Graft-versus-host disease (GVHD) is a commonly severe multifocal complication in patients undergoing allogeneic transplantation of hematopoietic progenitors. Its chronic form reflects a complex immune response with different degrees of inflammation, immune dysregulation and fibrosis. In some chronic graft-versus-host disease (cGVHD) patients, positive antibodies have been detected, which represent the presence of immune activity and suggest the different degrees of inflammation, immune dysregulation and fibrosis. In some chronic graft-versus-host disease (cGVHD) patients, positive antibodies have been detected, which represent the presence of immune activity and suggest the possibility of involvement of B lymphocytes in the disease etiopathogenesis, but their clinical utility is controversial.

Objectives: To describe the clinical characteristics of a group of cGVHD patients with positive autoimmunity treated in a multidisciplinary consultation of Rheumatology-Dermatology-Hematology of GVHD.

Methods: Observational and retrospective study to describe the clinical characteristics of the patients with positive autoimmunity collected in the database of the multidisciplinary consultation of GVHD. The variables reviewed for this study, in addition to the demographic ones, were type of antibody, disease causing the transplant, presentation, severity and type of involvement. The statistical analysis was done with Epi-info 7.2.2.6.

Results: Only 16% (16) of the 100 patients included in the database had positive autoimmunity. Twelve (75%) tested positive to ANA, although 5 (31.25%) in a lower titer (1/80). The most common immunofluorescence pattern was the nuclear in 88.9% and 22.22% nuclear + cytoplasmic. Other antibodies detected were: 6 anti-Ro52, 2 anti-dsDNA, 1 anti-RP155, 1 anti-Fibrillarin, 1 anti-SAE1, 1 p-ANCA and 1 anti-NOR-90. The mean of age was 51.31±14.03 years. As for sex 4 (25%) were female and 12 (75%) were men. The most frequent disease that caused the transplant was acute myeloid leukemia (58.3%). Ten (62.5%) patients presented de novo cGVHD, 1 (6.25%) progressive and 5 (31.25%) quiescent. The time since receiving the transplant until the first visit was 14 to 79 months. Ten (62.5%) patients had nonspecific symptoms (arthralgia and myalgia), 2 (12.5%) edema, 8 (50%) contractures, 8 (50%) fascitis and 6 (37.5%) eosinophilia. Eight (50%) patients had ocular involvement and 6 (37.5%) of the oral mucosa in the form of dry syndrome (Sjögren-like syndrome). Ten (62.5%) patients had limitation of joint mobility detected by the range of motion scale (ROM), of which 6 were mild and 4 moderate. Only 5 (31.25%) patients had general condition impairment. As for the skin involvement 10 (62.5%) patients had sclerodermiform involvement (8 of them being eosinophilic fasciitis-like), 2 (12.5%) lichenoid, and 3 (18.5%) mixed (sclerodermiform + lichenoid). Only 1 patient didn’t meet diagnostic criteria for GVHD. The sclerodermiform was the most common type of involvement in the positive ANA patients. Regarding the severity according to the of the American National Institute of Health (NIH) classification: 8 (50%) had serious affection, 5 (31.25%) moderate and 2 (12.5%) mild, with 4 (25%) exitus.

Conclusion: In our cohort of patients with cGVHD, serum detection of autoantibodies is uncommon, being the ANA with nuclear pattern the most frequent. Although the small sample size does not allow correlations with the clinical variables it’s worth highlighting a greater positivity of autoantibodies in the sclerodermiform skin forms.

References:

Disclosure of Interests: Maria Elisa Acosta: None declared, Luís Gómez-Lechón: None declared, Olga Compán: None declared, Sonia Pastor: None declared, Carlos A. Montilla-Morales: None declared, Olga Martínez González: None declared, Ana Isabel Turrión: None declared, Javier del Pino Grant/ research support from: Roche, Bristol, Consultant of: Gedeon, Cristina Hidalgo: None declared

DOI: 10.1136/annrheumdis-2020-eular.1871

Scleroderma, myositis and related syndromes

AB0543

CLINICAL CHARACTERISTICS OF A GROUP OF CHRONIC GRAFT-VERSUS-HOST DISEASE PATIENTS WITH POSITIVE AUTOIMMUNITY.

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Background: Graft-versus-host disease (GVHD) is a commonly severe multiorgan complication in patients undergoing allogeneic transplantation of hematopoietic progenitors. Its chronic form reflects a complex immune response with different degrees of inflammation, immune dysregulation and fibrosis. In some chronic graft-versus-host disease (cGVHD) patients, positive antibodies have been detected, which represent the presence of immune activity and suggest the possible involvement of B lymphocytes in the disease etiopathogenesis, but their clinical utility is controversial.

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Conclusion: In our cohort of patients with cGVHD, serum detection of autoantibodies is uncommon, being the ANA with nuclear pattern the most frequent. Although the small sample size does not allow correlations with the clinical variables it’s worth highlighting a greater positivity of autoantibodies in the sclerodermiform skin forms.

