



# **Interhospital Conference**

## **Case 4**

**Sasipim Chaijaras, MD**

**Theerapong Rattananukrom, MD**

**Penpun Wattanakrai, MD**



# A 16-year-old female

- A rash on back and upper chest for 1 month



# Present illness

## 1 month prior

- A pruritic erythematous rash on her back
- Gradually turned into brownish rash and progressed to chest
- Unresponsive to topical steroids

# Personal history

- No underlying disease or current medications
- Was currently on ketogenic diet for weight loss  
(↓20 kg/3mos)

A microscopic view of skin tissue, showing various cellular structures and fibers in shades of pink and orange. The image is used as a background for a title box.

# **Dermatological examination**



Multiple brownish and some erythematous scaly papules coalescing into reticulated patches on chest wall



Multiple brownish and some erythematous scaly papules coalescing into reticulated patches on back

## Problem list

- 1 month history of progressive pruritic reticulated hyperpigmentation on trunk
- 3 months history of ketogenic diet



# Differential diagnosis for Reticulate hyperpigmentation

- Truncal distribution
- Acral distribution
- Flexural distribution
- Facial distribution
- Nonspecific distribution

# Differential diagnosis for Reticulate hyperpigmentation

- Truncal distribution
- Acral distribution
- Flexural distribution
- Facial distribution
- Nonspecific distribution



- Prurigo pigmentosa
- Confluent and reticulated papillomatosis
- Erythema ab igne
- Ashy dermatosis
- Systemic sclerosis

# Differential diagnosis

	<b>Prurigo pigmentosa</b>	<b>CARP</b>
<b>Demographic</b>	Common in Asians	All ethnicities
<b>Age of onset</b>	Late teens-20s	Puberty
<b>Clinical</b>	<b>Early:</b> Markedly pruritic erythematous papules and papulovesicles <b>Late:</b> Reticulated hyperpigmentation	Multiple brown, verrucous, thin papules or plaques in a central confluence / peripheral reticulation
<b>Pruritus</b>	Severe	Absence to mild

# Differential diagnosis

	<b>Prurigo pigmentosa</b>	<b>CARP</b>
<b>Demographic</b>	Common in Asians	All ethnicities
<b>Age of onset</b>	Late teens-20s	Puberty
<b>Clinical</b>	<b>Early:</b> Markedly pruritic erythematous papules and papulovesicles <b>Late:</b> Reticulated hyperpigmentation	Multiple brown, verrucous, thin papules or plaques in a central confluence / peripheral reticulation
<b>Pruritus</b>	Severe	Absence to mild

# Differential diagnosis

	<b>Prurigo pigmentosa</b>	<b>CARP</b>
<b>Demographic</b>	Common in Asians	All ethnicities
<b>Age of onset</b>	Late teens-20s	Puberty
<b>Clinical</b>	<b>Early:</b> Markedly pruritic erythematous papules and papulovesicles <b>Late:</b> Reticulated hyperpigmentation	Multiple brown, verrucous, thin papules or plaques in a central confluence / peripheral reticulation
<b>Pruritus</b>	Severe	Absence to mild

# Differential diagnosis

	Prurigo pigmentosa	CARP
Histology	<p><b>Acute:</b> Neutrophilic spongiosis, necrotic keratinocytes, hyperkeratosis</p> <p><b>Chronic:</b> Parakeratosis, acanthosis, hyperpigmentation of the epidermis</p>	<p>Hyperkeratosis, papillomatosis, basilar hyperpigmentation, follicular plugging, flattening of rete ridge</p>

# Differential diagnosis

	Prurigo pigmentosa	CARP
Histology	<p><b>Acute:</b> Neutrophilic spongiosis, necrotic keratinocytes, hyperkeratosis</p> <p><b>Chronic:</b> Parakeratosis, acanthosis, hyperpigmentation of the epidermis</p>	<p>Hyperkeratosis, papillomatosis, basilar hyperpigmentation, follicular plugging, flattening of rete ridge</p>

# Differential diagnosis

	Prurigo pigmentosa	CARP
Histology	<p><b>Acute:</b> Neutrophilic spongiosis, necrotic keratinocytes, hyperkeratosis</p> <p><b>Chronic:</b> Parakeratosis, acanthosis, hyperpigmentation of the epidermis</p>	<p>Hyperkeratosis, papillomatosis, basilar hyperpigmentation, follicular plugging, flattening of rete ridge</p>

