Interhospital Conference
Case 2

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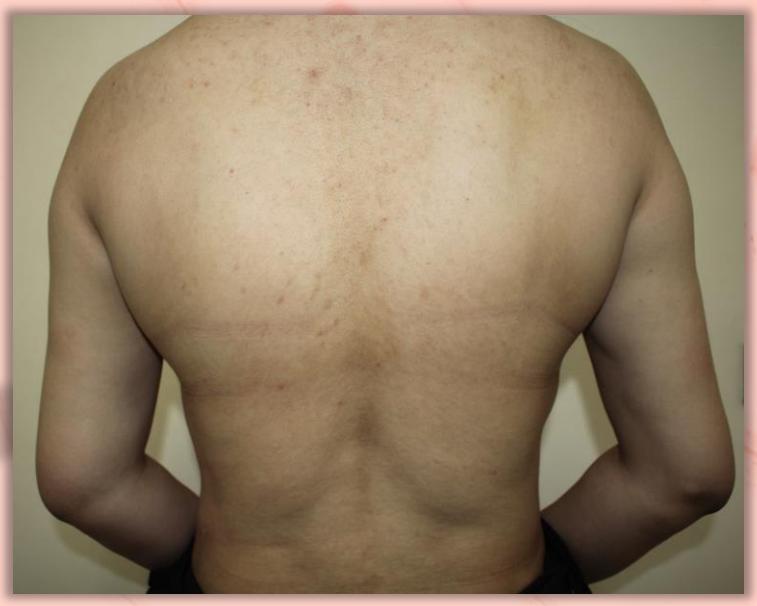
Ploysyne Rattanakaemakorn, MD

A 24-year-old female

Generalized,
ill-defined depressed areas on trunk and extremities for 2 years



Present illness

May 2015

Dx: SLE (Malar rash, polymyalgia, alopecia, ANA 1:1280)

Rx:
prednisolone 60 mg/day
HCQ 200 mg/day

Present illness

May 2015

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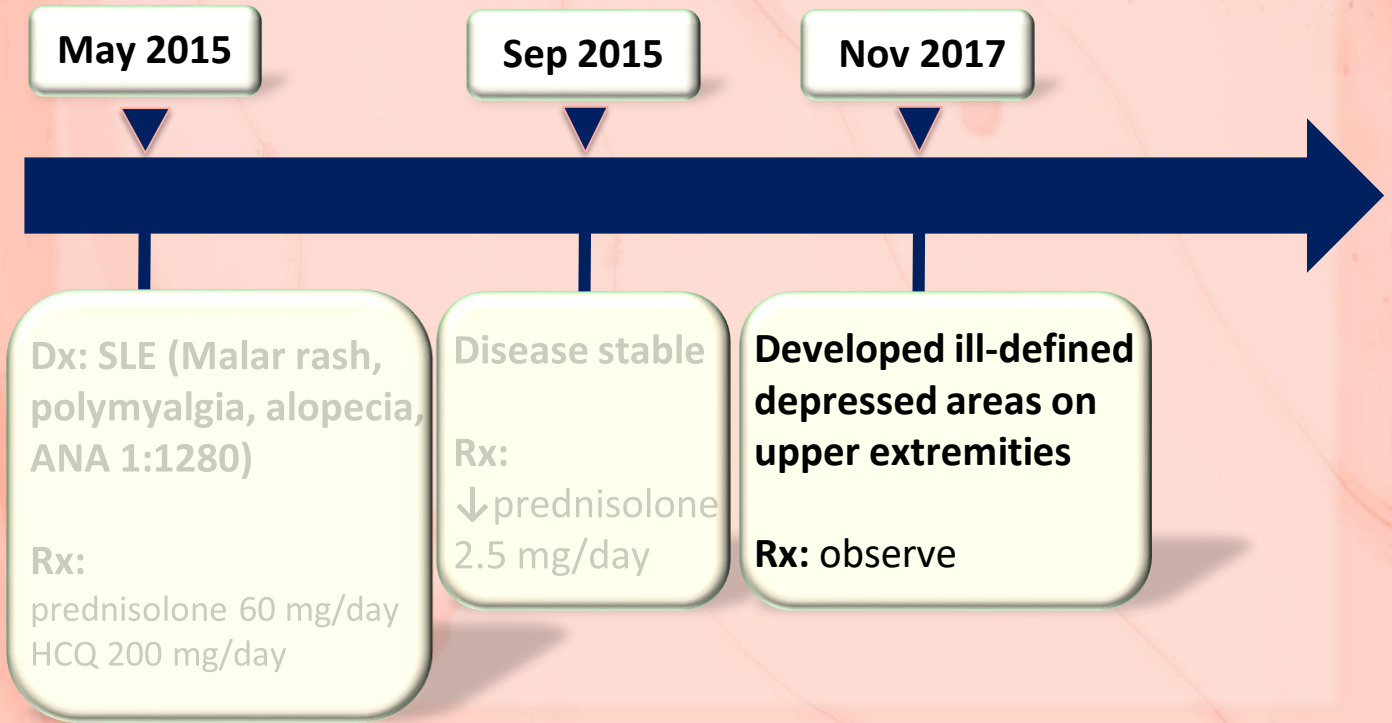
Rx:
prednisolone 60 mg/day
HCQ 200 mg/day

Sep 2015

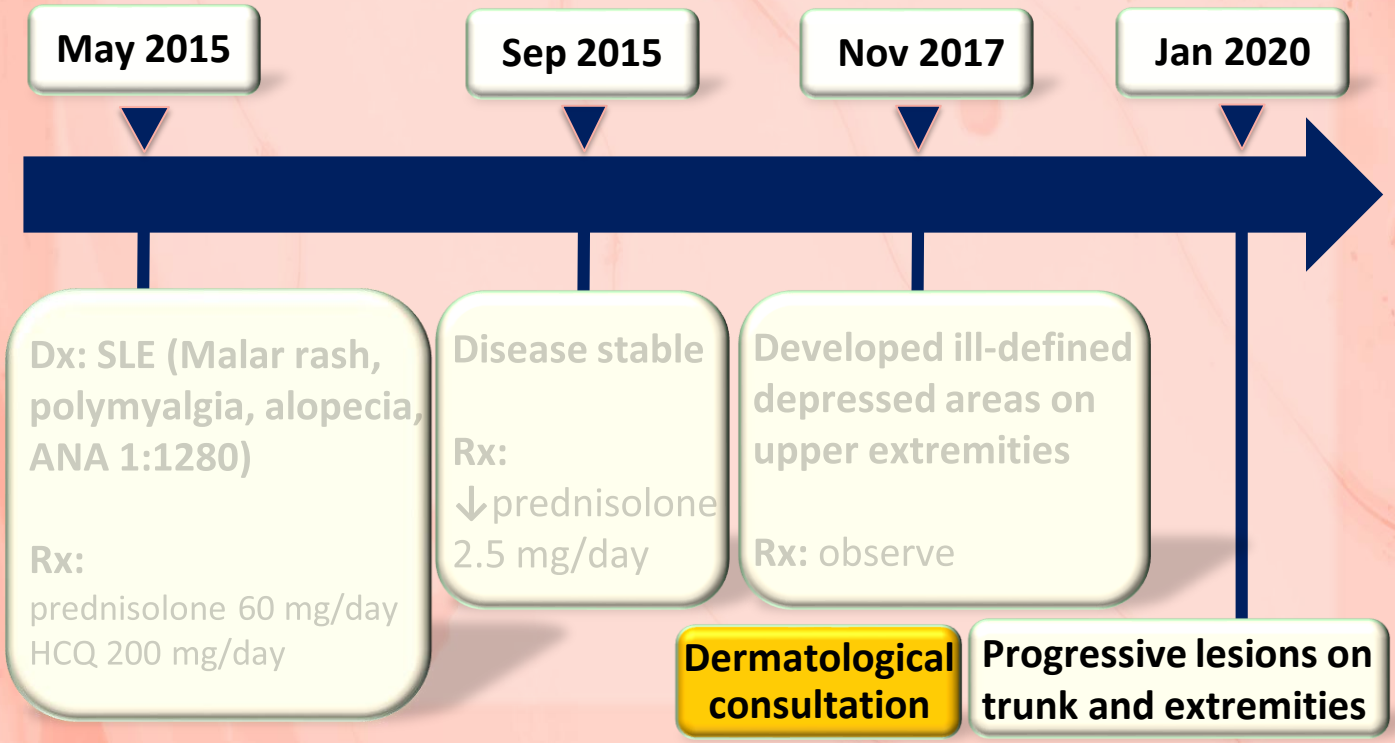
Disease stable

Rx:
↓prednisolone 2.5 mg/day

Present illness



Present illness



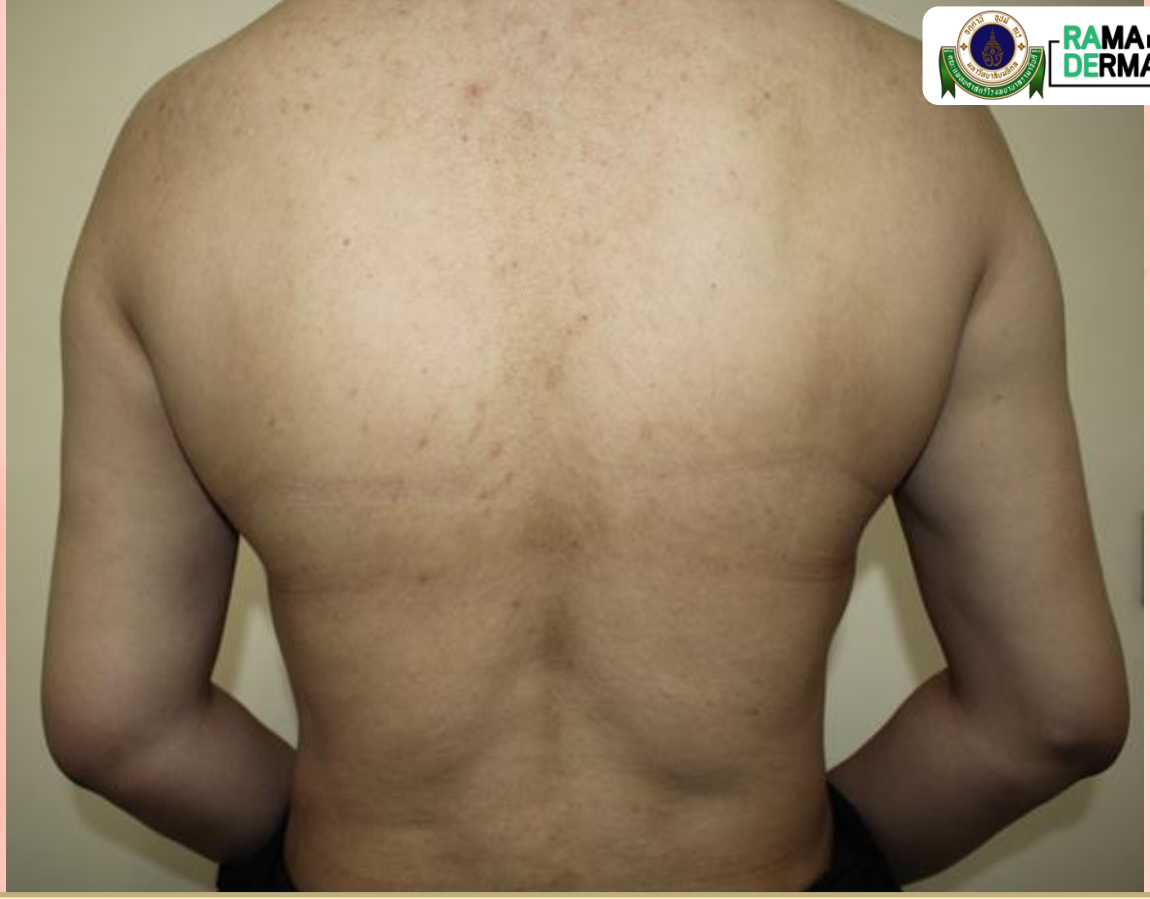
Past history

- Underlying disease
  - ♦ Stable SLE
- Current medications
  - ♦ Prednisolone 2.5 mg/day
  - ♦ Folic acid 5 mg/day

Physical examination

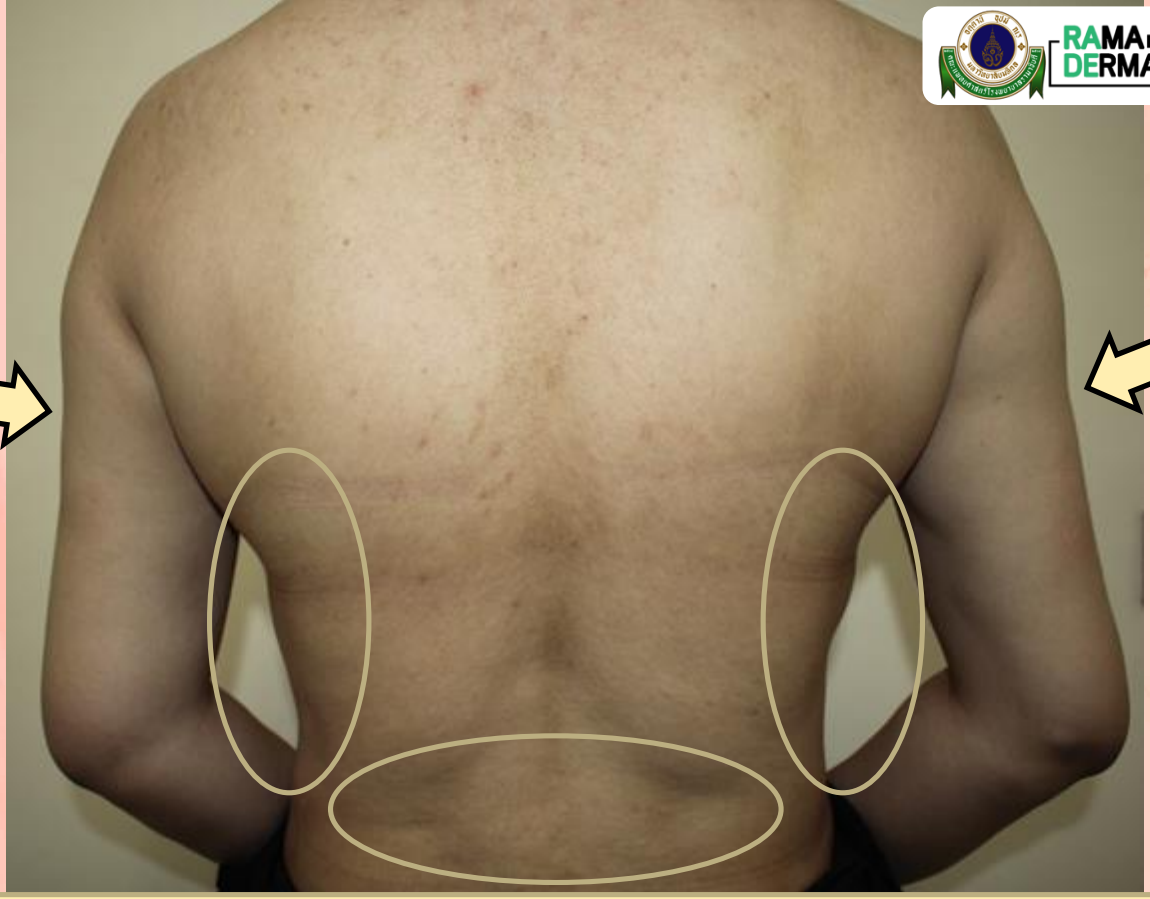
- Vital signs: BT 36 °C BP 120/80 mmHg
  HR 70 bpm RR 20 /min
- BW 60 kg, Ht. 165 cm (BMI 23 Kg/m2)
- HEENT: no pale conjunctivae, anicteric sclerae
- Heart & lungs: WNL
- Abdomen: no hepatosplenomegaly

