with iRPF from four tertiary medical units in Greece and b) to evaluate factors potentially associated with disease relapse.

Methods: Medical records of patients diagnosed with RPF from 2000-2018 in four rheumatology units (Laiko, Euroclinic, Sismanoglion and Sotiria Hospitals) were retrospectively evaluated. Sixty-seven patients with iRPF were included in the study.

Results: The median age at diagnosis was 56 years (IQR:52.0–60.0), with median disease duration 8.0 years (IQR:3.0–11.0), 58% were smokers and 73% males. Patients more often presented with constitutional symptoms (57%), low back pain (63%), raised inflammatory markers (78%), anemia (43%) and compromised renal function (15%). Commonest imaging findings were periaortic-periliac mass (46%), periaortic mass (33%), periaortitis (25%) and hydronephrosis (36%) with envelopment of one (31%) or both ureters (18%). Tissue biopsy was requested in all patients, but was performed in only 10, with 3 having marked numbers of IgG4-positive plasma cells. Serum IgG4 was measured in 36/67 and 36% had elevated levels at diagnosis (median 224 mg/dL, IQR:174–328). Clinical/laboratory/radiological presentation did not differ between patients with elevated and normal serum IgG4 levels. Steroids were first-line treatment in 93% of patients. Other immunosuppressives used as steroid-sparing agents were azathioprine (70%), cyclophosphamide (19%), mycophenolate-mofetil (18%), D-penicillamine (12%) and methotrexate (8%). Relapse occurred in 19% of patients at a median of 36 months (IQR:18–66) after diagnosis with 69% of them being under therapy. Relapse did not correlate to initial imaging findings or to any treatment modality, yet patients with increased serum IgG4 tended to have higher relapse rate (26% vs 11%, $p=0.071$).

Conclusion: Diagnosis of iRPF was mostly based on imaging studies in our cohort. Steroids were used as first-line treatment. Relapse occurred in one-fifth of patients independently of initial clinical/radiographic presentation or treatment modality used. RPF patients with initially elevated serum IgG4 levels tended to have a higher relapse rate.

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Disclosure of Interests: Aliki Venetsanopoulou: None declared, Evangelia Zampeli Speakers bureau: Roshe, Astrazeneca, Sophia Christaki: None declared, Ourania Argyropoulou: None declared, Kyriaki A. Boki: None declared, Menelaos N. Manoussakis: None declared, Fotini N. Skopouli: None declared, Athanasios Tzioufas Grant/research support from: ABBVIE, PFIZER, AMGEN, NOVARTIS, GSK, Haralampos M. Moutsopoulos: None declared

DOI: 10.1136/annrheumdis-2019-eular.4231

FRI0298

ABNORMALITY OF PERCENTAGES AND ABSOLUTE NUMBERS OF CD4+ T SUBSETS IN PATIENTS WITH ANCA-ASSOCIATED VASCULITIS AND ITS CORRELATIONS WITH CLINICAL INDICATORS

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Background: ANCA-associated vasculitis (AAV) is a heterogenous autoimmune disease with unknown etiology [1-2]. During the last decade, a panel of CD4+ T subsets have been identified. However, the exact role and quantitative status of these subsets in AAV patients remains unclear. **Objectives:** We therefore investigated these T cell subsets in AAV patients.

Methods: AAV patients ($n = 54$) and healthy controls (HCs) ($n = 19$) were enrolled. Of them, ten patients initially presenting with active disease were assessed again after remission was achieved. In addition, 38 patients were renal vasculitis. Proportions and absolute numbers of peripheral CD4+ T cell subsets and expression of multiplex cytokines were determined by flow cytometry (FCM). Correlations of clinical indicators with the CD4+ T cell subsets were systematically analyzed.

Results: Percentages of naive T cells (TN) ($p<0.001$), terminally differentiated effector (TEMRA) T cells ($p=0.027$) and activated Treg cells (aTreg) in AAV patients were decreased, but those of effector memory T-cell

subpopulation (TEM) ($p<0.001$), regulatory T cell (Treg) cells and FoxP3lowCD45RA- T cells were increased. Similar results were observed when we compared absolute numbers of the above corresponding cells in AAV patients and HCs, except TEM. Furthermore, the percentage of aTreg ($p=0.043$) was decreased while that of Th17 cells ($p=0.027$) was increased in renal vasculitis patients. A significant correlation was observed between the ratio of Th17 to Treg subset and creatinine or BUN, as well as the ratio of Th17/aTreg was significantly increased in active and renal vasculitis patient. In addition, we found that cytokine IL-2 and IL-4 exhibited a downward while IL-6, IL-10, TNF, IFN- γ and IL-17A trend upward in AAV patients.

Conclusion: There were abnormally quantitative changes in CD4+ T subsets and cytokines in AAV patients, especially the decrease in the relative and absolute number of aTreg (activated Tregs), which indicates an imbalance of pro- and anti-inflammatory T cells. These T subsets might be associated with the ANCA-related autoimmune response and can be used as diagnosis markers for disease activity and as targets for potentially powerful therapy of AAV.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.4667

FRI0299

THE PREVALENCE OF NON-VASCULAR PULMONARY MANIFESTATIONS IN PATIENTS WITH TAKAYASU' ARTERITIS: A RETROSPECTIVE MULTI-CENTERED COHORT STUDY

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Background: Takayasu' arteritis (TAK) is a rare vasculitis characterized by inflammation and obliteration of intermediate to large-size arteries. Even though more than 50% of patients with TAK have pulmonary artery involvement, non-vascular involvement and symptoms are uncommon.

Objectives: We aimed to investigate the frequency of non-vascular pulmonary involvement in TAK.

Methods: We assembled a retrospective cohort of patients with TAK from six different centers in Turkey. The demographics, clinical characteristics, treatment and outcomes of patients were abstracted from medical records, and the computed tomography findings were evaluated for pulmonary manifestations.

Results: As of January 2019, 197 TAK patients were recruited (mean age: 42.7±13.9 years [min-max: 17-75]) to the cohort, and 88.3% of them were female. Twenty-four patients had cough and/or dyspnea and four had hemoptysis as pulmonary symptoms. In CT assessment, parenchymal infiltrations were present in four (2%), pleural effusion in five (2.5%), nodule/cavity in one (0.5%), and pulmonary hemorrhage in one patient (0.5%). The patient who had pulmonary hemorrhage had also pleural effusion at the same time. In the whole cohort, 11.2% of patients ($n=22$) had pulmonary hypertension (PAH), three of them had cough and/or dyspnea and four of them had hemoptysis as a pulmonary symptom. Among patients with PAH, any pulmonary involvement in CT was more frequent compared to the rest of the patients (22.7% vs 5.1%, $p<0.0001$) (Table 1).

Conclusion: In this first assessment of Turkish TAK cohort, we observed non-vascular pulmonary involvement in about 5% of our patients and half of them were pleural effusions. The second most common manifestation was parenchymal infiltration with a frequency of 2%. Although rare, non-vascular pulmonary manifestations should also be investigated in TAK patients, especially in patients with pulmonary hypertension.