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SAT0514 MRI – GUIDED THERAPY FOR SYSTEMIC SCLEROSIS ASSOCIATED MYOSITIS

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Background: Muscle involvement in systemic sclerosis (SSc) has a significant impact on morbidity, functional capacity, and mortality. The muscle histopathology is heterogeneous including inflammatory and fibrotic changes. Currently there are no satisfactory means to diagnose inflammatory myopathy in SSc pts with normal creatine kinase (CK) and to assess the response to therapy.
Objectives: Our aim was to evaluate whether muscle magnetic resonance imaging (MRI) might be a tool to diagnose inflammatory myopathy in SSc patients (pts) and to assess the effect of muscle oriented- immunomodulatory therapy.
Methods: We retrospectively analysed the clinical data of 290 consecutive SSc pts seen at our centre between the years 2012–2017. Our cohort is part of the EUSTAR registry (centre 042). Pts with muscle weakness as defined by the Medsger muscle severity score of $\geq$1 and at least one MRI study were included. Clinical data analyzis included SSc subtype, disease duration, modified Rodnan skin score (mRSS), Medsger muscle severity score, CK, autoantibody profile, MRI and immunomodulatory treatment.
Results: 26 pts with muscle weakness answered the criteria of Medsger muscle severity score of $\geq$1 MRI data were available, in 17 of the pts. Muscle oedema and fasciitis were seen in MRI in 13 pts (10 diffuse subset, median: age 40, disease duration 1.25 years, mRSS 13.5). MRI was normal in 4 pts (2diffuse SSc, median: age 50 years, disease duration 6 years, mRSS 4). CK was normal in 10 pts with pathologic MRI. Anti-topoisomerase was positive in 6 pts, RNA polymerase 3 – in 3 pts, anti-centromere – in 2 pts and 6 pts were only ANA positive. Muscle biopsy results were available in 4 pts: Biopsy was compatible with myositis in 3 pts with pathologic MRI and revealed fibrosis in 1 pt with normal MRI. 14 pts received immunomodulatory treatment: rituximab (3 pts), rituximab and intravenous immunoglobulins (IVIG) (3 pts), IVIG and methotrexate/azathioprine/mycophenolate mofetil.[8] A second MRI was performed in 6 pts with first pathologic MRI, after 12 months of treatment. Significant regression of muscle oedema and perifasciitis was observed in 5 pts and correlated with clinical amelioration, with improvement of muscle strength. No clinical and imaging improvement occurred in one patient, despite the treatment. No change in muscle strength was seen in the patient with normal MRI and evidence of fibrosis on muscle biopsy, although the skin score improved.
Conclusions: MRI might serve as a non-invasive tool for diagnosis of inflammatory myopathy in SSc pts with early disease, Medsger muscle severity score of $\geq$1 and normal CK and for assessment of treatment efficacy.
Disclosure of Interest: None declared

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SAT0515 CLINICAL CHARACTERISTICS OF PATIENTS WITH CANCER-ASSOCIATED MYOSITIS COMPLICATED BY INTERSTITIAL LUNG DISEASE

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Background: Cancer-associated myositis (CAM) is believed to be rarely complicated by interstitial lung disease (ILD), and, thus, detailed clinical characteristics of CAM in patients with polymyositis (PM)/dermatomyositis (DM)-associated ILD are not well known.
Objectives: To clarify the incidence, risk factors, and impact on survival of malignancy in patients with PM/DM-associated ILD, using a large cohort data.
Methods: We used 497 patients with PM/DM-associated ILD enrolled in a multicentre retrospective cohort of incident cases from 44 institutions across Japan (JAMI). CAM was defined as malignancy diagnosed within 3 years before or after PM/DM diagnosis. Demographic data and clinical characteristics were recorded at the time of diagnosis, and follow-up survival and malignancy data were collected prospectively.
Results: Thirty-two patients with CAM (6.4%) were identified. Patients in the CAM group were older (64.3 vs 55.1 years, p<0.001), had shorter disease duration at onset (4.1 vs 7.0 months, p=0.01), and presented with arthritis less frequently (24.1 vs 48.5%, p=0.01), in comparison with those with non-CAM group. All patients with CAM were older than 40 years, and the proportion of CAM increased along with the age. Patients who were $\geq$59 years at diagnosis and lacked arthritis were at 12 times higher risk for concomitant malignancy than those without such features (figure 1). Frequencies of autoantibodies, including anti-MDA5, anti-ARS, and anti-TIF1gamma, were not different between the groups. In 19 patients (59%), malignancy was diagnosed within 3 months before or after PM/DM diagnosis. Eleven CAM patients were died, and cause of deaths included ILD in 6 and malignancy in 5. Survival analysis by Kaplan-Miere method demonstrated that CAM patients had a poorer survival than did non-CAM patients (p=0.016).
Conclusions: In patients with PM/DM-associated ILD, older age at diagnosis and lack of arthritis are predictors for concomitant malignancy, which leads to a reduced survival.
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Vasculitis

SAT0516 TWO DISTINCT SUBSETS OF LOW DENSITY GRANULOCYTES IN ANCA ASSOCIATED VASCULITIS

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Background: Low density granulocyte (LDG), a proinflammatory population of neutrophils, was first described in systemic lupus erythematosus (SLE) and has