been shown to contribute to lupus pathogenesis. It has been suggested that LDGs have a pathogenic role in ANCA associated vasculitis (AAV) based on the data of gene expression signature in AAV and the ability of excessive neutrophil extracellular traps (NETs) formation by LDGs in AAV. However, more detailed analysis of LDG in AAV patients has not been reported.

Objectives: In this study we investigated the characteristics of LDG in AAV patients using flow cytometry and proteomics approach and examined the correlations with disease activity.

Methods: We examined the presence of LDGs in peripheral blood of 10 AAV patients before treatment and followed them for 4 months with immunosuppressive therapy. Normal-density granulocytes (NDGs) were isolated by dextran sedimentation and PBMCs were isolated by Ficoll gradient. LDGs were assessed using cell surface expression of CD14 and CD15 by flow cytometry and isolated by magnetic bead procedure from PBMCs. We performed comparative proteomic analysis among LDGs and autologous NDGs and healthy controls (HC)-NDGs.

Results: LDG frequencies were 9.8-fold higher in AAV patients before treatment relative to HC. There were two distinct populations of LDGs showing different cell surface expressions of CD10, CD14, and CD15 in AAV patients. One population of LDGs was mainly CD10 positive and another one was CD10 negative. Although the frequency of CD10 positive-LDG decreased along with decrease of disease activity, the frequency of CD10 negative-LDG increased in 4 months (17.9-fold higher than before treatment). Comparative proteomic analysis revealed these two populations of LDGs were distinct from each other and had common differences from autologous NDGs and HC-NDGs.

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Conclusions: We identified two distinct subsets of LDGs in AAV. It is possible that they play different roles in the pathogenesis of AAV.

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SAT0517

GENDER DIFFERENCES INFLUENCES CLINICAL PRESENTATION AND VASCULAR PATTERN IN PATIENTS WITH TAKAYASU ARTERITIS: AN ITALIAN MONOCENTRIC STUDY

A. Tomelleri, C. Campochiaro, S. Sartorelli, A. Cariddi, G. Cavalli, E. Baldissera, L. Dagna. *Unit of Rheumatology, Immunology, Allergy and Rare Diseases (UnIRAR), San Raffaele Scientific Institute, Milan, Italy*

Background: Takayasu arteritis (TA) is a large-vessel vasculitis affecting aorta and its major branches¹. Vascular inflammation may cause luminal narrowing, occlusion or aneurysms². TA affects more frequently young women, but occurrence in patients >40 years is not rare. Female:male ratio is 9:1³

Objectives: To compare age at disease onset, age at diagnosis and diagnostic delay between male and female Italian TA patients from our cohort. To compare symptoms presentation and vascular involvement

Methods: Data from 10 male and 104 female TA patients (fulfilling ACR criteria) from our tertiary centre were retrospectively collected. Age at diagnosis, age at first symptom onset, diagnostic delay, signs and symptoms subsequently attributed to TA, and arteries involved before the diagnosis were compared. Non-parametric statistic tests were used

Results: In male patients mean age at diagnosis was 33.5±14.64 years and mean age at onset was 25.8±13.01 years. In female patients mean age at diagnosis was 36.26±12.6 years and mean age at onset was 31±13.1 years. The differences not being statistically significant. The highest percentage of male patients disease onset was <18 years (40%), in female patients it was between 18–40 years (61.5%). Mean diagnostic delay was 92.9±80.78 months in male and 63.03

±85.63 in female patients (p=0.050). Features at disease onset are summarised in table 1. Frequency of limbs claudication was not significantly different in the two groups (50% vs 56.7%), 10% of male patients had upper limbs claudication, compared to 43.3% of female patients, p=0.047. Arterial hypertension was more frequent in male patients (70% vs 35.6%, p=0.044). Acute coronary syndrome was more frequent in male patients (30% vs 5.8%, p=0.031). Frequency of cardiovascular risk was not significantly different in the two groups (40% vs 32.7%). Coronary angiography was more frequently performed in male patients (30% vs 2.9%, p=0.009). At disease onset, compared to female patients, male patients coronary (30% vs 6.7%, p=0.042), iliac (50% vs 15.4%, p=0.018) and femoral arteries (40% vs 6.6%, p=0.008) were more frequently involved. In female patients subclavian arteries were more frequently involved (72% vs 40%, p=0.066).

