others were lymphadenopathy, orbital pseudotumor, pancreas and salivary glands in decreasing order (Table). Twenty-four (46.1%) of patients had localized involvement. Corticosteroids were mainstay of treatment in 92.5% of patients, and in 57.5% with any immunosuppressive agents as first line treatment. Rituximab has been used for cases resistant to previous treatment or with relapses in 19 (47.5%) of patients. A complete response was achieved in 52.5% of patients and partial response (<50% of regression) in 40%. Two patients deceased due to IgG4-RD attributed problems and no malignancy was observed (median follow up: 18 months).
**Conclusions:** We observed similar features with previous European cohorts however no male predominance was seen. Even though conventional immunosuppressives were used in more than half of patients, treatment had switched to rituximab ~50% patients owing to resistance or relapses.
**Disclosure of Interest:** None declared
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## FRI0613 H-FERRITIN AND PRO-INFLAMMATORY CYTOKINES ARE INCREASED IN THE BONE MARROW OF ADULT PATIENTS AFFECTED BY MACROPHAGE ACTIVATION SYNDROME

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**Background:** During macrophage activation syndrome (MAS), an inflammatory life-threatening syndrome, extremely high levels of serum ferritin may be observed [1]. Ferritin is an intracellular iron storage protein comprising 24 subunits that may be divided in heavy (H) subunits and light (L) subunits, based on their molecular weight [2]. The H-/L-subunits ratio may change, depending on the specific tissue and the physiologic status of the cell. In the normal condition, ferritin enriched in L subunits (L-ferritin) has been found in the liver and in the spleen, whereas the ferritin enriched in H subunits (H-ferritin), may be mainly observed in the heart and kidneys [2].
**Objectives:** We investigated the tissue expression of both H-and L-ferritin as well as the macrophage subsets expressing these molecules, in the inflammatory BM infiltrate of MAS patients. In addition, the co-expression of IL-1β, TNF, IFN-γ and H- or L ferritin, within the inflammatory cells, was assessed. Finally, we explored if the imbalance between H-ferritin and L-ferritin as well as the number of ferritin positive cells may be considered helpful bio-markers to assess the severity of these patients.
**Methods:** We analysed the bone marrow (BM) biopsies, by immunofluorescence of 10 adult MAS patients affected by rheumatic disease to assess the presence of: i. both H- and L-ferritin; ii. the number of CD68+/H-ferritin+ and CD68+/L-ferritin+; iii. the tissue pro-inflammatory cytokines, IL-1β, TNF, IFN-γ; and we correlated these data with clinical and laboratory data. Furthermore, the presence of ferritins was assessed in the sera of the same patients by western blot analysis.
**Results:** We observed an increased tissue expression of H-ferritin and of pro-inflammatory cytokines (IL-1β, TNF, IFN-γ). Western blot analysis, in the sera, of H-ferritin mirrored data on the tissue. Furthermore, an increased number of CD68+/H-ferritin+ cells and an infiltrate of cells co-expressing H-ferritin and IL-12, suggesting an infiltrate of M1 macrophages, were observed.
Tissue H-ferritin levels correlated with the decreased counts of WBC (p=0.01) and PLT (p=0.0001); with the increased values of serum ferritin (p=0.012) and C-reactive protein (CRP) (p=0.0058); and with the tissue expression of IL-1β (p=0.006). The number of the CD68+/H-ferritin+ cells correlated with the decreased counts of WBC (p=0.03) and PLT (p=0.0007), and with the increased serum ferritin levels (p=0.0088) and CRP (p=0.049). The analyses concerning tissues L-ferritin as well as the number of CD68+/L-ferritin+ cells and the same parameters failed to show any significant result.
**Conclusions:** We observed an increased tissue expression of H-ferritin associated with an increased expression of IL-1β. Interestingly, in the BM inflammatory infiltrate an increased number of CD68+/H-ferritin+ cells was shown. Of note, tissue expression of H-ferritin as well as the number of CD68+/H-ferritin+ significantly were associated with the hematological involvement of the disease, suggesting possible bio-markers to assess the severity of these patients.
**References:**
[1] Ramos-Casals M, et al. Lancet. 2014;383:1503–16.
[2] Rosário C, et al. BMC Med. 2013;11:185.
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## FRI0614 FREQUENCY OF ORGAN MANIFESTATIONS IN CHRONIC SARCOIDOSIS

