



# **Interhospital Conference Case 6**

**Wimolsiri Iamsurang, MD**  
**Poonkiat Suchonwanit, MD**



# A 22-year-old female



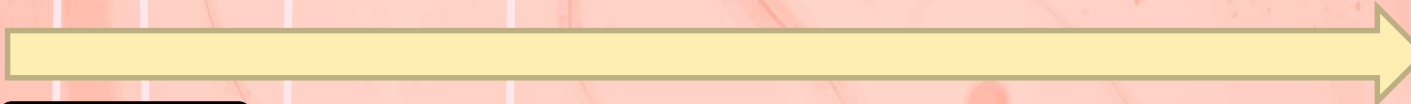
Biopsy-proven pityriasis lichenoides et varioliformis acuta (PLEVA)

**Jun  
2018**

**Jul  
2018**

**Sep  
2018**

**Dec  
2018**



**Doxycycline**

**NBUVB**

**MTX**

**Cyclosporine**

**Jun  
2018**

**Jul  
2018**

**Sep  
2018**

**Dec  
2018**

**Apr  
2019**

**Remission**

**Doxycycline**

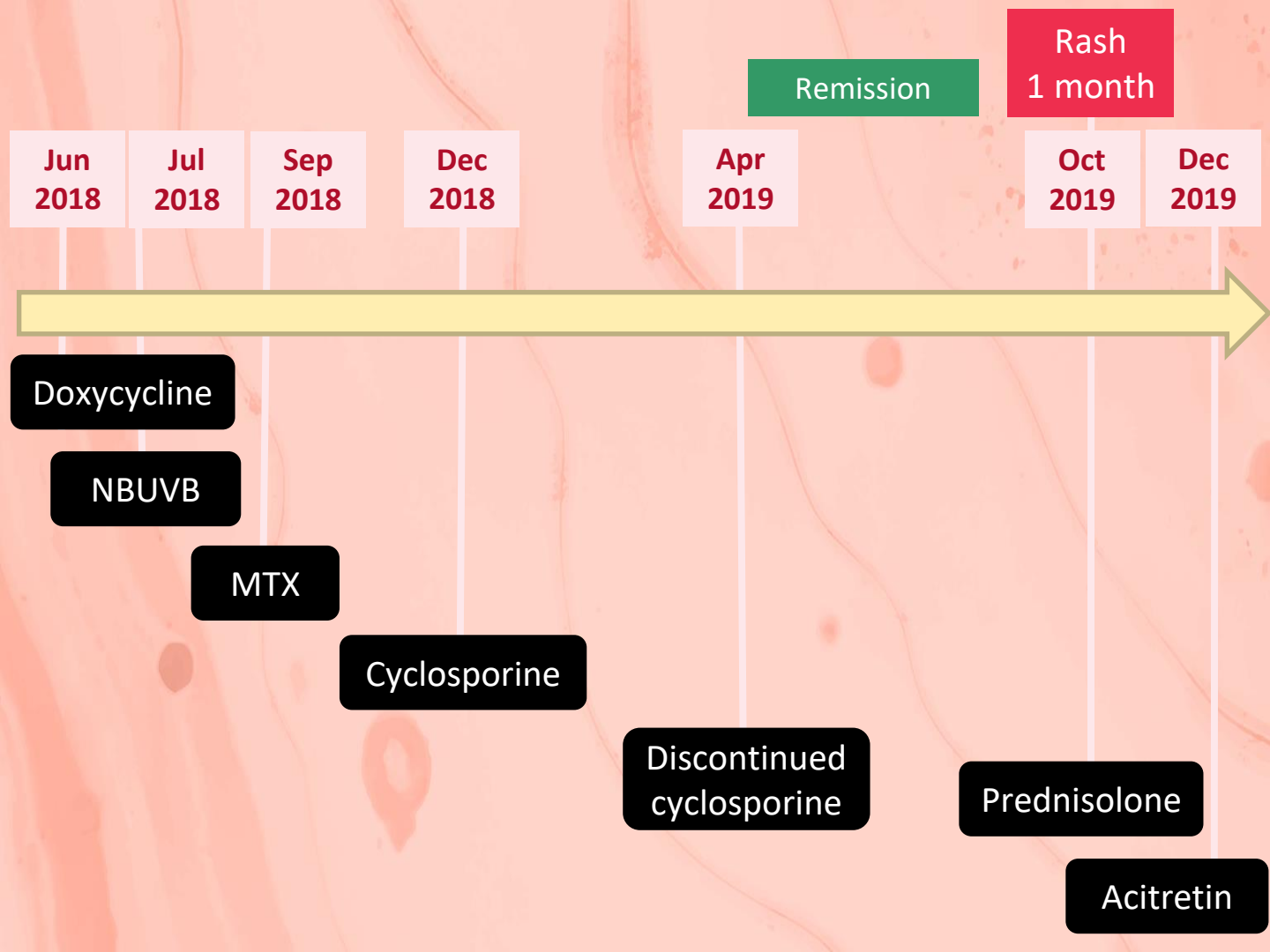
**NBUVB**

**MTX**

**Cyclosporine**

**Discontinued  
cyclosporine**





Jun  
2018

Jul  
2018

Sep  
2018

Dec  
2018

Apr  
2019

Oct  
2019

Dec  
2019

Doxycycline

NBUVB

MTX

Cyclosporine

Discontinued  
cyclosporine

Prednisolone

Acitretin

Remission

Rash  
1 month



Multiple discrete erythematous to brownish papules with crusted lesions and some brownish patches on the trunk and extremities

# Physical examination

- Vital signs: normal
  - HEENT: no pale conjunctivae, anicteric sclerae
  - LN: not palpable
  - Lungs:
  - Heart:
  - Abdomen:
  - Neurologic: grossly intact
- } WNL

