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AB1029

MYOCARDIAL DYSFUNCTION DETECTED BY USING SPECKLE TRACKING ECHOCARDIOGRAPHY IN FAMILIAL MEDITERRANEAN FEVER PATIENTS; IS THERE ANY DIFFERENCE REGARDING RESISTANT DISEASE?

U. Ozkerim¹, S. Guçenmez², S. V. Emren³, E. Ozdemir³, Ö. Gerçik⁴, D. Solmaz⁴, S. Akar⁴. ¹Izmir Katip Celebi University, Faculty of Medicine, Department of Internal Medicine, Izmir, Turkey; ²Izmir Katip Celebi University, Ataturk Education and Research Hospital, Department of Rheumatology, Izmir, Turkey; ³Izmir Katip Celebi University, Faculty of Medicine, Department of Cardiology, Izmir, Turkey; ⁴Izmir Katip Celebi University, Faculty of Medicine, Department of Internal Medicine, Division of Rheumatology, Izmir, Turkey

Background: Familial Mediterranean fever (FMF) is an auto-inflammatory disease commonly affects people from Mediterranean basin. It is characterized by acute self-limiting inflammatory attacks of serous membrane. Some recent studies showed diastolic dysfunction in patients with FMF however systolic function was rarely evaluated before.

Objectives: To assess cardiac functions by using speckle tracking echocardiography (STE) in addition to routine measurements and to evaluate whether there is any difference between colchicine responsive patients and those with colchicine-resistant or severe disease.

Methods: Seventy-four FMF patients (57 responsive [60% female and median age 35 [18-63] years], 17 resistant [53% female and median age 31 [20-49] years]) and 74 healthy controls ([53% female and median age 37 [22-55] years]) were included in the study. Patients with cardiac disease or risk factors affecting STE were excluded. Patients who had ≥ 1 attacks in three months despite ≥ 2 mg/day of colchicine or who were treated with IL-1 blocking agents were defined as colchicine resistant disease and who had amyloidosis (whom had proteinuria ≥ 500 mg/day in at least 2 occasion or biopsy-proven amyloidosis) defined as severe disease. Demographic and disease related characteristics were obtained by using a structured form. A detailed echocardiographic examination including the M-mode, Doppler, and STE was applied to whole study population.

Results: There was no significant difference between groups in terms of age and sex. Disease duration was not different between colchicine responsive and resistant patients (median duration in resistant disease was 11 years (1-27), in responsive patients 6 years (0-33) and, p: 0.133). Although ejection fraction was similar among groups, global longitudinal peak systolic strain; a marker of systolic function, was significantly lower in FMF patients in comparison with healthy subjects and this difference was due to colchicine resistant FMF patients (p<0.05). There was no significant difference in echocardiographic parameters between non-resistive and resistive FMF patients (p>0.05) (Table).

Table. Comparison of standard and speckle tracking echocardiographic findings among groups

	Responsive FMF (n: 57)	Resistant FMF (n: 17)	Healthy Controls (n: 74)
LA	31 (23-41)	30 (26-35)	31 (22-44)
LVD	43 (35-86)	41 (38-48)	43 (20-50)
LVS	26 (20-32)	24 (18-31)	26 (19-34)
EF	62 (40-77)	64 (46-73)	60 (52-70)
E	95 (23-148)	98 (9-130)	93 (57-130)
A	83 (26-153)	79 (8-134)	81 (57-120)
DT	187 (100-303) *	183 (100-296)	161 (100-254)
ELateral	14 (8-29)	13 (10-23)	14 (8-23)
ALateral	11 (5-18)	10 (7-18)	10 (6-17)
ESeptal	11 (6-20)	9 (5-16)	11 (5-16)
ASeptal	10 (5-17)	10 (5-17)	9 (4-23)
GLS	20 (8-55) *	20 (15-32)	22 (16-33)
GCS	21 (10-29)	20 (11-42)	20 (13-30)

*Significant difference among groups, p<0.05. LA: Left atrium, LVD: left ventricular diastolic diameter, LVS: Left ventricular systolic diameter, EF: Ejection fraction, E: Early diastolic filling A: Atrial filling, DT: Deceleration time, GLS: Global Longitudinal strain GCS: Global circumflex strain

Conclusion: This study shows that there is subclinical myocardial dysfunction in FMF patients. Those patients with severe or colchicine resistant disease has a numerical but statistically different trend toward systolic dysfunction.

