Figure 1. Flowchart of the results of PT in (suspected) BS patients

In 22 patients with two ISG criteria, grade I uveitis was diagnosed in 1 patient (4.5%, 95% CI [0.8,21.8]) and retinal vasculitis in 1 patient (4.5%, 95% CI [0.8,21.8]). He had shortly experienced photophobia 3 months earlier. Both patients (9%, 95% CI [2.5,27.8]) changed from “probable” to “true” BS, the latter also had a positive PT.

47 patients assessed the results of their own PT. The dermatologist scored PT as positive in one patient. This patient scored the results as unsure on all six PT points. In 46 patients, the dermatologist scored PT as negative, a total of 26 patients (56.5%, 95% CI [42.2,69.7]) scored their results negative as well.

Conclusion: The added diagnostic value of PT in patients fulfilling 2 features of the ISG criteria was limited in our study, i.e. 5.7% 95% CI [1.6,18.6]. Furthermore, half of patients scored their true negative PT result as negative. The added diagnostic value of routine eye examination in probable BS patients was limited in our study.

REFERENCES:

- Criteria for diagnosis of Behçet's disease. International Study Group for Behçet's Disease. *Lancet* 1990;335(8697):1078–8

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POS1362

NEUROSARCOIDOSIS. STUDY OF 30 PATIENTS FROM A SERIES OF 384 SYSTEMIC SARCOIDOSIS FROM A TERTIARY UNIVERSITY HOSPITAL

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Background: Neurosarcoidosis is a serious and infrequent complication of sarcoidosis. Data on Biologic Therapy (BT) efficacy is scarce.

Objectives: To assess in neurosarcoidosis a) its prevalence and associations of clinical clusters and b) efficacy and safety of BT.

Methods: Study of neurosarcoidosis from a large cohort (n=384) of all consecutive patients diagnosed with sarcoidosis from January 1, 1999 to December 31, 2019 in a single University hospital.

Results: 30 (19 women/ 11 men) out of 384 (7.8%) patients had neurosarcoidosis. Mean age at diagnosis was 41.8±15.8 years. Clusters of clinical associations were lungs (n=23, 76.7%) and joints (n=15, 50.0%) (Figure 1). The underlying neurological manifestations were chronic headache (n=13, 43.4%), peripheral neuropathy (n=6, 20.0%), cranial neuropathy (n=5, 16.7%), spinal cord abnormalities (n=3, 10.0%) and aseptic meningitis (n=3, 10.0%). 26 (86.7%) patients received oral corticosteroids (mean maximum dose 50±19.2mg) and 7 (23.3%) iv corticosteroids. In addition, 18 (60.0%) patients received conventional immunosuppressants and 12 (40.0%) BT. 4 (13.3%) did not have any treatment. Table 1 shows the main clinical features and treatment.

Figure 1. Clusters of clinical associations in neurosarcoidosis

12 patients received treatment with 14 BT. Monoclonal anti-TNF α (n=10, 83.3%) was the most used BT. 10 (83.3%) patients treated with BT achieved clinical remission. No serious adverse effects were observed.

Conclusion: Neurosarcoidosis was observed in 7.8% of systemic sarcoidosis. Most patients treated with BT, especially monoclonal anti-TNF α , achieved clinical remission.

Table 1. Main clinical features and treatment of 30 patients with neurosarcoidosis.

		Conventional immunosuppressant, n (%)	monoclonal anti-TNF α , n (%)	Etanercept, n (%)	Tocilizumab, n (%)	Secukinumab, n (%)	Rituximab, n (%)
Chronic headache	13 (43.4)	5 (38.5)	2 (15.4)	0 (0)	0	1 (7.7)	1 (7.7)
Peripheral neuropathy	6 (20.0)	4 (66.7)	3 (50.0)	1 (16.7)	0	0	0
Cranial neuropathy	5 (16.7)	4 (80.0)	3 (60.0)	0	0	0	0
Spinal cord abnormalities	3 (10.0)	1 (33.3)	1 (33.3)	0	0	0	0
Aseptic meningitis	3 (10.0)	2 (66.7)	2 (66.7)	0	1 (33.3)	0	0

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POS1363

CHONDROCALCINOSIS AS PREDICTOR OF DEVELOPMENT OF PRIMARY HYPERPARATHYROIDISM: RESULTS OF THE FOLLOW-UP OBSERVATION

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Background: The association between chondrocalcinosis and primary hyperparathyroidism (PHPT) is known, but chondrocalcinosis is considered as a late complication of PHPT.

Objectives: To investigate whether calcium pyrophosphate crystal deposition is an early clinical sign of PHPT.

