Abstract AB0986 – Table 1

Patient	Sex	Diagnostic age	Onset age	Arthritis	Location	Other musculoskeletal manifestations	Extra-articular manifestations	MEFV Mutation	Homo/ Heterocytosis
1	M	47	40	Yes	Knee	No	No	p Glu148Gln	Heterocytosis
2	M	14	13	Yes	Knee	No	No	R202Q	Homocytosis
3	M	22	22	No		No	Lymphocytic myocarditis	p Glu148Gln	Heterocytosis
4	M	52	47	No		Arthralgias	Abdominal pain. Pleuritis	R202Q	Heterocytosis
5	M	39	20	Yes	PIPs	Arthralgias	Abdominal pain	R202Q	Heterocytosis
6	M	61	54	No		Tendinopathy	Recurrent pericarditis	R202Q	Homocytosis
7	M	17	16	No		No	Abdominal pain	R202Q	Heterocytosis

Results: Seven patients with MEFV gene mutations were reviewed, all of them were women, ranging in age from 14 to 61 years old. Two of them had recurrent knee monoarthritis, one had a history of arthritis in the hands and erratic arthralgias, one had erratic arthralgias and two had no musculoskeletal manifestations. The 2 patients with intermittent hydrartrosis responded satisfactorily to colchicine, but not the other patients with other musculoskeletal manifestations.

Conclusions: Genetic testing of the common mutations of the MEFV gene should be considered in patients with recurrent episodes of monoarthritis without justifying cause (palindromic rheumatism, intermittent hydrartrosis, etc.)

Disclosure of Interest: None declared

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AB0988 BEHÇET'S DISEASE IN A DEFINED AREA OF NORTHWESTERN SPAIN

M. Estévez, A. Argibay, R. Lorenzo, M. Freire, J. Fernandez-Martín, B. Gimena, C. Vázquez-Triñanes, A. Rivera. *Systemic Autoimmune diseases and Thrombosis Unit, University Hospital Complex of Vigo, Vigo, Spain*

Background: Behçet's disease (BD) is a multisystemic inflammatory chronic disease. There is a wide variation in the clinical features of BD among geographical groups.

Objectives: To determine the demographic and clinical characteristics of BD in a defined area of northwestern Spain (Vigo).

Methods: Patients with BD (International Criteria BD) and seen in the University Hospital of Vigo in Spain, from 1994 to 2016, were retrospectively enrolled. Demographic, clinical, treatment and evolution data were recorded and analyzed using SPSS 22.0.

Results: Our patients were 26 male and 25 female. The mean age at the onset of the disease was 33±11.95 years (11–62). Oral and genital ulcers were seen in 100% and 84.3% respectively and skin lesions in 88.2%. Ocular involvement occurred in 35.3%, neurological disease in 39.2% and gastrointestinal involvement in 29.4% (the area worst affected was colon and small bowel). Vascular disease was present in 33.3%. See table 1. Pathergy test was performed in 18 patients (35.29%) and 10 (55.5%) were positives. HLA B51 was studied in only 13 patients (25.5%) and 8 (61.5%) were positives. 62.7% of patients had no cardiovascular risk factors (CVRF), 27.4% were smokers, 7.8% were hypertensive and 3.9% were hyperlipidemic and diabetic respectively. CVRF were not related to thrombotic events (p>0.05). In regard to gender influence, only pseudofolliculitis was significantly more frequent in men (p<0.001). There was a trend for increased prevalence of ocular disease and elevated erythrocyte sedimentation rate/serum C-reactive protein in men, and anemia in women, which, however, did not reach statistical significance. Treatment consisted of corticosteroids (92.2%), colchicine (68.6%) and another immunosuppressive agent (35.3%). During the disease course 78.4% of the patients had an outbreak, 45.1% initiated or changed to immunosuppressive agent and 19.6% used biological drugs. Most of our patients (92.1%) were admitted to hospital and these constitute an evident bias. Two patients died during the followup period, but only one in relation of BD (upper gastrointestinal bleeding and seizures).

Conclusions: Our series has some particular aspects especially the high frequency of gastrointestinal lesions and neurologic involvement. CVRF do not seem to play a role in the development of thrombotic events. Our results confirm the ethnic and geographic variation of BD expression.

