

A deadly case of rash



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Case presentation

- 45 year old female caucasian
- Prior medical history:
 - 6 miscarriages
 - Many year history of scaly psoriasis-like rash on the back of the elbows
- Family history:
 - Crohn's disease (brother)
 - Hypothyrosis (mother)
 - B12 deficiency (mother)
 - Psoriasis (brother)
- Social history irremarkable

Symptoms

- Beginning of symptoms in January 2009:
 - Oral ulcers
 - Red rash in nailbeds bilaterally with raised inflammation
 - Joint pain and stiffness in the small joints of hands, later also in the shoulders and knees
 - Scaling rash on the right shin
 - No history of Raynaud's phenomenon, serositis or pleuritis/pericarditis and no dyspnea

Clinical examination

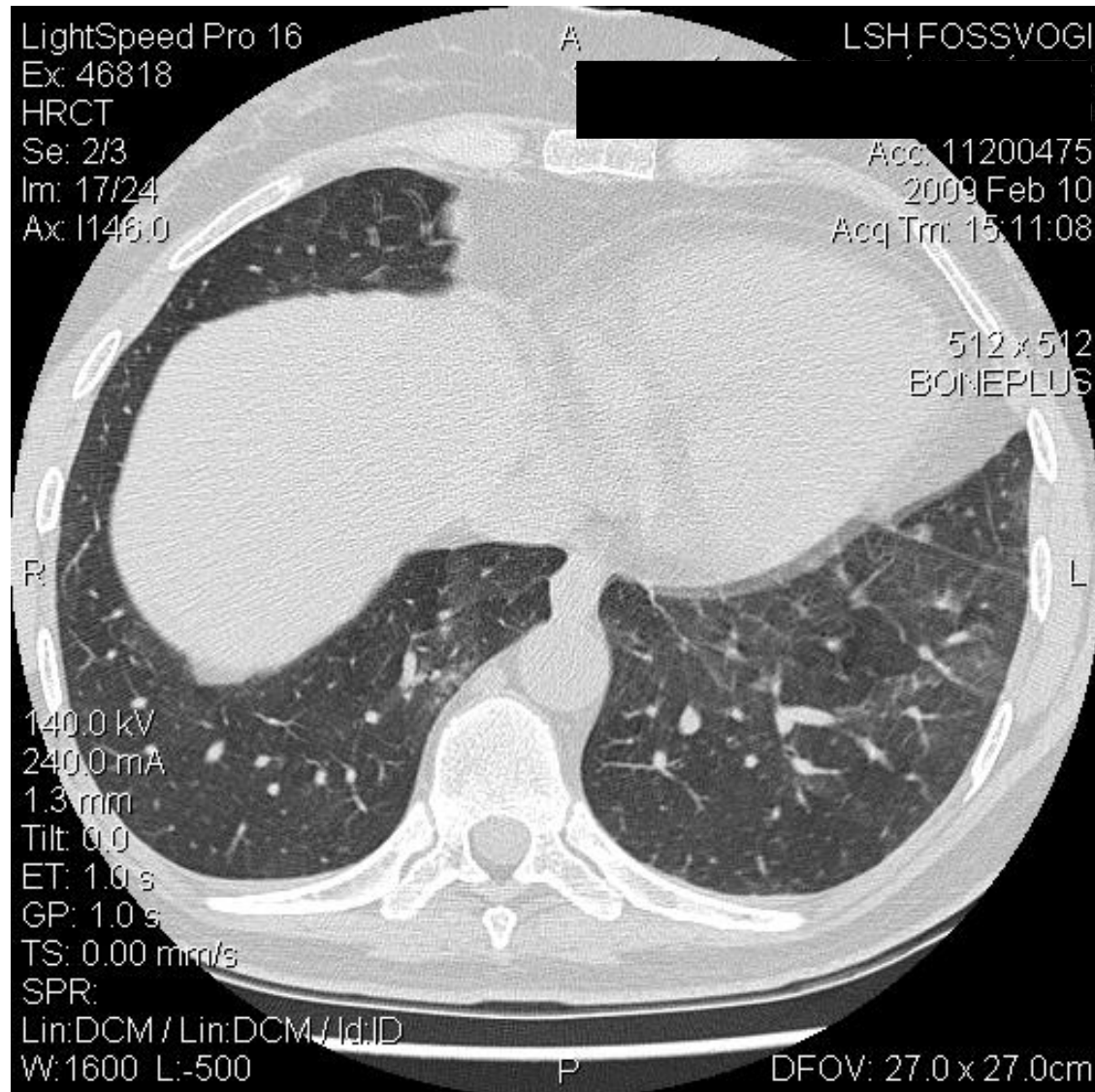
- Clinical examination at onset of symptoms:
 - Oral stomatitis
 - Dilated capillaries in the nailbeds according to capillary microscopy
 - Scaling rash on the elbows and the right shin
 - Little red spots in fingers and palms
 - Slight erythema around the eyes
 - No arthritis, no other clinical symptoms
- A week later she had developed a heliotrop rash in the face and red nodules on the knuckles and the PIP joints. Resembled Gottron's papules