Dermatological examination



Bilateral, ill-defined, depressed areas

on both arms and trunk



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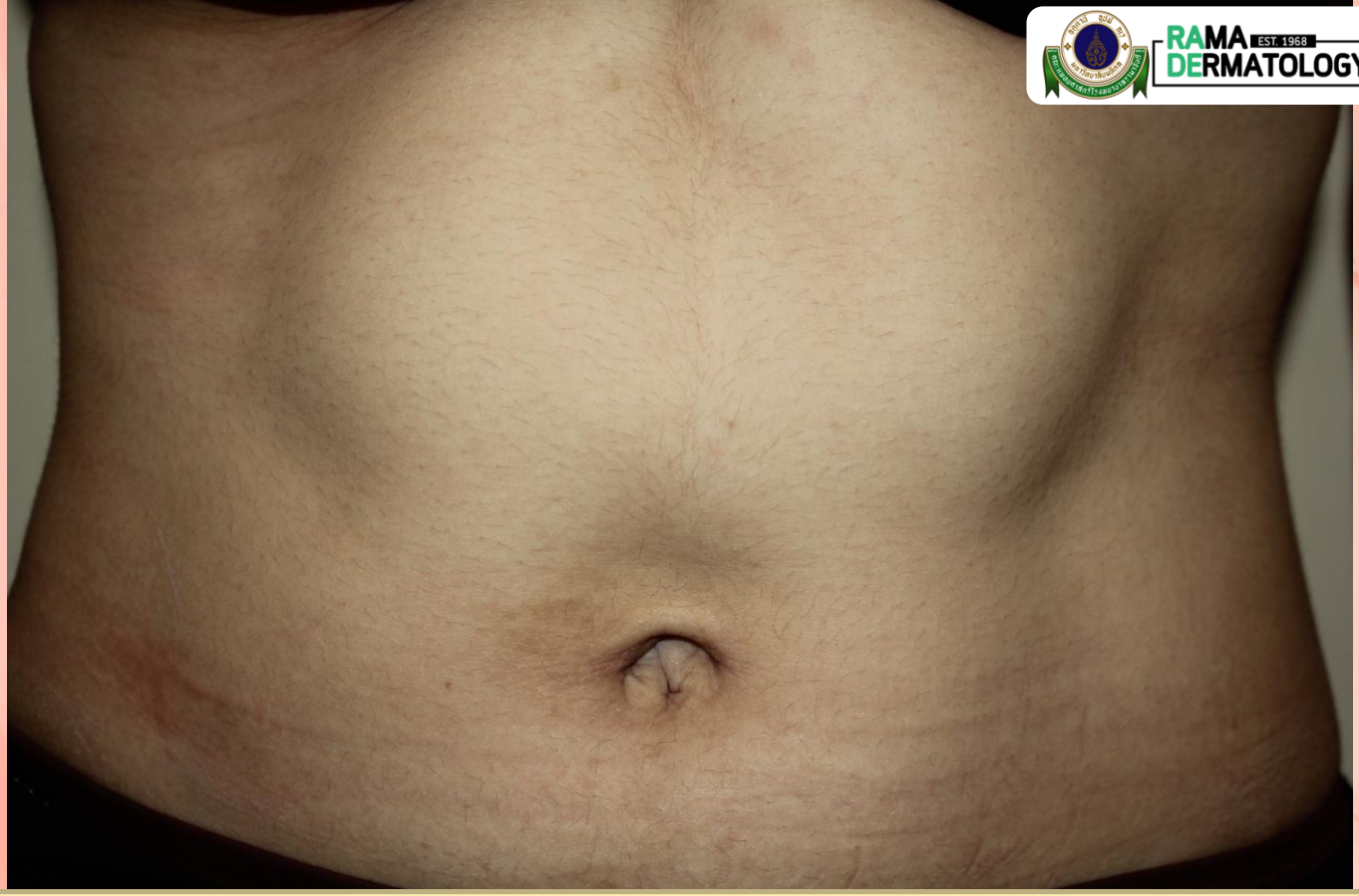
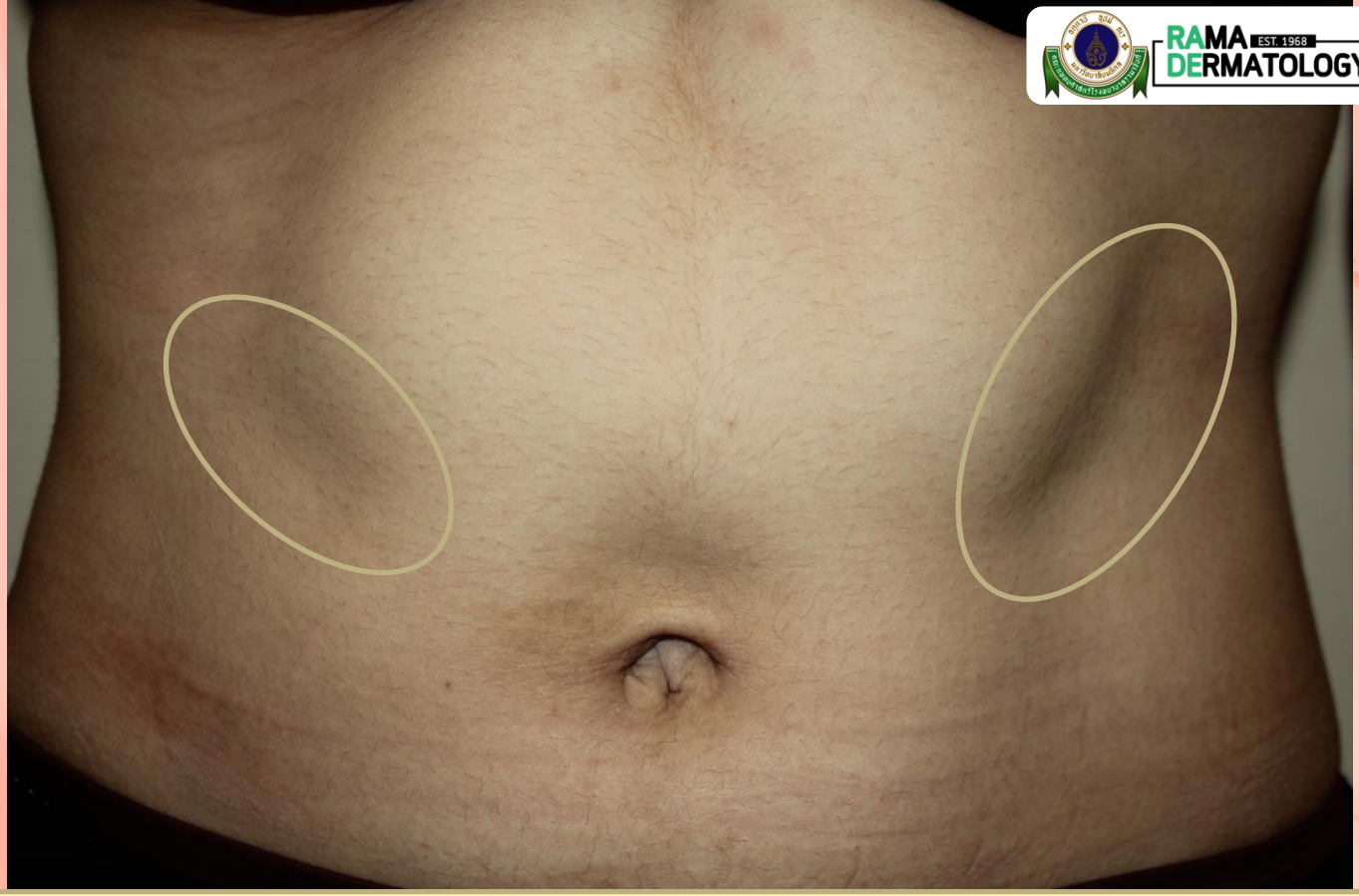
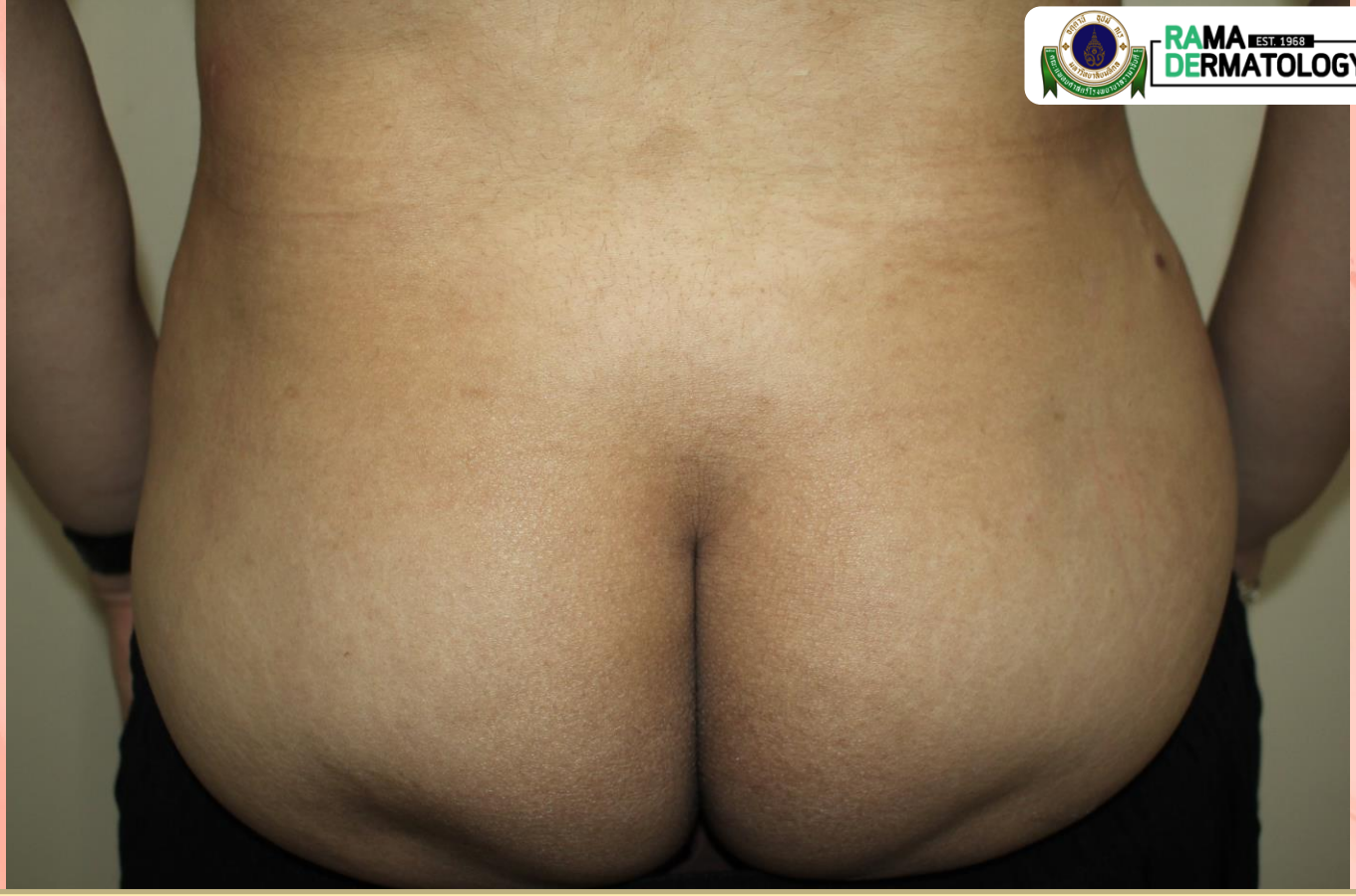


