The mean disease duration was 6.4±5.8 years (0.5-30). The observation period was 12 years. All pts received prednisolone at a dose of 11.7±4.8 mg/day, and 73 patients (49%) received immunosuppressants at inclusion. The indications for the appointment of RTX were ineffectiveness or impossibility of standard therapy and a severe course of the disease with high activity and unfavorable prognosis factors. AE were assessed and recorded by a physician at a hospital immediately after the infusion of RTX, then by patient reported outcome during the observation period. Severe AE were defined as those that required hospitalization for more than 24 hours, exacerbation of the disease requiring therapy, malignancies, life-threatening situations. All causes of death were considered, regardless of treatment.

Results: The mean follow-up period after the first infusion of RTX was 5.6±2.6 years [834.4 patient-years (PY)]. Pts received a mean of 3.4 courses of RTX (1–10). The cumulative mean dose of RTX was 3.2±2.4 gr (0.5–11). AE were reported in 77 patients (52%), the overall frequency of AE was 9.3/100 PY (95% Confidence Interval 6.2–12.4). The highest frequency of all AE was observed in the first 2-6 months after the first infusion of RTX, however these were mainly mild AE (71%). There was a decrease of AE in the follow-up period (3.4/100 PY, 95% CI 2.4–4.9 – at the period from 3 to 10 course of RTX). The overall incidence of serious AE was 2.2/100 PY (95% CI 1.4–3.5). The specter of serious AE included: pneumonia in 7 pts, infusion reactions in 5, as well as in one case: cerebral ischemia, acute pancreatitis, allergic pneumonitis, lymphoma of pharynx, purulent arthritis, lower limb vein thrombosis, pulmonary embolism of small arteries. The most frequent AE were infections (95% CI 1.4–3.5). The specter of serious AE included: pneumonia in 7 pts, infusion reactions in 5, as well as in one case: cerebral ischemia, acute pancreatitis, allergic pneumonitis, lymphoma of pharynx, purulent arthritis, lower limb vein thrombosis, pulmonary embolism of small arteries. The most frequent AE were infections (95% CI 1.4–3.5). The specter of serious AE included: pneumonia in 7 pts, infusion reactions in 5, as well as in one case: cerebral ischemia, acute pancreatitis, allergic pneumonitis, lymphoma of pharynx, purulent arthritis, lower limb vein thrombosis, pulmonary embolism of small arteries. The most frequent AE were infections (95% CI 1.4–3.5). The specter of serious AE included: pneumonia in 7 pts, infusion reactions in 5, as well as in one case: cerebral ischemia, acute pancreatitis, allergic pneumonitis, lymphoma of pharynx, purulent arthritis, lower limb vein thrombosis, pulmonary embolism of small arteries. The most frequent AE were infections (95% CI 1.4–3.5).
AB0425
CLINICAL AND PATHOLOGICAL FEATURES OF BREAST CANCER IN PATIENTS WITH SYSTEMIC SCLEROSIS: PRELIMINARY DATA FROM THE SCLERO-BREAST STUDY

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Background: Systemic Sclerosis (SSc) is a rare and life-threatening connective tissue disease characterized by vascular dysfunction, specific autoimmune abnormalities and fibrosis of the skin and internal organs. Previous studies have shown a 1.5-fold increase in cancer risk in SSc patients compared with the general population, including breast cancer (BC). The relationship between BC and SSc has long been discussed but past research has been contradictory and inconclusive on this topic.

Objectives: The aim of our project was to analyze clinical and pathological characteristics of BC developed by SSc subjects and possible correlations with scleroderma features. Here we present the preliminary data from the Sclero-Breast Study.

Methods: Our observational retrospective multicenter study enrolled 33 SSc women with a personal history of BC identified at two rheumatology/SSc Units in the north of Italy between January 2017 and December 2019 (1cdScSsc 23/39, 1 unknown, mean age at SSc onset 57 years, range 32-73). All patients underwent general and instrumental assessment: smoking habits, presence of skin ulcer, calcinosis, Raynaud phenomenon, gastro-oesophageal reflux disease, interstitial lung disease, but not with BC status. 93.1% of patients were diagnosed with any type of myositis except for inclusion body myositis. Cancer screening strategy was individualized according to clinical and serological data, including PET/CT as the main test to detect occult cancer (OC). Procedures derived from a positive PET/CT were registered. Qualitative data expressed as percentages, and quantitative data as the median with the interquartile range were analyzed. A ROC curve was used to estimate the reliability of PET/CT for CAM diagnosis.

Results: Seventy-seven out of 131 patients underwent a PET/CT for OC screening. The performance of the PET/CT in patients with myositis at disease onset yielded an area under the curve ROC of 0.87 (0.73-0.97) for CAM diagnosis. Invasive procedures in 7 (9%) patients without a final diagnosis of cancer did not cause derived complications. Patients not evaluated for OC did not develop cancer after a median follow-up of 3.3 years (1.7-6.7). Conclusion: Cancer screening strategy should be individualized. PET/CT at myositis onset seems to be an efficient approach to rule out CAM. This practice does not seem to significantly increase harm to patients related to the additional tests needed to clarify inconclusive results.

Disclosure of Interests: None declared

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AB0426
CANCER SCREENING IN IDIOPATHIC INFLAMMATORY MYOPATHIES: TEN YEARS EXPERIENCE FROM A SINGLE CENTER

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Background: There is a well-recognized association between cancer and myositis, so cancer screening at diagnosis is recommended.

Objectives: We aim to report the results of our cancer screening strategy and to ascertain the reliability of using PET/CT to identify cancer-associated myositis (CAM) in a large cohort of patients with myositis from a single center over 10 years.

Methods: This retrospective observational study included all patients diagnosed with any type of myositis except for inclusion body myositis. Cancer screening strategy was individualized according to clinical and serological data, including PET/CT as the main test to detect occult cancer (OC). Procedures derived from a positive PET/CT were registered. Qualitative data expressed as percentages and quantitative data as the median with the interquartile range were analyzed. A ROC curve was used to estimate the reliability of PET/CT for CAM diagnosis.

Results: Seventy-seven out of 131 patients underwent a PET/CT for OC screening. The performance of the PET/CT in patients with myositis at disease onset yielded an area under the curve ROC of 0.87 (0.73-0.97) for CAM diagnosis. Invasive procedures in 7 (9%) patients without a final diagnosis of cancer did not cause derived complications. Patients not evaluated for OC did not develop cancer after a median follow-up of 3.3 years (1.7-6.7). Conclusion: Cancer screening strategy should be individualized. PET/CT at myositis onset seems to be an efficient approach to rule out CAM. This practice does not seem to significantly increase harm to patients related to the additional tests needed to clarify inconclusive results.

Disclosure of Interests: None declared

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AB0427
ANTI-NOR90 AUTOANTIBODIES: FAVORABLE OR UNFAVORABLE PROGNOSIS?

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Background: Anti-NOR 90 autoantibodies (anti-NOR90 Ab) are autoantibodies that target nucleolar transcription factor 1 or HUFI, involved in transcription of RNA polymerase I. These autoantibodies have been detected in 6.1% of patients with Systemic Sclerosis (SSc), but their clinical or prognostic significance has not been clearly defined. Anti-NOR90 Ab have been mostly associated with limited scleroderma with mild organ involvement and can also be found in other rheumatic diseases such as rheumatoid arthritis, systemic lupus erythematosus or Sjögren’s syndrome.

Objectives: The aim of this study was to identify the main clinical characteristics of patients with positive anti-NOR90 in our Centre.

Methods: This is a retrospective, descriptive, cross-sectional study of all patients with positive anti-NOR90 Ab between January 2013 and December 2020 in a single center. Autoantibodies testing was performed using EuRomun EuROLINE SSc profile IgG autoAb assay kit. Patient demographics, clinical characteristics, associated diagnoses, laboratory and immunological findings were collected.

Results: We identified a total of 26 patients with at least a positive value for anti-NOR90 Ab (Table 1). In most cases anti-NOR90 patients were ANA positive, predominantly with nucleolar pattern and coexisted with other SSc autoantibodies. 12 patients had rheumatic diseases and two had SSc, both with limited cutaneous SSc and absence of organ involvement. 14 patients had no definite diagnosis. Clinical features of anti-NOR90 patients are represented in Figure 1. Five patients presented Raynaud’s phenomenon, two cases with pathological nailfold capillaroscopy and one patient had SSc. There was no patient with skin ulcers, calcinosis, interstitial lung disease or pulmonary hypertension. Four patients had gastroesophageal reflux disease and one patient presented antral vascularity. Six patients developed some neoplasm.

Disclosure of Interests: None declared

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