Primary results

- Normal:
 - Blood status and differential count, electrolytes, creatinin, calcium, CRP, ferritin, liver function tests, TSH, protein electrophoresis
 - Immunologic tests (ANA, RF etc.)
 - Arterial blood gases
 - Electrocardiogram
 - Electromyogram
 - Heart ultrasound
- Spirometry: Mild restrictive pattern
- DLCO (lung diffusing capacity) : 53% of normal
- CT-thorax: Minor interstitial changes in both lungs
- Muscle biopsy: Very mild type II atrophy
- Skin biopsy: Mild perivascular chronic inflammation, unspecific
- Further malignancy search was negative

CT thorax 10.2.2009



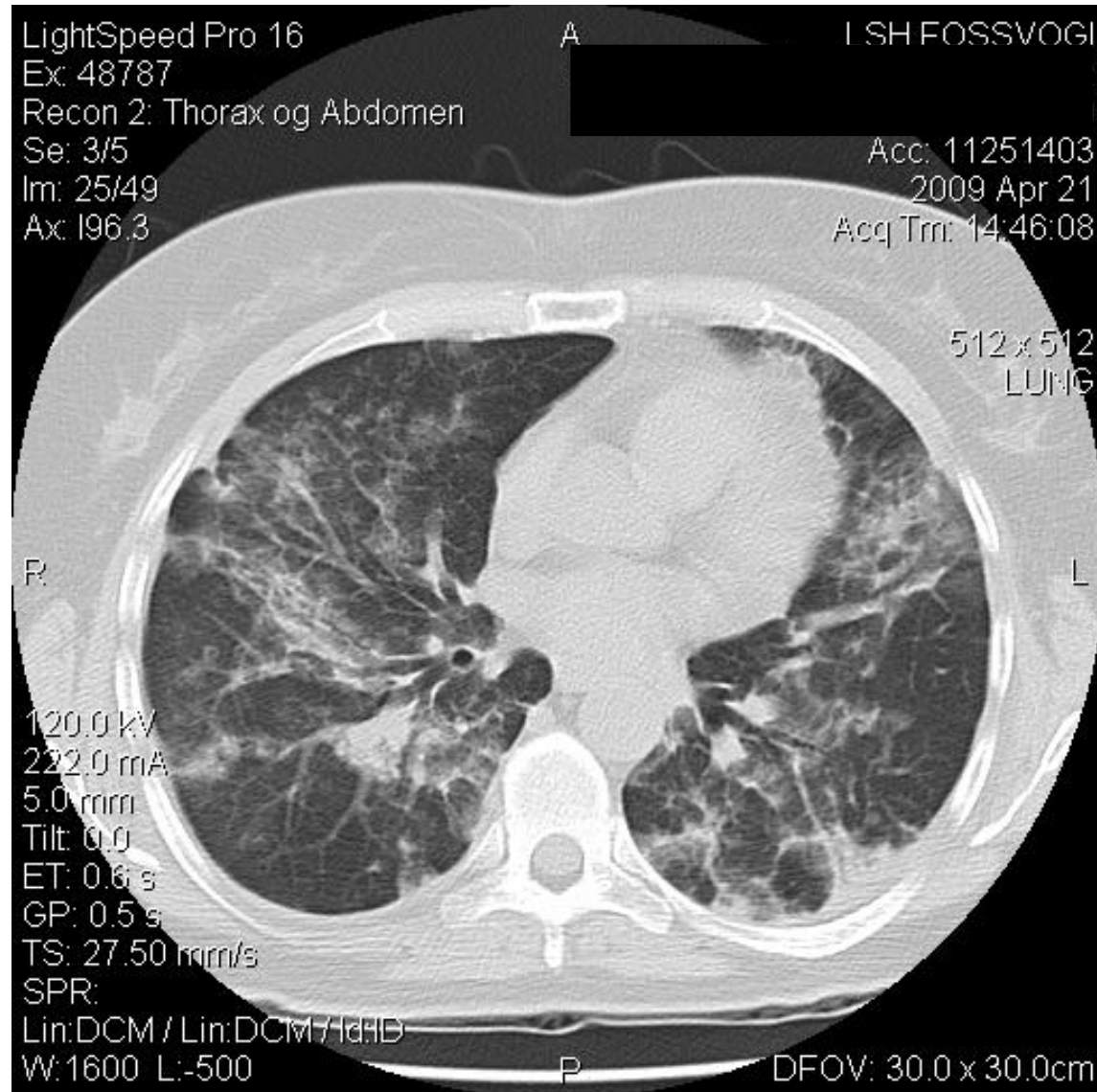
Progression of symptoms

- The symptoms change in March 2009
 - Increased tiredness
 - Increased rash and arthralgias
 - Nightly fever
 - Non-productive cough, dyspnea, pleuritic pain
- Clinical examination:
 - Decreased breath sounds in the basal zones of both lungs, inspiratory crackles

Secondary results

- Normal:
 - Blood status, electrolytes, CK, CRP, APTT, PT
- Immunologic tests
- Arterial blood gases: pH 7,57/ pCO₂ 27/ pO₂ 90/HCO₃ 27/SpO₂ 96%
- CT-thorax: Progression for the earlier CT scan, increasing ground glass changes
- DLCO: 41%, sharp decrease from former testing

CT thorax 17.3.2009



Clinical diagnosis at the time

