Finally, a higher percentage of AOSD patients with MAS showed a significant lymph node enlargement, either mediastinal or abdominal, than others on CT scan (AOSD: 36.0% vs MAS 71.4% $p=0.048$). The presence of lymphadenomegaly correlated with the systemic score (coefficient 0.368, $p=0.032$).

Conclusion: Our findings showed a higher prevalence of multiorgan involvement in AOSD patients with MAS, suggesting imaging-based differences, although other studies are needed to fully assess this issue. Pulmonary disease, hepatomegaly, splenomegaly, lymph nodes enlargement, and abdominal effusions were associated with these more aggressive patients.

REFERENCES:

[1] Giacomelli R, Ruscitti P, Shoenfeld Y. A comprehensive review on adult onset Still's disease. *J Autoimmun.* 2018 Sep;93:24-36.

Disclosure of Interests: None declared

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POS1345 PULMONARY ARTERY PSEUDOANEURYSMS IN BEHCET'S DISEASE

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Background: Pulmonary artery pseudoaneurysms (PAP) is a serious complication of Behcet's Disease (BD).

Objectives: The aim of this work is to analyze, through 4 observations, the clinical, para-clinical and therapeutic aspects of PAP.

Methods: Matériels et méthodes:

Retrospective study including 4 cases of PAA among 150 patients who satisfied the criteria of the International Study Group on BD, followed in the Internal Medicine and Radiology Departments at Tahar Sfar Hospital Mahdia TUNISIA.

Results: Résultats:

Four men, with an average age of 27 years (20-34). PAPs were inaugural, and multiple (>3) in 3 cases and associated with pulmonary embolism in 2 cases. Hemoptysis was the main clinical symptom. All patients were treated with high-dose corticosteroid therapy combined with monthly boluses of cyclophosphamide in addition to colchicine. Two patients had undergone arterial embolization. Surgery was indicated for 2 patients, one of whom died after surgery as a result of massive hemoptysis. The others had a favorable outcome.

Conclusion: PAP severity justifies the necessity of early diagnosis and management. The thoracic angio-CT is the imaging of choice for diagnosis. The treatment is based on a corticosteroid therapy and immunosuppressive association with possibly a selective embolization.

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POS1346 CLINICAL ASSOCIATIONS OF ANTI-RO52 ANTIBODIES IN PATIENTS WITH SYSTEMIC AUTOIMMUNE RHEUMATIC DISEASES

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Background: Anti-SSA/Ro antibodies (Abs) can target Ro60 and Ro52 antigens. The presence of anti-Ro60 Abs has been widely described in patients with systemic autoimmune rheumatic diseases (SARDs). However, the clinical implication of anti-Ro52 Abs for the diagnosis and management of SARDs remains unclear.

Objectives: To assess the clinical associations of anti-Ro52 antibodies in patients with high clinical suspicion of SARDs.

Methods: We retrieved the clinical records of all patients with positive anti-Ro52 Abs tested in our hospital between November 2017 and September 2020. Patients were divided into 3 groups: 1) anti-Ro52+Ro60+2) antiRo52+Ro60-3) antiRo52+Ro60+ with other Abs. A comparative study between groups was performed.

Results: 57 patients (43 women/14 men; mean age 62.1±13.6 years) with antiRo52+ Abs were identified. Final diagnosis were: undifferentiated connective tissue disease (UCTD) (n=13), anti-synthetase/overlap myositis (n=12), Sjögren's syndrome (n=7), interstitial pneumonia with autoimmune features (IPAF) (n=6), scleroderma (n=4), systemic lupus erythematosus (n=2), dermatomyositis (n=2), other systemic inflammatory diseases (n=3). In 8 (14%) patients the diagnosis of inflammatory diseases was finally ruled out. 27 patients were classified in the Ro52+Ro60+ group, 11 in the Ro52+Ro60- group and 19 Ro52+ with other Abs. Patients with Ro52+Ro60- were younger and more often women than patients with Ro52+Ro60+. Interstitial lung disease (ILD) was less frequent in patients with Ro52+Ro60- (Table 1). Isolated Ro52 Abs were more frequently

associated with UCTD, while IPAF was more commonly found in patients with anti-Ro52+Ro60+ Abs (Table 1 and Figure 1).

Figure 1.

Conclusion: Anti Ro52 Abs determination has clinical implications in the diagnosis of SARDs.

Table 1.

