

ESIM Clinical Case Presentation

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Clinical Case

Woman, age 59, caucasian

History

3 months dry cough

Progressive dyspnea with exercise and in rest

Weight loss 2kg

Diffuse pains

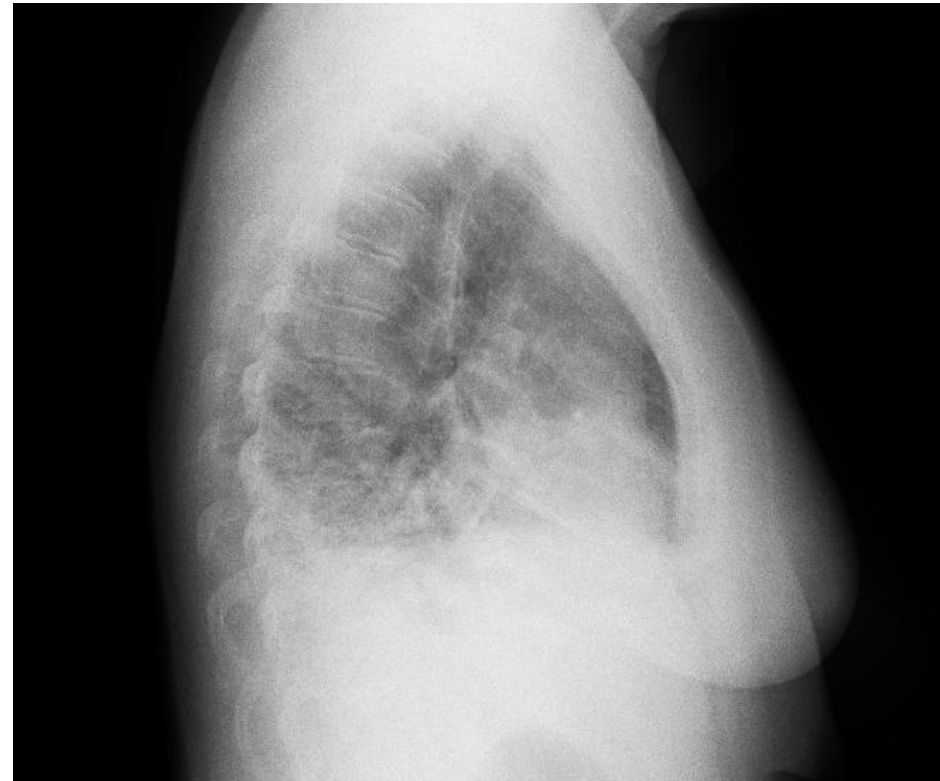
