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.

Disclosure of Interest: None declared

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SAT0519

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

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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.

Disclosure of Interest: None declared

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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

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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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