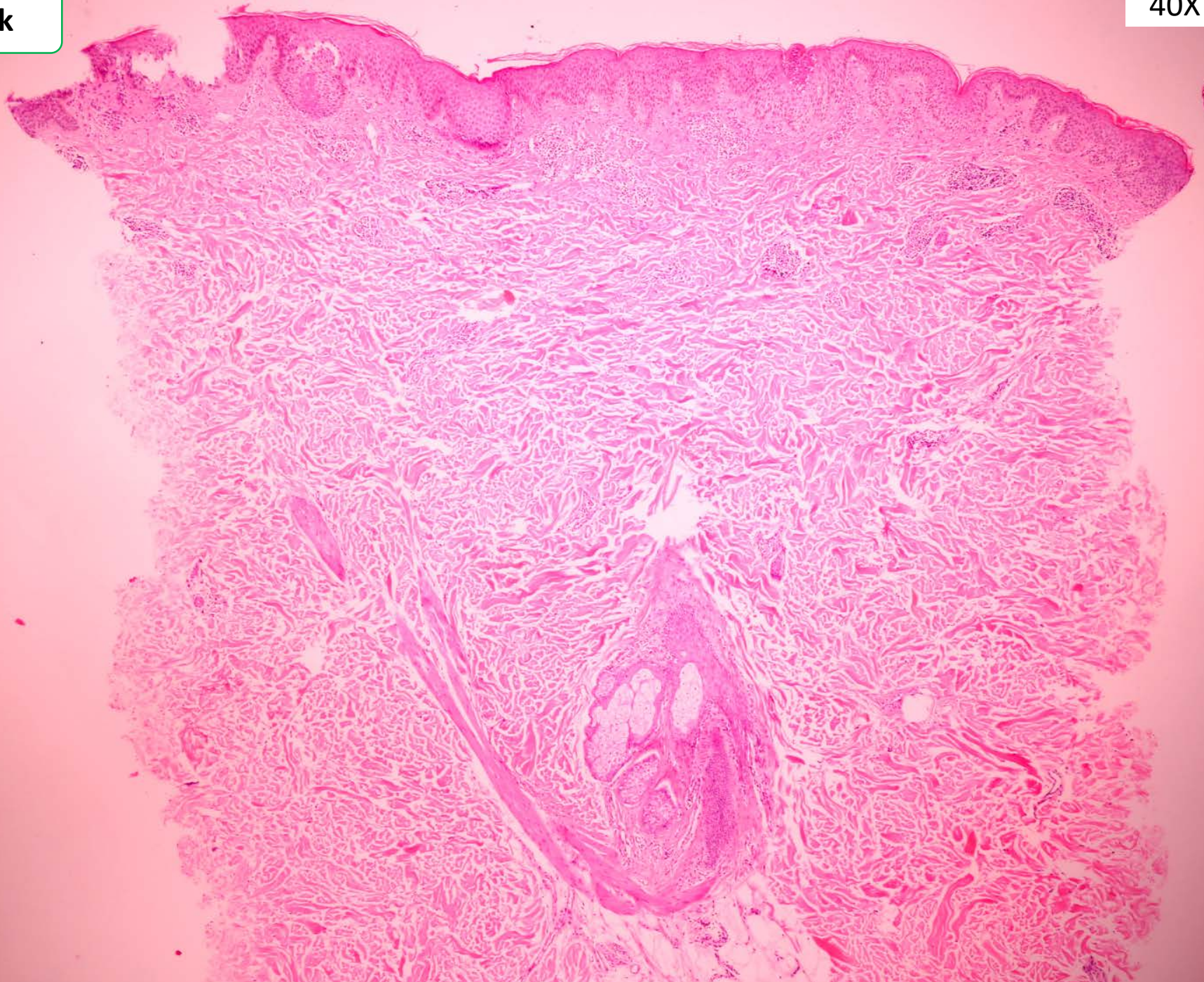


The background of the slide is a microscopic image of tissue, likely stained with hematoxylin and eosin (H&E). It shows various cellular structures, including elongated, spindle-shaped cells and some larger, more rounded cells with distinct nuclei. The overall color palette is a mix of light pink, orange, and brown tones. A prominent black rounded rectangle is centered on the slide, containing white text.