Methods: The prospective study included 113 patients aged ≥ 18 years, 42 (37.2%) men and 71 (62.8%) women, with a crystal-verified diagnosis of calcium pyrophosphate crystal deposition disease (CPPD) (McCarty criteria). Exclusion criteria were the presence of other rheumatic diseases with symptoms of arthritis, the presence of hyperparathyroidism (HPT). Patients were examined at their enrollment and over time, the median follow-up was 3.86 [2.07; 8.36] years. The examination of the patients included assessment of anthropometric parameters, information about the affected joints and the time of onset of symptoms.

Table 1. Results of patients with PHPT at screening and in dynamics.

	Crystals of calcium pyrophosphate in synovial fluid	Chondrocalcinosis by ultrasound	Chondrocalcinosis by X-ray	Convulsions	ECG changes (shortening of the QT interval)	Arthralgias / arthritis	PTH, pg/ml (15,0-65,0)	Ca, µmol/l (2,10-2,62)	Ca ⁺⁺ , µmol/l (1,10-1,33)	vitamin D (25-OH), ng/ml (30-100)
screening / dynamics										
Patient A.	yes/yes	yes/yes	not/yes	not/not	not/not	yes/yes	61,6/96,2	2,73/2,67	1,37/1,24	-/31
Patient B.	yes/yes	yes/yes	yes/yes	not/yes	not/yes	yes/yes	40,7/252	2,63/2,56	1,34/1,77	19,1/14,2
Patient C.	yes/yes	yes/yes	not/yes	not/not	not/not	yes/yes	26,6/102,4	2,60/2,65	1,25/1,26	-/40

Laboratory tests included determination of the following in blood serum: creatinine (with estimating glomerular filtration rate (eGFR) according to the MDRD formula), total and ionized calcium (Ca and Ca⁺⁺), phosphorus (P), parathyroid hormone (PTH), vitamin D (25-OH); ultrasound and X-ray investigations of the target joints (hands, knee joints) were made for all patients. Scintigraphy and ultrasound of the parathyroid glands were performed if medically required. Statistica 12.0 package was used for statistical data processing.

Results: 113 patients were examined, the average age was 58.4±12.4 years. 10 patients were excluded due to diagnosed HPT at screening. 41 patients dropped out of observation: 5 - died, 36 were not available for dynamic examination. 62 patients were examined in dynamics, 4 (6.5%) of those had HPT. In 1 (1.6%) case, HPT was associated with chronic renal failure, 3 (4.8) patients were diagnosed PHPT. The results of the examination of patients who developed PHPT during screening and in dynamics are presented in Table 1 above.

In all three patients, chondrocalcinosis was revealed by ultrasound and Crystals of calcium pyrophosphate in synovial fluid during the screening examination, in one patient chondrocalcinosis was revealed by X-ray. In two out of three cases, the level of serum Ca and Ca⁺⁺ was minimally increased at a normal level of PTH, no other disorders of calcium metabolism were observed during the screening.

Interestingly, the first clinical manifestation of HPT in all patients was damage of the musculoskeletal system - mainly arthralgia, as well as arthritis of large and small joints. Moreover, all patients had a different CPPD phenotype: patient A - asymptomatic chondrocalcinosis, patient B - chronic arthritis (pseudo-rheumatoid form) and patient C - chronic arthritis (pseudosteoarthritis).

Conclusion: The deposition of calcium pyrophosphate crystals may be an early predictor of the development of PHPT and precedes other manifestations.

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POS1364

MUSCULOSKELETAL VARIANT OF FAMILIAL MEDITERRANEAN FEVER MAY HAVE MORE SEVERE DISEASE

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Background: Familial Mediterranean fever (FMF) is one of the prototypes of auto inflammatory diseases. Besides recurrent serositis, some patients also have musculoskeletal involvement similar to spondyloarthropathies (SpA) (1). These patients usually have more severe disease (2). In addition, some experts think FMF can be considered a variant of SpA.

Objectives: There is handful of studies in literature that evaluated the disease severity, FMF complications and treatment variations in FMF patients with individual musculoskeletal features separately (3). However, these patients can be considered as musculoskeletal/SpA variants of FMF. Here, we evaluated the validity of hypothesis: "The patients with any of musculoskeletal symptoms or SpA variant of FMF have more severe disease."

