THE MANAGEMENT OF INTERSTITIAL LUNG DISEASES: THE IMPORTANCE OF THE RHEUMATOLOGIC EXPERTISE IN MULTIDISCIPLINARY MEETINGS

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Background: Multidisciplinary Team (MDT) meetings are the current gold standard in interstitial lung disease (ILD). Descriptive studies report that respiratory physicians, radiologists and pathologists are involved in MDT. A rheumatologist is not routinely involved in MDT even if up to 20% of ILD are related to connective tissue disease, rheumatoid arthritis or systemic vasculitis.

Objectives: The aim of this study is to evaluate the prevalence and predictors of systemic rheumatologic diseases in a cohort of patients with ILD, evaluated by a rheumatologist on the advice of MDT in a university hospital.

Methods: Thirty-two patients with ILD, evaluated at dedicated MDT were referred to a rheumatologist in 2018, usually for autoantibodies positivity or clinical history suspected for a rheumatological disorder. Rheumatological evaluation included physical examination, routine blood and urine tests, serum levels of C3 and C4, ANA, Rheumatoid Factor (RF), ANCA, anti-Sm, anti-RNP, anti-Ro/SSA, anti-La/SSB, anti-Jo1, anti-dsDNA and anti-CCP antibodies. Family history of autoimmune diseases, presence of rheumatologic red flags (Raynaud phenomenon, photosensitivity, inflammatory skin manifestations, sicca syndrome, recurrent fever, inflammatory arthralgias, paresthesias, oral or genital aphthosis, thrombosis and recurrent miscarriages), respiratory symptoms and pulmonary function test were also evaluated. When indicated, capillaroscopy, joint imaging and salivary gland biopsy were performed.

Results: Twenty-one patients (65.6%) were female. At the time of rheumatological evaluation, the patients had a mean age of 64.4±12.6 years with a mean period of 21.1±39.6 months since the first identification of ILD on a CT scan. Twenty-five (78.1%) and 18 (56.3%) patients respectively complained exertional dyspnea and cough, 6 patients (18.8%) were on oxygen therapy and 13 (40.6%) had a FVC<80%. The pattern on CT scan suggested a UIP, NSIP and OP respectively in 18 (56.3%), 6 (18.8%) and 2 (6%) patients. Ten patients (31.3%) were diagnosed with a defined rheumatological condition (4 with Sjögren Syndrome, 3 with scleroderma, 1 with Rheumatoid Arthritis, 1 with Mixed Connective Tissue Disease and 1 with Granulomatosis with Polyangiitis); 7 (21.9%) were diagnosed with Interstitial Pneumonia with autimmune feature (IPAF). A rheumatological diagnosis was not statistically associated with any specific rheumatologic red flag, routine laboratory abnormalities or family history of systemic autoimmune disease. Three out of 13 capillaroscopies and 3 out of 9 salivary glands biopsies performed had diagnostic findings. The only immunologic abnormalities associated to a rheumatological diagnosis were RF positivity (20.0%±54.5%, p=0.024) and p-ANCA positivity (10.0%±40.9%, p=0.042), both with low specificity (respectively 15.0% and 14.3%) and sensitivity (respectively 55.6% and 42.9%). After diagnosis, 11 patients (34.4%) started immunosuppressive therapy and 7 (21.9%) started biologic therapy.

Conclusion: Single clinical or laboratory abnormalities are not strongly associated to a rheumatological diagnosis in patients with ILD, suggesting that only a comprehensive rheumatological evaluation allows correct classification of the disease associated with ILD and is mandatory to make an accurate diagnosis. In this study, we evaluated only preselected patients by ILD-MDT, but the results indirectly suggest that direct participation of rheumatologist to MDT is advisable to increase accuracy and reduce delay in diagnosis and treatment.

Disclosure of Interests: Enrico De Lorenzis: None declared, Gerlando Natalello: None declared, Lucrezia Verardi: None declared, Silvia Laura Bosello: None declared, Elisa Gremese: Consultant for: AbbVie, BMS, Celgene, Janssen, Lilly, MSD, Novartis, Sanofi, UCB, Roche, and Pfizer.

Conclusion: CD26 is a potential novel histological marker of idiopathic inflammatory myopathies

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Background: Idiopathic inflammatory myopathies (IM) are a heterogeneous group of acquired skeletal muscle disorders including polymyositis (PM), dermatomyositis (DM) and immune-mediated necrotizing myopathy (IMNM). They are characterized by immune-mediated muscle damage. Activated T cells are the predominant inflammatory infiltrates in muscle biopsies of PM and DM patients and the lack of T regulatory cells (Treg) has been implicated in the persistence of muscle damage. CD26 is an intrinsic membrane glycoprotein and a serine exopeptidase involved in the activation of T lymphocytes and amplification of inflammatory cytokines production. The enzymatically active form of CD26 is selectively expressed by activated T cells and has been described as a negative selection marker for human Treg.5-7

Objectives: The aim of this study was to evaluate the expression of CD26 in muscle biopsies of IM patients and to correlate it with patients' clinical and histological features.

Methods: Immunofluorescence was used to evaluate CD26 expression and co-localization with CD3 and CD31, markers of T cells and endothelial cells respectively, in muscle biopsies of 6 DM, 6 PM, 3 IBM and 3 IMNM patients and of 6 healthy controls.

