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Predictors of Response to Methotrexate in Patients with Eosinophilic Fasciitis (Shulman's Disease)

Walter A. Sifuentes-Giraldo[1], Dolors Grados Canovas[2], Marina De Los Riscos Alvarez[3], María Pascual Pastor[4], Pablo Moreno Fresneda[5], Estibaliz Loza[6], María J. García Yébenes[6], Alejandro Olivé[2], Patricia Carreira Delgado[3], Francisco J. Narvaez Garcia[4], Rosario García-Vicuña[7] and Antonio Zea Mendoza[1], [1]Rheumatology, Hospital Universitario Ramón y Cajal, Madrid, Spain, [2]Rheumatology, Hospital Universitario Germans Trias i Pujol, Barcelona, Spain, [3]Rheumatology, Hospital Universitario 12 de Octubre, Madrid, Spain, [4]Rheumatology, Hospital Universitario de Bellvitge, Barcelona, Spain, [5]Rheumatology, Rheumatology Service, Hospital Universitario de La Princesa, IIS-IP, Madrid, Spain, [6]Instituto de Salud Musculoesquelética (InMusc), Madrid, Spain, [7]Rheumatology, Hospital Universitario de La Princesa. IIS La Princesa, Madrid, Spain

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SESSION INFORMATION

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Session Title: Systemic Sclerosis, Fibrosing Syndromes, and Raynaud's – Clinical Aspects and Therapeutics - Poster II

Session Time: 9:00AM-11:00AM

Background/Purpose: Eosinophilic fasciitis (EF) is a rare scleroderma-like disorder described in 1974 by Shulman. It is characterized by the acute onset of edema and induration of the skin and the subcutaneous tissue associated with peripheral blood eosinophilia. Deep skin biopsy shows characteristic alterations in the muscle fascia. There is no consensus regarding the treatment of the EF. Oral or intravenous glucocorticoids (GC) are usually the initial treatment with significant improvement in most cases. Immunosuppressive drugs may be associated when the response is insufficient and the most used is methotrexate (MTX). The response to MTX is usually favorable, especially in cases with concomitant morphea lesions[1]. In this study we investigated which demographic, clinical and laboratory baseline features are associated with remission during MTX treatment in patients with EF.

Methods: We performed an observational, retrospective (1983–2014) and multicentric study of patients with EF from 5 Spanish university hospitals. Inclusion criteria: 1) characteristic cutaneous manifestations; 2) deep biopsy with consistent changes in muscle fascia, and 3) treatment with MTX. Response to treatment was divided into: 1) complete remission (absence of symptoms and disappearance of lesions); 2) partial response (patients who develop limitation despite treatment); and 3) lack of response (persistence of symptoms and findings on examination)[1]. Statistical non-parametric tests were used for the data analysis, Kruskal Wallis for continuous variables and χ^2 for categorical variables.

Results: 33 patients were included, 18 women (54.5%), with a mean age of 54.6 years (range 22–81) and median duration of disease until diagnosis of 4 months (range: 1–25). Most of these patients (97%) had previously been treated with GC with insufficient response, 8 (24.2%) had previously received other immunomodulatory drugs (hydroxychloroquine, azathioprine, cyclosporine) and 5

(15.2%) photochemotherapy (PUVA). MTX median dose was 15 mg/week (range: 10–25); 16 cases (48.5%) achieved complete remission, 15 (45.5%) partial response and 2 (6%) lack of response. Patients who achieved complete remission had a mean age at diagnosis slightly higher (64), presented more frequently induration ≥ 50% of body surface, myalgia and associated malignancies but C-reactive protein (CRP) levels were lower. Of all the variables analyzed, only low CRP level was significantly associated with complete remission (P=0.004). Two patients in remission relapsed after discontinuation of MTX, with a favourable response to re-treatment with GC.

Conclusion: The only variable that seems to be associated with remission during treatment with MTX in our series is the absence of elevated CRP. All other variables showed no significant differences, although the statistical power may be small due to the limited sample size.

	Complete remission (N=16)	Partial response (N=15)	Lack of response (n=2)	P value
Age at diagnosis	64 (40-73)	49,5 (42-57)	54 (42-69)	0.640
Female gender	8 (44%)	8 (44%)	2 (11%)	0.405
Smoking	3 (25%)	8 (67%)	1 (8.3%)	0.204
Time to diagnosis (months)	4,0 (2-7)	4,0 (2.5-7.5)	6,5 (3-10)	0.867
Induration ≥ 50%	10 (56%)	7 (39%)	1 (5%)	0.437
Erythema	4 (40%)	5(50%) 1	1 (10%)	0.79
Pruritus	3 (50%)	2 (33%)	1 (17%)	0.419
Edema of extremities	8 (35%)	13 (56%)	2 (9%)	0.151
“Orange peel”	3 (37%)	5 (62%)	–	0.545
Hyperpigmentation	4 (40%)	5 (50%)	1 (10%)	0.791
Other morphea lesions	6 (50%)	5 (42%)	1 (8%)	0.808
Arthritis	3 (43%)	4 (57%)	–	0.709
Joint contractures	3 (43%)	4 (57%)	–	0.709
Muscle weakness	6 (75%)	2 (25%)	–	0.144
Myalgia	10 (53%)	8 (42%)	1 (5%)	0.628
Carpal tunnel syndrome	3 (50%)	3 (50%)	–	–
Raised ESR	8 (50%)	7 (44%)	1 (6%)	0.926
Raised CRP	7 (39%)	11 (61%)	–	0.004*
Eosinophilia	10 (42%)	13 (54%)	1 (4.2%)	0.500
Hipergammaglolinemia	5 (45%)	6 (54%)	–	0.570
ANA	4 (36%)	6 (54%)	1 (9%)	0.714
Glucocorticoids	14 (44%)	16 (50%)	2 (6%)	0.539
Other previous treatments	3 (37%)	4 (50%)	1 (12%)	0.646

PUVA	3 (60%)	1 (20%)	1 (20%)	0.207
Neoplasm	4 (80%)	1 (20%)	–	0.236

References: 1. Lebeaux D, et al. Rheumatology (Oxford). 2012;51:557-61

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