Signs/symptoms	N°male patients (%)	N°female patients (%)	P-value
Constitutional symptoms	5 (50)	61 (58.7)	0.740
Arthralgias	3 (30)	28 (26.9)	1
Carotidynia	1 (10)	25 (24.0)	0.451
Syncope/vertigo	2 (20)	22 (21.2)	1
Neurological signs	1 (10)	8 (7.7)	0.576
Overall limbs claudication	5 (50)	59 (56.7)	0.746
Upper limbs claudication	1 (10)	45 (43.3)	0.047*
Lower limbs claudication	5 (50)	25 (24.0)	0.125
Arterial hypertension	7 (70)	37 (35.6)	0.044*
Discrepancy in BP	1 (10)	29 (27.9)	0.287
Reduced/absent pulses	3 (30)	46 (44.2)	0.511
Vascular bruits	4 (40)	32 (30.8)	0.723
Angina abdominis	0 (0)	16 (15.4)	0.353
Chest pain	3 (30)	9 (8.7)	0.071
Acute coronary syndrome	3 (30)	6 (5.8)	0.031*
Serositis	0 (0)	7 (6.7)	1

Conclusions: Male patients with TA have more frequent involvement of the coronary arteries and of the iliac and femoral arteries. As a consequence, they suffer more frequently acute coronary syndrome and lower limbs claudication. Besides, they have more frequently arterial hypertension

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SAT0518

THE ROLE OF LEFLUNOMIDE IN THE TREATMENT OF GIANT CELL ARTERITIS

A. Hocevar, R. Ješe, Ž. Rotar, M. Tomšič. *Dpt. of Rheumatology, University Medical Center Ljubljana, Ljubljana, Slovenia*

Background: Glucocorticoids (GC) have been the mainstay treatment in giant cell arteritis (GCA) for decades. Recently tocilizumab and abatacept have been proven to be effective alternatives to glucocorticoids. However, not all GCA patients are eligible for a biologics.

Objectives: We aimed to evaluate the role of leflunomide (LEF) as a steroid sparing agent in GCA.

Methods: This prospective observational study included newly diagnosed GCA patients followed at least 48 weeks at a single secondary/tertiary rheumatology centre.

Patients were treated with GC in line with the EULAR recommendations.¹ In short, patients with uncomplicated GCA initially received oral methylprednisolone (MP) 32–48 mg qd, while those with ischaemic complications or large vessel disease first received MP 250 mg on 3 consecutive days intravenously, followed by oral MP. MP tapering was started 2–4 weeks after treatment initiation slowly to 4 mg qd which was continued for at least 6 months. At week 12, LEF 10 mg qd was recommended as an add-on therapy to those GCA patients without contraindications for it. The final decision to add LEF was patient dependent. Follow-up visits with predetermined clinical and laboratory tests were performed 4, 12, 24, 48, 96 and 144 weeks after diagnosis. In patients who relapsed during the MP tapering unscheduled visits were arranged and treatment was adjusted (GC dose was

increased and LEF 10 mg or LEF dose increase to 20 mg or alternative steroid sparing agent in case of LEF ineffectiveness recommended). The number of relapses and a cumulative GC dose during follow-up were recorded.

Results: Between July 2014 and December 2016 we identified 76 (65.8% female, median (IQR) age 73.7 (66.1–78.8) years) new GCA cases with a follow-up of at least 48 weeks (median (IQR) 75^{51–104} weeks). 30/76 patients (39.5%) received LEF at W12 ("LEF" group), the others continued with GC only. During the follow-up 22 patients relapsed, 4 in "LEF" group (13.3%) and 18 (39.1%) in "GC only" group. The difference was statistically significant ($p=0.02$; NNT 3.9 (95%CI 2.2–17.4)). Furthermore, 17/30 GCA cases (56.7%) in "LEF" group managed to stop GC at W48 (with 1 relapse (5.9%) shortly afterwards), but none in GC only group. Patients tolerated LEF relatively well. Adverse events (AEs) were usually mild. 8/30 patients (26.7%) discontinued LEF (1 due to ineffectiveness and 7 due to one/more AEs – hair loss developed in 4/7 cases, diarrhoea in 2/7 patients, weight loss in 2/7 cases, and elevated transaminases in 1/7). The occurrence of infections requiring antibiotics and/or hospital admission was lower in LEF group compared to "GC only" group (10% vs. 26.5%).

Conclusions: We found in our prospective observational study in GCA a steroid sparing action and a rather good tolerability of LEF.

