



POSTER PRESENTATION

Open Access

PReS-FINAL-2008: Juvenile dermatomyositis: a report of 22 cases

A Souto^{1*}, N Alvarez², A Barros¹, L Fernandez¹, B Lopez³, I Calvo⁴

From 20th Pediatric Rheumatology European Society (PReS) Congress Ljubljana, Slovenia. 25-29 September 2013

Introduction

Juvenile dermatomyositis is a chronic inflammatory muscular disease that affects every year 1.5-3 children per million and also produces systemic, skin, joint, digestive and respiratory manifestations.

Objectives

To describe the main epidemiological, clinical and analytical aspects of Juvenile dermatomyositis diagnosed in La Fe Hospital (Valencia, Spain).

Methods

Patients diagnosed of Juvenile dermatomyositis in La Fe Hospital (Valencia, Spain) since June 1992 to March 2013. Dates are shown in median and interquartile range for non-parametric variables, and for parametric variables is given as mean and standard deviation. To assess response to treatment, we used Wilcoxon test for paired measures of non-parametric variables and t-student for parametric variables.

Results

We included 22 patients, 45.5% women, age at diagnosis 7.6 (3.7-10.8) years and evolution time 2.75 (1-5) months.

The most frequent initial manifestation were muscle weakness (40%) and skin alterations (40%). Also constitutional symptoms (15%) and myalgia (5%). They presented muscle weakness (100%), heliotrope rash (100%), Gottron papules (90.9%), calcinosis (27.3%), other skin lesions such as telangiectasias, oral ulcers, livedo reticularis and purpura (55.5%), mialgia (55.5%), arthralgia (55.5%), arthritis (50%), constitutional symptoms (50%), esophageal involvement (31.8%), gastrointestinal (28.6%), Raynaud (18.2%),

fever (27.3%), anasarca (18.2%), dysphonia (18.2%), and lung disease (18.2%).

Patients were treated with corticosteroids, immunoglobulins and immunosuppressants such as methotrexate, tacrolimus, cyclosporine and antimalarial, alone or in combination, with significant improvement.

CPK at diagnosis was 1114 UI/l (280-4952), and 73 (46-106) after 3 months, with significant improvement ($p = 0.0010$). Aldolase at diagnosis 15.45 U/L (12-37) and 6 (4-10) after 3 months ($p = 0.0148$). GOT at diagnosis 149 U/L (73-180) and 28 (25-38) after a year ($p = 0.0005$). GPT at diagnosis 102 U/L (44-187) and 23 (17-36) after a year ($p = 0.0003$). ESR at diagnosis 18 mm (16-36.5) and 13 (7-18) after a year ($p = 0.0139$). Strength at diagnosis was 2.5 ± 1 and 4.25 ± 0.87 after 3 months ($p < 0.00001$).

Capillaroscopy was realised in most of patients at diagnosis and showed pathological capillary patterns (decrease in the number of capillaries, enlargement, megacapillaries, and bleeding), with improvement in subsequent assessments.

Conclusion

Dermatomyositis is a rare disease of childhood. Children we presented had muscle weakness and heliotrope erythema (100%) and in varying proportions Gottron's papules, calcinosis, Raynaud, artromialgias and general symptoms. We propose a comprehensive patient monitoring, assessing muscle strength and laboratory findings as CPK, aldolase, GOT, GPT and ESR but also by capillaroscopy. Capillaroscopy may be indicative of dermatomyositis outbreak, has diagnosis value and may observe an early improvement pattern when patients are in remission.

Disclosure of interest

None declared.

¹Hospital Clinico Santiago Compostela, Santiago de Compostela, Spain
Full list of author information is available at the end of the article

Authors' details

¹Hospital Clinico Santiago Compostela, Santiago de Compostela, Spain.
²Hospital Universitario A Coruña, A Coruña, Spain. ³Hospital La Fe Valencia, Valencia, Spain. ⁴Hospital La Fe Valencia, Santiago de Compostela, Spain.

Published: 5 December 2013

doi:10.1186/1546-0096-11-S2-P21

Cite this article as: Souto *et al.*: PReS-FINAL-2008: Juvenile dermatomyositis: a report of 22 cases. *Pediatric Rheumatology* 2013 11(Suppl 2):P21.

**Submit your next manuscript to BioMed Central
and take full advantage of:**

- Convenient online submission
- Thorough peer review
- No space constraints or color figure charges
- Immediate publication on acceptance
- Inclusion in PubMed, CAS, Scopus and Google Scholar
- Research which is freely available for redistribution

Submit your manuscript at
www.biomedcentral.com/submit