# Problem list

Recurrent crusted papular lesions  
on the trunk and extremities



# Differential diagnosis

- PLEVA

# Differential diagnosis

- PLEVA
- Lymphomatoid papulosis (LyP)

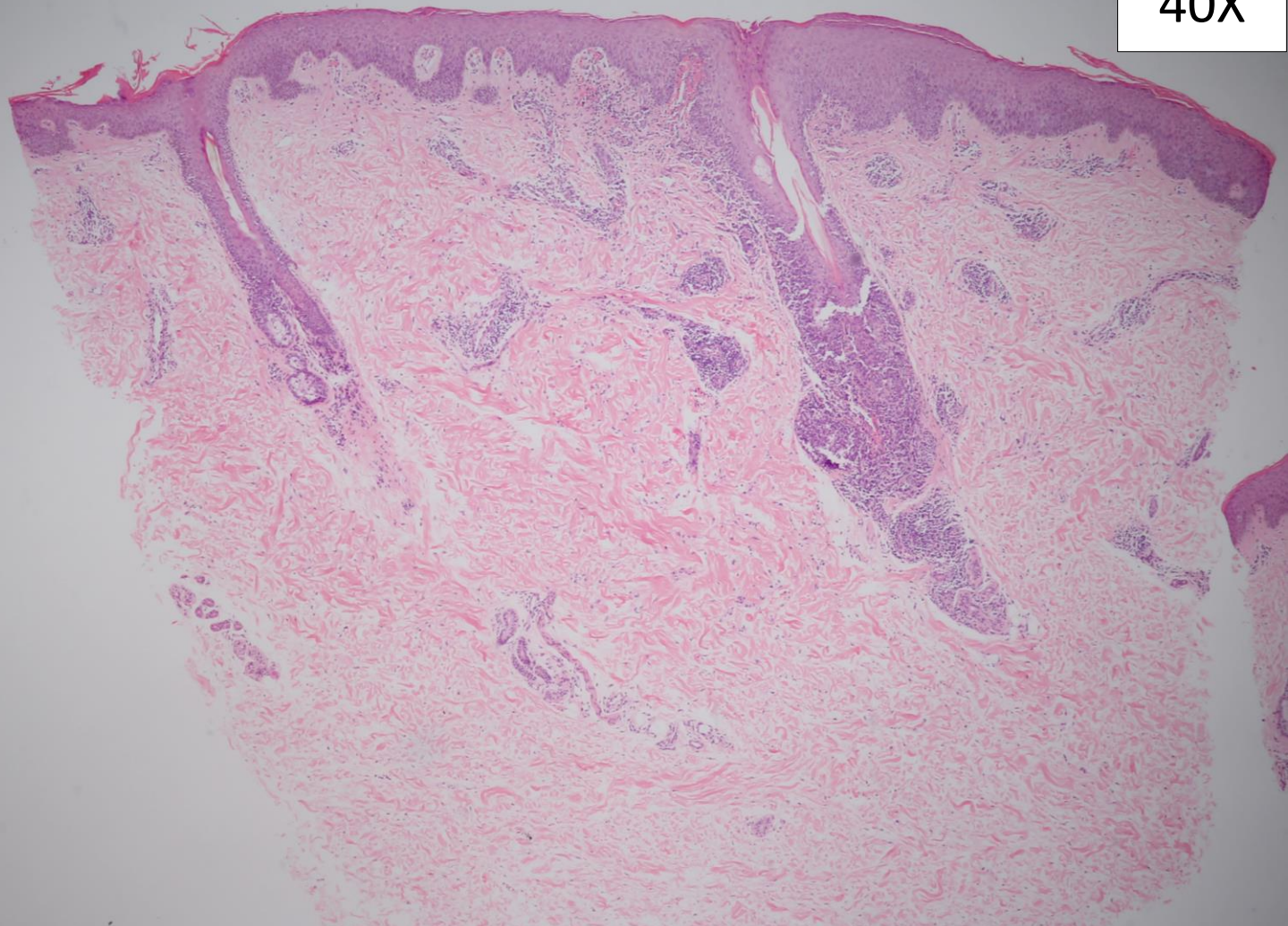
# Differential diagnosis

- PLEVA
- Lymphomatoid papulosis (LyP)
- Mycosis fungoides (MF)
  - Papular variant
  - Folliculotropic variant