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DOI: 10.1136/annrheumdis-2020-eular.1871

AB0544

EVALUATION OF MACULAR AND OPTIC DISC MICROVASCULAR NETWORK IN PATIENTS WITH SYSTEMIC SCLEROSIS: AN OPTICAL COHERENCE TOMOGRAPHY ANGIOGRAPHY STUDY

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Background: Systemic sclerosis (SSc) is characterized by fibrosis of the skin, internal organs and vasculopathy. Invivo, the retina provides a unique opportunity to assess the microcirculation in the eye. Previous studies have been evaluated the changes in the retinal and choroid layer and showed thinning of the choroid layer and reduced retinal microvascular density. The objective of this study was to describe the ch
layer (GC-IPL), vessel density (VD), perfusion density (PD), optic disc PD, reflux index and foveal avascular zone (FAZ) parameters were measured by the same experienced operator.

Results: There was no significant difference between SSC and controls in terms of age, sex, spherical equivalent (SE), intraocular pressure (IOP), and axial length (AL). Central and mean macular thickness, nasal and inferior RNFL thicknesses were significantly thinner in SSC patients (Table). Additionally GC complex thicknesses were significantly thinner in all quadrants compared to controls. 

Central vessel density (CVD) and central perfusion density (CPD) values were found significantly decreased in all regions in patients with SSC. Optic disc perfusion density values were also decreased in SSC group. An inverse correlation was found between central macular thickness, FAZ area and perimeter values (rho: -0.300, p = 0.007; rho: -0.276, p = 0.013, respectively). There was no relationship between the disease duration and the OCTA measures.

Conclusion: Vascular and perfusion density were found decreased in patient with SSC at the results of OCTA measures. These findings may help to understand vasculopathy in the pathogenesis of the disease and OCTA may be a new method providing objective and non-invasive information about capillary network in SSC.

Table. Demographic and ocular parameters of study population

<table>
<thead>
<tr>
<th>Parameters</th>
<th>SSc (n = 40)</th>
<th>Control (n = 40)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, years; mean (SD)</td>
<td>472.8 (6.4)</td>
<td>475.8 (8.1)</td>
<td>0.631</td>
</tr>
<tr>
<td>Male sex, n (%)</td>
<td>24 (60)</td>
<td>16 (40)</td>
<td>0.087</td>
</tr>
<tr>
<td>Disease duration, months; mean (SD)</td>
<td>813.8 (38.0)</td>
<td>812.9 (40.5)</td>
<td>0.585</td>
</tr>
<tr>
<td>Foveal MT(µ)</td>
<td>246.3 ± 19.4</td>
<td>252.6 ± 15.3</td>
<td>0.033</td>
</tr>
<tr>
<td>Average MT(µ)</td>
<td>280.8 ± 12.4</td>
<td>286.5 ± 8.7</td>
<td>0.008</td>
</tr>
<tr>
<td>Vessel density (mm⁻¹), 6 mm total area; mean (SD)</td>
<td>1760 ± 13.1</td>
<td>1866 ± 0.64</td>
<td>0.006</td>
</tr>
<tr>
<td>Perfusion density, 6 mm total area; mean (SD)</td>
<td>432.3 ± 33.2</td>
<td>459.4 ± 1.52</td>
<td>0.002</td>
</tr>
<tr>
<td>Circularity index; mean (SD)</td>
<td>0.72 ± 0.09</td>
<td>0.73 ± 0.06</td>
<td>0.049</td>
</tr>
<tr>
<td>RNFL nasal (µ); mean (SD)</td>
<td>74.37 ± 12.96</td>
<td>74.05 ± 8.48</td>
<td>0.011</td>
</tr>
<tr>
<td>RNFL inferior (µ); mean (SD)</td>
<td>122.62 ± 17.87</td>
<td>127.40 ± 12.63</td>
<td>0.023</td>
</tr>
<tr>
<td>Inferior nasal GCC(µ); mean (SD)</td>
<td>85.72 ± 8.53</td>
<td>85.82 ± 4.84</td>
<td>0.001</td>
</tr>
<tr>
<td>Inferior temporal GCC(µ); mean (SD)</td>
<td>82.70 ± 8.62</td>
<td>84.95 ± 4.15</td>
<td>0.011</td>
</tr>
<tr>
<td>Superior nasal GCC(µ); mean (SD)</td>
<td>86.35 ± 17.73</td>
<td>86.8 ± 5.70</td>
<td>0.012</td>
</tr>
<tr>
<td>Superior temporal GCC(µ); mean (SD)</td>
<td>472 ± 8.6</td>
<td>475.8 (1.8)</td>
<td>0.631</td>
</tr>
</tbody>
</table>

Conclusion: Almost two thirds of our cohort of SSC have symptomatic gastrointestinal disease. GIT manifestations are heterogeneous. Symptoms are non-specific and overlapping for a particular anatomical site. Esophagus is the most commonly affected. More than seventy-five per cent of patients experience symptoms of gastroesophageal reflux. We did not find differences among patients with and without SSC GIT disease. 17% of patients had a Reynold’s syndrome.

References:

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.4055