Background: Interstitial Lung Diseases (ILD) may present features suggesting an underlying autoimmune process, which seem to differentiate them from idiopathic interstitial pneumonias, although without fully meeting the classification criteria (CC) for a specific connective tissue disease. Different terms had been used to describe these conditions and, to reach a consensus, the European Respiratory Society/American Thoracic Society proposed the CC for an entity named Interstitial Pneumonia with Autoimmune Features (IPAF). Clinical evolution and prognosis of this entity are still poorly understood.

Objectives: To evaluate clinical evolution and prognosis of a population of patients with IPAF.

Methods: Retrospective analysis of clinical files of patients followed by the Pulmonology Department since 02/2012 until 06/2019, who met the CC for IPAF regarding clinical, functional and radiological evolution. Patients were considered to have a progressive phenotype in 24±3 months from their 1st evaluation if they fulfilled 1 of the 4 criteria: relative decline in FVC ≥10% predicted; relative decline in FVC ≤5–10% predicted and worsened respiratory symptoms; relative decline in FVC ≤5–10% predicted and increased effort of breathing on High-resolution Computed Tomography (HRCT); worsened respiratory symptoms and increased extent of fibrosis on HRCT.

Results: 22 (7.4%) of 296 ILD patients met IPAF CC. 50.9% were female with a median age of 1 at the 1st evaluation of 66.7±12.4 years. They were all non-smokers (53.6%) or ex-smokers (36.4%). Serologic and morphologic criteria were both present in 21 (95.4%) and clinical criteria in 5 patients (22.7%). Antinuclear antibodies (ANA) were identified in 19, rheumatoid factor in 4, SAA in 3 and anti-jo-1 in 1 patient. HRCT patterns were identified in 21 patients: 15 nonspecific interstitial pneumonia (NSIP), 5 organizing pneumonia (OP) and 2 lymphocytic interstitial pneumonia (LIP). One NSIP and 1 LIP identified on HRCT were confirmed by histopathology. Three patients had inflammatory arthritis and 2 had Raynaud’s phenomenon. Immunosuppressive therapy was introduced in most cases (18 patients, including systemic corticotherapy in 17, azathioprine in 4, mycophenolate mofetil in 1), azithromycin was prescribed in 2 patients and 3 remained without therapy. Regarding the follow up at 24±3 months from the 1st evaluation (3 patients were excluded due to too recent follow-up), 4 patients (18.2%) had progressive phenotype, 7 (31.8%) had a favourable evolution and 3 (13.6%) patients had died. During a follow-up of 31.1±19.8 months, this number rose to 6 patients (27.3%), all of them died by respiratory cause and had NSIP pattern. No differences were found in age, last FVC, therapy and time of disease evolution between those who died and the others.

Conclusion: Our study showed that a small proportion of IPAF patients had a progressive phenotype and the NSIP pattern seemed to be a poor prognosis factor for survival.

References:

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FRI0497 THERAPEUTIC STRATEGIES AND LONG-TERM OUTCOME IN PATIENTS WITH INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES: A SINGLE CENTER LARGE-SCALE OBSERVATIONAL COHORT STUDY

O. Murata1,2, K. Suzuki1, N. Sasaki1, T. Takeuchi1, M. Maemondo2,1 Keio University School of Medicine, Division of Rheumatology, Department of Internal Medicine, Tokyo, Japan; 1Wata Medical University School of Medicine, Division of Pulmonary medicine, Allergy, and Rheumatology, Department of Internal Medicine, Morioka, Japan

Background: Patients with idiopathic interstitial pneumonia (IIP) may have features of connective tissue diseases (CTDs). The term interstitial pneumonia with autoimmune features (IPAF) has been recently proposed for such patients [1]. To date, only few studies have comprehensively described outcomes over a long-term period and choices of treatment [2-4].

Objectives: The aim of this study was to investigate the therapeutic strategies and long-term outcome among patients with IPAF, IIP, and CTD-ILD.

Methods: Six-hundred and seventy-two patients who had visited our department between April 2009 and March 2019 and were evaluated by chest HRCT scan. They were clinically and radiologically diagnosed as having interstitial lung disease (ILD), including IIP, CTD-ILD, and undifferentiated connective tissue diseases associated ILD or other AD. Then, we applied IPAF criteria to these patients, 68 patients were diagnosed as IPAF. We extracted the patients classified by ACR/EULAR 2010 criteria and SLE patients classified by ACR/EULAR 2019 criteria. SLE and RA patients were compiled consecutively from a rheumatology clinic of the Regional University hospital of Malaga. Controls: subjects without rheumatologic autoimmune diseases (AD) from the same population area. Protocol: All subjects filled out a pre-designed questionnaire for the collection of polyautoimmunity data on the cut-off date. Main variables: polyautoimmunity was defined as co-occurrence of SLE or RA and other AD. Secondary variables: Rheumatologic, cutaneous, endocrine, digestive and neurological AD. MAS was defined as presence of three or more AD. Family history of SLE, RA and other autoimmune diseases were also collected. Statistical analysis: descriptive analysis, bivariate analysis and multivariable analysis were done. (Dependent variable: Polyautoimmunity).