Disclosure of Interest: None declared

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AB0989 RHEUMATOLOGIC COMPLICATIONS OF THALASSEMIA: SHOULD RHEUMATOLOGISTS JOIN THE MANAGEMENT TEAM?

M.H.A. Noureldine¹, A.T. Taher², A.A. Haydar³, A. Berjawi², M.A. Khamashta^{4,5}, I. Uthman². ¹Department of Neurosurgery, Lebanese American University; ²Department of Internal Medicine, American University of Beirut; ³Gilbert and Rose-Marie Chagoury School of Medicine, Lebanese American University, Beirut, Lebanon; ⁴Lupus Research Unit, St Thomas Hospital, London, United Kingdom; ⁵Department of Rheumatology, Dubai Hospital, Dubai, United Arab Emirates

Background: Beta-thalassemia major is accompanied by progressive multiple organ systems involvement due to the disease pathophysiology as well as iron overload from blood transfusions on a regular basis. Rheumatologists are not frequently involved in the multidisciplinary management of the disease, in which rheumatologic complications are relatively common.

AB0987 THE RELATIONSHIP BETWEEN ANTI-INTERLEUKIN-1 THERAPIES AND MEFV GENE MUTATIONS IN FAMILIAL MEDITERRANEAN FEVER PATIENTS: A SINGLE CENTER EXPERIENCE

M.E. Derin¹, S. Gültekin², M. Şahin³, A. Şahin¹. ¹Rheumatology - Internal Medicine; ²Internal Medicine; ³Biochemistry, Cumhuriyet University Medical Faculty, Sivas, Turkey

Background: Familial Mediterranean fever (FMF) is an autoinflammatory disease characterizing recurrent self-limiting attacks of inflammation mainly placed in serosal surfaces of the body with fever (1). Colchicine is an important weapon of the humanity in FMF treatment yet. However, the puzzle of inflammation was solved, and so, inflammasome complex and interleukin-1 were discovered. Eventually, new era started with anti-interleukin-1 agents in colchicine resistant and/or intolerant FMF patients (2).

Objectives: The aim of this study is to evaluate the effectiveness of anti-interleukin-1 (anti-IL-1) agents on the characteristics of attacks, the adverse effects associated with anti-IL-1 treatment, and relation between therapy and MEFV gene mutations in 23 FMF patients with resistant and/or intolerant to colchicine.

Methods: Between January 2015 and December 2016, twenty-three-FMF patients that following-up at Cumhuriyet University Medical Faculty Rheumatology-Internal Medicine Department were included in to the study. Anakinra (69,6%), and canacinumab (30,4%) were used in 23 FMF patients. 20 cases were resistant to colchicine, 3 were intolerant to colchicine.

Results: The median age of the patients was 28 years,(18–54) and the median age at diagnosis was 20 years (3–50). Of the FMF patients, nine (39.1%) were female and fourteen (60.9%) were male. The distribution of MEFV gene mutation frequencies in the FMF patients was no mutation in one (4,3%) patient, M694V heterozygous in nine (39,1%), M694V homozygous in 7 (30,4%), E148Q heterozygous in one (4,3%), and compound heterozygous mutation in 3. (13%) patients. 16 patients used anakinra (100mg/day) for 6 mounths (median) and 7 used canacinumab (150mg/2 months) for 3 mounths (median). Four of 7 patiens were resistant to anakinra. After a median follow up 6 months overall clinical response [no attack (18 patients) or decreased frequency of attacks (5 patients)] was %100. In an FMF patient with no mutation, his brother and mother had FMF with no mutations on MEFV gene. We achieved good clinical and laboratory responses in the patient by canacinumab.

Conclusions: Anti-IL-1 agents can be got involved in a new place safely and effectively in FMF patients along with colchicine. In addition, whole gene analysis should be done in refractory FMF patients with no mutations in order to could investigate new mutations, epigenetic mechanisms or other unexplained reasons.

