**FRI0495**

**FOLLOW UP OF INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES – THE EXPERIENCE OF ONE CENTRE**

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**Background:** Interstitial Lung Diseases (ILD) may present features suggesting an underlying autoimmune process, which seem to differ from those in idiopathic interstitial pneumonias, although without fully meeting the classification criteria (CC) for a specific connective tissue disease. Different terms have 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 extent of fibrosis 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. 59.0% were female with age at the 1st evaluation of 66.7±12.4 years. They were all non-smokers (53.6%) or ex-smokers (36.4%). Serological 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, SjS 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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**FREQUENCY OF POLYAUTOIMMUNITY IN RHEUMATOID ARTHRITIS AND SYSTEMIC LUPUS ERITEMATOSUS**

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**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 a 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 outcomes among patients with IPAF, IIP, and CTD-ILD.

**Methods:** Six hundreds and seventy-two patients who had visited our department between April 2009 and March 2019 were evaluated by chest HRCT scan. They were clinically and radiologically diagnosed as having interstitial lung disease (ILD), including IIP, CTD-ILD, undifferentiated connective tissue diseases associated ILD or other ILD. Then, we applied IPAF criteria to these patients, 68 patients were diagnosed as IPAF. We extracted the

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**FRI0497**

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

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**Background:** Interstitial Pneumonia with Autoimmune Features (IPAF) have been recently proposed for such patients [1]. To date, only a 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 outcomes among patients with IPAF, IIP, and CTD-ILD.

**Methods:** Six hundreds and seventy-two patients who had visited our department between April 2009 and March 2019 were evaluated by chest HRCT scan. They were clinically and radiologically diagnosed as having interstitial lung disease (ILD), including IIP, CTD-ILD, undifferentiated connective tissue diseases associated ILD or other ILD. Then, we applied IPAF criteria to these patients, 68 patients were diagnosed as IPAF. We extracted the

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FRI0498 DOES TESTING FOR SAA IS MORE BENEFICIAL THAN CRP FOR THE FOLLOW-UP OF FMF PATIENTS WITH M694V HETEROZYGOUS OR M694V HOMOZYGOUS MUTATIONS?

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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 <6,8 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 31 from 43 patients. Twenty-three measurements were obtained during the attack period in M694V homozygous group and the remaining 160 measurements were obtain in attack-free period. The figure describes 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.

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Figure:

P<0,001, n=0,745

Figure. The folds CRP and SAA in whole M694V homozygous and heterozygous mutant population

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