Rede
Hospitalar Federal
no Rio de Janeiro

FEMALE ADOLESCENT WITH PLEURAL EFFUSION, SHOCK, AND CYTOPENIAS

ADRIANA RODRIGUES FONSECA

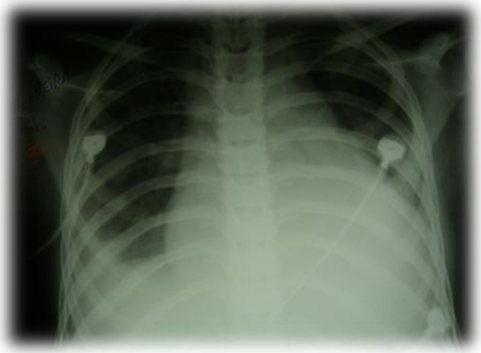
Pediatric Rheumatology Unit

Instituto de Puericultura e Pediatria Martagão Gesteira

Universidade Federal do Rio de Janeiro

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/mm^3, anemia (hemoglobin 8g/dL), thrombocytopenia 90.000/mm^3, high C-reactive protein 20 mg/dL (< 0.5mg/dL) and ESR 80mm/h



EVOLUTION

- Despite intensive support + broad spectrum antibiotics → **refractory shock**, **renal failure (**creat 2.7mg**/**dL**)**, **hepatic dysfunction**, worsening **thrombocytopenia** (60.000/mm^3), **bleeding** at puncture sites
- **↓ ESR 6mm/h, AST 120U/L, ↓ fibrinogen 150mg/dL, ↑ triglycerides 350 mg/dL, ↑ ferritin 1.500mg/dL**

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MACROPHAGE ACTIVATION SYNDROME

Methylprednisolone pulse therapy and IV cyclosporin

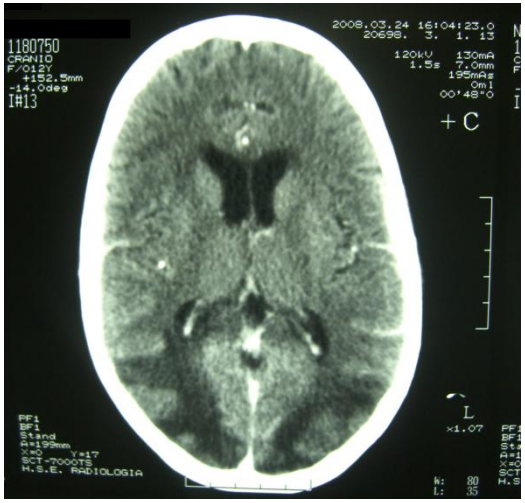
OTHER EXAMS

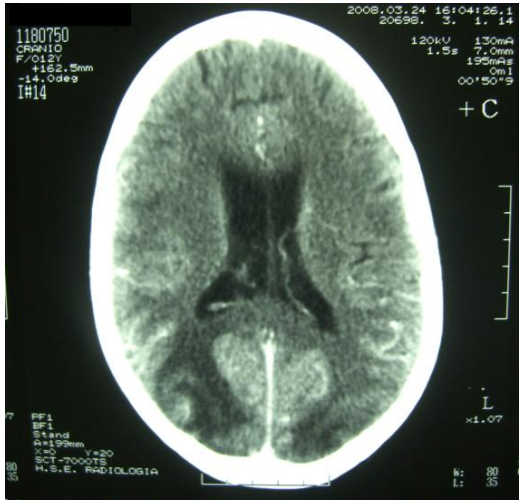
- Low C3, 24h urine protein 1.5g
- Positive direct Coombs test, ANA, anti-dsDNA
- Negative: Anti-Sm, lupus anticoagulant, anticardiolipin IgM/IgG, anti-β2 glycoprotein 1, ANCA
- Diagnosis of cSLE: serositis, anemia, leukopenia, thrombocytopenia, low complement, nephritis, ANA, anti-dsDNA

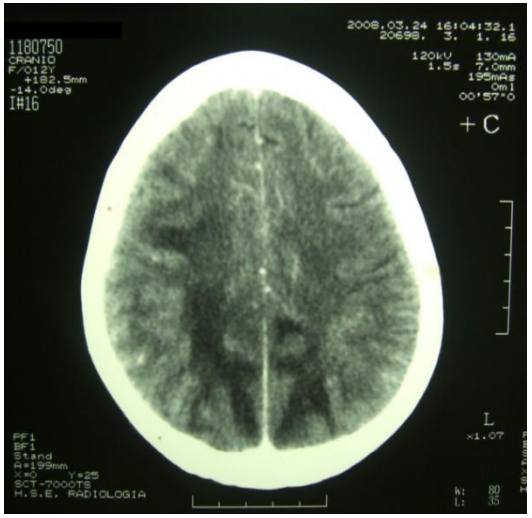
IMPROVING! (BUT…)

