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Scientific Abstracts

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None declared
treatments in this subset of fragile patients.
to improve BC diagnosis and prognosis and to personalize oncological targeted
detection in SSc patients. Further investigations on larger numbers of patients
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treated with an early-stage tumor, 70.8% of invasive carcinomas with a low
association (p<0.05) was observed between the use of immunosuppressive
(treatment); type of systemic treatment (neoadjuvant/adjuvant chemotherapy and
receptor status; MIB1, HER2 expression; clinical and pathological stage at
diagnosis; metastatic sites; type of loco-regional treatment (surgery and radio-
cholangiocarcinoma; presence of gastro-duodenal ulcers, calcinosis, interstitial
lung disease or pulmonary hypertension. Four patients had

Background: Systemic Sclerosis (SSc) is a rare and life-threatening connec-
tive 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 gen-
eral 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 char-
acteristics of BC developed by SSc subjects and possible correlations with scler-
oderma 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 (lc/dcSSc
23/9, 1 unknown; mean age at SSc onset 57 years, range 32-73). All patients
underwent general and instrumental assessment: smoking habits; presence of
skin ulcers, calcinosis, interstitial lung disease; presence of gastro-duodenal
ulcers; presence of gastro-intestinal and kidney involvement; interstitial lung disease (at HR-CT); pulmonary function
tests; ECG abnormalities; echocardiographic assessment of pulmonary arterial hypertension (PAH); videocapillaroscopic pattern; autoantibody profile; exposure
to immunosuppressive and vasoactive therapies; status at last follow-up
evaluation and cause of death. Clinical and pathological characteristics of BC
were also evaluated: age at diagnosis; menopausal status; histotype; hormone
receptor status; MB1, HER2 expression; clinical and pathological stage at
diagnosis; metastatic sites; type of loco-regional treatment (surgery and radio-
therapy); type of systemic treatment (neoadjuvant/adjuvant chemotherapy and
docrine treatment); other cancers and time from diagnosis of the first disor-
der to the second one.

Results: A total of 54.5% of subjects developed BC before SSc (median
interval of 5 years), whereas 45.5% of patients developed BC after SSc
(median delay of 8 years). 54.5% of patients showed interstitial lung disease and the cause of death of the 6 deceased subjects was PAH. A significant
association (p<0.05) was observed between the use of immunosuppressive
therapy and diffuse skin extension, negative ACA, positive Anti-Scl-70 and
interstitial lung disease, but not with BC status. 93.1% of patients were diag-
nosed with an early-stage tumor, 70.8% of invasive carcinomas with a low
MB1-1, 8.3% with a tubular histotype, while 42.8% presented with a Luminal
A-like tumor. 66.6% underwent breast conserving surgery and 55.5% RT
after surgery. 40% of patients developed interstitial lung disease after RT
and 20% dSSc.

Conclusion: According to our preliminary data, SSc patients developed BC at
good prognosis, suggesting a de-escalation strategy of cancer therapies. On
these grounds, a proper screening is mandatory in order to allow for early cancer
detection in SSc patients. Further investigations on larger numbers of patients
are needed. First of all, they would further clarify the intriguing relationship
between BC and SSc. Secondly, they would help to explore the common bio-
chemical and molecular pathways at the basis of these two disorders, with the aim
to improve BC diagnosis and prognosis and to personalize oncological targeted
treatments in this subset of fragile patients.

Disclosure of Interests: None declared

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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 screen-
ing. 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.

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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 HUFB, 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 rheu-
matic diseases such as rheumatoid arthritis, systemic lupus erythematosus or
Sjogren’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 sin-
gle center. Autoantibodies testing was performed using Euroimmun 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, pre-
dominantly with nucleolar pattern and coexisted with other SSc autoantibodies.
12 patients had rheumatic diseases and two had SSc, both with limited cutane-
ous SSc and absence of organ involvement. 14 patients had no definite diag-
nosis. 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 vascular ectas-
ia. Six patients developed some neoplasm.
Background: Intersitial lung disease (ILD) is the most frequent pulmonary manifestation of scleroderma and is seen in 80% of the patients with diffuse and in 20% of the limited forms of scleroderma. Even seen in early stages of the disease with varying degrees of severity, it does not correlate with the clinical manifestations. Hence, it is difficult to select patients for ILD screening based on symptoms. Digital clubbing is a common feature of ILD and has been associated with poor prognosis. Methods: A prospective observational study involving 15 patients each of scleroderma with ILD, without ILD and controls visiting our clinic from August 2020 to December 2020 were included. The presence of ILD was confirmed by HRCT thorax. Patients with a history of smoking, alcohol intake or pre-existing cardio-respiratory illnesses were excluded. Ultrasound assessed NBT was calculated using Philips affiniti 70 eL18-4 probe. A total of 436 nail beds were recorded (142 of scleroderma with ILD, 144 of scleroderma without ILD and 150 of control group). 14 nails were excluded due to trauma. Statistical analysis was done using ANOVA test. Results were considered significant at p value < 0.05.

