Systemic symptoms can be life-threatening in cystic fibrosis

Sophie Gohy, Antoine Froidure & Patrick Lebecque

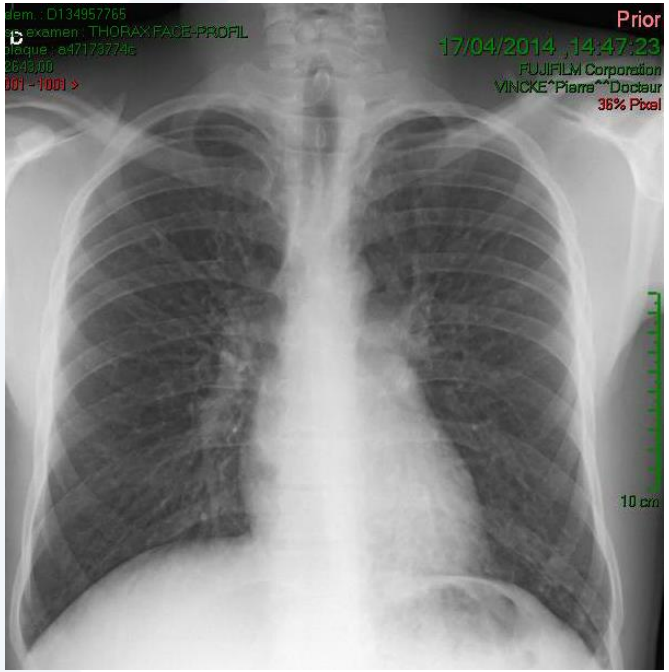
Cliniques universitaires
SAINT-LUC
UCL BRUXELLES

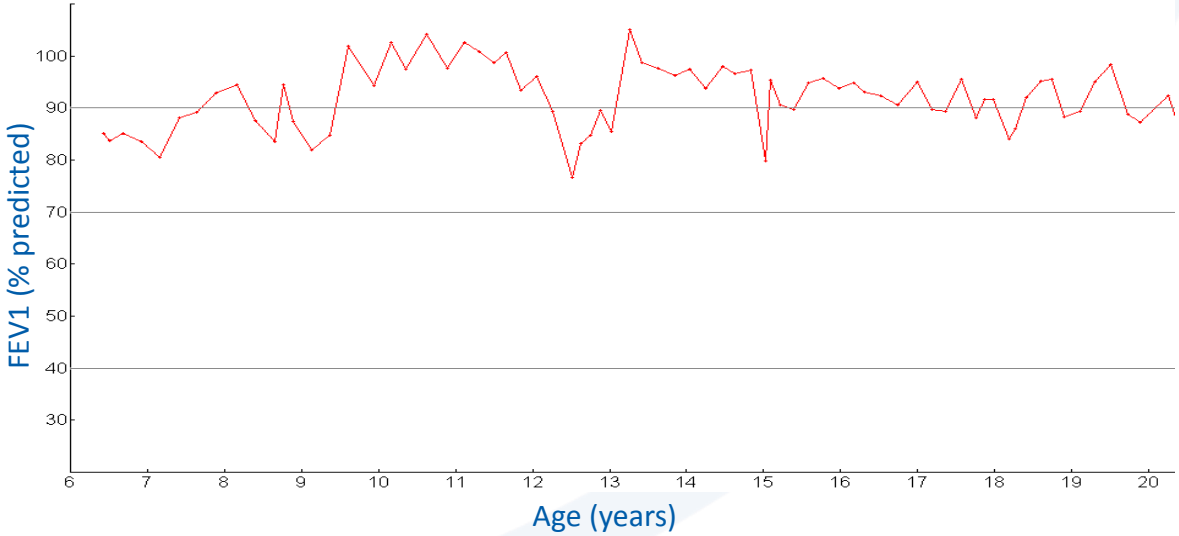
FM: stable & very reliable young CF

- 20 y, M
- F508del/F508del
- Engineering student
- FEV1 ~ 90% predicted
- BMI (Z score) ~ 0
- MSSA (scv)

- Treatment:
  - AB: inhaled colimycin, azithomycin, rifampicin, moxifloxacin
  - Physiotherapy, DNase, acetylcystein, montelukast, inhaled corticosteroids/LABA, pancreatic enzymes, vitamines.

FM: stable & very reliable young CF





First assessment

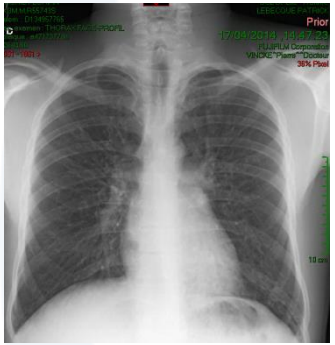
- High fever (40°C)
- Fatigue
- Headache
- Hoarseness
- Polyadenopathy

- Blood tests:

	26/09/2014 = j1
CRP (mg/dl	163
WBC (/µl)	35680
Neutrophils (/µl)	28200
Eosinophils(/µl)	700
IgG (g/L)	NA
Platelets (/µ)	227000
IgE (kUI/L)	5760
Creatinin (mg/dl)	1.05
GOT/GPT/LDH (UI/L)	28/17/NA
INR	NA