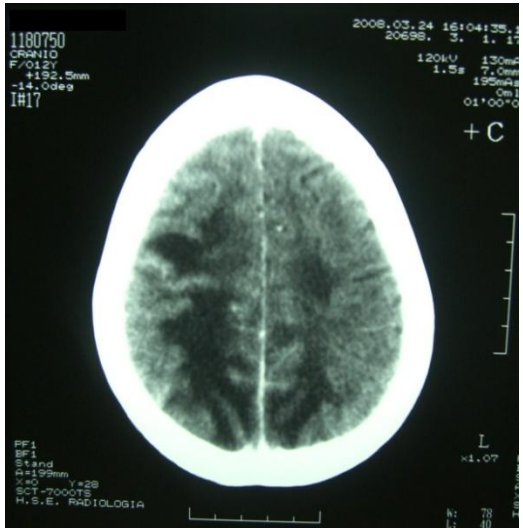
- High BP=140x100mmHg (slightly elevated MAP=113)
  - normal echocardiogram and fundus
- Sudden onset of confusion, headache, reduced visual acuity, nystagmus, conjugate gaze deviation

CONTRAST AXIAL COMPUTED TOMOGRAPHY





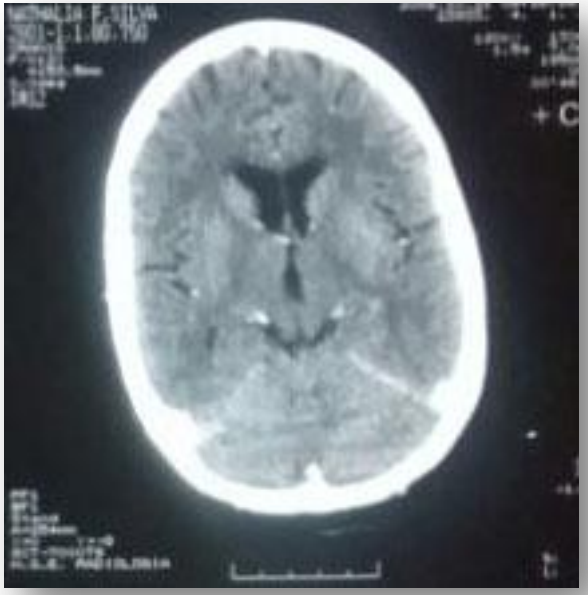


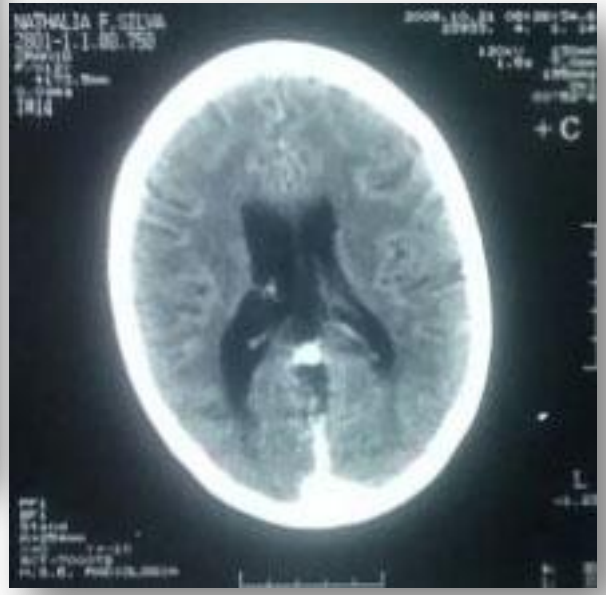


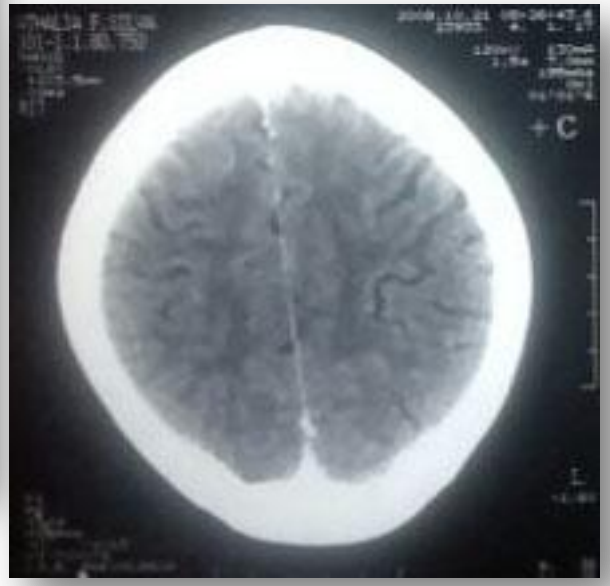
ill-defined, hypodense lesions at parietal-occipital white matter

PRES

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**
- **Immunosupressants (Cyclosporin)**
- **Autoimmune diseases**
- New SLE diagnosis, high disease activity, nephritis, immunosuppressants
- Hypoalbuminemia (<2g/dL), thrombocytopenia ($\leq$ 30.000/mm^3), SLEDAI > 18 $\Rightarrow$ 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?