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Disclosure of Interest: None declared

Abstract AB0988 – Table 1. Clinical manifestation of our BD patients

Skin lesions 41/55 (88.2%)	Ocular disease 18/51 (35.3%)	Neurologic disease 20/51 (39.2%)	Vascular disease 17/51 (33.3%)	Gastrointestinal involvement 15/51 (29.4%)
Pseudofolliculitis 58.8%	Uveitis 23.5%	Aseptic meningitis 11.7%	Venous thrombosis 21.6%	Pain 11.7%
Erythema nodosum 39.2%	Retinal vasculitis 5.9%	Seizures 7.8%	Arterial thrombosis 7.8%	Bleeding 9.8%
Leukocytoclastic vasculitis 5.9%	Episcleritis 3.9%	Cerebral venous thrombosis 5.9%	Mixed thrombosis 2%	Diarrhea 5.9%
	Vascular occlusion 2%	White matter lesions 5.9%	Aneurysm 2%	Fever 2%
		Stroke 3.9%		
		Paresthesias 3.9%		
		Others 5.9%		

Objectives: To highlight the rheumatologic complications of beta-thalassemia, which include musculoskeletal manifestations such as arthritis and arthropathies, joint effusions, osteoporosis, bone fractures, and arthralgias as well as connective tissue diseases such as pseudoxanthoma elasticum (PXE).

Methods: We searched the literature through Google Scholar and PubMed for all publications reporting on rheumatologic manifestations in beta-thalassemia. The following keywords were used: thalassemia, rheumatoid arthritis, osteoarthritis, iron-overload, septic arthritis, deferiprone-related arthropathy, crystal arthritis, effusions, osteoporosis, fractures, bone marrow hyperplasia, scoliosis, aseptic necrosis, arthralgia, myalgia, and pseudoxanthoma elasticum. All reports published between 1970 and 2016 were included.

Results: Arthralgia and low back pain are among the most common musculoskeletal manifestations of thalassemia. Many studies are reporting an increased rate of RA in hemoglobinopathies such as thalassemia, the association of which may be attributed to genetic factors. Arthritis may develop secondary to iron deposition in the synovial tissue or due to iron chelators such as deferiprone, which may provoke a self-limited arthritis due to synovial destruction sustained by free radicals production during iron interchange. Several studies reported increased incidence (16%–30%) of arthropathy in beta-thalassemia patients on deferiprone therapy. Multiple etiologies contribute to osteoarthritis in thalassemia patients such as bone marrow expansion, iron deposition within the joint, and hypoparathyroidism. Only few cases of Salmonella enteritidis septic arthritis were described in thalassemia patients. The associations between osteoporosis and hypogonadism, diabetes, and vitamin D and calcium deficiency were found significant. Skin lesions of PXE were reported in up to 16% in beta-thalassemics.

Conclusions: The pathophysiology of thalassemia and subsequent blood transfusions affect almost every organ system. Rheumatologic manifestations ranging from musculoskeletal complications to connective tissue diseases are common among thalassemia patients. Rheumatologists are highly encouraged to be involved in the multidisciplinary approach for the management of beta-thalassemia.

Disclosure of Interest: None declared

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AB0990 PERI-ARTERITIS AS A NOVEL PARANEOPLASTIC PRESENTATION OF MYELODYSPLASTIC SYNDROME: A CASE SERIES AND REVIEW OF THE LITERATURE

N. Droz, C. Gota. *Rheumatology, Cleveland Clinic Foundation, Cleveland, United States*

Background: Myelodysplastic syndromes (MDS) are characterized by ineffective and dysplastic hematopoiesis resulting in peripheral blood cytopenias with varying risk for progression to acute myelogenous leukemia (AML).¹

Autoimmune manifestations have been well described in association with MDS.² Peri-arteritis has rarely been reported.

Objectives: Here, we present a novel association of MDS with peri-arteritis.

Methods: Three patients within our institution and one in the literature were identified. Patients were diagnosed with MDS according to 2008 World Health Organization classification with concomitant occurrence of peri-arteritis on imaging.³

Results: See Table 1.

Conclusions: Autoimmune manifestations associated with MDS occur in 7–25% of patients. The most commonly reported conditions include vasculitis, inflammatory arthritis and connective tissue disease. There is a paucity of literature describing peri-arteritis associated with MDS.² Review of the literature revealed only one other case of peri-arteritis associated with MDS outside of those identified at our facility.⁴

With this case series, we report peri-arteritis as a novel presentation of MDS. Peri-vascular inflammation may mimic idiopathic retroperitoneal fibrosis, however, persistent cytopenias, lack of response to immunosuppressive therapy and persistence of elevated inflammatory markers should prompt the physician to evaluate for an underlying diagnosis of MDS.