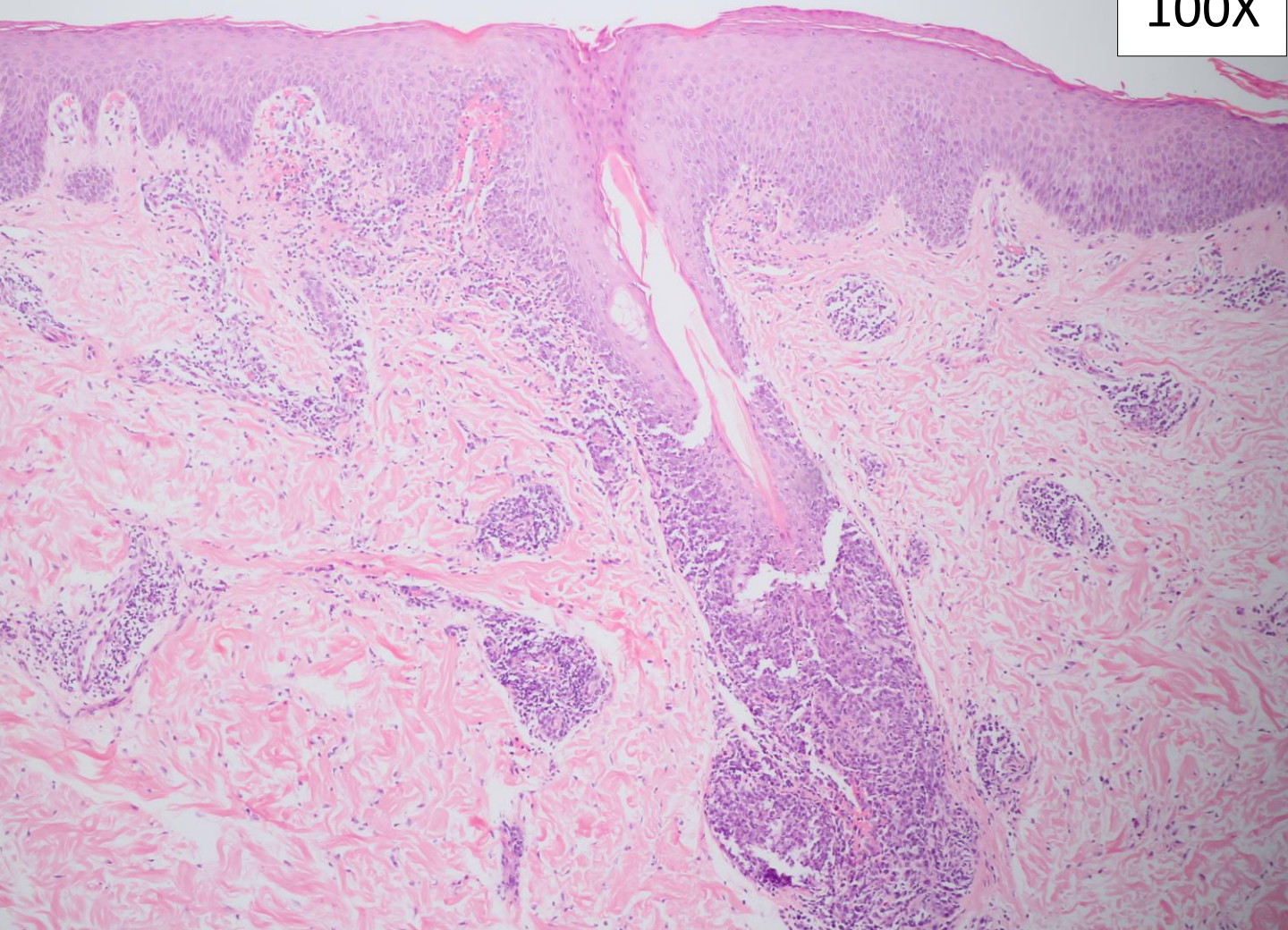


# Histopathology

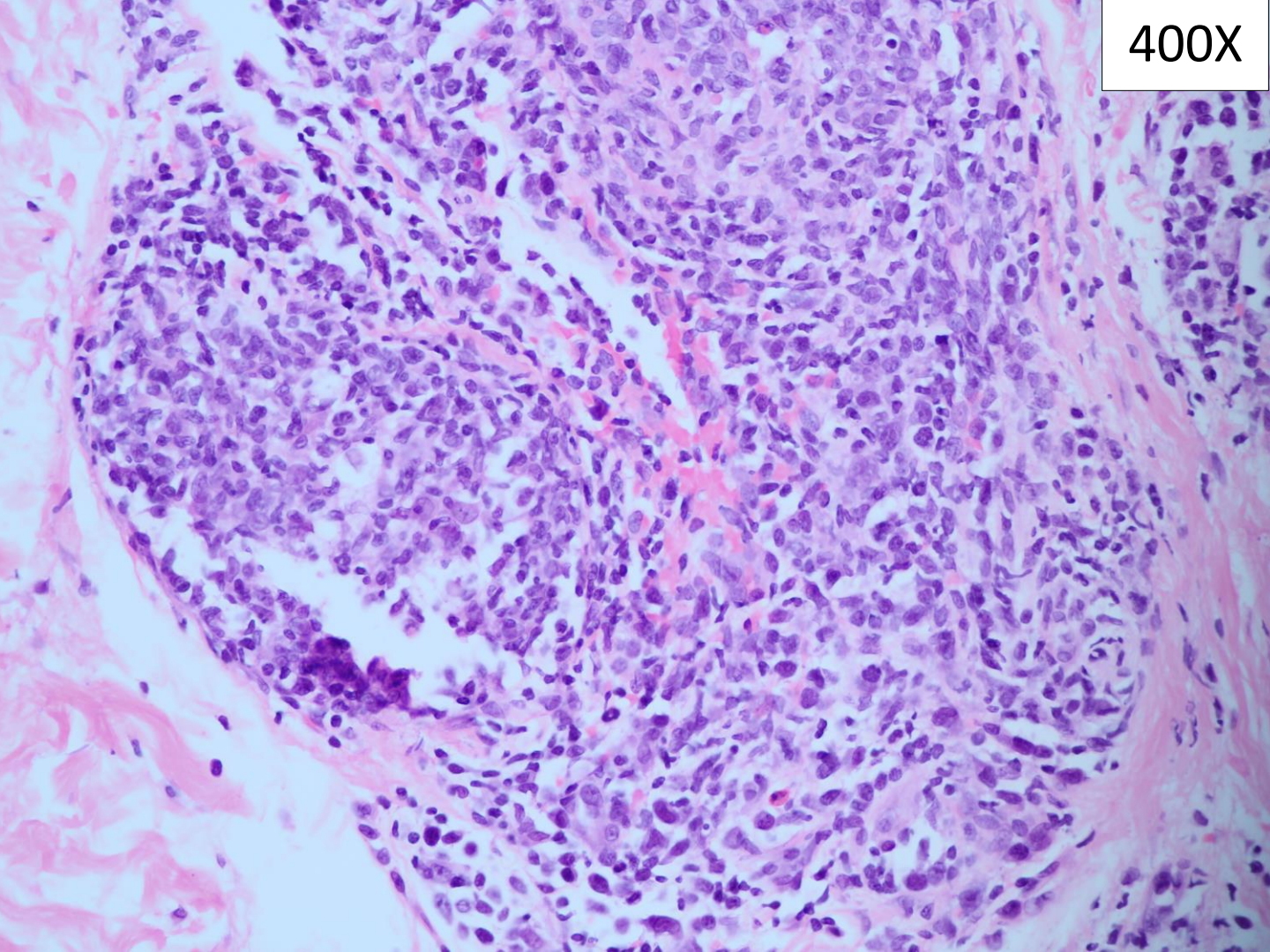
40X



100X



400X

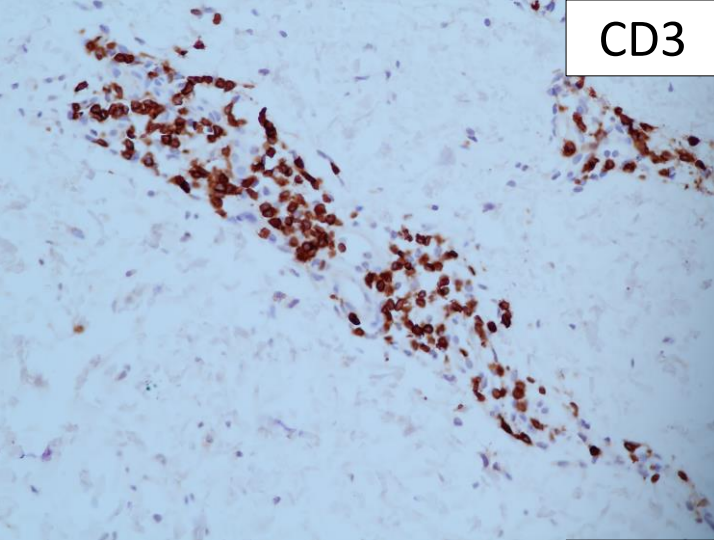


The image features a background of a light pinkish-orange color with a subtle, wavy, marbled texture. Overlaid on this background is a solid black rounded rectangle. Inside this rectangle, the word "Immunohistochemistry" is written in a bold, white, sans-serif font, centered horizontally and vertically.

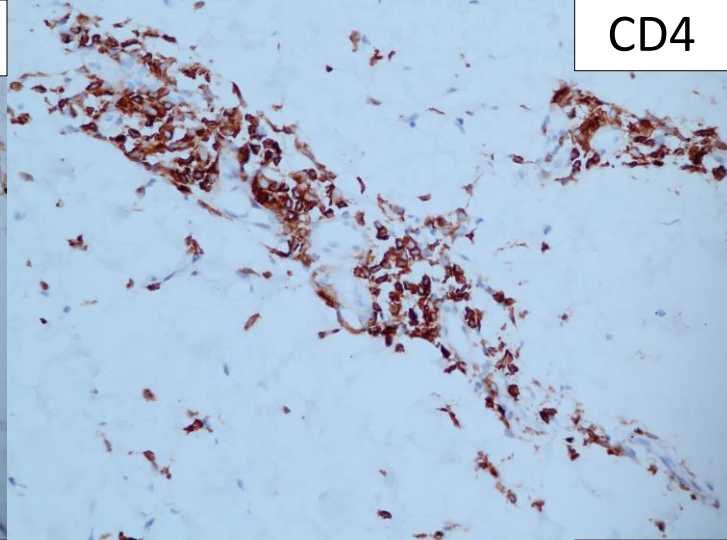
# Immunohistochemistry



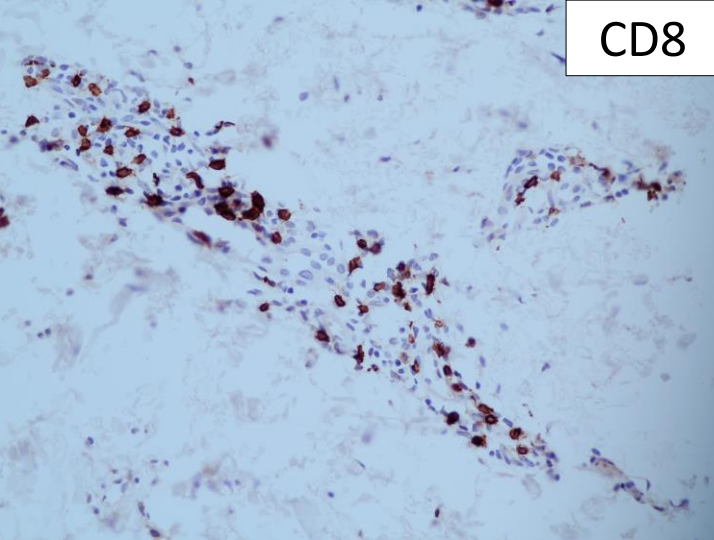
CD3



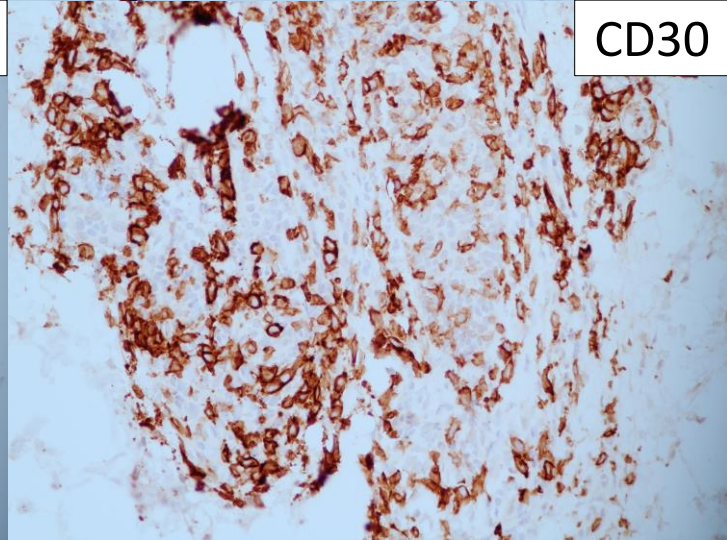
CD4



CD8



CD30



	<b>PLEVA</b>	<b>Papular MF</b>	<b>Folliculotropic MF (Early variant)</b>	<b>LyP (Type F)</b>
<b>Clinical features</b>	Crusted erythematous papules and vesicles	Monomorphic scaly erythematous papules	Papules or patches with follicular accentuation	Papulonodular or papulonecrotic lesions
<b>Distribution</b>	Trunk and extremities	Trunk and extremities	Trunk and extremities, head and neck	Trunk and extremities
<b>Histologic findings</b>	-Parakeratosis -Exocytosis of lymphocytes -Wedge-shaped infiltrate with lymphocytes -Necrotic keratinocytes -Red blood cells	-Lichenoid infiltrate with atypical lymphocytes -Epidermotropism	-Mild lymphocytic perifollicular infiltrate -Atypical lymphocytes -±Eosinophils -±Plasma cells	-Perifollicular infiltrates -Folliculotropism of atypical lymphocytes -±Neutrophils
<b>CD30</b>	Negative	Negative	Negative	Positive
<b>Clinical course</b>	Spontaneous resolution within weeks	Persistent	Persistent	Chronic relapsing with spontaneous resolution

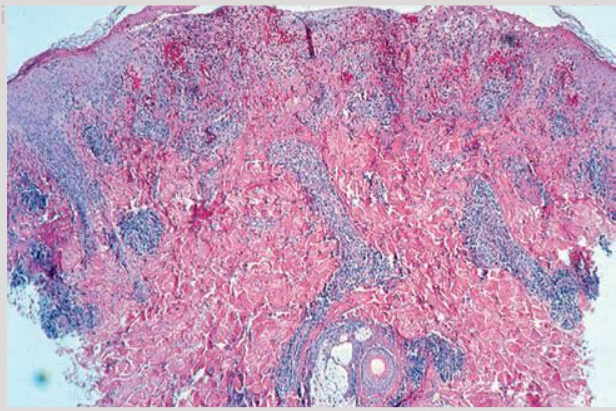
Fitzpatrick's Dermatology, 9<sup>th</sup> edition  
 J Am Acad Dermatol. 2005;52:694-8.  
 J Am Acad Dermatol. 2013;68:809-16.  
 Am J Dermatopathol. 2016;38:e1-10.  
