FEMALE ADOLESCENT WITH PLEURAL EFFUSION, SHOCK, AND CYTOPENIAS

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ADMISSION

- 12-year old female adolescent
- 2-month history of intermittent fever, wrists and knees arthralgia, and malaise
- Admission in the ICU with bilateral pleural effusion, respiratory failure and uncompensated shock
- Baseline exams: leukopenia 2.800/mm³, anemia (hemoglobin 8g/dL), thrombocytopenia 90.000/mm³, high C-reactive protein 20 mg/dL (< 0.5mg/dL) and ESR 80mm/h
Despite intensive support + broad spectrum antibiotics
→ refractory shock, renal failure (creat 2.7mg/dL),
hepatic dysfunction, worsening thrombocytopenia
(60.000/mm³), bleeding at puncture sites

↓ ESR 6mm/h, AST 120U/L, ↓ fibrinogen 150mg/dL,
↑ triglycerides 350 mg/dL, ↑ ferritin 1.500mg/dL
↓

MACROPHAGE ACTIVATION SYNDROME

Methylprednisolone pulse therapy and IV cyclosporin
OTHER EXAMS

• Low C3, 24h urine protein 1.5g

• Positive direct Coombs test, ANA, anti-dsDNA

• Negative: Anti-Sm, lupus anticoagulant, anticardiolipin IgM/IgG, anti-β2 glycoprotein 1, ANCA

• Diagnosis of cSLE: serositis, anemia, leukopenia, thrombocytopenia, low complement, nephritis, ANA, anti-dsDNA
IMPROVING! (BUT...)  

- High BP=140x100mmHg (slightly elevated MAP=113)
  
  – normal echocardiogram and fundus

- Sudden onset of confusion, headache, reduced visual acuity, nystagmus, conjugate gaze deviation
CONTRAST AXIAL COMPUTED TOMOGRAPHY

ill-defined, hypodense lesions at parietal-occipital white matter
LATER...

- Pressure control, anticonvulsants, steroids, CSA → cyclophosphamide - clinical recovery in 7 days
- Renal biopsy – class IV nephritis
PRES SYNDROME

- Clinical-radiological condition characterized by seizures (75%), mental status changes, headache, visual abnormalities, and focal neurological signs.
- Multifactorial pathogenesis: breakdown of cerebral autoregulation and endothelial dysfunction $\Rightarrow$ vasogenic edema.
Most frequent associated conditions

- Infection, sepsis, shock
- Immunosuppressants (Cyclosporin)
- Autoimmune diseases
- New SLE diagnosis, high disease activity, nephritis, immunosuppressants
- Hypoalbuminemia (<2g/dL), thrombocytopenia (≤30.000/mm³), SLEDAI > 18 ⇒ poor prognosis
QUESTIONS

• PRES: a consequence of active SLE or its treatment?
  – neuropsychiatric SLE manifestation?
• What clinical and laboratory parameters would better discriminate macrophage activation syndrome from SLE activity?
• Treatment of MAS in the context of SLE?