Results: On comparing nail bed thickness among scleroderma with ILD, scleroderma without ILD and control groups, the difference in NBT was statistically significant in all the three groups, except for left 1st finger (Table 1). NBT’s among scleroderma patients with ILD and control group did not show significant difference between the two groups. Whereas, when scleroderma patients with ILD were compared with control group, statistical significance were found (p<0.001) in all the fingers of both the hands, except 1st and 2nd finger of left and right hand respectively. On comparing NBT’s among individual fingers of right hand in patients of scleroderma with ILD and without ILD, a statistically significant difference among the two groups. Whereas, when scleroderma patients with ILD were compared with control group, statistical significance were found (p<0.001)

Conclusion: In our case series anti-NOR90 Ab were associated with multiple rheumatic diseases with heterogeneity of clinical manifestations. We did not observe a further progression to SSC or presence of organ involvement or severe scleroderma, so these autoantibodies could be related with a favorable prognosis. In contrast with previous reports, a striking association with cancer has been detected in our population.

Table 1. Demographic characteristics and main diagnoses of anti-NOR90 positive patients.

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Total Anti-NOR90: 26 patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex, n</td>
<td>Male: 11; Female: 15</td>
</tr>
<tr>
<td>Age, mean (years)</td>
<td>58.9 (IQR [46.7-72.2])</td>
</tr>
<tr>
<td>Race, n</td>
<td>Asian: 1; Hispanic: 7; Caucasian: 18</td>
</tr>
<tr>
<td>Smoker, n</td>
<td>3</td>
</tr>
<tr>
<td>Positive ANA (&gt;1/160), n</td>
<td>24</td>
</tr>
<tr>
<td>Pattern, n</td>
<td>7 Homogeneous, 4 Nucleolar, 4 Speckled, 1 Centromere, 5 Speckled-nucleolar, 3 Homogeneous-nucleolar.</td>
</tr>
<tr>
<td>Positive ENA, n</td>
<td>3</td>
</tr>
<tr>
<td>Systemic sclerosis autoantibodies, n</td>
<td>Anti-Ku: 7, Anti-SS-A-Ro52 and Ro60, 1 anti-RNP and anti-Sm</td>
</tr>
<tr>
<td>Neoplasm, n</td>
<td>6: 2 solid organ cancer (bladder, kidney), 1 lung adenocarcinoma, 1 multiple myeloma, 1 acute myeloid leukemia: 1 basal cell carcinoma.</td>
</tr>
</tbody>
</table>

Table 2. Nail bed thickness in patients of scleroderma with ILD, without ILD and control groups.

<table>
<thead>
<tr>
<th>Finger</th>
<th>Mean</th>
<th>SD</th>
<th>Mean</th>
<th>SD</th>
<th>Mean</th>
<th>SD</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right 1st</td>
<td>1.86</td>
<td>0.239</td>
<td>1.71</td>
<td>0.081</td>
<td>1.69</td>
<td>0.107</td>
<td>0.021</td>
</tr>
<tr>
<td>2nd</td>
<td>1.73</td>
<td>0.303</td>
<td>1.50</td>
<td>0.079</td>
<td>1.61</td>
<td>0.108</td>
<td>0.003</td>
</tr>
<tr>
<td>3rd</td>
<td>1.89</td>
<td>0.200</td>
<td>1.75</td>
<td>0.091</td>
<td>1.50</td>
<td>0.131</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>4th</td>
<td>1.80</td>
<td>0.263</td>
<td>1.39</td>
<td>0.147</td>
<td>1.35</td>
<td>0.078</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>5th</td>
<td>1.80</td>
<td>0.119</td>
<td>1.24</td>
<td>0.095</td>
<td>1.29</td>
<td>0.069</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Left 1st</td>
<td>1.98</td>
<td>0.442</td>
<td>1.72</td>
<td>0.084</td>
<td>1.68</td>
<td>0.050</td>
<td>0.078</td>
</tr>
<tr>
<td>2nd</td>
<td>1.82</td>
<td>0.123</td>
<td>1.57</td>
<td>0.125</td>
<td>1.57</td>
<td>0.092</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>3rd</td>
<td>2.05</td>
<td>0.309</td>
<td>1.76</td>
<td>0.078</td>
<td>1.42</td>
<td>0.109</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>4th</td>
<td>1.92</td>
<td>0.246</td>
<td>1.36</td>
<td>0.130</td>
<td>1.35</td>
<td>0.107</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>5th</td>
<td>1.81</td>
<td>0.158</td>
<td>1.15</td>
<td>0.368</td>
<td>1.23</td>
<td>0.103</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Abbreviations: LES: systemic lupus erythematosus; MCTD: mixed connective tissue disease

References:

Disclosure of Interests: None declared

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