

## Medical History



26yo

- Caucasian
- No relevant past medical history
- **2-month history of fever (max. 39°C), malaise, weight loss and inflammatory arthralgia of knees, wrists and hands**

## Brought to the Emergency room



- Normotensive, sinus tachycardia 150bpm, temperature 34.°C, oxygen saturation 94% ambient air
- **Glasgow Coma Scale 8 points** (Eyes 2, Verbal 2, Motor 4)
- **Pale, icteric, purpuric lesions** on the lower limbs
- Cardiac auscultation unremarkable (except for tachycardia)
- Pulmonary auscultation: **rhonchi and dispersed “dry” rales**
- **Hepatomegaly** 3cm below costal margin
- **Palpable axillary lymph nodes ~2cm**
- No signs of arthritis

## Workup

Hemoglobin	6.1 g/dL
Platelets	36 000 /uL
Leucocytes	11 600 /uL
Neutrophils	10 790 /uL
Lymphocytes	390 /uL
Sedimentation Rate	54 mm/h
Haptoglobin	<10 mg/dL
Peripheral blood smear	No schistocytes
Prothrombin	54 %
aPTT	39,3 s
D-dímer	4,79 ug/mL
Fibrinogen	56 mg/dL

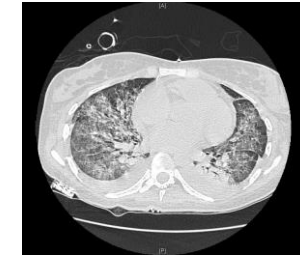
AST	2 207 U/L
ALT	234 U/L
Bilirubin T/D	9.3 / 6.84 mg/dL

LDH	1 569 U/L
Triglycerides	346 mg/dL
Ferritin	11 719 ng/dL

Creatinine	2 mg/dL
Urinalysis	Unremarkable

Arterial gasometry: pH 7.27; PaO2 62mmHg; PaCO2 17.8mmHg; HCO3- 8.1 mM; Lactate 9.6 mM

C3	24 mg/dL
C4	8.1 mg/dL
ANA	1/2560
Anti-dsDNA	263 UI/mL
Anti-Smith	Positive



CT scan of the chest showing diffuse alveolar hemorrhage on axial plane

- Computed tomography: signs of Diffuse Alveolar hemorrhage; **hepatomegaly; multiple adenopathies**
- Bronchofibroscopy: **Bronchoalveolar lavage showing Alveolar Hemorrhage**
- Axillary lymph node biopsy: **necrotizing lymphadenitis**

- Bone marrow aspirate: unremarkable (hemophagocytosis not seen)
- PCR to EBV DNA: negative
- RT-PCR to EBV DNA: negative
- **CD25 / soluble IL-2 receptor alpha: 11 533 pg/mL**
- (Reference values 458-1997)

## Diagnosis

### Systemic Lupus Erythematosus with Macrophage Activation Syndrome (MAS) and Diffuse Alveolar Hemorrhage (DAH)

#### Treatment

- Plasmapheresis - 5 sessions
- Methylprednisolone 1g/day for 3 days
  - Prednisolone 1mg/Kg/day
- Cyclophosphamide 500mg
- Hydroxychloroquine 5mg/Kg/day

#### Outcomes

- Resolution of the pulmonary hemorrhage and MAS markers, allowing for extubation
- Complete remission without signs of disease activity after 5 months

#### KEY POINTS

- ✓ Plasmapheresis's optimum role isn't established in the setting of MAS, as of 2019 ASFA guidelines, but has been considered an acceptable second-line therapy for the treatment of severe complications of SLE;
- ✓ A decision to proceed with plasmapheresis was primarily made on the setting of Diffuse Alveolar Hemorrhage, with positive end results;