ASSOCIATION BETWEEN INTERSTITIAL LUNG DISEASE AND RHEUMATIC DISEASE: IMPLICATIONS IN CLINICAL PRACTICE

Vera Ortiz-Santamaría, Noeıl Reguart Otó, Pau Sendra de Dios, Noemí Busquets, Maria Pascual, Andrés Ponce, Xavier Suris, Yolanda Galea, Anna Mola, Marta Torrella, Julia Tarrega, Enric Barbé. "Granollers General Hospital, Rheumatology, Granollers, Barcelona, Spain; 2Granollers General Hospital, Internal Medicine, Granollers, Barcelona, Spain; 3Granollers General Hospital, Family and Community Medicine, Granollers, Barcelona, Spain; 4Granollers General Hospital, Pulmonaty, Granollers, Barcelona, Spain.

Background: The relationship between interstitial lung disease (ILD) and rheumatic diseases is well known. Recently, a new clinical entity has been described that relates ILD with autoimmune findings, IPAF (Interstitial Pneumonia with Autoimmune Features), which allows identifying and condensing non-cataloged diseases with pulmonary and rheumatic involvement.

Objectives: Our aim is to establish which patients with ILD have a rheumatic disease, and which of them meet the IPAF criteria. We analyzed the role of nailfold capillaroscopy in these patients.

Methods: This is a prospective descriptive observational study. The observation period was 2 years (2016-2018), in a county university hospital. Patients diagnosed with ILD of unknown etiology, derived from Pneumology Service, were included. A medical history was obtained focused on autoimmune disease, rheumatological evaluation, rheumatological blood markers and nailfold capillaroscopy, in order to establish whether they had a concomitant rheumatic disease.

Results: Thirty patients with ILD were evaluated, with a mean age of 70.5 years (53 - 88). Of the 30 patients, 12 (40%) had usual interstitial pneumonia (UIP), 11 (36.67%) had organizing pneumonia (OP) and 7 (23.33%) had nonspecific interstitial pneumonia (NSIP). Eleven (36.67%) of the 30 patients had an altered capillaroscopy, 6 of them (54.5%) had a rheumatic disease. In our sample, 5 cases meet diagnostic IPAF criteria, 4 OP and 1 UIP, three of them with altered capillaroscopy.

Conclusion: In our serie, one third of the patients were diagnosed with rheumatic disease associated with interstitial lung disease. Of the thirty evaluated patients, 36.67% had altered capillaroscopy, 54.5% presenting concomitant rheumatic disease. Following the IPAF classification criteria, we obtained 5 cases in our sample: four OP and one UIP, 3 of them with altered capillaroscopy. It is important to be aware of this association with a multidisciplinary approach for the adequate diagnosis and follow-up of these patients.

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EXPERIENCE WITH BIOLOGIC AGENTS FOR THE TREATMENT OF CARDIAC SARCOIDOSIS IN A U.S. ACADEMIC MEDICAL CENTER

Anjani Pillarisetty1, Mithun Devraj2, Farooq Sheikh3, Florina Constantinescu1.
1MedStar Washington Hospital Center, Department of Rheumatology; 2MedStar Washington Hospital Center, Department of Internal Medicine, Washington, United States of America; 3MedStar Washington Hospital Center, Advanced Heart Failure, Washington, United States of America.

Background: Sarcoidosis is a multisystem granulomatous disease of unclear etiology characterized histologically by non-caseating granulomas. Lungs are the most common organs affected but sarcoidosis can affect almost any organ system. While clinically manifest cardiac involvement occurs in only about 5% of patients with sarcoidosis, a significant proportion have clinically silent disease. Symptomatic cardiac involvement portends a poorer prognosis with manifestations varying from heart failure and conduction abnormalities to ventricular arrhythmias including sudden death. Immunosuppression with corticosteroids and DMARDs such as methotrexate and mycophenolate mofetil has been the mainstay of treatment despite a paucity of data. There is a subset of patients that are either non-responders to these agents or in whom the side effect profile is prohibitive for their long term use. Biologic agents, mainly TNF alpha antagonists, have been used as salvage therapies in these patients. However, the evidence regarding their efficacy and safety is limited to a few case reports. In fact, there remains much apprehension regarding the use of TNF alpha antagonists in patients with systolic heart failure due to concerns that they can exacerbate heart failure.

Objectives: To study the efficacy and safety of using biologics for the treatment of cardiac sarcoidosis.

Methods: We conducted a retrospective and prospective observational study of all adult patients with cardiac sarcoidosis treated with biologics at an academic medical center in Washington D.C, USA between 2013 and 2018.

Results: We identified 9 patients (3 men and 6 women) diagnosed with cardiac sarcoidosis at our institution. The mean age at diagnosis was 49.9 (SD 8.6). 1 patient was Caucasian and the rest (n=8) were African American. Lungs were the most common extra cardiac organ involved (n=7) followed by CNS (n=4), liver (n=4) and skin (n=3). 5 of the patients presented with systolic heart failure (EF<50%), 3 with atrial and
ventricular arrhythmias and 1 was found to have incidental abnormal myocardial uptake on PET imaging. 8 of the 9 patients had abnormal myocardial uptake on PET imaging. All but 1 patient had been initially treated with oral steroids (1 refused) and 7 of the 9 patients had also been given oral DMARDs; methotrexate (n=6), azathioprine (n=2), hydroxychloroquine (n=2) and mycophenolate mofetil (n=1). Biologics used were adalimumab (n=5), infliximab (n=3) and rituximab (n=1). The most common indication for biologics was progression of disease despite optimal doses of standard therapy, followed by intolerance or contraindication to standard therapy. 72% of the patients were noted to have marked clinical improvement with the addition of a biologic. 4 out of 9 patients had decreased myocardial uptake on PET following treatment with a biologic. One patient had no change on PET and 4 have not had repeat imaging done yet. None of the patients had worsening of left ventricular systolic function with the addition of a TNF alpha antagonist. There were no reported major infections or significant adverse events that were attributable to the use of biologics.

Conclusion: Based on our small cohort, biologics (mainly TNF alpha antagonists) appear to be safe and efficacious as salvage therapy for cardiac sarcoidosis. However, there is a need for prospective studies to further validate these findings as well as to identify the subset of patients that would benefit from early initiation of these therapies.

REFERENCES: