"Life-threatening systemic symptoms in a cystic fibrosis patient"

Gohy, Sophie ; Froidure, Antoine ; Lebecque, Patrick

ABSTRACT

Case report A 20-year-old male patient with cystic fibrosis (CF) (F508del/F508del, last FEV1: 92% predicted) was admitted for the sudden occurrence of high-grade fever, FEV1 drop (72% predicted) without increased cough, malaise, diffuse pain and generalized lymphadenopathies. Initial check-up revealed the presence of very recent bilateral hilar lymphadenopathies along with a marked inflammatory syndrome (WBC: 35,680/µl, neutrophils: 28,220/µl, CRP: 163mg/L). Within 24 hours, the patient developed a whole body non-pruritic purpuric rash. Despite broad-spectrum IV antibiotherapy (Tobramycin and Ceftazidim), daily fever persisted (up to 40°C). On the 6th day, the clinical picture worsened and became life-threatening with evidence of impaired renal function (glomerulopathy), multiple serositis, need for supplemental oxygen, hepatic cytolysis and coagulation abnormalities. Both C3 and C4 levels were drastically low. At this point, all non-essential drugs were stopped with the exception...

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Systemic symptoms can be life-threatening in cystic fibrosis

Sophie Gohy, Antoine Froidure & Patrick Lebecque
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:

<table>
<thead>
<tr>
<th>Test</th>
<th>Value</th>
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<tbody>
<tr>
<td>CRP (mg/dl)</td>
<td>163</td>
</tr>
<tr>
<td>WBC (/µl)</td>
<td>35680</td>
</tr>
<tr>
<td>Neutrophils (/µl)</td>
<td>28200</td>
</tr>
<tr>
<td>Eosinophils (/µl)</td>
<td>700</td>
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<tr>
<td>IgG (g/L)</td>
<td>NA</td>
</tr>
<tr>
<td>Platelets (/µl)</td>
<td>227000</td>
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<tr>
<td>IgE (kUI/L)</td>
<td>5760</td>
</tr>
<tr>
<td>Creatinin (mg/dl)</td>
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</tr>
<tr>
<td>GOT/GPT/LDH (UI/L)</td>
<td>28/17/NA</td>
</tr>
<tr>
<td>INR</td>
<td>NA</td>
</tr>
</tbody>
</table>
First assessment

- Chest X-ray

- 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, thrombocytopenia, 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,

• 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