

# 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



# Suplemmentary Information

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

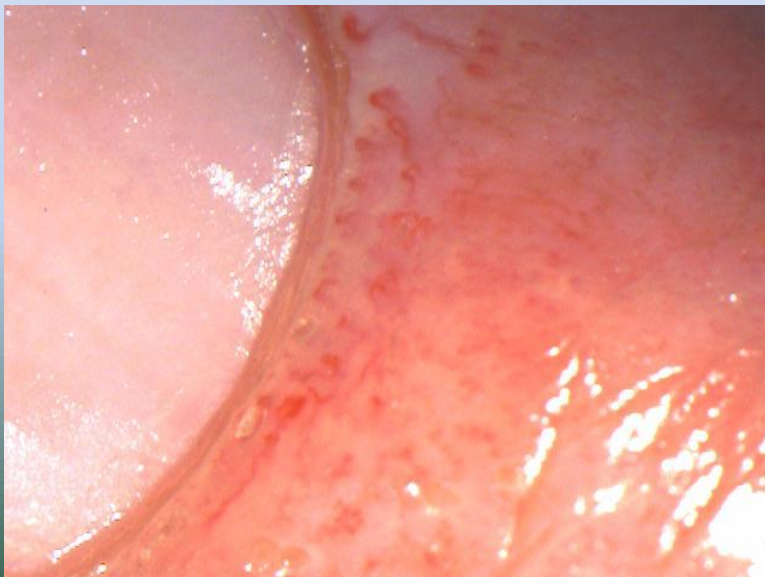
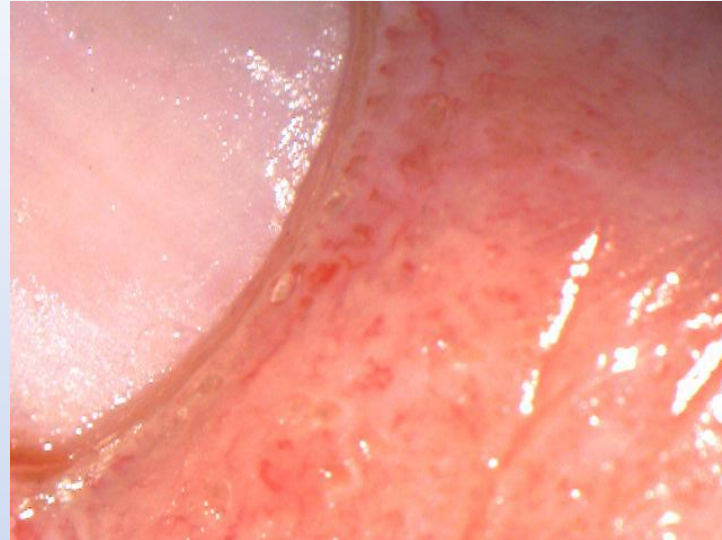
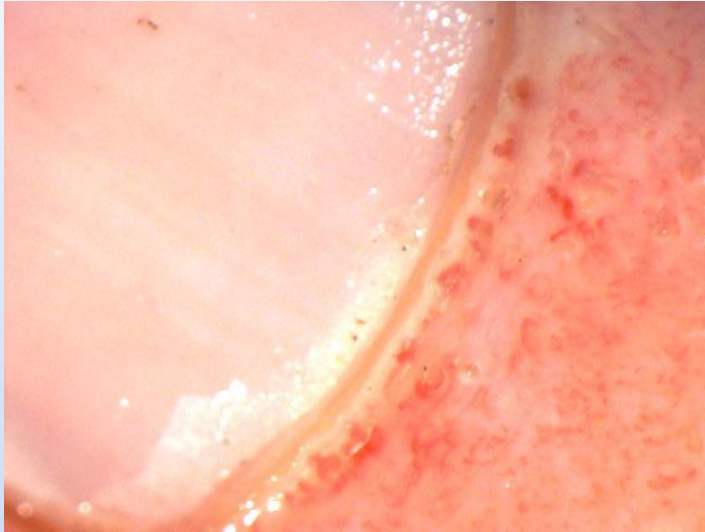
# Treatment

- 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
  - Methotrexate
  - 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<sup>nd</sup> Rituximab dose



# Skin Lesions after 4<sup>th</sup> 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 5<sup>th</sup> 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