Results: We recruited 109 patients with RA, 105 with SLE and 88 controls. Fifteen patients with RA (18.3%), 43 with SLE (41%) and 2 controls (2%) reported polyautoimmunity. Table 1 describes the epidemiological characteristics, comorbidities and polyautoimmunity in study population. The most frequent AD associated with RA was Sjögren’s syndrome (SS) (53.3%) and SS (55.8%) followed by the antiphospholipid syndrome (30.2%) were associated with SLE. Hashimoto’s thyroiditis and pernicious anemia were the next most frequent AD. According to family history, 5 patients with RA (33.3%) and 12 with SLE (27.9%) had a family history of first degree of other AD. Obesity was associated with polyautoimmunity in RA (OR = 3.362, p = 0.034). In SLE, joint damage (2.282, p = 0.039) and anti-RNP antibodies (OR = 5.095, p = 0.028) were factors associated with polyautoimmunity and taking hydroxychloroquine was a protective factor (OR = 0.190, p = 0.004).

Conclusion: Polyautoimmunity in RA and especially in SLE is frequent. It was associated with obesity in RA and in SLE with joint damage and anti-RNP antibodies. The hydroxychloroquine appeared as a protective factor.

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DOES TESTING FOR SAA IS MORE BENEFICIAL THAN CRP FOR THE FOLLOW-UP OF FMF PATIENTS WITH M694V HETEROZYGOUS OR M694V HOMOZYGOUS MUTATIONS?

M Oztas1, S. Ugur1, O. Selvi1, B. Ergezen1, H. Ozdogan1,1 Istanbul University-Cerrahpasa, Department of Medicine, Division of Rheumatology, Istanbul, Turkey; 1Okmeydani Training and Research Hospital, Istanbul, Turkey

Background: In order to follow subclinical inflammation and adjust the therapy for an optimal Familial Mediterranean Fever (FMF) disease control, clinicians seek for readily accessible, affordable and reproducible markers. C-reactive protein (CRP) is widely used for this purpose. Some suggest that CRP measures are not conclusive in all cases, especially at initial stages of inflammation. It is suggested that Serum Amyloid A (SAA) may be more reliable and sensitive in predicting an ongoing inflammation.

Objectives: In order to evaluate and to compare the sensitivity of SAA and CRP in FMF patients with M694V homozygous and M694V heterozygous mutations respectively.

Methods: Blood samples from 28 patients with M694V homozygous mutation and from 15 patients with M694V heterozygous mutation were obtained during a mean follow-up of 1 year. Multiple samples were drawn in both attacks and attack-free periods of FMF (153 from M694V homozygous and 31 from M694V Heterozygous). For the analysis of the correlation, the folds of normal CRP and SAA levels were used. Serum levels of the given markers were measured with nephelometric kits (normal CRP levels <5 mg/L and SAA levels <8 mg/L). More than one-and-a-half-fold increase of CRP and SAA was defined as an active inflammation.

Results: Except in one patient, all patients in the whole cohort were on prophylactic colchicine. Among 28 patients with M694V homozygous mutation, one patient was treated with adalimumab, and 12 patients with anti-IL-1 regimens. Among 15 patients with M694V heterozygous mutation, 4 were under anti-IL-1 treatment. There were a total of 183 measurements of CRP and 283 measurements of SAA from 43 patients. Twenty-three measurements were obtained during the attack period in M694V homozygous group and the remaining 160 measurements were obtaine in attack-free period. The figure demonstrates the correlation between CRP and SAA results (r=0.745, p<0.001). Both acute phase reactants were increased in 69 measurements, while in 13, CRP was high but SAA was normal and in 31, SAA was high however CRP was within normal limits. The mean increase in CRP of the whole cohort was 2.37 ± 3.22-fold of the normal, whereas mean increase in SAA was 6.77 ± 13.23-fold of the normal.

Conclusion: According to these results, serial testing of SAA does not provide any additional advantages over CRP. As it is readily accessible and affordable, CRP seems to be sufficient for the follow-up of FMF patients.

Figure: