most common malignancy in patients with Sjogren's syndrome was thyroid cancer. The risk of overall malignancy was not higher than that of the control. However, the incidence of prostate cancer (HR; 1.339, 95% CI; 1.076~1.667), thyroid cancer (HR; 1.320, 95% CI; 1.093~1.594) and lymphoma (HR; 1.620, 95% CI; 1.066~2.462) were higher and hepatocellular carcinoma (HR; 0.591, 95% CI; 0.455~0.767) was lower than that of control. Lymphoma developed in 305 patients (3.1% of total malignancy); 10 cases of Hodgkin lymphoma, and 290 cases of non-Hodgkin lymphoma. Gender, age, smoking, drinking, body mass index, fasting blood glucose, and proteinuria were not significantly different from the control group in lymphoma development.

Conclusions: Sjogren's syndrome patients' overall malignancy risk was not higher than that of control group. However, risk of lymphoma, prostate cancer, and thyroid cancer was higher than control group.

Disclosure of Interest: None declared

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SAT0415 THE MRZ REACTION HELPS TO DISTINGUISH RHEUMATOLOGIC DISORDERS WITH CENTRAL NERVOUS INVOLVEMENT FROM MULTIPLE SCLEROSIS

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Background: Some rheumatologic disorders (RD) may initially manifest with central nervous system (CNS) affection, mimicking the clinical, magnetic resonance imaging, and cerebrospinal fluid (CSF) findings of multiple sclerosis (MS). Vice versa MS might be difficult to separate from some RD because of the presence of autoantibodies (e.g. ANA) in up to 50%. The MRZ reaction (MRZR), composed of the three respective antibody indices (AI) against measles, rubella, and varicella zoster virus, has been found positive frequently in MS patients. However, it is unclear whether the MRZR is helpful to distinguish rheumatologic disorders with CNS involvement (RDwCNS) from MS.

Objectives: To investigate the MRZ reaction as a diagnostic tool to distinguish patients with RDwCNS from patients with MS.

Methods: The MRZR was evaluated in 35 patients with RDwCNS and compared to 70 sex- and age- matched MS patients. An AI result ≥ 1.5 was indicative for intrathecal IgG production against the respective pathogen. Two previously established stringency levels, MRZR-1 (≥ 1 of 3 AIs positive) and MRZR-2 (≥ 2 of 3 AIs positive), were applied. CNS involvement of RDwCNS was defined as clinical manifestation with neurological symptoms and signs of inflammation in CSF analysis and/or cerebral/spinal magnetic resonance imaging (MRI). MRZR results were compared using the Fisher's exact test with $p < 0.05$ for statistical significance.

Abstract SAT0415 – Figure 1. Frequency of positive MRZ reaction (MRZR-1 and MRZR-2) in patients with RDwCNS compared to MS patients.

Results: Within the RDwCNS group, 31 patients suffered from systemic lupus erythematosus, four had a small vessel vasculitis. In both groups 77.1% were female, mean age ($\pm$ SD) was 43.2 years ($\pm$ 18.7) in RDwCNS and 47.5 years

($\pm$ 7.8) in MS ($p = n.s.$). All RDwCNS patients showed clinical symptoms indicative for CNS involvement and signs of inflammation in CSF analyses and/or MRI of the brain. In 52 MS patients autoantibody screening was performed. 42% were positive for ANA ($n=20$) or ANCA ($n=2$) in indirect immunofluorescence. Only 14.3% of RDwCNS patients had a positive MRZR-1 compared to 85.7% within the MS group ($p < 0.0001$). The more specific MRZR-2 was positive in 60% of the MS patients compared to only 8.5% of the RDwCNS patients ($p < 0.0001$). By using a higher threshold of >2.0 for a positive AI, the prevalence of positive MRZR-2 dropped to 5.7% ($n=2$) in the RDwCNS group compared to 54.3% ($n=38$) in the MS group ($p < 0.0001$). Oligoclonal bands were found in 94.3% of the MS and 28.6% of the RDwCNS patients ($p < 0.0001$).

Conclusions: Considering the high specificity of the MRZR-2 for MS confirmed in this study, this laboratory test may be a helpful diagnostic tool to distinguish RDwCNS from MS.

Disclosure of Interest: None declared

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SAT0416 LIFE-THREATENING PRIMARY SJÖGREN SYNDROME: CLINICAL CHARACTERISATION AND OUTCOMES IN 1535 PATIENTS (GEAS-SS REGISTRY)

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Objectives: To analyse the clinical features and outcomes of patients presenting with life-threatening systemic disease in a large cohort of Spanish patients with primary Sjogren syndrome (SjS).

Methods: The GEAS-SS multicenter registry was formed in 2005 with the aim of collecting a large series of Spanish patients with primary SS. By January 2018, the database included 1535 consecutive patients fulfilling the 2002/2016 criteria. Life-threatening systemic disease was defined as an activity level scored as "High" in at least one ESSDAI domain.

Results: 209 (14%) were classified as presenting with a life-threatening systemic disease: 194 presented one ESSDAI domain classified as high, 14 two domains and only one presented three high activity domains. The high-ESSDAI domains included lymphadenopathy in 78 (37%) cases, CNS in 28 (13%), PNS in 25 (12%), pulmonary in 25 (12%), renal in 22 (10%), cutaneous in 18 (9%), articular in 18 (9%), haematological in 7 (3%) and muscular in 4 (2%); the most frequent clinical presentations in each domain were, respectively, parotid lymphoma ($n=41$), focal neurological deficit ($n=20$), ganglionopathy ($n=11$), usual interstitial pneumonitis ($n=9$), renal failure ($n=11$), ulcerated cutaneous vasculitis ($n=9$), symmetric polyarthritis ($m=17$), severe thrombocytopenia ($n=3$) and severe myositis ($n=3$). With respect to therapeutic approach, 144 (69%) required glucocorticoids, 65 (31%) immunosuppressive agents and 42 (20%) biological therapies. During the follow-up, 36 (17%) patients died, mainly due to lymphoma ($n=16$), pulmonary fibrosis ($n=5$), end-stage renal failure ($n=4$), CNS progressive disease ($n=3$) and systemic vasculitis ($n=3$).

Conclusions: A 14% of patients with primary SjS develop a potentially life-threatening systemic disease (mainly lymphoma, but also severe internal organ involvements including nervous system, the lungs and the kidneys). This subset of patients requires intensive therapeutic management with a mortality rate of nearly 20% of cases.

Disclosure of Interest: None declared

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