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SAT0519

INTERSTITIAL LUNG DISEASE IN ANCA-ASSOCIATED VASCULITIS PATIENTS: COMPARISON WITH IDIOPATHIC PULMONARY FIBROSIS AND INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES

A. Abri¹, M. Kwon², C. Rojas³, A. Khoo⁴, I. Mira-Avendano². ¹Rheumatology; ²Pulmonary medicine; ³Radiology; ⁴Pathology, Mayo Clinic, Jacksonville, FL, USA

Background: Patients with ANCA-positive vasculitis may develop interstitial lung disease (ILD), it is an uncommon but increasingly recognised manifestation. Clinical characteristics and prognosis are not well known in these patients. The largest report to date is from East Asia describing microscopic polyangitis (MPA) as the most common association with ILD.

Objectives: To describe the clinical manifestations and response to therapy of patients with AAV and ILd compared with patients with idiopathic pulmonary fibrosis (IPF) and interstitial pneumonia with autoimmune features (IPAF)

Methods: We conducted a retrospective 10 year chart review at the Mayo Clinic Florida, including patients with confirmed diagnoses of both AAV and ILD done by expert rheumatology and pulmonology clinical evaluations. Clinical characteristics of ANCA-ILD in 24 patients were compared to 29 patients with idiopathic pulmonary fibrosis (IPF) and 22 patients with interstitial pneumonia with autoimmune features (IPAF), which were confirmed by applying the IPF diagnostic criteria based on the most recent ATS guidelines and IPAF criteria defined by Fischer and colleagues, respectively.

Results: We identified 24 ANCA-ILD patients. 14 patients had MPA, 12 patients had granulomatosis with polyangitis (GPA), and 2 patients had eosinophilic granulomatosis with polyangitis (EGPA). 54% were female and mean age was 70. In half of the ANCA-ILD patients, vasculitis presented prior to ILD, mainly MPA, 36% of patients presented with ILD first, most of them with GPA. The rest presented with ILD and vasculitis at the same time. Usual interstitial pneumonia (UIP) was the most common radiographic pattern. Honeycombing was more common in MPA compared to GPA patients. Ground glass opacity was present in 5 (63%) of GPA and in 5 (36%) of MPA patients. Most MPA patients had positive anti-MPO antibody and p-ANCA. Only one GPA patient had positive anti-MPO antibody and two were p-ANCA positive. The majority of the GPA patients were positive for anti-proteinase-3 antibody and c-ANCA. The mainstay of treatment was corticosteroids. Rituximab was used in 14 patients. The decline in functional vital capacity (FVC) and diffusing capacity (DLCO) was most marked in IPF group, followed by ANCA-ILD and then the IPAF group (Δ FVC, -0.5 , -0.5 , and 0.3 L/s; Δ DLCO, -3.7 , -3.6 , and -0.1 , respectively). In a similar manner, survival was poorest in IPF, followed by ANCA-ILD and was best in the IPAF group.

Conclusions: This is, to our knowledge, the largest case series of clinically confirmed AAV with ILD in North America. Sizable number of GPA with c-ANCA positive patients presenting concomitant ILD is a novel observation for the clinical characteristics of ANCA-ILD that contradicts previous epidemiology of ANCA-ILD. Our data is also of value by adding prognostic information in an era of newer therapeutics, such as rituximab. In addition, the intermediate prognosis of ANCA-ILD, between IPF and IPAF, is very interesting especially after the new classification IPAF.

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SAT0520

ANTI-IL6-RECEPTOR TOCILIZUMAB IN REFRACTORY UVEITIS ASSOCIATED TO EXTRAOCULAR MANIFESTATIONS IN PATIENTS WITH BEHÇET'S DISEASE. MULTICENTER STUDY OF 11 PATIENTS

B. Atienza-Mateo¹, J.L. Martín-Varillas¹, V. Calvo-Río¹, E. Beltrán², L. Martínez³, E. Valls⁴, M. Hernandez⁵, A. Atanes⁶, M. Cordero⁷, J.M. Nolla⁸, C. Carrasco⁹, J. Loricera¹, N. Palmou-Fontana¹, E. Pons¹, M.C. González-Vela¹, R. Demetrio-Pablo¹, M.A. González-Gay¹, R. Blanco¹. ¹Rheumatology, Anatomical Pathology and Ophthalmology, Hospital Universitario Marqués de Valdecilla. IDIVAL, Santander; ²Rheumatology, Hospital General Universitario de Valencia; ³Ophthalmology; ⁴Rheumatology, Hospital Peset; ⁵Ophthalmology, Hospital General Universitario de Valencia, Valencia; ⁶Rheumatology, Hospital Universitario de A Coruña, A Coruña; ⁷Rheumatology, Hospital de León, León; ⁸Rheumatology, Hospital Universitari de Bellvitge, Barcelona; ⁹Rheumatology, Hospital de Mérida, Mérida, Spain

Objectives: To assess the efficacy of Tocilizumab (TCZ) in refractory uveitis associated to extraocular manifestations due to Behçet's disease (BD).