 J Dtsch Dermatol Ges. 2018;16:543-557.

# PLEVA

<b>Clinical features</b>	Crusted erythematous papules and vesicles
<b>Distribution</b>	Trunk and extremities
<b>Histologic findings</b>	<ul style="list-style-type: none"> <li>-Parakeratosis</li> <li>-Exocytosis of lymphocytes</li> <li>-Wedge-shaped infiltrate with lymphocytes</li> <li>-Necrotic keratinocytes</li> <li>-Red blood cells</li> </ul>
<b>CD30</b>	Negative
<b>Clinical course</b>	Spontaneous resolution within weeks

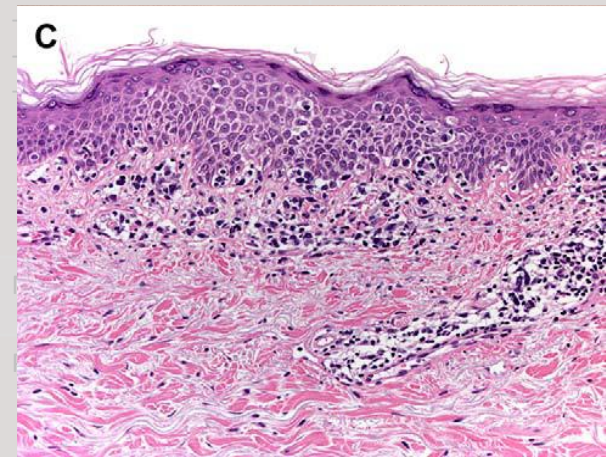
# Papular

Monomorphous erythematous papules
Trunk and extremities
<ul style="list-style-type: none"> <li>-Lichenoid infiltrate with atypical lymphocytes</li> <li>-Epidermotropism</li> </ul>
Negative
Persistent



Fitzpatrick's Dermatology, 9th edition  
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CD30	Negative	Negative
Clinical course	Spontaneous resolution within weeks	Persistent



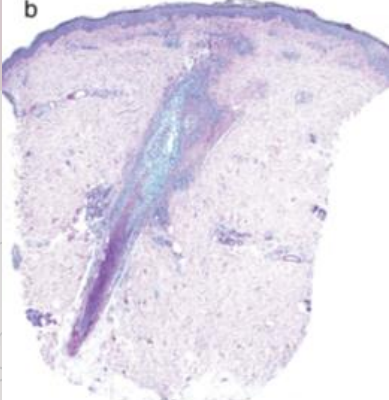
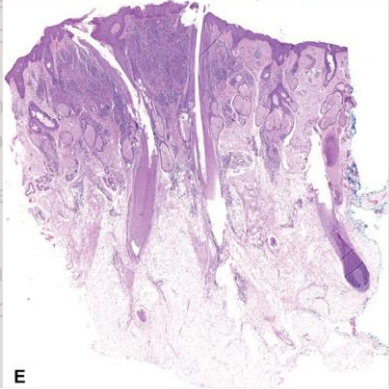
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	PLEVA	Papular MF	Folliculotropic MF (Early variant)	LyP (Type F)
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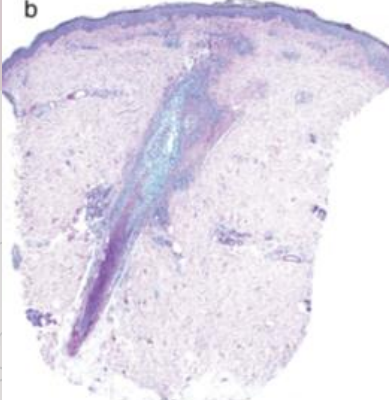
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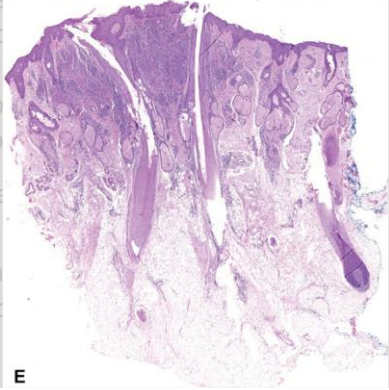
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**Folliculotropic MF**