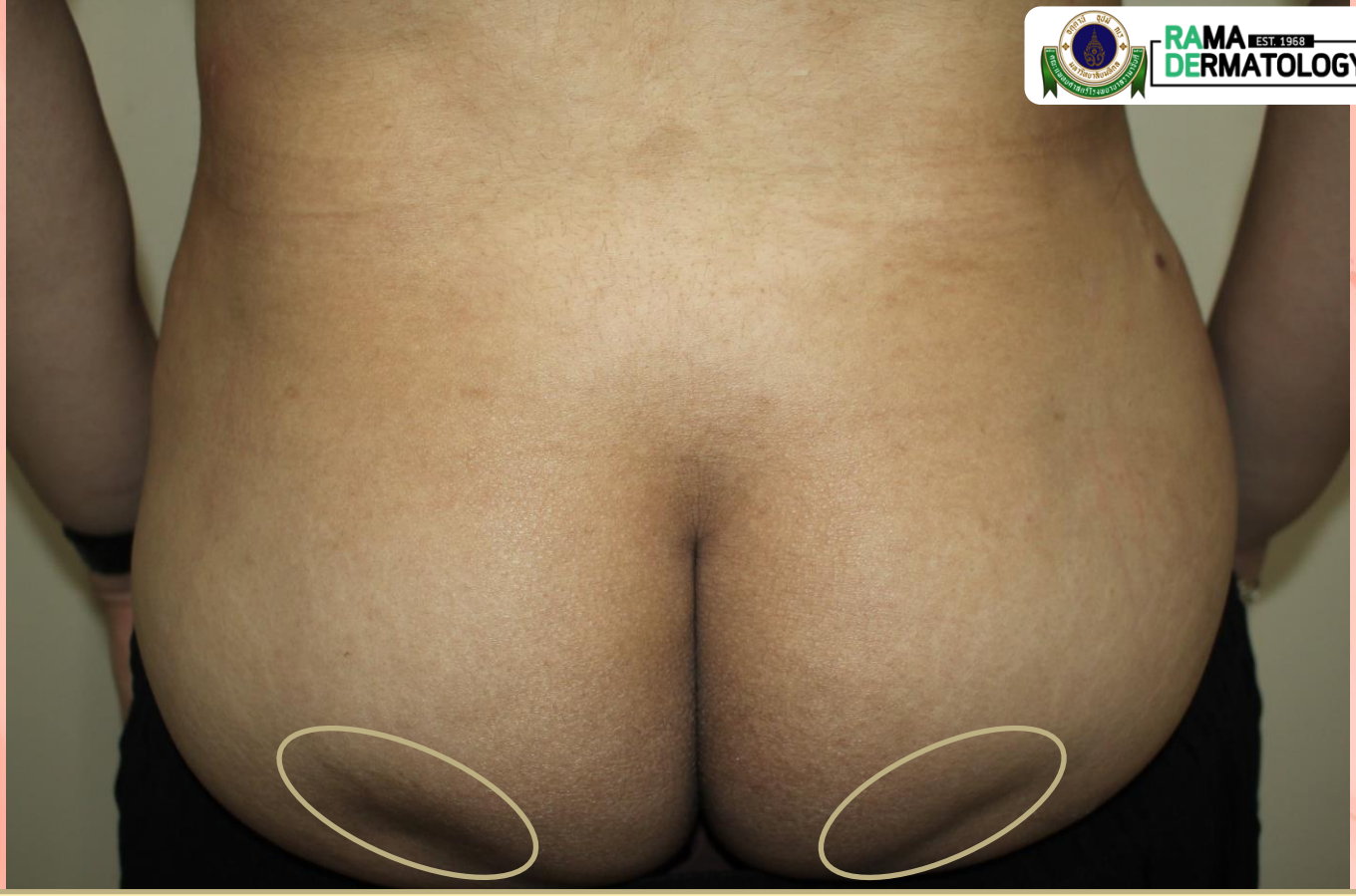
Ill-defined, depressed areas on abdomen



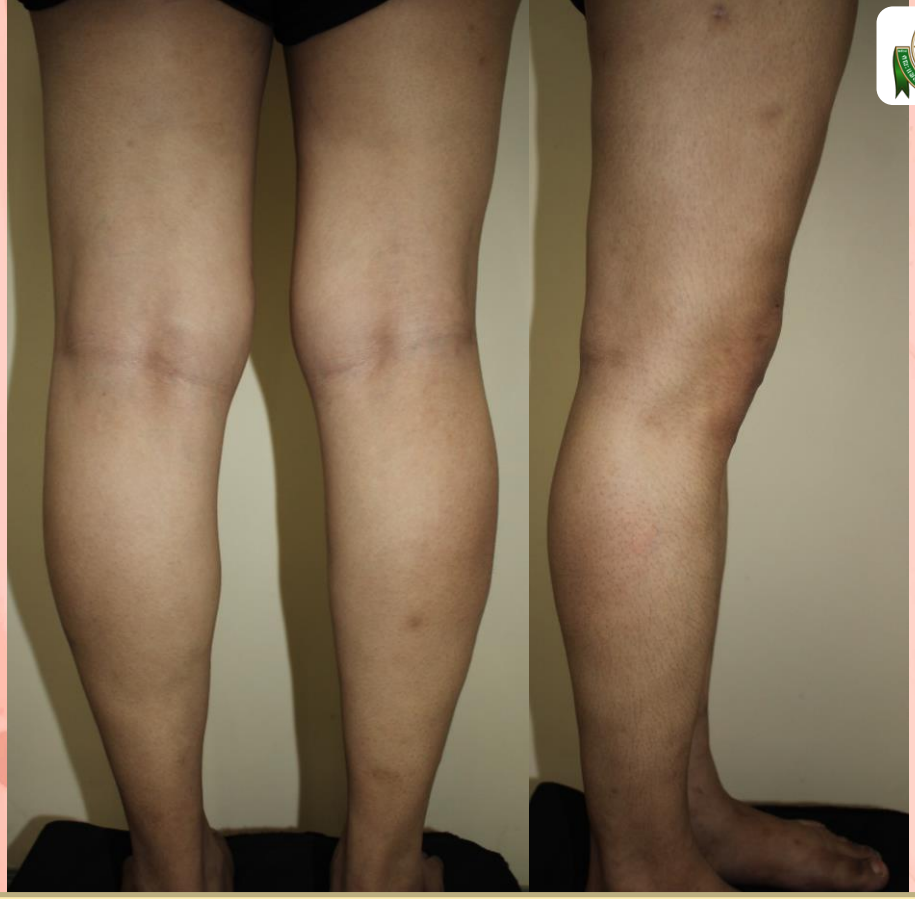
Ill-defined, depressed areas on abdomen



Bilateral, ill-defined, depressed areas on buttock

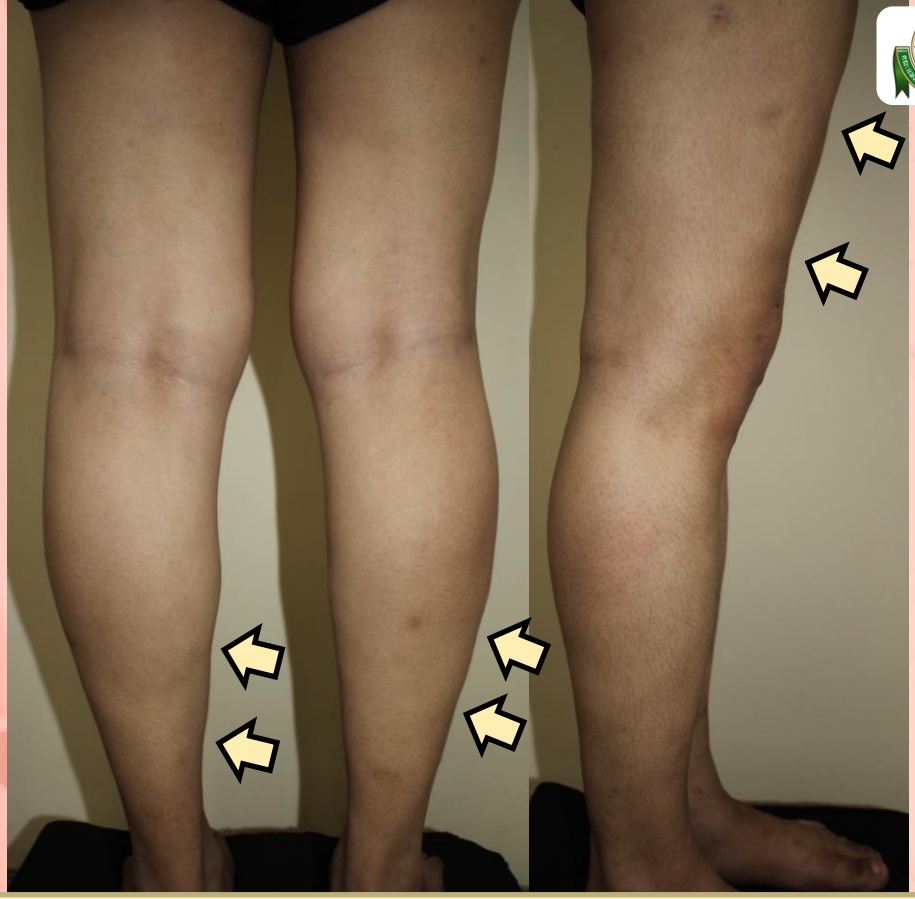


Bilateral, ill-defined, depressed areas on buttock