Methods: Multicenter study of patients with BD refractory to standard systemic treatment (conventional immunosuppressive drugs and/or anti-TNF- α agents).

Results: We studied 11 patients (7 men/4 women) (20 affected eyes); mean age 38.4 $\pm$ 20.4 years. Uveitis was bilateral in 9 patients. The pattern of ocular involvement was: panuveitis (n=8; with retinal vasculitis in 4), anterior uveitis (n=2) and posterior uveitis (n=1). Cystoid macular oedema (CME) was present in 7 patients. The clinical course was recurrent (n=7) or chronic (n=4). Apart from the visual complications, at TCZ onset the following extraocular manifestations were present: oral and/or genital ulcers (n=7), arthritis (n=4), folliculitis/pseudofolliculitis (n=4), erythema nodosum (n=2), livedo reticularis (n=1), intestinal affection (n=1), and neurological involvement (n=2).

Before TCZ, they had received systemic corticosteroids, conventional immunosuppressive drugs and biologic agents, adalimumab (n=8), infliximab (n=4), golimumab (n=3), canakinumab (n=1), or etanercept (n=1). TCZ was used in monotherapy or combined with conventional immunosuppressive drugs at 8 mg/kg/iv/4 weeks (n=10) or 162 mg/sc/week (n=1). TCZ yielded rapid and maintained improvement in all ocular parameters (TABLE). After a mean follow-up of 9.5 $\pm$ 8.05 months using TCZ, all patients experienced ocular improvement, with complete remission in 8 of them. However, TCZ was only effective in 3 of the patients with extraocular manifestations. This biologic agent had to be withdrawn in 2 cases, 1 due to a severe infusion reaction and 1 due to arthritis impairment.

Abstract SAT0520 – Table 1. Epidemiological and clinical characteristic of the sample.

Case	Sex / Age (years)	Ocular pattern and course	Extraocular manifestations	Extraocular manifestations with no response to TCZ
1	Male / 27	Bilateral posterior uveitis + unilateral CME Chronic	Oral ulcers, asymptomatic white matter lesions on MRI, arthritis, folliculitis	-
2	Female / 42	Bilateral panuveitis + unilateral CME Chronic	Oral and genital ulcers, erythema nodosum	Oral and genital ulcers, erythema nodosum
3	Male / 50	Bilateral panuveitis/papillitis + unilateral CME Relapsing	Papillitis, arthritis	-
4	Male / 35	Bilateral panuveitis + retinal vasculitis Relapsing	Oral ulcers, folliculitis	Oral ulcers, folliculitis
5	Female / 67	Bilateral panuveitis + retinal vasculitis + bilateral CME Relapsing	Livedo reticularis	Livedo reticularis
6	Male / 31	Unilateral panuveitis + retinal vasculitis + unilateral CME Relapsing	Oral and genital ulcers, folliculitis	Oral and genital ulcers, folliculitis
7	Female / 22	Bilateral panuveitis + bilateral CME Chronic	None	-
8	Male / 75	Bilateral panuveitis + retinal vasculitis + unilateral CME Relapsing	Oral and genital ulcers, arthritis, folliculitis	Oral and genital ulcers, folliculitis
9	Male / 10	Bilateral anterior uveitis Relapsing	Oral and genital ulcers, hemorrhagic stroke, erythema nodosum	Hemorrhagic stroke
10	Female / 48	Bilateral anterior uveitis Relapsing	Oral and genital ulcers, arthritis, pseudofolliculitis, erythema nodosum, intestinal involvement	Oral and genital ulcers, arthritis, pseudofolliculitis, erythema nodosum, intestinal involvement
11	Male / 16	Unilateral panuveitis Chronic	Oral ulcers, arthritis	Oral ulcers, arthritis

Abbreviations: CME, cystoid macular edema; MRI, magnetic resonance imaging; TCZ, tocilizumab.

Abbreviations:

CME, cystoid macular oedema; MRI, magnetic resonance imaging; TCZ, tocilizumab.

Conclusions: TCZ appears to be useful in highly refractory BD-related uveitis. However, there are controversial results regarding its efficacy in the treatment of extraocular manifestations of BD.

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