AB1089

CLINICAL FEATURES OF FAMILIAL MEDITERRANEAN FEVER PATIENTS IN NORTH-WESTERN PART OF TURKEY: ANALYSIS OF 139 PATIENTS

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Background: Familial Mediterranean fever (FMF) is known the most common monogenic autoinflammatory disease. Its prevalence is reported high from the eastern Mediterranean areas (1) The disease is characterized by episodes of fever, serositis, arthritis, renal complications and other different clinical manifestations (2)

Objectives: Here, we aimed to present our data of our 139 FMF patients for demonstrating the demographic and clinical features of the study group, from North-western part of Turkey.

Methods: A total of 139 FMF patients who were diagnosed and treated in the department of Internal medicine/Rheumatology, Sakarya University (North-western area of Turkey) were included in the study and the demographic and clinical characteristics of the patients were examined.

Results: The mean age of the patients was 39.02 ± 11.3. Male gender was 42 (30.2%) and female gender was 97 (69.8%). 107 (77%) of patients had fever and 32 (23%) had no history of fever. 127 (91.4%) patients complained about peritonitis, 27 (19.4%) patients had pleuritic pain, 19 (13.7%) patients had erysipelas like erythema and 53 (38.1%) patients had arthritis attack. 34 (24.5%) patients also had sacroiliitis. The ratio of resistance of treatment response to colchicine drugs that can be available in Turkey (Colchicum dispersum®), 6 (4.3%) was determined. Interestingly these patients responded to the colchicine drugs available from some other countries from Europe (as Colchicine-opocalcium® and Colchicina-lirca®) None of our patients needed anti-IL1 therapies. The rate of amyloidosis was 5 (3.6%).

Conclusion: FMF is a disease with high morbidity and mortality, 95.7% of the patients in our region have response to colchicine drugs which is available in our country. The remaining patients have also response to colchicine available from some other countries. None of our patients had anti-IL1 therapies.

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AB1090

CECR1/ADA2 MUTATION IN A BRAZILIAN FAMILY

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Background: Deficiency of adenosine deaminase 2 (DADA2) is a recently identified disease caused by mutations in the CECR1/ADA2 gene, encoding for adenosine deaminase 2 protein. Clinical presentation is variable and includes early-onset polyarteritis nodosa, hemorrhagic and ischemic strokes, hypogammaglobulinemia and cytopenia

Objectives: To present the clinical cases of two Brazilian siblings with early stroke episodes carrying a homozygous CECR1/ADA2 mutation.

Methods: Chart review of clinical data, laboratory tests and mutation analysis

Results: The index case is a 7-year-old boy who presented to the emergency unit with right lower limb weakness, rhyme deviation and palpebral ptosis, associated with recurrent and intermittent fever, mood change and hypertension. The physical exam revealed drowsiness, lateral and vertical ocular paresis, diplopia, facial palsy, and bilateral ataxia. The brain MRI showed acute left mesencephalic small vessel lacunar stroke, previous right mesencephalic subacute stroke, and cerebellar cavity related with anterior cerebellar artery segmental narrowing. Laboratory tests revealed increased inflammatory markers and anemia. Autoantibodies and viral screening were negative. Renal ultrasound showed a pattern of low

resistance in the intrarenal arteries bilaterally. At this time, he was diagnosed as polyarteritis nodosa (PAN). Despite of adequate treatment (cyclophosphamide and corticosteroids), two new stroke episode occurred at left head of caudate and right thalamus. His brother, a 9-year-old healthy boy at that time, had a previous history of ischemic stroke when he was 4 years old, after receiving a vaccine, with complete recover. Considering this family history and the fact that parents are consanguineous, mutation analysis of CECR1/ADA2 gene was performed and showed homozygosity for the p.Y453C mutation in exon 9 in both siblings and heterozygosity for the same mutation in both parents. Although the eldest boy remained asymptomatic for 5 years without any specific treatment, shortly after the identification of the gene mutation, he presented recurrent fever episodes, myalgia, livedo reticularis and increased inflammatory markers. Anti-TNF treatment was initiated for both patients with good disease control.

Conclusion: DADA2 should be suspected in patients with PAN-like phenotype and history suggestive of an inherited disease (eg.: affected siblings and consanguineous parents) or resistance to conventional treatment.

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AB1091

THE RELATIONSHIP BETWEEN NAILFOLD CAPILLAROSCOPY FINDINGS IN BEHÇET'S PATIENTS AND COURSE OF THE DISEASE

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Background: Behçet's disease is a chronic, recurrent and systemic vasculitis that may affect veins and arteries at all diameters. Small vessel involvement is responsible for most of its pathological signs.