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**Background:** Chronic sarcoidosis is a systemic disease of unknown etiology, characterized by the histological finding of granulomas in involved organ systems. The most often affected organ is the lung with approximately 90–95%. Systematic data of organ manifestations other than the lung are scarce and show a wide range from 1–2% up to 50% depending on the series.
**Methods:** We analyzed data of newly diagnosed chronic sarcoidosis in 3 tertiary hospitals. We analyzed data on organ manifestations (OM), type of OM and laboratory findings. The certainty of OM was classified as grade 0 (not investigated), grade 1 (no sign of OM), grade 2 (clinical sign of OM), grade 3 (signs of OM in laboratory findings or imaging) and grade 4 (histological proven OM), respectively.
**Results:** We included 151 patients with biopsy-proven chronic sarcoidosis. Mean age was 50.8±15 years with a male predominance (87 [57.2%] vs. 65 [42.8%] patients).
Except for 3 patients, all demonstrated pulmonary involvement. The predominant type of lung involvement was type I (mediastinal lymph node enlargement) in 54.2% and type II (mediastinal lymph node enlargement and interstitial involvement) in 27.7%.
96.5% of patients were investigated for an affection of the kidneys, 97.3% for hepatic, 92.7% for skin involvement, 68.8% for ocular manifestations, 67.5% for ear, nose, throat (ENT) manifestations and 92% for cardiac manifestations, respectively.
Grade 3 (imaging/laboratory) and grade 4 (histology) findings were seen in the kidneys in 7.6/22.8%, in the liver in 13.3/11.9%, in the heart in 10.6/0.7%, in the eyes in 6.6/
**Conclusions:** OM in chronic sarcoidosis are more frequent than suggested in the current literature, especially renal and hepatic. About 20% of patients with chronic sarcoidosis suffered from moderate to severe CKD due to sarcoidosis, which is a major organ complication contributing to overall morbidity.
We recommend a systematic screening for OM in all patients with chronic sarcoidosis as it is performed in other systemic rheumatic disease.
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## FRI0615 SARCOIDOSIS AND CANCER: DIFFERENT PATTERNS OF ASSOCIATION IN A MULTICENTER COHORT FROM SOUTHERN EUROPE

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**Objectives:** To evaluate the temporal association between the diagnosis of neoplasia and sarcoidosis in a large cohort of Spanish patients with sarcoidosis.
**Methods:** In January 2016, the Autoimmune Diseases Study Group (GEAS-SEMI) created a national registry (SARCOGEAS) of patients with sarcoidosis. Sarcoidosis was diagnosed with the criteria proposed by the ATS/ERS/WASOG 1999 statement, and extrathoracic disease by the 2014 WASOG instrument. Diagnosis of neoplasia was recorded before and after the diagnosis of sarcoidosis.
**Results:** The cohort included 1082 patients (82% biopsy-proven, 618 women, mean age 47yrs). Association with neoplasia was detected in 135 (13%) patients who developed 140 neoplasms (110 solid and 30 hematological neoplasia). The neoplasia more frequently reported were breast (n=18), lymphoma (n=16), non-melanoma skin (n=15) and colon (n=15). Association with neoplasia was more frequent in patients born in Spain (97% vs 86%, p<0.001, OR 4.06), older patients (55 vs 46yrs, p<0.001, OR 1.03) and those with bone marrow involvement (14% vs 4%, p<0.001, OR 3.64). Patients in whom cancer preceded the diagnosis of sarcoidosis had a higher frequency of sarcoidosis diagnosed incidentally (20% vs 4%, p=0.011) and a lower frequency of ocular sarcoidosis (3% vs 16%, p=0.016). Patients with associated hematological neoplasia had a higher frequency of ENT (13% vs 1%, p=0.009) and bone marrow (33% vs 9%, p=0.002) involvements in comparison with patients with associated solid neoplasia.
**Conclusions:** Association between sarcoidosis and cancer was found in 13% of patients (80% solid and 20% hematologic malignancies). Elderly patients and those born in Spain were at high risk of having associated cancer. Asymptomatic