Abstract AB0990 – Table 1

Patient	1	2	3	4 ^a
Age	67	37	67	70
Sex	Female	Female	Female	Male
MDS type	CMML-1	MDS with multi-lineage dysplasia; progression to AML	MDS Unclassifiable	MDS with multi-lineage dysplasia
Presenting Symptoms	Hydronephrosis, abdominal pain, weight loss	Serositis, dyspnea, fevers, rash	Rapidly evolving peri-arteritis, abdominal pain	Organizing pneumonia, fever, arthralgias
Bone Marrow Biopsy	Hypercellular, features of CMML-1	Normocellular, multilineage dysplasia, 4% blasts, 20q deletion	Hypercellular, granulocytic hyperplasia, 2% blasts	Normocellular, multilineage dysplasia
ANCA and ANA	Negative	Negative	Negative	Negative
ESR (0–15mm/hr)	21	76	57	
CRP (<0.9mg/dL)	5.4	31.2	4.9	9
Radiographic extent of peri-arteritis	Infra-renal abdominal aorta and common iliac arteries	Thoracic aorta and branch vessels	Carotid arteries, thoracic and abdominal aorta, femoral vessels	Internal carotid artery
Treatment	Corticosteroids	Corticosteroids, anakinra, methotrexate, IVIG	Corticosteroids	Corticosteroids
Clinical Course	Ureteral stenting and nephrostomy tube placement improved symptoms. Inflammatory markers remained elevated despite treatment.	Multiple treatment modalities failed to improve symptoms or inflammatory markers. MDS transformed to AML requiring stem cell transplant. Her disease was then quiescent.	Despite treatment, disease was complicated by aortic dissection which was managed non-operatively. Azacitidine was initiated but she died of infectious complications.	Epipharyngeal mass improved with corticosteroids. Disease course was complicated by infection.

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AB0991 PREVALENCE OF FIBROMYALGIA AND DEPRESSION IN PATIENTS WITH AUTOIMMUNE /INFLAMMATORY SYNDROME INDUCED BY ADJUVANTS COMPARED TO PATIENTS WITH SYSTEMIC SCLEROSIS

O. Vera-Lastra, P. Blas Hernandez, A. Sánchez-Rodríguez, L.J. Jara. *Internal Medicine, Hospital Especialidades CMN la Raza. Instituto Mexicano Seguro Social, Mexico City, Mexico*

Background: Autoimmune/inflammatory syndrome induced by adjuvants (ASIA) has been associated with previous exposure to various agents such as mineral oil, silicone and vaccines, which act as adjuvants eliciting chronic stimulation of the immune system and manifestation of autoimmune diseases. There is little information about fibromyalgia (FM), depression and the deterioration of the quality of life in ASIA patients.

Objectives: To determine the prevalence of FM, depression and both in ASIA patients associated to mineral oil versus systemic sclerosis (SSc) patients.

Methods: A comparative cross-sectional study was performed in patients with ASIA according to Shoenfeld's criteria associated to injection of mineral oil with cosmetic purposes. The prevalence of fibromyalgia according to criteria of ACR, depression by Beck questionnaire and quality of life with FS-36 in patients with ASIA versus patients with systemic sclerosis matched by age were evaluated.

Results: There were 100 women, 50 ASIA patients (mean age 49±9.7 years) compared to 50 SSc patients (mean age 49±10 years). Of patients with ASIA, 40 patients had non-specific rheumatic disease and 10 met ACR criteria for SSc in 4 patients, SLE 3 and overlap syndrome 1 (SLE plus SSc) and rheumatoid arthritis 2. In ASIA vs. SSc we found depression in 72% vs 46% respectively, p<0.005, fibromyalgia in 66 vs 48%, p<0.008 and fibromyalgia plus depression in 56% vs. 28, p<0.005. Patients with ASIA had a worse quality of life than SSc by SF36 and the most affected parameter was the emotional one.

Conclusions: ASIA patients associated to injection of mineral oil had higher prevalence of depression, fibromyalgia and greater deterioration of quality of life compared to SSc patients. There is a possible link between ASIA, depression and FM.

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