Objectives: We aimed to compare the nailfold capillaroscopy findings of patients with Behçet's disease to a healthy control group and examine the relationships, as well as revealing the relationships with the sub-type, activity and other characteristics of Behçet's disease.

Methods: We conducted a cross-sectional analysis of 153 patients with Behçet's disease and 165 healthy volunteers in a single center. The capillaroscopic findings of the 2nd-5th fingers of both hands of the participants in the Behçet's patients and control groups were included in the analysis. Capillaroscopic findings were evaluated by two different experts who were experienced in this field by using the scoring at Atlas of Capillaroscopy in Rheumatic diseases by Maurizio Cutolo (1).

Results: There was no statistically significant difference between the two groups in terms of age or sex (respectively p=0.189 and p=0.585). There was no difference between the Behçet's patients and healthy volunteers in the qualitative analysis on capillary density, capillary visibility, aneurism, capillary tortuosity, capillary enlargement and presence of avascular areas (p values respectively: 0.610, 0.147, 0.481, 0.057, 0.514 and 0.110). In the Behçet's patients, bushy capillaries (24.2%, 37/153), capillary dilatation (32%, 49/153) and microhemorrhage (39.2%, (60/153) rates were significantly higher than those in the healthy control group (p<0.001). In the quantitative analysis, total capillaroscopy score was significantly higher in the Behçet's patients than those in the healthy control group (p<0.001) (Table 1). No statistically significant relationship was found between the presence of clinical signs and capillaroscopy scores, except for erythema nodosum.

Conclusion: the Behçet's patients had significantly higher total capillaroscopy scores in comparison to those in the healthy control group. Based on these data, we believe that the capillaroscopic changes found in Behçet's patients, though unspecific, may support clinical diagnosis in uncertain cases where Behçet's disease is considered as a probability. There is a need for well-planned prospective studies to support this our thought.

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Abstract AB1091 Table 1. The results of the quantitative evaluation of nailfold capillaroscopic findings

	Behçet's patients N=153	Healthy controls N=165	p
Irregularly enlarged capillary score*	0 (0-0.44)	0 (0-0.25)	<0.001
Giant capillary score*	0 (0-0.13)	0 (0-0.13)	0.517
Microhemorrhage score *	0 (0-0.63)	0 (0-0.19)	<0.001
Capillary number score *	0 (0-0.88)	0 (0-0.25)	<0.001
Capillary ramification score *	0 (0-0.75)	0 (0-0.25)	0.224
Disorganize capillary score*	0 (0-0.44)	0 (0-0.25)	<0.001
Total capillaroscopy score*	0.16 (0-1.75)	0 (0-0.50)	<0.001

*Data were given as median (minimum - maximum)

Abstract AB1091 Table 2. The relationship between clinical findings and total capillaroscopy scores

	Total Capillaroscopy Score		p
	Present	Absent	
Vascular involvement (n=54/99) *	0.19 (0-1.48)	0.13 (0-1.75)	0.814
Ocular involvement (n=64/89) *	0.19 (0-1.48)	0.13 (0-1.75)	0.358
Neurological involvement (n=12/141) *	0.19 (0-1.48)	0.13 (0-1.75)	0.222
Genital ulcer (n=131/22) *	0.19 (0-1.75)	0.13 (0-1.48)	0.121
Erythema nodosum (99/54) *	0.13 (0-1.75)	0.19 (0-1.48)	0.026
Populopustular eruption (n=119/34) *	0.19 (0-1.75)	0.13 (0-1.2)	0.238

N, number of patients, *Data were given as median (minimum - maximum)

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AB1093

INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES: A SINGLE CENTER EXPERIENCE AND THE IMPORTANCE OF A MULTIDISCIPLINARY APPROACH

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Background: Interstitial lung disease (ILD) remains a significant cause of morbidity and mortality in patients with connective tissue diseases (CTD). Interstitial pneumonia with autoimmune features (IPAF) is a subset of ILD with clinical features suggestive of but do not definitive for a CTD. IPAF is a new concept relatively unknown to rheumatologists. A multidisciplinary approach to diagnosis and management of IPAF patients is essential, involving close interaction between pulmonologists, rheumatologists, radiologists and pathologists.

Objectives: Revision of ILD patients followed at a specialized tertiary hospital's ILD department and description of their clinical characteristics and multidisciplinary approach.

Methods: The study was conducted according to the declaration of Helsinki. All patients who met the Fischer criteria for IPAF in 2000-2018 were identified. Clinical characteristics, comorbidities, ILD subtype, pulmonary function tests, baseline serologies and treatment strategies were collected. The consents from a multidisciplinary meeting and Rheumatology referral and evaluation were also recorded.

