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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?

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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

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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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- Khosroshahi A, CarruthersMN, Deshpande V, Unizony S, Bloch DB, Stone JH. Rituximab for the treatment of IgG4-related disease: lessons from 10 consecutive patients. *Medicine* 2012; 91(1):57-66.

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)

Table 2. Distribution of patients according to organ involvement.

Organ involvement	Number (%)
Primary sclerosing cholangitis	1 (1.4)
Musculoskeletal	1(1.4)
Submandibular gland	1(1.4)
Central nervous system	1 (1.4)
Multisystem involvement	9 (12.86%)

Figure 1. Right eye proptosis **Figure 2.** CT abdomen showing hydronephrosis due to retroperitoneal fibrosis

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AB1031 DYSKERATOTIC CELLS IN PERSISTENT PRURITIC SKIN LESIONS AS A PROGNOSTIC FACTOR IN ADULT-ONSET STILL'S DISEASE.

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Background: Adult-onset Still's disease (AOSD), a systemic inflammatory disorder, is characterized by high fever, evanescent rash, arthritis, and hyperferritinemia. AOSD is also reported to be associated with other skin lesions, including persistent pruritic papules and plaques.

Objectives: This study aimed to assess the significance of dyskeratotic skin lesions in Japanese AOSD patients.

Methods: We retrospectively assessed the histology of persistent pruritic skin lesions and evanescent rashes and the relationship between dyskeratotic cells, serum markers, and outcomes in 20 Japanese AOSD patients, comparing AOSD histology with that of dermatomyositis (DM), drug eruptions, and graft-versus-host disease (GVHD).

Results: Persistent pruritic lesions were characterized by scattered single keratinocytes with an apoptotic appearance confined to the upper layer of the epidermis and horny layer without inflammatory infiltrate. In contrast to AOSD, the histology of DM, drug eruption, and GVHD demonstrated dyskeratotic cells in all layers of the epidermis with inflammatory infiltrate. AOSD with evanescent

rash showed no dyskeratotic cells. The dyskeratotic cells in pruritic AOSD lesions stained positive for ssDNA and terminal deoxynucleotidyl transferase-mediated dUTP nick end-labeling, indicating apoptosis. Serum IL-18 was significantly higher in AOSD patients with dyskeratotic cells than those without, and generally required higher doses of glucocorticoids, immunosuppressants, and biologic agents. Two of ten AOSD patients with dyskeratotic cells died from haemophagocytic lymphohistiocytosis.

Conclusion: Persistent pruritic AOSD skin lesions are characterized by dyskeratotic cells with apoptotic features, involving the upper layers of the epidermis. There may be a link to elevated IL-18. This dyskeratosis may be a negative prognostic indicator.

Disclosure of Interests: None declared

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AB1032 CONTRIBUTION OF BONE BIOPSY DURING REVELATORY BONE METASTASES

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Background: Bone metastases (BM) are tumor cells that originate in a primary malignant tumor and are localized remotely in bone tissue. They more or less faithfully reproduce the morphological and biological characteristics of the primary tumor. Histological analysis is essential to confirm the diagnosis of BM and to identify the primary tumor if possible and sometimes to help in the selection of treatment.

Objectives: The aim of this work is to study the contribution of bone biopsy during revealing BM in diagnostic strategy and therapeutic decision.

Methods: We retrospectively studied the files of 105 patients hospitalized in a Rheumatology department for BM revealing from January 2000 until December 2015. For each patient we collected epidemiological and anatomopathological data to arrive at the diagnosis of primary neoplasm and histological type.

Results: The patients were divided into 86 men (81.9%) and 19 women (18.1%) with a sex ratio (M / F) of 4.52. The average age of our patients was 64.91 ± 13.29 years. Pain was the most frequent reason for consultation found in 97.1%. This pain was either of bone site (61.9%) or of radicular topography (41.9%). Bone swelling or a pathological fracture revealed BM in 4.8% and 8.6% of the cases, respectively. The onset of neurological damage was noted in 13.3% of the cases.

Histologically, the bone biopsy performed in 64 patients made it possible to specify the histological type (carcinoma, adenocarcinoma) in 64% of the cases and to lead to primary cancer in 57.8%. A non-radio-guided percutaneous bone biopsy was performed in 44 patients (68.75%) including 41 osteo-medullary biopsy in iliac crest (BOM) and 3 in the sternum, a bone biopsy directed under scanner in 16 cases (25%) and a surgical bone biopsy in 4 cases.

The BOM was positive in 21 cases (51.2%) showing a poorly or moderately differentiated adenocarcinoma or carcinoma. It allowed referral to a primitive in 20 cases: a prostatic origin in 11 cases, a pulmonary origin in 5 cases, a digestive origin in 2 cases, a mammary origin in one case and a neuroblastoma in one case.

Conclusion: Thanks to improved sampling and immunohistochemistry techniques, the precise histological type and location of the primary tumor could be identified, thereby improving the quality of care for patients with increased life expectancy.

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AB1034 DEPRESSION AND ANXIETY IN FAMILIAL MEDITERRANEAN FEVER

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Background: Familial Mediterranean Fever (FMF) is a hereditary autoinflammatory disease characterized by recurrent attacks of fever, peritonitis, pleuritis, arthritis, and skin eruption (1). It is shown by studies that chronic diseases like diabetes mellitus, chronic heart disease, hypertension which other than inflammatory – rheumatologic disease increase depression and anxiety (2). There are a few studies evaluating depression and anxiety in FMF patients, and these results are conflicting (3,4).