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PEDIATRIC RHEUMATOLOGY

POSTER PRESENTATION Open Access

Subclinical pulmonary abnormalities in juvenile dermatomyositis

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Introduction

Pulmonary involvement in juvenile dermatomyositis (JDM) is frequent and associated with poor outcome. However, a systematic assessment of pulmonary function and high-resolution computed tomography (HRCT) was rarely reported in this population.

Objectives

To assess pulmonary function and HRCT in JDM patients and to evaluate possible associations between pulmonary abnormalities and disease activity, cumulative damage and health-related quality of life (HRQL) scores.

Methods

A cross-sectional study was performed in 20 JDM patients. Pulmonary function test included spirometry, body plethysmography and diffusion capacity for carbon monoxide (DLCO). They were also carried out six-minute walk test (6MWT) and HRCT scan. Disease activity score (DAS), childhood myositis assessment scale (CMAS), myositis damage index (MDI) and HRQL (Pediatric Quality of Life Inventory - PedsQL) data were also assessed

Results

The mean age was 11.6 years (6-18). Subclinical mild/moderate obstruction according to American Thoracic Society criteria was observed in 35% and DLCO was reduced in 20% of JDM patients. Spirometric and/or DLCO abnormalities were observed in 45% of patients. In plethysmography, reduced total lung capacity (TLC) and conductance were observed in 25% and 50% of JDM patients, respectively. In contrast, increased resistance and residual volume (RV)/TLC were evidenced in 10% and 35% of JDM patients, respectively. Thirteen patients underwent HRCT and 8 had alterations: interstitial lung disease in 6 and a mixed pattern in two. A positive correlation was observed between DAS and ratio between forced expiratory volume in one second and vital capacity (VEF1/CV) (r=+0.50, p=0.003), conductance (r=+0.46, p=0.045) and HRCT score (r=+0.60, p=0.003). A positive correlation was observed between CMAS and VEF1/CV (r=+0.47, p=0.042), DLCO (r=+0.67, p=0,002) and 6MWT (r=+0.54, p=0.048), and negative correlation between DAS and HRCT score (r=-0.63, p=0.021). Correlations were identified between MDI and conductance (r=+0.72, p=0.0004), DLCO (r=-0.46, p=0.042) and HRCT score (r=+0,81, p=0.0008); and between PedsQL and VEF1/CV (r=+0.45, p=0.046), conductance (r=-0.60, p=0.006) and HRCT score (r=+0.62, p=0.024). Correlations were also observed between HRCT score and VEF1/CV (r=-0.64, p=0.017), forced expiratory flow between 25 and 75% of vital capacity (FEF25%>75 %) (r=-0.59, p=0.035) and conductance (r=+0.78, p=0.0018).

Conclusion

Subclinical pulmonary abnormalities were frequent in this rare idiopathic inflammatory myopathy. Importantly, these findings may be related to disease severity and activity, and may influence HRQL of these patients.

Disclosure of interest

None declared.

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