# **Histopathology in this patient**

Back

40X



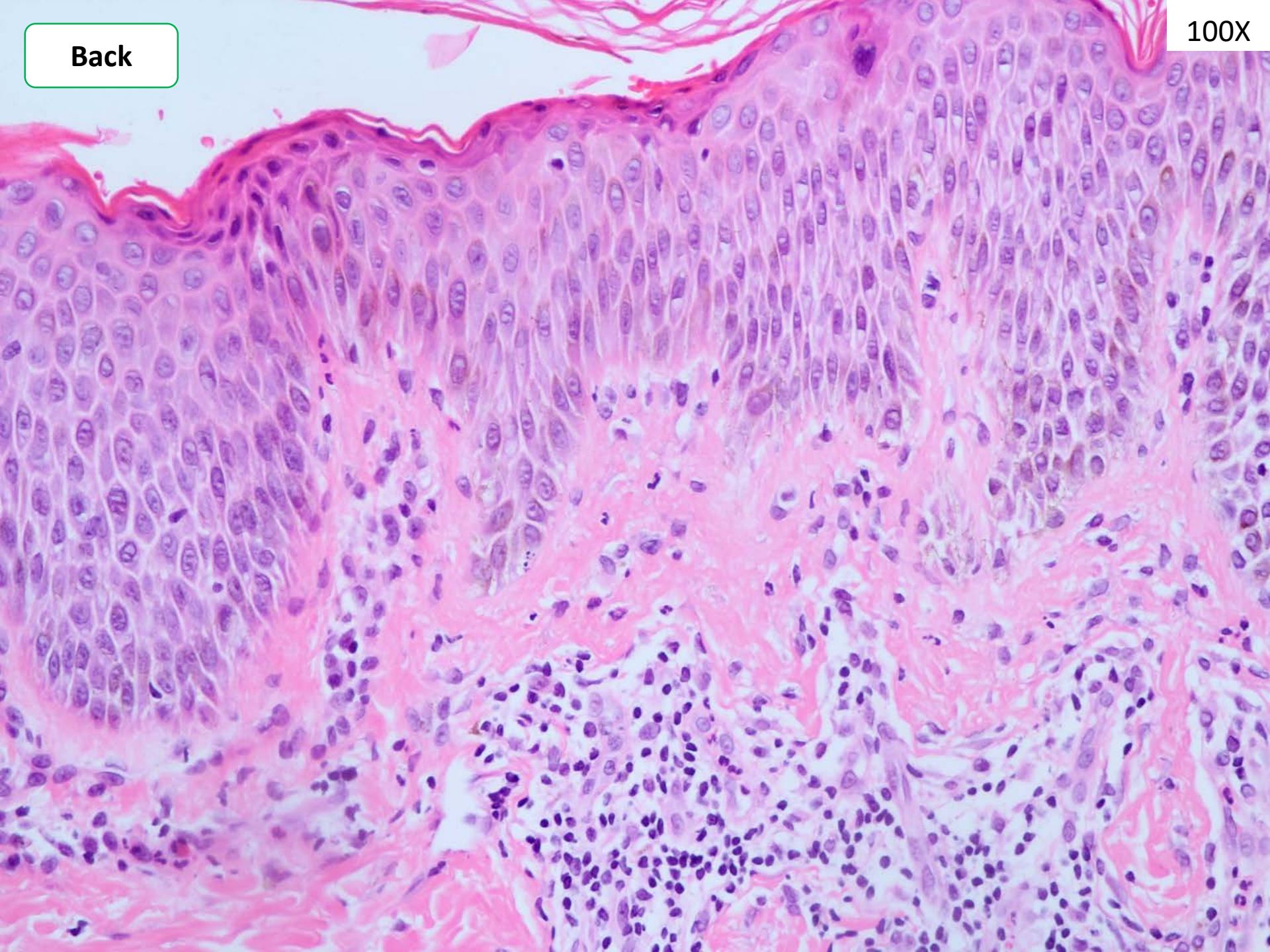
Back

100X



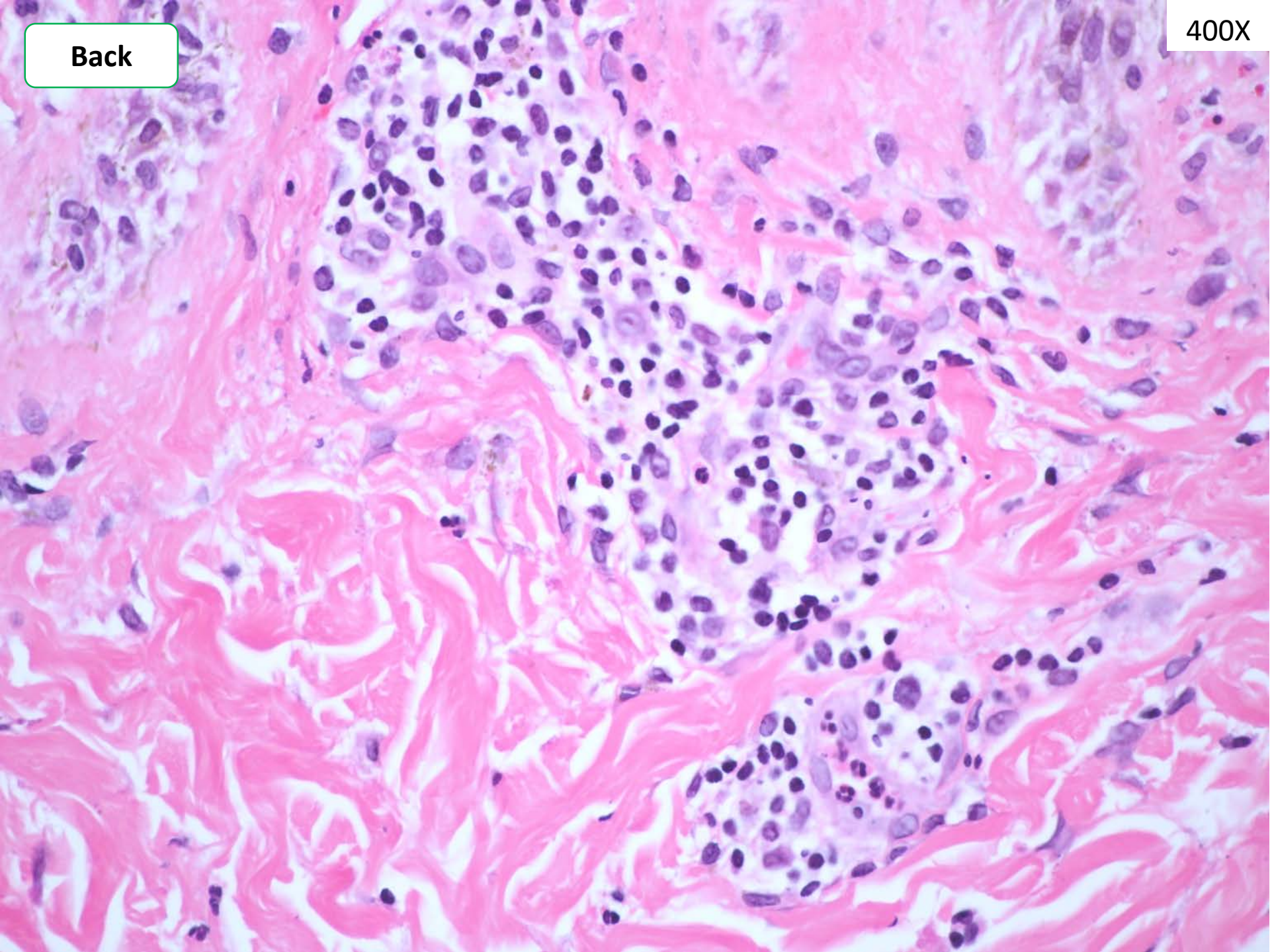
Back

100X



Back

400X



# Prurigo pigmentosa

- Chronic, recurrent inflammatory skin diseases
- First described by Nagashima in 1971
- Most cases reported from Japan
- Etiology: unknown cause

# Prurigo pigmentosa

- F:M = 2:1
- Late teens to early 20s
- Most commonly on back, chest, shoulder, neck, and lumbosacral region

# Aggravating factors

- Ketosis
- Allergy/atopy
- Autoimmune/inflammatory diseases
- Infection - *Helicobacter pylori*, *Borrelia* spp.
- Hot climate
- Mechanical irritation



# Laboratory investigations

- Urinalysis
  - Ketone: marked positive
  - Protein: trace
- FBS/Lipid - normal

# **Diagnosis**

**Prurigo pigmentosa  
associated with ketogenic diet  
“Keto rash”**

# Management

- Dietary modification
  - Increase dietary carbohydrate intake
- Doxycycline 200 mg/day

# Ketogenic diet

- Restriction in carbohydrate < 20-50 g/d or 10% of energy intake
- Increase in protein/fat