- **A variant of Dermatomyositis or Scleroderma**

Treatment

- After the primary clinical diagnosis was made
 - corticosteroids and cyclophosphamide
- In the end of March she had developed infiltrations in both lungs
 - Amoxicillin – Clavulanic acid and Azithromycin added

Disease progress

- 15.4. she had worsening dyspnea and hypoxia
 - Underwent open lung biopsy which showed unspecific inflammatory changes
- Rapid deterioration the next day, worsening hypoxia and fever
- Underwent workup for possible lung transplant
 - Cardiac catheterization showed mild pulmonary hypertension
- BiPAP treatment began 29.4., intubation the following day
- She died on the ICU on 7.5.2009, cause of death hypoxic respiratory failure and hypotension
- No autopsy done according to her family's wishes

Post-mortem clinical diagnosis

**Clinically amyopathic
dermatomyositis**

Adult clinically amyopathic dermatomyositis (CADM) with rapid progressive interstitial lung disease (ILD)

- CADM is a subgroup of dermatomyositis, without substantial myositis
- 75% have ILD compared to 50% of patient with DM
- 6 month survival rate of 40%
- Rapidly progressive respiratory failure is the main cause of death
- Positive ANA is protective
- CADM-ILD is most common in Asia
 - 14% of all DM/PM

