Case Presentation
PReS – Latin America

Catherine Gusman Anelli M.D.
Pediatric Rheumatologist from UNIFESP
Case Presentation

- OGS, 11y male
- Diagnosed with **JDM** 6 mo prior to reference to our service
- One year of the beginning of the symptoms

**Features**

1. Symmetrical proximal muscle weakness
2. Muscle biopsy evidence of myositis
3. Elevation in serum skeletal muscle enzymes
4. Characteristic electromyogram pattern of myositis
5. Typical rash of dermatomyositis

Bohan and Peter

- **CMAS** 12/52 – seriously ill
- Skin ulcers on the buttocks
Case Presentation

- Fever and cough with posterior oropharyngeal discharge

- Also:
  - ANA: positive (unspecific pattern)
  - Anti-Jo1: negative
  - Capillaroscopy with SD pattern
Skin Lesions

All images used in this presentation were authorized by the patient and responsible
Skin Lesions
Supplementary Information

- No esophageal impairment
- HR torax CT without lung involvement
- Echocardiogram: normal
- Abdominal USG: normal
- No calcinosis at all
- Normal immunoglobulin levels
Treatment

- Due to concomitant infection he was started on IVIG and metilprednisolone pulse after 48 hours of antibiotic therapy

- Resolution of skin ulcers and a better CMAS

- After the discharge: metothrexate, prednisone + skin care
  - sunscreen
  - moisturizing creams
  - anti-histaminic drugs
Follow-up

- He got a lot better with regard to the muscle (CMAS 45) and all the labs turned into absolutely normal.

- The skin was always a concern.

- In one year:
  - IVIG
  - Prednisone / metilprenisolone
  - Metothrexate
  - Hydroxicloroquine
Videocapillaroscopy

Still showing SD pattern with no improvement
Treatment

• Rituximab

• Talidomide – some reports showed improvement in vasculopathy lesions (until we got approval for Rituximab)
Skin lesions after 2\textsuperscript{nd} Rituximab dose
Skin Lesions after 4th Rituximab dose
Any questions?

1. How to measure the skin improvement?
   – Physician and patient VAS
   – CDASi

2. How to differentiate cronic findings from active lesions?

3. Since we are using Rituximab every 6 months – and he’s going soon for the 5th dose – for how long should we use this treatment?

4. What other procedures or treatments do you suggest?
Thank you

“Everybody has to start somewhere. You have your whole future ahead of you”.
Haruki Murakami