAL STAGES

1

Early

Pruritic, erythematous papules in a reticular pattern - back, neck, chest

2

Fully developed

Crusted papulovesicles

3

Late

Reticulated, pigmented macules

Case Presentation



Fig. 1 — Erythematous papules coalescing into plaques, bilateral flanks

20-year-old healthy Caucasian male

HISTORY

- 1 week of intensely pruritic, well-demarcated rash on bilateral flanks; sleep-disrupting
- No new meds/products, no sick contacts, no bug bites, no dietary restriction
- Urgent care: empiric permethrin for presumed scabies resulted in no improvement

EXAM

Erythematous papules coalescing into plaques, bilateral flanks (Fig. 1)

LABS

Isolated, mild ↑ total bilirubin – likely incidental

Otherwise unremarkable CBC and CMP

Diagnostic Journey

DIFFERENTIALS

Arthropod assault • Allergic contact dermatitis • Hypersensitivity reaction

FAILED TREATMENTS

Prednisone + clobetasol + hydroxyzine

Worsened

Ciclopirox

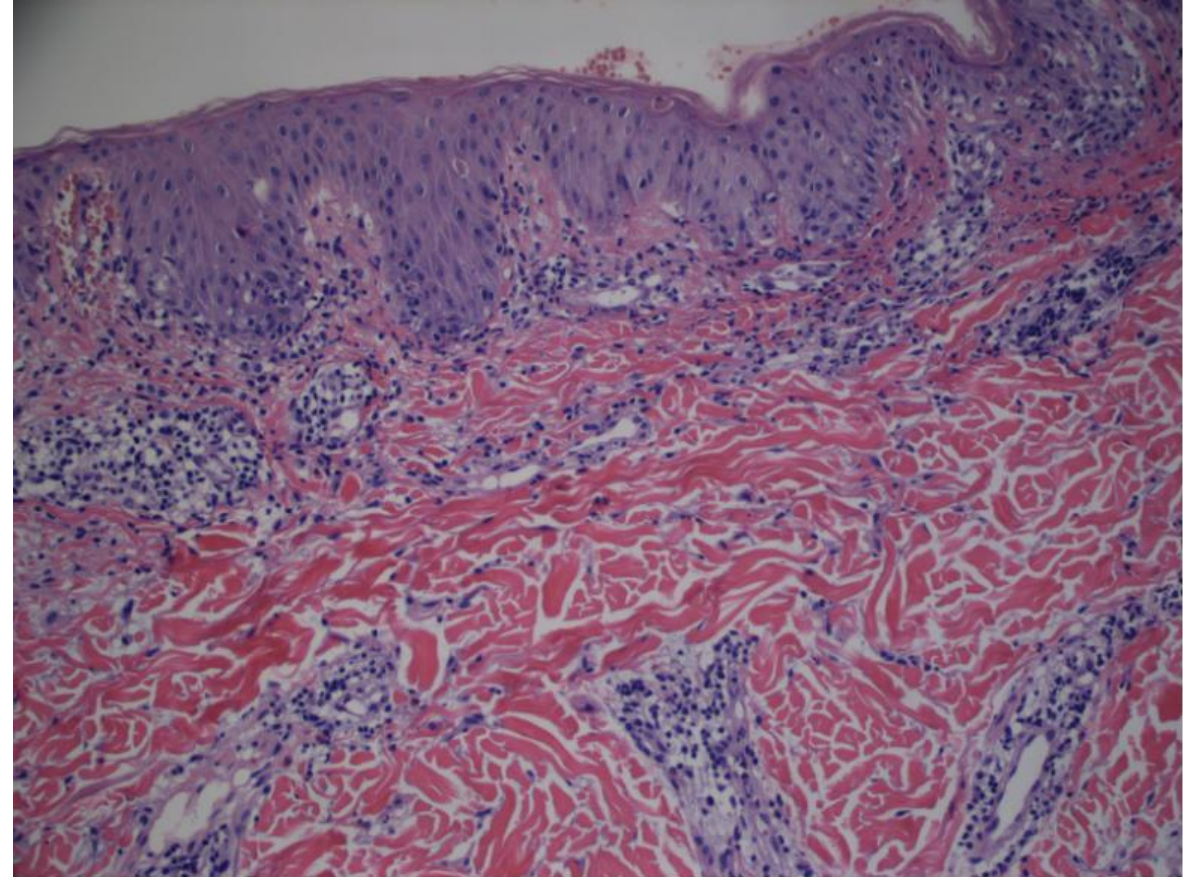
No change

PUNCH BIOPSY

Interface dermatitis with apoptotic keratinocytes; mixed infiltrate with rare eosinophils (Fig. 2)

Initially suggested erythema multiforme / fixed drug eruption

Fig. 2 — H&E (200x): interface dermatitis with scattered suprabasilar apoptotic keratinocytes. The dermis is characterized by a mixed inflammatory infiltrate of lymphocytes, eosinophils and neutrophils.



Clinicopathologic correlation → Prurigo Pigmentosa

Treatment & Outcome

TREATMENT

- Started doxycycline
- All other agents discontinued
- Prednisone tapered off

Pruritus resolved promptly after starting doxycycline

OUTCOME

At ~ 3 weeks: rash faded to a light brown, reticulated patch of hyperpigmentation



Fig. 1 & Fig. 3 — Evolution to a reticulated hyperpigmented patch after 3 weeks of doxycycline

Discussion

1 Atypical demographic

Healthy young Caucasian male - no ketosis, no diabetes, no dietary trigger

2 A possible viral precipitant?

Sore throat on day 7: A prior SARS-CoV-2-associated case had concurrent diabetes; a viral trigger without metabolic disease has not been described.

3 The mild hyperbilirubinemia

Isolated, unconjugated-pattern, and mild - most consistent with an incidental finding. Not a previously reported association with prurigo pigmentosa.

4 Mimicker to rule out: CARP

Confluent and reticulated papillomatosis is typically non-pruritic, with hyperkeratosis, acanthosis, and papillomatosis - helps distinguish from PP.

Key Takeaways

1

Prurigo pigmentosa can occur outside its classic demographics, and without any metabolic, dietary, or autoimmune trigger

2

Suspect it with pruritic papules coalescing into plaques that evolve into reticulated hyperpigmentation

3

First-line therapy is an oral tetracycline (doxycycline or minocycline); topical steroids are ineffective

4

Early recognition prevents diagnostic delay and rapidly restores quality of life

References

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