Bilateral, ill-defined, depressed areas

on both thighs and legs



Bilateral, ill-defined, depressed areas

on both thighs and legs

Problem list

1. Acquired, generalized, depressed areas for 2 years
2. Stable SLE

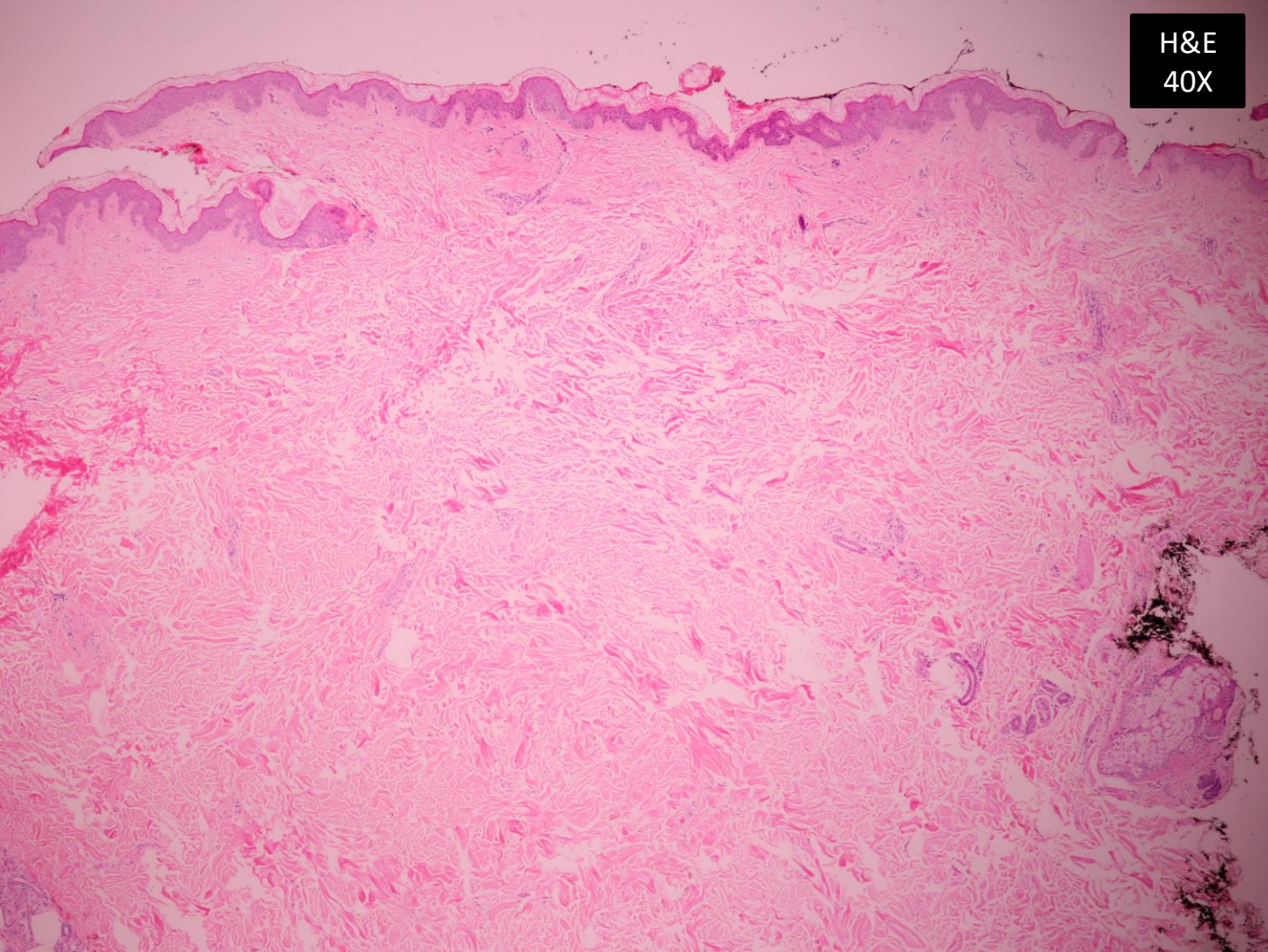
Differential diagnosis

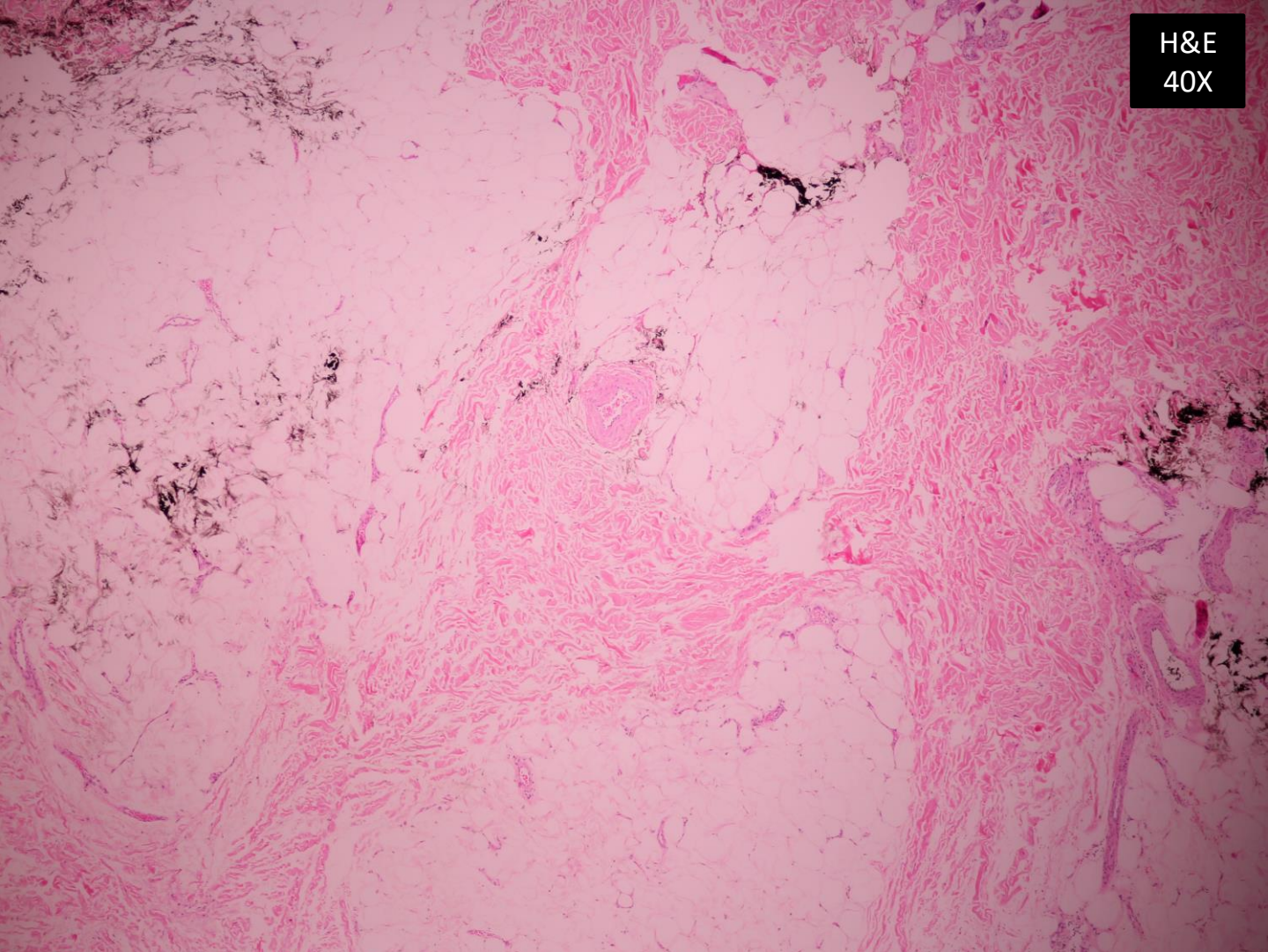
