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-periiliac 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 IgG_4 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 IgG_4 levels tended to have a higher relapse rate.

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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 CD^+ 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 AVV 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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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.

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Table 1. The frequencies of non-vascular pulmonary manifestations in patients with Takayasu' arteritis

n (%)	All Patients n=197	Patients with PAH n=22	p
Symptoms	24 (12.2)	3 (13.6)	0.049
Cough/Dyspnea	4 (2)	3 (13.6)	0.000
Hemoptysis			
Pulmonary involvement in	10 (5.1)	5 (22.7)	0.000
CT	4 (2)	1 (4.5)	0.000
Pulmonary infiltrates	1 (0.5)	1 (4.5)	0.000
Nodules/cavities	1 (0.5)	1 (4.5)	0.000
Pulmonary hemorrhage	5 (2.5)	2 (9.1)	0.000
Pleural effusion			

(PAH: pulmonary arterial hypertension, CT: computed tomography)

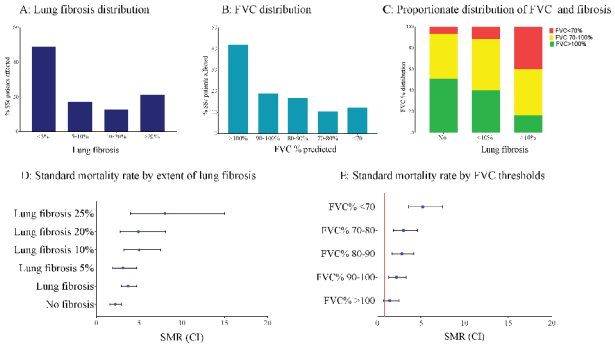
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Scleroderma, myositis and related syndromes

FRI0300 IMPACT OF INTERSTITIAL LUNG DISEASE IN SYSTEMIC SCLEROSIS IN A COMPLETE, NATIONWIDE COHORT

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Background: Interstitial lung disease (ILD) represents a clinical challenge in systemic sclerosis (SSc) and associates with high mortality. The presence of severe lung fibrosis is a strong predictor for early mortality. There is substantial progress in SSc-ILD research, but precise, population-based data on cumulative incidence, range of severity and predictive value of clinical risk factors are lacking. Such data are vital for clinical decision making, and highly warranted as background information for appropriate development of screening and management strategies for SSc-ILD.
Objectives: To assess cumulative incidence of ILD, range of ILD severity and mortality risk predicted by baseline pulmonary function tests (PFT) and ILD extent by CT in a complete, nationwide SSc cohort.
Methods: The Norwegian SSc cohort study (Nor-SSc) includes all the 630 incident and 185 prevalent SSc patients from 2000-2012 meeting SSc classification criteria. A baseline PFT was recorded in 703 (86%) patients, and 650 (80%) had high resolution computed tomography (HRCT) images available for analyses. Extent of fibrosis was scored on 10 sections from every HRCT and expressed as percentage of total lung volumes. For the survival and mortality analyses, all Nor-SSc patients diagnosed from 2000-2012 (the 630 incident cases) were included and compared with 15 age- and gender matched controls per patient drawn from the national population registry. Vital status was available for all patients and controls at study end (January 2018). Descriptive statistics and standardized mortality rates (SMR) were estimated.
Results: Of the 815 patients in the total Nor-SSc cohort, 682 (84%) were female and 629 (77%) had limited cutaneous SSc. Mean age at SSc diagnosis was 53 yrs, with mean time from SSc onset to diagnosis of 3.8 yrs. We observed ILD on HRCT in 324/650 patients (50%), and the majority of these had <5% lung fibrosis (Figure 1A). Mean FVC at baseline was 94% of expected value, and nearly half of the patients (42%) had an FVC>100% (Figure 1B). Proportionate distribution of FVC values in patients with no lung fibrosis, <10% lung fibrosis of total lung volume and >10% lung fibrosis is shown in Figure 1C. During the mean 8.6 yrs observation period of this study, 148 of the 630 incident SSc patients died, corresponding to an overall SMR of 2.4. Separate analyses of the 650 patients with baseline HRCT data showed that the SMR correlated with presence and extent of lung fibrosis, from SMR 2.2 in patients with no fibrosis to SMR 8 in patients with >25% lung fibrosis (Figure 1D). Correspondingly, we found that the SMR changed across patient groups stratified by baseline FVC%, with increased mortality evident already in the FVC 90-100% group (Figure 1E).
Conclusion: The results from this population based SSc cohort study provide new, unbiased data regarding the impact of ILD. Our results indicate a dose-response relationship between lung fibrosis extent and SMR; and between FVC% and SMR. Importantly, this relationship was evident even in groups with limited lung fibrosis and groups with normal range FVC%, strongly suggesting that all SSc patients should be screened with PFT and HRCT at baseline, to diagnose ILD early and tailor further management.



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FRI0301 GASTROINTESTINAL ADVERSE EVENTS IN PATIENTS WITH SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE (SSC-ILD) TREATED WITH NINTEDANIB: DATA FROM THE SENSCIS TRIAL

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Background: In patients with idiopathic pulmonary fibrosis (IPF), nintedanib has a manageable adverse event (AE) profile characterised predominantly by gastrointestinal (GI) events. The efficacy and safety of

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