Clinical examination

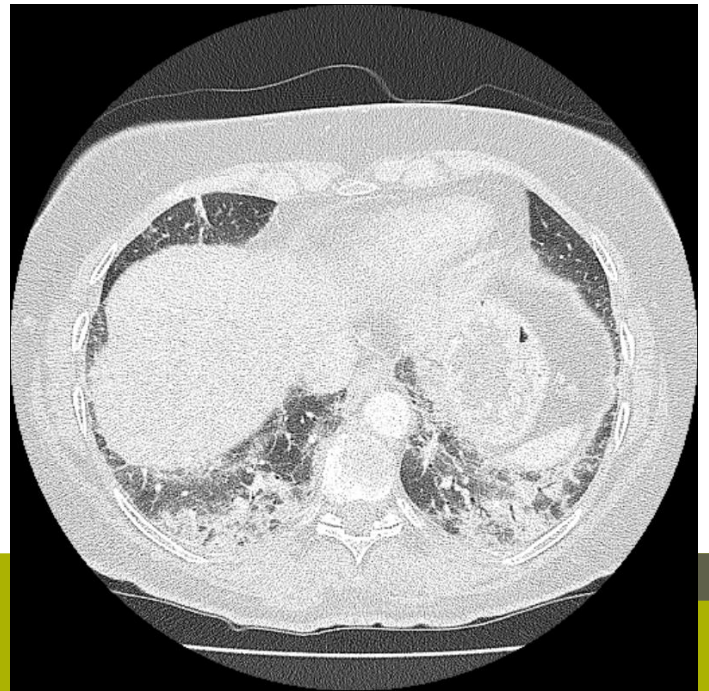
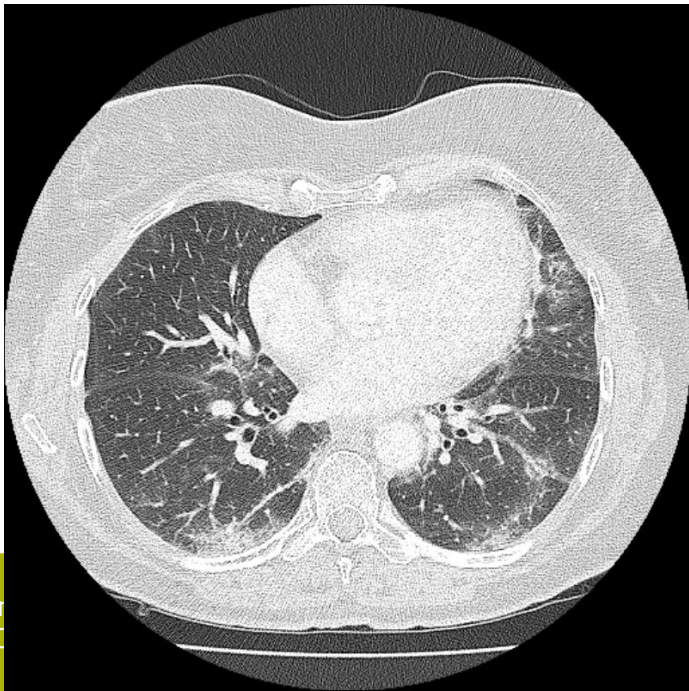
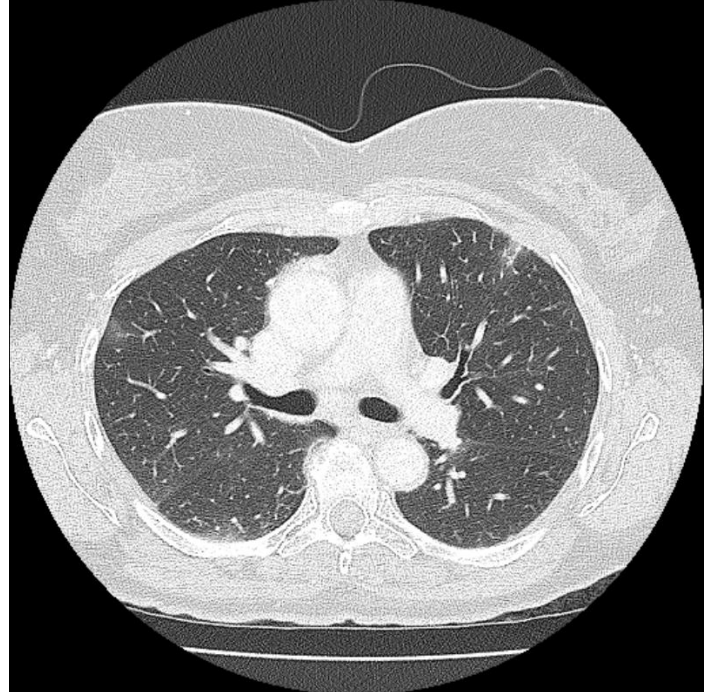
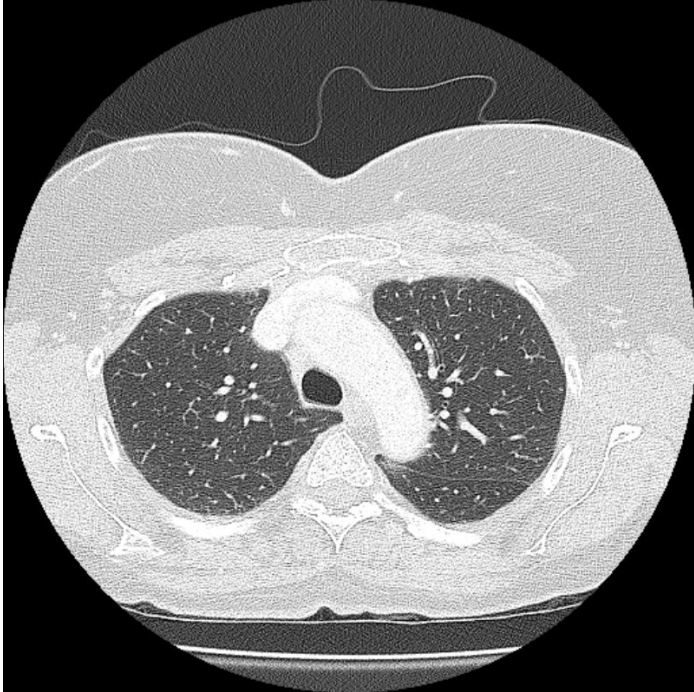
Weight 64kg, BP 140/80mmHg, HR 80/’, Sat 91%, subfebril