	Anti-Ro52+Ro60+ (n=27)	Anti-Ro52+Ro60- (n=11)	Anti-Ro52+Ro60+ with other Abs (n=19)
Age (years), mean ± SD	65.7 ± 10.0	53.8 ± 18.0*	61.7 ± 13.6
Sex (females), n (%)	19 (70.4)	10 (90.9) *	14 (73.7)
Clinical manifestations at anti-Ro52 determination, n (%)			
Arthralgias/arthritis	18 (66.7)	7 (63.6)	14 (73.7)
Raynaud's phenomenon	9 (33.3)	4 (36.4)	9 (47.4)
Myopathy	4 (14.8)	1 (9.1)	2 (10.5)
Final diagnosis, n (%)			
Systemic lupus erythematosus	0 (0)	0 (0)	2 (10.5)
Sjögren's syndrome	3 (11.1)	1 (9.1)	4 (21.1)
Scleroderma	1 (3.7)	0	3 (15.8)
Undifferentiated connective tissue disease	3 (11.1)	7 (63.6) **	3 (15.8)
Overlap myositis	7 (25.9)	1 (9.1)	4 (21.1)
Dermatomyositis	1 (3.7)	0	1 (5.3)
IPAF	5 (18.5) **	0	0
Other systemic inflammatory diseases	3 (11.1)	0	0
Non inflammatory disease	4 (14.8)	2 (18.2)	2 (10.5)
Comorbidities, n (%)			
ILD	11 (40.7)	1 (9.1) **	7 (36.8)
Malignancy	5 (18.5)	1 (9.1)	0 (0.0)

* $p < 0.05$ (Ro52+Ro60+ vs Ro52+Ro60-)

** $p < 0.05$ (Ro52+Ro60+ vs Ro52+Ro60- vs Ro52+Ro60+ with other Abs)

Disclosure of Interests: None declared

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POS1347 IMPACT OF CANAKINUMAB AND ANAKINRA ON PATIENT-REPORTED OUTCOMES IN ADULT-ONSET STILL'S DISEASE PATIENTS

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Background: Adult-onset still's disease (AOSD) is a rare systemic inflammatory disease. Interleukin-1 (IL1) blockade has shown to be crucial for the management of refractory AOSD patients. The IL1 inhibitors anakinra and canakinumab are both effectively used in clinical practice for the management of AOSD. No data are available on the impact of these therapies on patient-reported outcomes (PROs).

Objectives: To assess the impact of ANK and CNK therapies on PROs of AOSD patients.

Methods: Medical records of AOSD patients followed up at our Institution who had been treated with both ANK and CNK were identified. Disease features were retrospectively collected. All patients were initially treated with ANK and

subsequently with CNK. At the moment of CNK introduction and at their latest visit while on CNK, patients were asked to fill a questionnaire investigating their experience with either ANK or CNK. The questionnaire included 4 different domains that were assessed through means of Visual Analogue Scales (VAS) ranging from 0 to 10. The following domains were considered: injection-related pain, satisfaction with the frequency of administration, concern for drug-related adverse events including infections, and overall drug tolerability. VAS values for each of the domains were compared between ANK and CNK with Mann-Whitney U test.

Results: Ten AOSD patients treated with both ANK and CNK were included in the analysis. Disease characteristics are shown in Table 1 below. ANK was discontinued due to inefficacy in 6 patients and due to adverse reactions in 4. As shown in Figure 1, CNK overall tolerability significantly exceeded ANK one ($p=0.021$). Only 1 AOSD patient reported a greater tolerability with ANK compared to CNK because she felt more comfortable with pre-filled syringes. The reasons for higher CNK tolerability were the lower pain at injection site (median 1, IQR 1-1 vs median 4.5, IQR 1.25-7; $p=0.015$) and the greater level of satisfaction with the frequency of administration (median 10, IQR 9.25-10 vs median 3.5, IQR 2.25-4.75, $p<0.001$). The concern for drug-related adverse events was not significantly different between ANK and CNK ($p=0.549$) and overall low among AOSD patients. CNK led to a satisfying control of systemic inflammation and steroid-sparing effect in most of patients and it was discontinued in only one case (Patient 5), due to ineffective disease control.

Figure 1. Comparative tolerability of anakinra (blue line) and canakinumab (orange line) in our study population, expressed as Visual Analogue Scale values (range from 1 to 10). Reported p -values refer to the comparative analysis between the two drugs.