# Pathophysiology of keto rash

- Ketone bodies pass from circulating blood into tissue, around blood vessels

# Pathophysiology of keto rash

- Ketone bodies pass from circulating blood into tissue, around blood vessels
- ↑ Ketone bodies upregulate ICAM-1, LFA-1, other proinflammatory mediators  
→ leukocyte-mediated cytotoxicity
- Corticosteroids are ineffective in reducing ICAM-1

# Treatment

- Tetracyclines
  - Minocycline (50-200 mg/day)
  - Doxycycline (100-200 mg/day)
- Dapsone (50-100 mg/day)
- Others: potassium iodide, macrolides, isotretinoin, colchicine
- Dietary modification

Am J Clin Dermatol. 2015; 16: 533-43.

Am J Dermatopathol. 2017; 39: 267-74.

J Eur Acad Dermatol Venereol. 2016; 30: 1794-8.

J Eur Acad Dermatol Venereol. 2012; 26: 1149-53.

Pediatr Dermatol. 2018; 35: 202-3.

# Summary

- A 16-year-old female
- Pruritic reticulated hyperpigmentation on trunk
- Hx of ketogenic diet
- Dx: Prurigo pigmentosa  
“Keto rash”
- Mx: Doxycycline, dietary modification
- Improved within 1 week





**Before**



**After**



# Take home message

## Prurigo pigmentosa

Can be associated with ketosis and ketogenic diet  
“Keto rash”