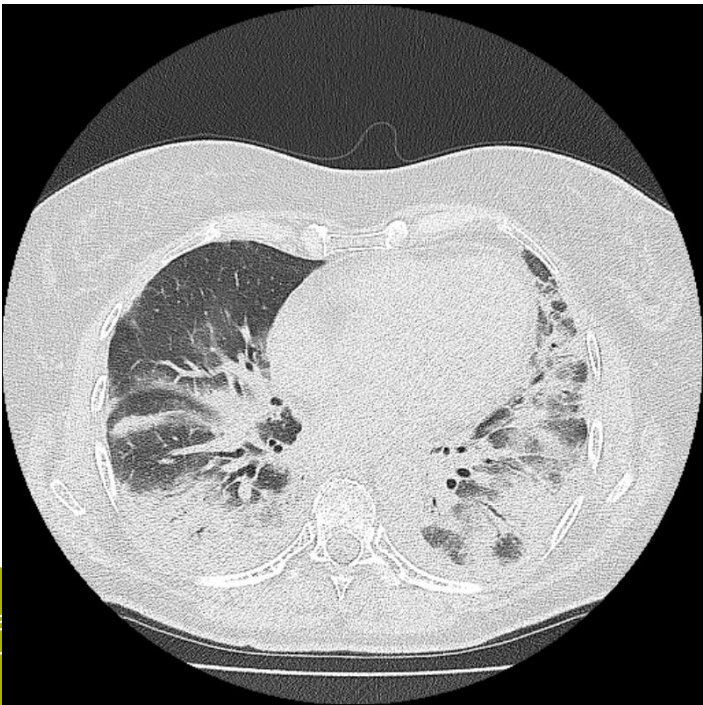
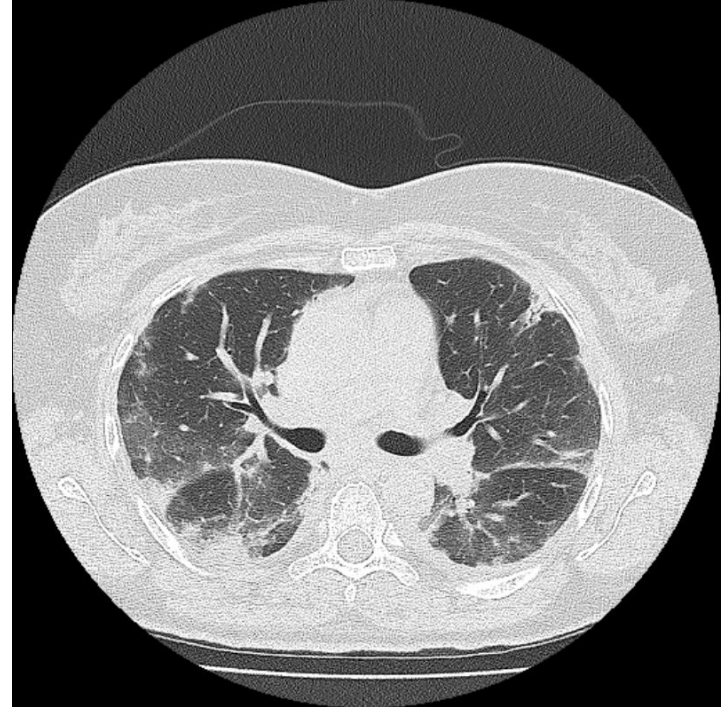
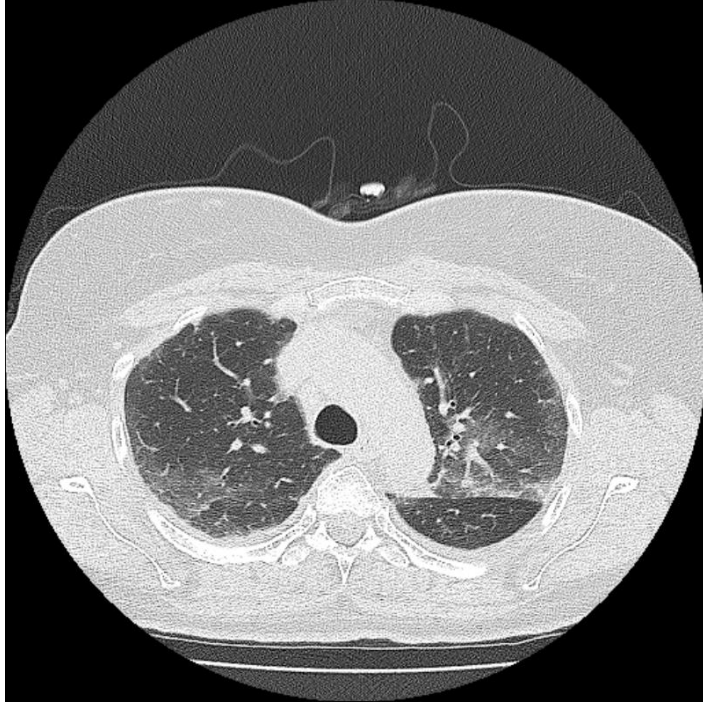
Lungauscultation: bibasilar inspiratory crackles