Table 1. Clinical and treatment characteristics of study population.

	Sex and age (years)	Disease duration (months)	Disease manifestations	Sequential biologic therapies	Steroid dosage variation (mg/day) [*]	ESR variation (mm/hour) [*]	CRP variation (mg/L) [*]
Patient 1	Male, 70	22	Arthritis, Fever	TCZ, ANK, CNK	25 → 5	120 → 13	55 → 1
Patient 2	Female, 72	11	Arthritis, Fever	TCZ, ANK, CNK	5 → 0	60 → 24	265 → 6
Patient 3	Male, 32	48	Fever, Rash	ANK, CNK	15 → 0	29 → 11	27 → 1
Patient 4	Male, 71	12	Fever	ANK, CNK	20 → 5	120 → 2	180 → 1
Patient 5	Female, 58	72	Fever	ANK, CNK	0 → 5	27 → 36	5 → 83
Patient 6	Female, 48	5	Arthritis, Fever	ANK, CNK	10 → 0	8 → 10	2 → 1
Patient 7	Female, 74	180	Arthritis, Fever, Rash, MAS	ETN, ANK, CNK	5 → 5	33 → 1	1 → 25
Patient 8	Male, 78	180	Arthritis, Fever	ANK, CNK	40 → 20	46 → 10	58 → 40
Patient 9	Male, 22	144	Rash, Myocarditis				
Patient 10	Male, 43	24	Pleuropericarditis	ETN, TCZ, ANK, CNK	20 → 10	86 → 94	135 → 1
			Arthritis, Fever	ANK, CNK	15 → 5	15 → 10	4 → 1
			Rash				

^{*} Before and after canakinumab start.

ANK, anakinra; CNK, canakinumab; CRP, C-reactive protein; ESR, erythrocyte sedimentation rate; ETN, etanercept; MAS, macrophage activation syndrome; MTX, methotrexate; PDN, prednisone; TCZ, tocilizumab.

Conclusion: In our cohort of AOSD patients, CNK proved not only to be effective in controlling systemic inflammation in ANK refractory cases, but its use was associated with greater patients' satisfaction and tolerability.

REFERENCES:

[1] Kluetz *et al*, Value Health, 2018;21(6):742-747.

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POS1348

MORTALITY AND ITS RELATED FACTORS IN PATIENTS WITH IGG4-RELATED DISEASE: A JAPANESE SINGLE-CENTER STUDY

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Background: In recent years, IgG4-related disease (IgG4-RD) has become a widely recognized disorder. However, mortality and its related factors in this disease are not well known.

Objectives: This study aimed to clarify mortality and its related factors in patients with IgG4-RD.

Methods: We retrospectively reviewed the medical records of patients with IgG4-RD diagnosed by experts based on fulfillment of the Japanese comprehensive diagnostic criteria and/or the 2019 ACR/EULAR classification criteria for IgG4-RD at a single center in Japan. Using the collected data, we calculated the crude mortality rate and the standardized mortality ratio (SMR) using national Japan mortality statistics and investigated the cause of death. We performed Cox regression analyses to assess mortality-related factors.

Results: A total of 179 patients with IgG4-RD were included: 124 were male (69.3%); the median age was 68 years (interquartile range [IQR] 60-75 years); and the median follow-up from diagnosis was 47 months (IQR 17-84). Ten patients (5.6%) in our cohort died during the follow-up period. Five died of malignancy, one of respiratory failure, two of infectious pneumonia, one of sudden cardiac event, and one of suspected aortic aneurysmal rupture. The crude mortality rate was 11.1 per 1,000 person-years. According to national Japan mortality statistics, 11.6 age- and sex-matched deaths were expected to occur within the follow-up period, resulting in a SMR of 0.86 (95% confidence interval [CI] 0.41-1.59). Univariate Cox regression analyses indicated that the number of affected organs at diagnosis (hazard ratio [HR] 1.45, 95% CI 1.02-2.05), serum creatinine levels at diagnosis (HR 1.82, 95% CI 1.06-3.12), and the presence of malignancy during the clinical course (HR 3.93, 95% CI 1.10-14.02) had a significant impact on the time to death, whereas the other factors including age at diagnosis and serum C-reactive protein and IgG4 levels at diagnosis did not.

Conclusion: Our findings suggest that the mortality rate of patients with IgG4-RD does not significantly differ from that of the Japanese general