Results: We identified 8 cases fulfilling classification criteria for IPAF (4 [50%] female); mean age 64.9 years (range 34-83); past smoking was referred in 5 (62.5%) patients with an average of 54.15 smoking pack years. Overall, 4 (50%) patients were exposed to organic dusts and 2 (25%) to inorganic dusts. Arterial hypertension was the most frequently

recorded comorbidity (50%). Among the 8 patients, 6 (75%) had at least 1 feature from the serologic and morphologic domains, 1 patient had at least 1 feature from clinical and serologic domains and 1 patient had at least 1 feature from all 3 domains. From those meeting "suggestive radiology pattern based on high resolution chest CT (HRCT)", 2 had nonspecific interstitial pneumonia (NSIP) and 1 had organizing pneumonia (OP). Biopsy (3 transbronchial cryobiopsies and 1 transthoracic biopsy) were conclusive in 4 patients (2 NSIP, 1 lymphoid interstitial pneumonia and 1 OP). Usual interstitial pneumonia (UIP) pattern was observed in two patients. Antinuclear antibodies were positive in 5 (62.5%) patients. Overall, 5 (62.5%) clinical cases were discussed in a multidisciplinary meeting including revision of imaging and biopsies. A Rheumatology appointment was requested in 5 patients to investigate a possible CTD diagnosis. Six (75%) patients had pulmonary function tests (PFT) and diffusing capacity of the lung for carbon monoxide (DLCO) results recorded at baseline: 4 patients had a DLCO below 70% (33.8 – 61.8%), 3 patients had normal PFT, 1 had restriction pattern and 2 had small airways obstruction. During a median of 2.7 years of follow-up, none of the patients progressed to a definitive diagnosis of CTD. Pharmacological treatment was prescribed in 6 patients, including corticosteroids (5), DMARDs (3), antifibrotic therapy (2) and azithromycin (2).

Conclusion: IPAF is a relatively new and developing concept. Rheumatologists and pulmonologists should share their experience to uniformize terms and classifications, recognize relevant clinical patterns and optimize management of those affected with CTDs and ILD and IPAF.

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AB1094

ARTERIOSCLEROSIS MAY AFFECT PERIAORTIC/PERIARTERIAL LESIONS AND THEIR ANEURYSMAL CHANGES IN PATIENTS WITH IGG4-RELATED PERIAORTITIS/PERIARTERITIS

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Background: IgG4-related periaortitis/periarteritis (IgG4-PA) is a major organ manifestation of IgG4-related disease (IgG4-RD) [1]. In the affected aorta/artery, arteriosclerotic changes are frequently observed [2]. However, the influence of arteriosclerosis on IgG4-PA and aneurysmal changes of the affected lesions has not been well evaluated.

Objectives: This study aimed to clarify the relationship of arteriosclerosis with periaortic/periarterial lesions and their aneurysmal changes.

Methods: We retrospectively investigated the medical data, including the presence of IgG4-PA at diagnosis, new development of IgG4-PA during the clinical course, arteriosclerotic and aneurysmal changes of the affected lesions, and classic risk factors for arteriosclerosis in 130 patients with IgG4-RD at a single center in Japan. The relationship of arteriosclerosis with the periaortic/periarterial lesions and their aneurysmal changes was statistically analyzed.

Results: Of the 130 patients with IgG4-RD, 44 comprising 39 men and 5 women (mean age 67.4 years) were diagnosed with IgG4-PA. The mean follow-up period was 72.8 months, and the mean serum IgG4 level at diagnosis was 851 mg/dL. The proportions of males (88.6% vs. 55.8%, $P<0.001$), positive smoking history (81.4% vs. 47.6%, $P<0.001$), and history of malignancy (36.4% vs. 14.0%, $P=0.006$) were significantly higher in the 44 patients with IgG4-PA than in the 86 without it. Arterial wall calcification and/or mural thrombus in the affected lesions were detected in 86.0% of the IgG4-PA patients, some of whom had had these arteriosclerotic changes before the development of periaortic/periarterial lesions. Of the 44 with IgG4-PA, 9 had aneurysmal changes of the affected lesions at the time of the first diagnosis of IgG4-RD. Notably, these 9 patients with aneurysmal changes had a significantly higher incidence of hypertension (77.8% vs. 34.3%, $P=0.027$) and mural thrombus in the affected lesions (100% vs. 64.7%, $P=0.044$) than the remaining 35 without aneurysmal changes. However, diabetes mellitus, dyslipidemia, smoking history, C-reactive protein level elevation, or arterial wall calcification of the affected lesions did not differ significantly between the two groups.

Conclusion: The present study suggests that arteriosclerotic changes and the classic risk factors for arteriosclerosis may be related to periaortic/periarterial lesions and their aneurysmal changes in IgG4-PA.