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AB1030

IGG4-RELATED DISEASE: A RETROSPECTIVE CLINICAL CASE SERIES FROM A TERTIARY CARE CENTRE IN INDIA

M. Gupta¹, L. Duggal², B. Gurbir Singh², J. Patel², N. Jain², A. K. Grover². ¹Sir Ganga Ram Hospital, Rheumatology and Clinical Immunology, New Delhi, India; ²Sir Ganga Ram Hospital, New Delhi, India

Background: IgG4-related disease (IgG4-RD) is an often unrecognized, rare fibro-inflammatory condition that can involve various organ systems.

Objectives: The aim of this study was to identify the different clinical patterns of this disease in a single centre in North India.

Methods: 70 patients were diagnosed on the basis of published diagnostic criteria for IgG4-RD. Patients' presenting complaints, epidemiological profiles, laboratory, radiological and histological findings along with the treatment and outcomes were collated and analyzed from 2011 – 2019.

Results: In a total of 70 patients who were diagnosed with the disease, the ratio of female to male ratio was 1:1. The mean age of patients was 41.4 years. Involvement of orbits and peri-orbital tissues was highest (52.9%) due to large number of referrals from ophthalmology services. 13% of patients had multiple organ involvement. Patients with involvement of retroperitoneal tissues and lymph nodes were 8.5% and 5.7%, respectively. Increased serum IgG4 levels were found in 74.3% of the patients. Histopathological examinations of the affected organs were performed in 44 (62.85%) patients and a diagnosis of possible (38.57), probable (32.85%) and definite (28.57%) IgG4-RD was made. Majority of the patients (94.3%) required immunosuppressive medications along with corticosteroids. Azathioprine was the most commonly used (72.8%) immunosuppressive medication. Rituximab was used in 17.1% of the patients, of whom one had multisystem involvement.

Conclusion: This study depicts the most common patterns of organ involvement along with the epidemiological, laboratory, histological, radiological data and response to treatment, in IgG4-RD, with a definite ophthalmology referral bias, in a tertiary care centre in North India.

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Table 1. Clinical and laboratory characteristics

Characteristics	Number of patients (N=70)
Age (years)	41.4
Age group in years, n (%)	
>50	24 (34.28)
<50	46 (65.71)
Sex, n (%)	
Male	36 (51.42)
Female	34 (47.22)
ESR (mm/hr)	24.37
CRP (mg/dL)	12.31
Serum IgG (mg/dL)	1214.2
Serum IgE (IU/L)	587.2
Type of IgG4 related disease, n (%)	
Definite	20 (28.57)
Probable	23 (32.85)
Possible	27 (38.57)

Data represented as mean, unless otherwise specified.

CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; Ig, immunoglobulin.

Table 2. Distribution of patients according to organ involvement.

Organ involvement	Number (%)
Orbital and periorbital	37(52.9)
Retroperitoneum	6 (8.5)
Laryngotracheal tissue	4 (5.7)
Aorta and branches	4 (5.7)
Lymph nodes	3(4.2)
Paranasal sinus	2 (2.8)
Paravertebral tissue	2 (2.8)
Pancreas	2 (2.8)
Lung and bronchus	2 (2.8)
Eye (Scleritis)	1 (1.4)
Prevesical Mass	1 (1.4)
Ear polyp	1 (1.4)
Small Bowel	1 (1.4)