Acquired, generalized, depressed lesions

Inflammation
- **Lipodystrophy syndromes**
- **Lupus panniculitis**
- **Subcutaneous morphea**

Infection
- **Infectious panniculitis**

Histopathology


H&E
40X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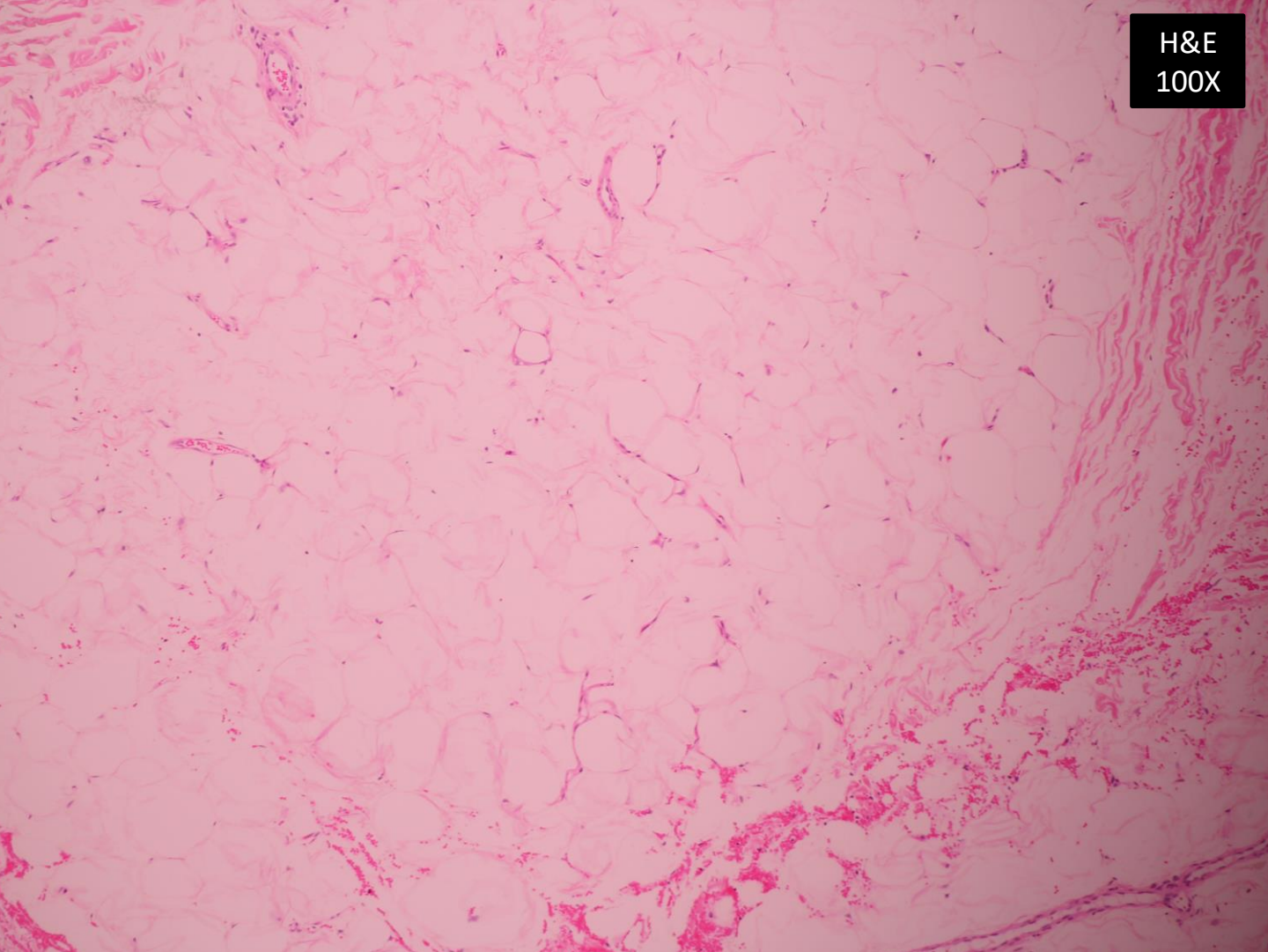
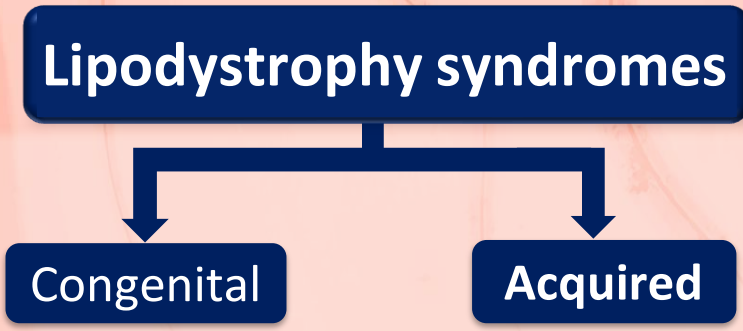

