



# Systemic Lupus Erythematosus with severe neuropsychiatric manifestations in an adolescent

Case Presentation

PRES-LA Basic Pediatric Rheumatology Course 2015

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Identification: ACAN, female, 12 years.

History:

- Patient presented with the diagnosis of SLE: hemolytic anemia, arthritis, pericarditis, nephritis, ANA, AntiDNA, lupus anticoagulant.
- Admitted to undergo pulse therapy with methylprednisolone. At this stage she presented with low blood pressure showing myocarditis at echocardiogram.
- Four days later she presented with lowered consciousness levels, diffuse muscle spasms, conjugate gaze deviation and decreased oxygen saturation. She then evolved with seizures, psychosis, and psychomotor agitation refractory to high doses of sedation.

- She was admitted at the ICU and underwent a dose of IVIG 2 g/kg.
- Treatment with cyclophosphamide was initiated as soon as clinical stability was achieved.
- Progressive decrease in sedation was made, but she persisted with no interaction, no spontaneous eye opening, showing shoulder girdle and upper limbs hypertonia, and was kept intubated for airways safety.

Brain contrasted CT: Normal

Brain and spinal cord angio MRI: Normal

- After two cycles of pulse therapy with methylprednisolone and cyclophosphamide (500mg/m<sup>2</sup>, three weeks apart) she maintained neurological symptoms and severe hemolytic anemia, but showed partial improvement in renal function.

|                                     | 23/02     | 29/03     |
|-------------------------------------|-----------|-----------|
| Urea (mg/dL)                        | 72        | 49        |
| Creatinine (mg/dL)                  | 1,3       | 0,8       |
| Hemoglobin (g/dL)                   | 7         | 5,8       |
| Hematocrit (%)                      | 20,8      | 17,6      |
| Leuko/Lympho (cel/mm <sup>3</sup> ) | 6.200/868 | 6.800/204 |
| Platelets (/mm <sup>3</sup> )       | 35.000    | 190.000   |
| Urinary protein/creatinine ratio    | 1,5       | -         |

- Rituximab (750mg/m<sup>2</sup>; max 1g) was recommended.
- After one week: normal renal function and partial improvement of neurological symptoms - she started making interactions, answering to demands and frequency of seizures was decreased.

| Immunoglobulin | Before Rituximab | After Rituximab |
|----------------|------------------|-----------------|
| A              | 40 mg/dl (N)     | 20 mg/dl (L)    |
| G              | 807 mg/dl (N)    | 325 mg/dl (L)   |
| M              | 27 mg/dl (N)     | 5,46 mg/dl (L)  |
| E              | 115 KU/L (N)     | 32,1 UI/ml (L)  |
| CD 20          | not measured     | 0,1%            |

Two weeks later, she underwent another cycle of pulse therapy with rituximab, cyclophosphamide and methylprednisolone. She was discharged three weeks after that when she showed complete improvement of the neuropsychiatric symptoms, no hemolytic anemia and normal function at the echocardiogram.

|                                     | 23/02     | 29/03     | 08/06       |
|-------------------------------------|-----------|-----------|-------------|
| Urea (mg/dL)                        | 72        | 49        | 35          |
| Creatinine (mg/dL)                  | 1,3       | 0,8       | 0,3         |
| Hemoglobin (g/dL)                   | 7         | 5,8       | 12,6        |
| Hematocrit (%)                      | 20,8      | 17,6      | 38,6        |
| Leuko/Lympho (cel/mm <sup>3</sup> ) | 6.200/868 | 6.800/204 | 15.000/3000 |
| Platelets (/mm <sup>3</sup> )       | 35.000    | 190.000   | 384.000     |
| Urinary protein/creatinine ratio    | 1,5       | -         | 0,01        |

# Questions for the audience



- What is your experience with neuropsychiatric symptoms in SLE?
- What is your experience in using rituximab for the treatment of SLE?
- At which time should it be introduced?
- Role of therapeutic plasma exchange?
- Should have brain auto-antibodies be searched in this case?