Methods: We included 218 FMF patients who met Modified Tel- Hashomer criteria. We divided the patients according to suffering from any of the musculoskeletal symptoms (arthritis, enthesitis, arthralgia/myalgia, sacroiliitis, standing myalgia). Here, arthritis, enthesitis were diagnosed with physical examination according to appropriate methods. Standing myalgia diagnosis was depending upon patients' history. Lastly, sacroiliitis was demonstrated in radiological screening. The patients with any of the musculoskeletal symptoms were considered musculoskeletal variants of FMF. In this study we compare disease characteristics and treatment differences between the groups. We used Mann-Whitney-U or chi-square tests where appropriate.

Results: There were 126 (57.7%) patients in the musculoskeletal group. FMF patients in both groups had similar demographic and disease characteristics except musculoskeletal signs. Musculoskeletal variant group had higher The International Severity Score for FMF (ISSF) (p<0.001); more frequent (p=0.009), intense (p=0.009) and longer attacks (p=0.001) than the others. Also, patients with musculoskeletal variant received higher colchicine dose (p=0.006).

Table 1. Demographic, disease and treatment characteristics of the patients

	Musculoskeletal variant n=126	Non-musculoskeletal group n=92	p
Gender (M/F)	45/81	34/58	0.85
Disease duration(year)	9.6±7.3	8.3±6.9	0.08
Fever (%)	81 (64.3)	53 (57.6)	0.37
Abdominal pain (%)	106 (84.1)	83 (90.2)	0.19
Pleuritis (%)	46 (36.5)	28 (30.4)	0.35
Number of attack region	1.7±0.8	1.3±0.5	<0.001
Attack frequency (/year)	3.4±5.0	2.1±3.5	0.009
Attack duration (day)	1.6±1.7	0.9±1.5	0.001
Attack VAS score (0-100)	33.3±32.7	23.1±31.5	0.009
ISSF score	1.6±1.3	0.6±0.9	<0.001
Exon 10 homozygote (%)	33 (26.2)	19 (20.7)	0.42
Amyloidosis (%)	6 (4.8)	2 (2.2)	0.47
Colchicine dose (mg/day)	1.4±0.4	1.2±0.4	0.006
IL-1 blocker (%)	1 (0.8)	2 (2.2)	0.57

M: Male; F: Female; ISSF: The International Severity Score for FMF; VAS: Visual analog score. All continuous variables shown as mean±SD. p<0.05 is significant.

Conclusion: We showed that FMF patients with musculoskeletal signs have more severe disease and received higher colchicine dose as compared to non-musculoskeletal group and can be considered as the SpA variant of FMF or a distinct variant of SpA.

REFERENCES:

- [1] Atas N, Armagan B, Bodakci E, et al. Familial Mediterranean fever is associated with a wide spectrum of inflammatory disorders: results from a large cohort study. *Rheumatol Int* 2020; 40:41-48.
- [2] Sen N, Yilmaz M, Mercan R, et al. Enthesitis may be one of the signs of severe disease in familial Mediterranean fever. *Clin Rheumatol* 2020
- [3] Eshed I, Rosman Y, Livneh A, et al. Exertional leg pain in familial Mediterranean fever: a manifestation of an underlying enthesopathy and a marker of more severe disease. *Arthritis Rheumatol* 2014; 66:3221-6.

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POS1365

ANALYSIS OF PRO-INFLAMMATORY BIOMARKERS IN A POLYMYALGIA RHEUMATICA COHORT DURING THE ACUTE ONSET OF THE DISEASE

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Background: Polymyalgia Rheumatica (PMR) is a common inflammatory disease found in elderly patients and results in a high burden of long-term CS-based therapies. Most patients diagnosed with PMR have a good evolution following the introduction of medium-dose glucocorticoid treatment. However, the need to maintain corticosteroid treatment for an extended period (around 12-15 months) is often the norm in standard clinical practice. Activation of Th-17 lymphocytes with the consequent action of pro-inflammatory cytokines such as IL-1, IL-6, IL-17, or IL-23 is well known in the acute phase of this disease and treated by glucocorticoids or IL-6 blockers satisfactorily. However, activation of Th-1 lymphocytes appears to play a leading role in a chronic phase of the disease through the activation of INF-gamma and IL-12.

Objectives: Our aim was to assess proinflammatory biomarkers in a cohort of patients with PMR during the acute onset of the disease and its possibly relationship with corticosteroid dose.

Methods: Prospective study of a wide and unselected series of patients classified as PMR following 2012 ACR/EULAR classification criteria for PMR. Patient evaluations were performed at baseline and at 3 and 6 months after initiation of corticosteroid therapy. Serum levels of the proinflammatory biomarkers interleukin-6 (IL-6), interleukin-8 (IL-8), CXC motif ligand 10 chemokine (CXCL10), CXC motif ligand 9 chemokine (CXCL9), CXC motif ligand 2 chemokine (CXCL2) and