H&E
100X


Lipodystrophy syndromes
Congenital
Acquired

Lipodystrophy syndromes

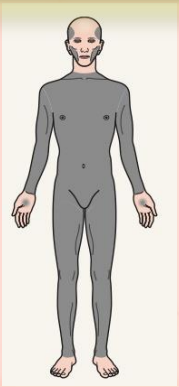
- Congenital
- **Acquired**
  - Localized
    - Trauma
    - Injection
    - Pressure

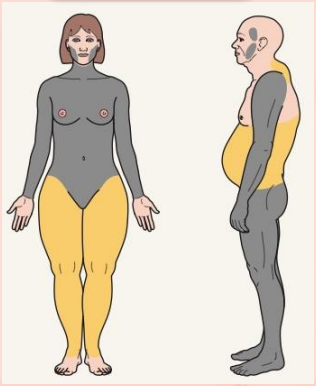
Lipodystrophy syndromes

- Congenital
- Acquired
  - Localized
  - Generalized
  - Partial

Lipodystrophy, often with muscular prominence

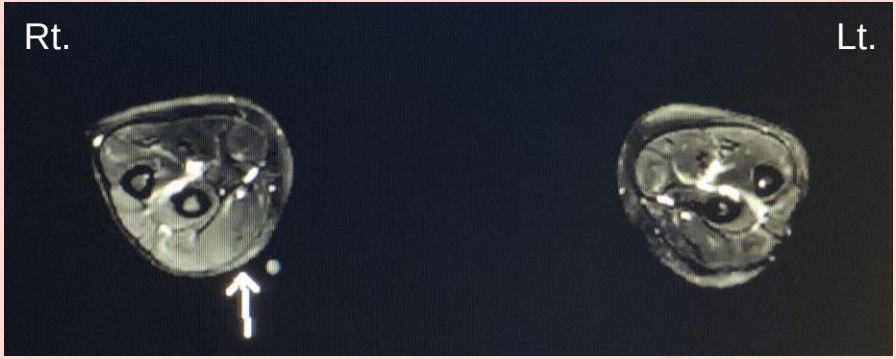
Lipohypertrophy





Bolognia, 4th edition.

MRI of forearms



- Mild inflammatory process at subcutaneous tissue of both forearms

MRI of forearms

- No muscular dystrophy

Investigation

- Liver and renal function tests
- Lipid profiles
- Serum glucose
- Serum uric acid

WNL

Acquired generalized lipodystrophy or Lawrence syndrome

Epidemiology

- Rare condition ~80 cases until now
- Childhood to adolescence
- F:M 3:1

Bolognia, 4th edition.
Fitzpatrick, 9th edition.

Etiology and pathogenesis

- Unknown
- 25% have a preceding panniculitis
- Localized → generalized fat loss

Bolognia, 4th edition.
Fitzpatrick, 9th edition.

Locations of fat loss

Common — Large areas of face, trunk, and extremity

Uncommon — Palms, soles, and intraabdominal regions

Not found — Bone marrow and retroorbital fat

Bolognia, 4th edition.
Fitzpatrick, 9th edition.
Medicine (Baltimore). 2003;82:129-146.

Subtypes

- Type 1: panniculitic variant (25%)
- Type 2: autoimmune variant (25%)
- Type 3: idiopathic (50%)

Bolognia, 4th edition.
Fitzpatrick, 9th edition.
Medicine (Baltimore). 2003;82:129-146.

Complications

- Severe hepatic steatosis and fibrosis
  - ♦ A significant cause of death
- Diabetes mellitus
  - ♦ Latency period ~ 4 years
- Hypertriglyceridemia
- Proteinuric nephropathy

Bolognia, 4th edition.
Fitzpatrick, 9th edition.
Medicine (Baltimore). 2003;82:129-146.

Prognosis

- Severe metabolic complications
- ↑ Risk of peripheral T-cell lymphoma
- Panniculitis-associated variety
  - ♦ Severe fat loss
  - ♦ Metabolic complications

Less

Fitzpatrick, 9th edition.

Suggested investigation

- Serum chemistry profiles
  - ♦ Glucose
  - ♦ Liver enzymes
  - ♦ Lipids
  - ♦ Uric acid
- Insulin resistance

Suggested investigation

- Serum leptin
  - ♦ Low level
  - ♦ Guide treatment decisions

  (metreleptin replacement therapy)
- Skin biopsy

Special tests

- Skinfold thickness
  - ♦ Anthropometry
- Body fat distribution
  - ♦ Dual X-ray absorptiometry
  - ♦ MRI

Management

- No specific treatment

1. Reassure
2. Supportive treatments
    - ♦ Cosmetic surgery
3. Management and surveillance of complications

Fitzpatrick, 9th edition.

Our case

- 3 subtypes
  - ♦ Type 1: panniculitic (25%)
  - ♦ Type 2: autoimmune (25%)
  - ♦ Type 3: idiopathic (50%)

Type 2: autoimmune variety

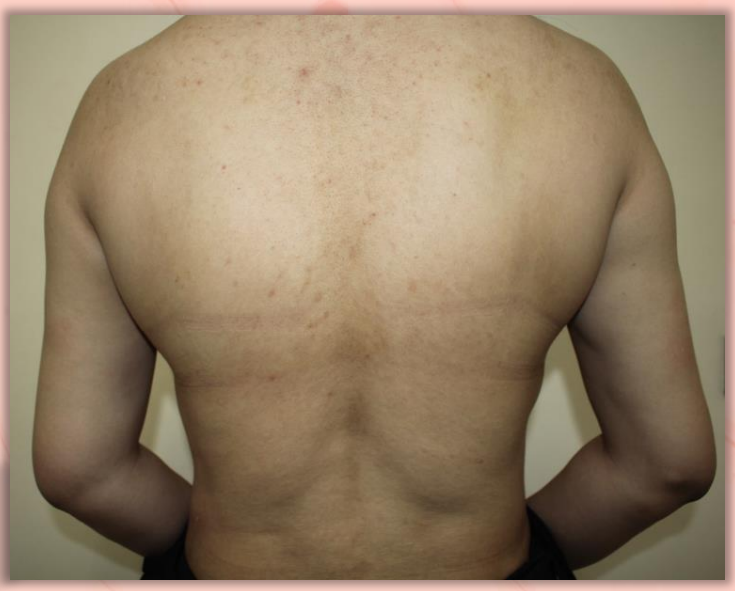
- **Organ-specific autoimmunity**
  - Type 1 diabetes mellitus
  - Autoimmune hepatitis
- **Systemic autoimmune diseases**
  - Juvenile dermatomyositis
  - Juvenile idiopathic arthritis
  - Sjogren syndrome

Bolognia, 4th edition.
Fitzpatrick, 9th edition.

Case summary

- A 24 y/o female with SLE
- **CC:** Generalized depressed areas on trunk and extremities for 2 years

- **Dx:** Acquired generalized lipodystrophy or Lawrence syndrome
- **Treatment:** Denied lipofilling
- **Plan:** Surveillance of complications



Take home message

Acquired generalized lipodystrophy (Lawrence syndrome)

Associated with autoimmune diseases

Surveillance of metabolic complication is suggested