Results: We found that CD26 is preferentially expressed in muscle biopsies of IM patients with respect to controls and that its level of expression is higher in DM patients. In muscle biopsies of IM patients, CD26 is distributed not only in the extracellular matrix surrounding myofibers and infiltrating leukocytes, but also at the level of T cell membranes and endothelial cells. Specifically, CD26 co-localization with CD3 is more prominent in DM muscle biopsies. We could not find any association between vessel morphology in terms of size and shape and CD26 endothelial expression, suggesting that CD26 is expressed at the perivascular level independently of the degree of vessel dysfunction. With regard to clinical features, we found that CD26 was more expressed in patients presenting the typical DM rash. Moreover, CD26 expression was found not to be significantly associated with the degree of muscle weakness nor with the presence of interstitial lung disease, dysphagia, myalgias or mechanic's hands. As for histological data, higher levels of CD26 expression were found in biopsies with perivascular inflammatory infiltrates, especially T lymphocytes and macrophages.

Conclusion: Our data suggest that CD26 may represent a suitable marker for the diagnosis of IM and a potential novel target for selective immune-therapies.

Disclosure of Interests: Rebecca De Lorenzo Consultant for: AbbVie, BMS, Celgene, Janssen, Lilly, MSD, Novartis, Sanofi, UCB, and Pfizer.

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Disclosure of Interests: Rebecca De Lorenzo Grant/research support from: Adienne Pharma & Biotech, Clara Sciorati: None declared, Antonella Monno: None declared, Francesco Bonomi: None declared, Silvia Cavalli: None declared, Antonio Francesco Di Naro: None declared, Stefano Privital: None declared, Patrizia Rovere-Querini Grant/research support from: Adienne Pharma & Biotech


AB1138

QUANTIFYING KNEE JOINT EFFUSIONS WITH CLINICAL TESTS, MUSCULOSKELETAL ULTRASOUND AND SYNOVIAL FLUID ASPIRATION: A PROSPECTIVE COHORT STUDY

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Background: Aspiration of knee joint effusions is an integral diagnostic and therapeutic intervention in many rheumatologic diseases. Clinical examination has traditionally involved tests including the “patella tap” or “bulge test”. The accuracy of these tests for determining effusion presence and size is not well established. Musculoskeletal ultrasound (MUS) is considered better for identification and quantification of knee effusions.

Objectives: To investigate the correlation between both clinical examination and MUS to aspirated knee effusion volume.

Methods: We performed a prospective cohort study of 37 osteoarthritis patients with symptomatic knee effusions. Clinical assessment with patella tap, bulge test and knee circumference measurement were carried out. MUS was used to measure effusion depth in the suprapatellar, lateral and medial parapatellar views. All knee effusion aspirations were performed by the same experienced clinician using a consistent, lateral approach. Linear regression analysis was used to assess correlations between clinical tests, MUS and aspiration volume.

Results: In patients with >3ml of fluid aspirated, patella tap and bulge test were positive in 67% and 80% respectively. The positive predictive value for bulge test was 80%. Where larger volumes were aspirated (i.e. >10ml), patella tap and bulge test were only positive in 52% and 65% respectively. There was a significant correlation between the measured circumference of the index and non-index knee and aspiration of fluid (coefficient=5.6, p=0.007). The relationship between fluid depth on MUS and aspirated volume showed a trend towards statistical significance, with a depth of 1mm equating to 1.57 ml of fluid (coefficient=1.57, p=0.06).

Conclusion: This pilot study demonstrates that a positive patella tap or bulge test is moderately predictive of knee effusion volume. However, this association is weaker when larger knee effusions are present. MUS showed promise at accurately predicting knee effusion volume. A larger study is underway to assess this relationship further.

Disclosure of Interests: Thilinie De Silva: None declared, Chris Pham: None declared, Adam Rischin: None declared, Kim Le Marshall: None declared, Sara Vogrin: None declared, Albert Leung: None declared, Keith Lim Consultant for: Advisor for role of hepatits and TB in Cimzia, UCB, Speakers bureau: Role of biological in pregnancy UCB

Disclosure of Interests: None declared


AB1137

BONE SARCOIDOSIS: USEFULNESS OF 18F-FDG PET/CT

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Background: Bone sarcoidosis is usually rare but more sensitive imaging procedures such as 18F-fluorodeoxyglucose Positron Emission Tomography/Computed Tomography (18F-FDG PET/CT) allow a better characterization of such lesions. We aimed to describe bone sarcoidosis involvement using 18F-FDG PET/CT.

Methods: We performed an observational retrospective study of patients with pulmonary sarcoidosis having a 18F-FDG PET/CT. As stated by ATS/ERS/WASOG criteria, diagnosis of sarcoidosis was established on the presence of clinical symptoms and/or imaging features of sarcoidosis, and evidence of non-caseating epithelioid granuloma in a biopsy sample after exclusion of other known etiology of granuloma. We assessed clinical and 18F-FDG PET/CT characteristics.

Results: A total of 85 patients (56.5% of female, median age 47 years) with sarcoidosis were analyzed. The median of disease follow-up was 4 years. Sarcoidosis occurred in more than three organs among 66% of cases. Using ATS/ERS/WASOG criteria, bone sarcoidosis was diagnosed in 12 (14%) patients. Spine was the most commonly affected bone (92%), followed by pelvis (67%), sternum (33%), humerus (25%) and fingers (17%). Only peripheral adenopathy was associated with bone lesions (p=0.04). Seven patients have benefited from a follow-up 18F-FDG PET/CT, which in 100% of cases showed an improvement of lesions.

Conclusion: Bone sarcoidosis occurred in 14% of patients, affecting multiple bones and mostly the axial skeleton. 18F-FDG PET/CT appears to be a sensitive imaging for diagnosis and follow-up of bone sarcoidosis.

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Disclosure of Interests: None declared


Abstract AB1137 Figure 1. Relationship between aspirated volume and USS detected knee effusion depth