**LyP type F**

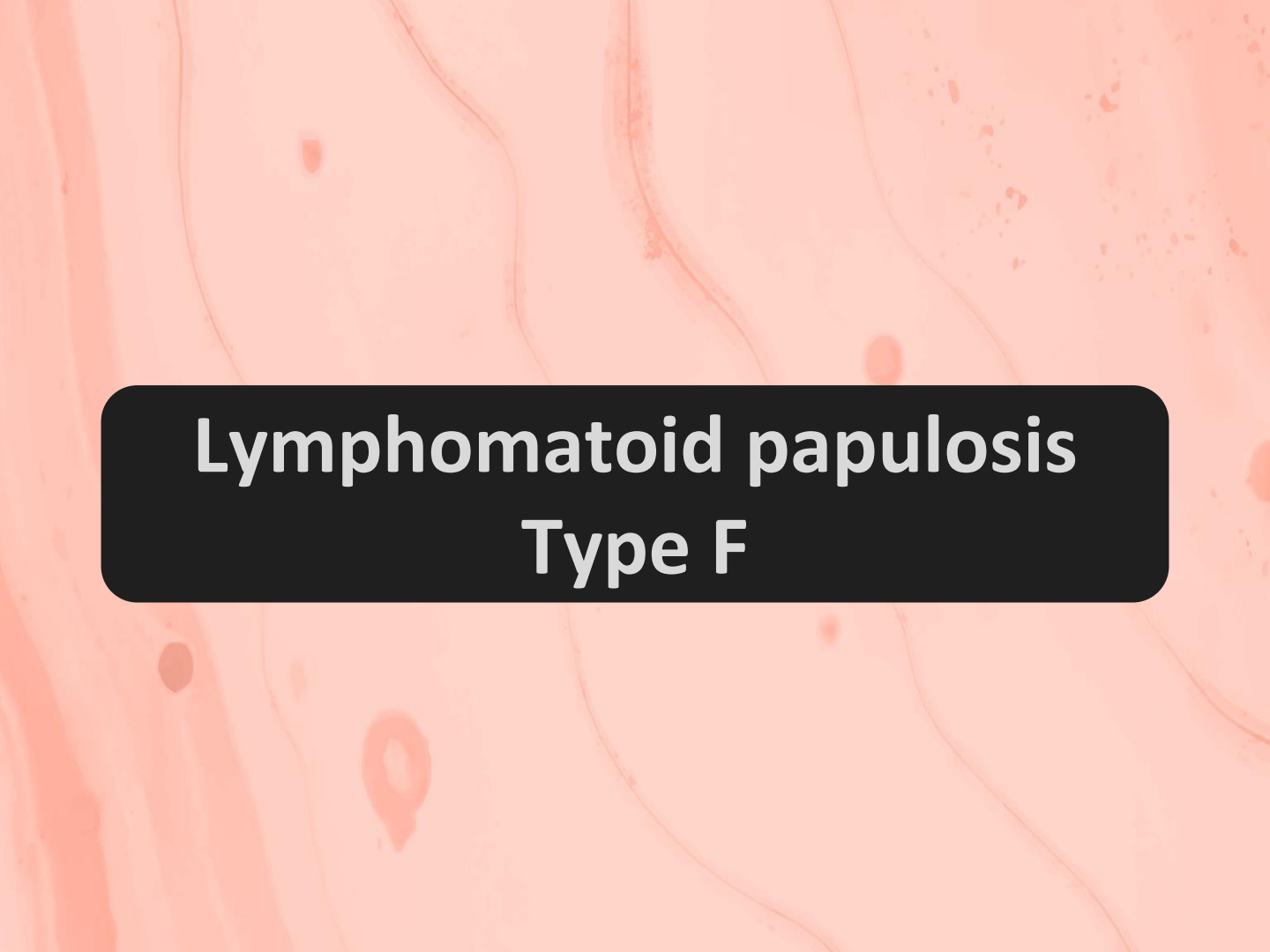


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The background of the slide is a microscopic image of a skin biopsy stained with hematoxylin and eosin (H&E). It shows a dense infiltrate of atypical lymphoid cells within the dermis, characteristic of lymphomatoid papulosis Type F. The cells are arranged in a diffuse pattern, and there are some larger, atypical cells with prominent nuclei. The overall appearance is that of a lymphoid infiltrate within the dermal tissue.

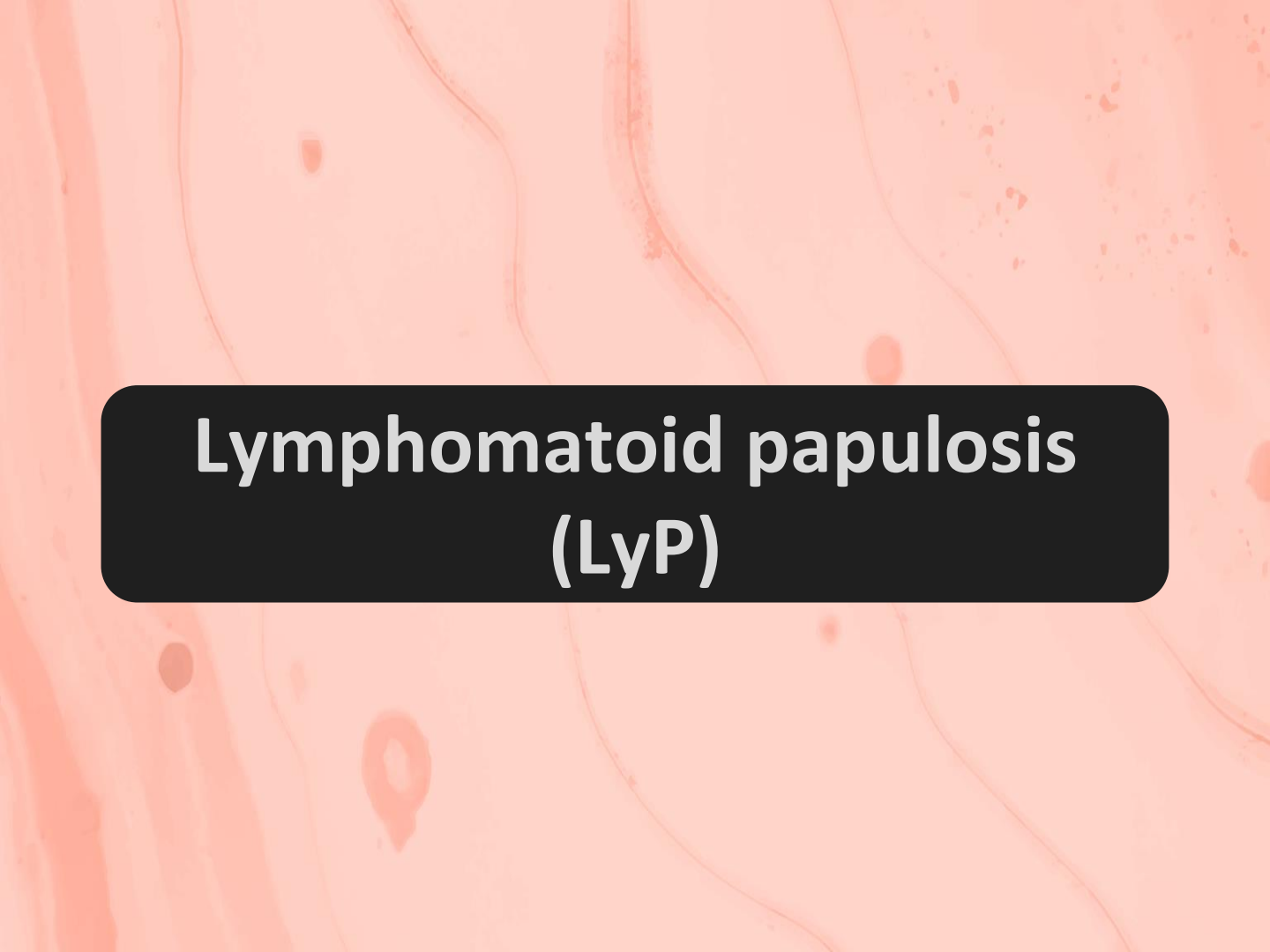
**Lymphomatoid papulosis  
Type F**

# Investigation

- CBC
  - Hb 12.8 g/dL, Hct 38.6%,
  - Plt 224,000/cumm,
  - WBC 5,110/cumm,  
N 43%, L 48%, M 5%, E 3%, B 1%
- Liver/renal function: WNL

# Management

- Acitretin (10) 1 tab PO BID
- 0.25% desoximetasone cream AP BID
- Plan to start PUVA

The background of the slide is a microscopic image of tissue, likely a skin biopsy, showing various cellular structures and fibers in shades of pink and orange. A prominent black rounded rectangle is centered on the slide, containing the text 'Lymphomatoid papulosis (LyP)' in white, bold, sans-serif font.

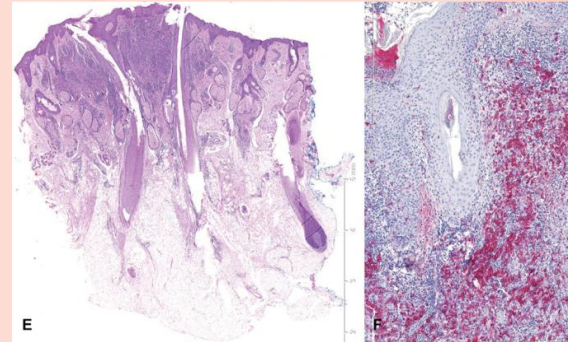
# Lymphomatoid papulosis (LyP)

LyP type	CD4	CD8	CD30	Histopathology	Lymphocyte morphology
A	+	-	+	Wedge-shaped infiltrate	Large
B	+	-	+/-	Epidermotropic infiltrate	Small to medium
C	+	-	+	Nodular cohesive infiltrate	Large
D	-	+	+	Epidermotropic infiltrate	Small to medium
E	-	+	+	Angioinvasive infiltrate	Medium
F	+	+	+	Perifollicular infiltrate and folliculotropism	Medium to large
DUSP22-IRF4	-	+/-	+	Epidermotropism by small to medium atypical lymphocytes Medium to large blast cell in the dermis	

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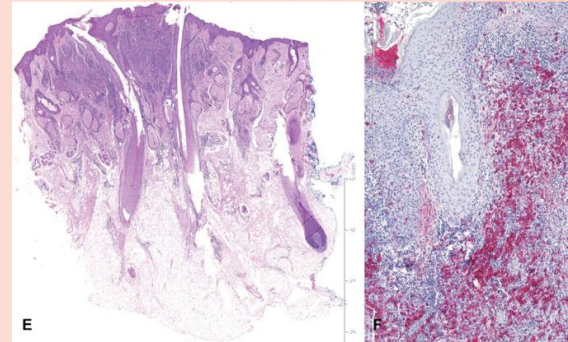
# Follicular lymphomatoid papulosis revisited: A study of 11 cases, with new histopathological findings

- Perifollicular infiltrate of CD30+ medium to large atypical lymphoid cells
- Variable degree of folliculotropism
- Less common features
  - Follicular epithelial hyperplasia
  - Ruptured hair follicle
  - Neutrophils in the follicle



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# LyP & PLEVA

## Controversial relationship

Artifact of sampling  
of LyP lesions  
at various stages

vs.

Biologically related  
disorders

# Prognosis

- LyP lasts for months to years
- Self-healing
- 10-year survival rate nearly 100%
- 20% of patients may develop a second lymphoid neoplasm
  - Mycosis fungoides
  - Anaplastic large-cell lymphoma
  - Hodgkin lymphoma

# Treatment

Few lesions

- Observation
- Topical/intralesional corticosteroids

# Treatment

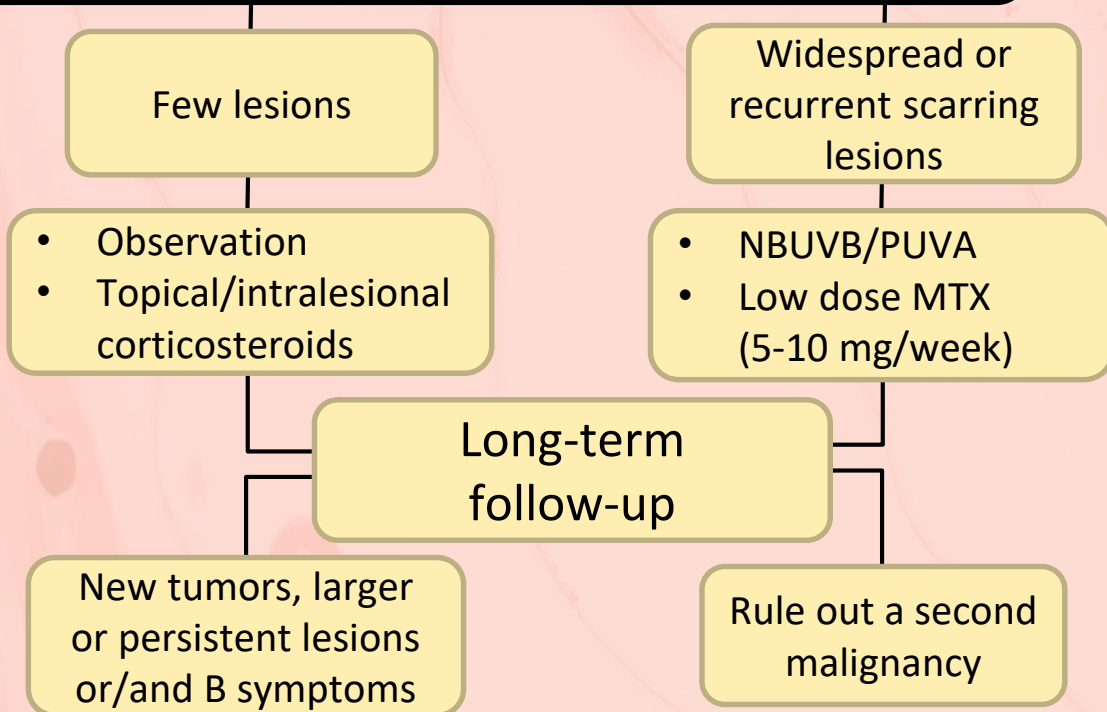
Few lesions

- Observation
- Topical/intralesional corticosteroids

Widespread or recurrent scarring lesions

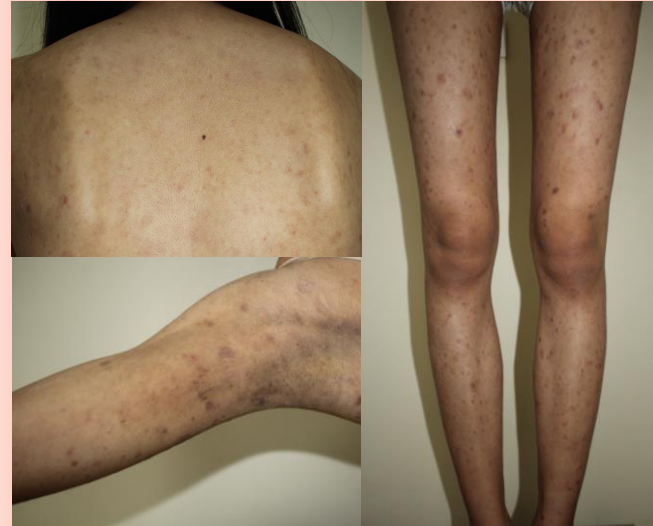
- NBUVB/PUVA
- Low dose MTX (5-10 mg/week)

# Treatment



# Case summary

- 22-year-old female
- Recurrent crusted papules for 1.5 years
- Diagnosis: LyP type F
- Treatment
  - Acitretin 20 mg/day
  - Topical corticosteroids
  - Plan for PUVA
  - Long-term follow-up



# Take home messages

## Lymphomatoid papulosis

Should be suspected in recalcitrant PLEVA

Long-term follow-up is necessary