First assessment

- Chest X-ray



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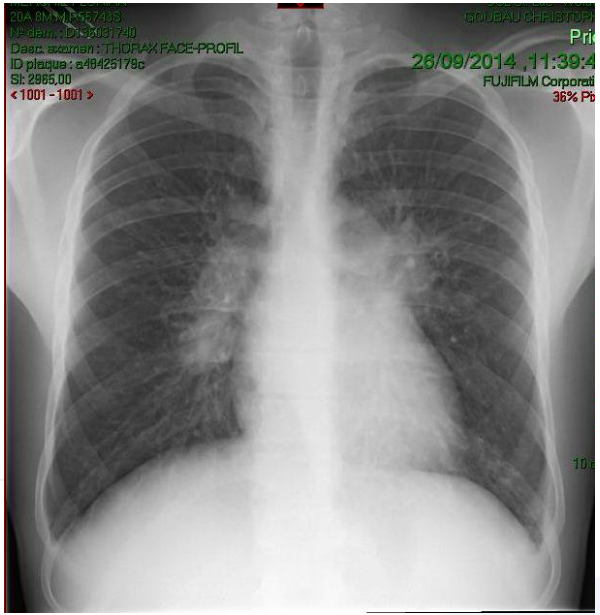


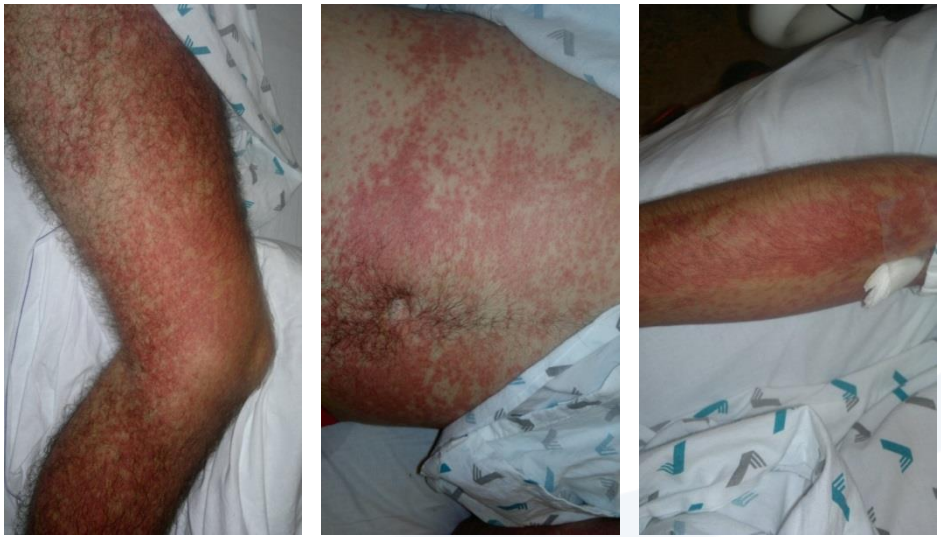
26/09/2014

- Spirometry:
  - FEV1: 2,82L, 79% predicted (previous 92%)
  - FVC: 4,5L, 108% predicted (previous 128%)

Evolution Day 2

- Hospitalization with ceftazidim/tobramycin + home treatment except AB.

- BUT…



Shift to meronem

Evolution day 4-5

- High fever
- Dyspnea, needs for supplemental oxygen
- Blood gaz: pH: 7,54; pCO2: 30 mmHg; pO2: 71 mmHg; HCO3: 26 mmol/L; Lactate: 3,7 mmol/L
- Multiple organ failure
  - Coagulopathy, renal failure, cytolysis and cholestasis, thrombocytopathy, polyserositis

We stopped all medications except ceftazidim

Evolution from day 6 to 13

- Decrease of dyspnea, adenopathies, rash
- Normalization of the blood tests

  - Go back home
  - Take usual his treatment

Back 2 days later…

- With a similar (milder) picture but with high eosinophilia (2223/µl)!

- We stopped rifampicin et moxifloxacin and symptoms resolved themselves in 24hrs.

Drug Rash, Eosinophilia and Systemic Symptoms (DRESS) syndrome

(probably with rifampicin)

DRESS syndrome

- Rare, potentially life-threatening, adverse drug reaction with cutaneous manifestations and internal organ involvement, usually within 2 months of a new drug.
- Physiopathology:
  - Drug detoxification enzyme abnormalities,
  - Reactivation of Herpes and EBV viruses,
  - Genetic predisposition.
- Symptoms:
  - Erythematous morbilliform rash,
  - Organs manifestations (hepatic - lymphatic – hematologic - neurologic – gastrointestinal – endocrine – pulmonary – cardiac)
- Management:
  - Patch test and lymphocyte transformation test
  - Skin biopsy
- Treatment:
  - **Withdrawal of the drug**
  - H1-antihistamines
  - Topic steroids
  - No AB or NSAID
  - If severe: Corticosteroids 1mg/kg/d, 3-6 months
  - If life-threatening: add IV IG

Husain *et al.* 2013. J Am Acad Dermatol
Romano *et al*. 2011. JACI
Philips *et al*. 2011. JACI