Swollen hands

Chest x-ray – CT thorax







Work-up

Laboratory testing

CRP 1.7 mg/dl (<1), ESR 30 mm/h (<20), CK 220 IU/l (<110), ANA 1/160 (speckled) (<1/160), anti-ENA + (anti-Jo-1 +)

ABG

pH 7.4 (7.35-7.45), pCO₂ 35 mmHg (35-45), pO₂ 65 mmHg (80-100), Bic 23 mmol/l (22-28)

Pulmonary function test

restriction, low diffusion

Bronchoscopy

BAL: lymphocytes, majority CD8

Evolution

- Fast respiratory deterioration
- Transfer to intensive care: intubation, mechanical ventilation
- R/ large spectrum AB and high dose corticotherapy
- Extubation after 11d

Work-up part 2

Skeletal scintigraphy
normal

EMG
mixed myopathic and neuropathic pattern

MRI quadriceps
nl

Muscle biopsy
waiting for result

Diagnosis

Anti-synthetase syndrome:

- **interstitial pneumonia**
- **anti-synthetase antibody (Jo-1)**
- inflammatory myopathy
- polyarthritis
- Raynaud syndrome
- skin anomalies

Treatment

Prednisolon 1mg/kg

Azathioprine 2mg/kg

Anti-synthetase syndrome

1. Part of myositis and arthritis patients have antibodies anti-Jo-1
2. These patients have more severe lung disease
3. Interstitial lung disease can be the first and most important manifestation
4. Trend to classify patients with anti-Jo-1 under the name of anti-synthetase syndrome