Methods: We enrolled SSC patients without PH from January 2016 to December 2019 in our institution. SSC patients were diagnosed according to EULAR classification criteria in 2013.ILD was detected by chest CT scans. We assessed severity of ILD with pulmonary function tests (PFT). Abnormal PFT was defined as vital capacity (%VC) or diffusion capacity (DLCO) < 70%. NFC abnormalities were detected with “OptiPiX capillaroscopy Clinic 1.x” and the number of capillaries was measured per 1mm in 2nd to 5th fingers of both hand. We defined enlarged and giant capillaries as >30 µm and >50 µm, respectively.

Results: We enrolled 59 SSC patients (64 females, 5 males). Mean age is 65.0 ± 8.0 years. Thirty-one patients (52.5%) were complicated with ILD. Mean capillary counts are 6.6/mm. The number of patients with each NFC abnormalities (enlarged capillaries, giant capillaries, microhemorrhages, ramified, avascular areas) are 42, 32, 38, and 33, respectively. Two cases did not have NFC abnormalities. SSC patients with giant capillaries had fewer ILD complications (p <0.05, odds ratio 0.183 [0.059 – 0.57]). Other NFC abnormalities were not associated with ILD in SSC patients. We inspected %VC of 23 patients and DLCO of 20 patients with ILD. Eleven patients had abnormal PFT (5 patients had abnormal %VC and 9 patients had abnormal DLCO). Most of them had not enlarged capillaries than patient with normal PFT (odds ratio 0.11 [0.016 – 0.81]). Other NFC abnormalities including giant capillaries were not associated with abnormal PFT.

Conclusion: We investigated the relationship between NFC abnormalities and ILD complications in SSC patients. NFC abnormalities are associated with ILD complications and severity of ILD. It was suggested that no giant capillary in SSC patients may predict ILD complication. Moreover, no enlarged capillary may predict the severe ILD. NFC abnormalities are associated with ILD complications in SSC patients.

References:

Table 1. NFC abnormalities in SSC patients

<table>
<thead>
<tr>
<th>With ILD (n=31)</th>
<th>Without ILD (n=28)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>enlarged capillaries</td>
<td>19</td>
<td>23</td>
</tr>
<tr>
<td>giant capillaries</td>
<td>11</td>
<td>21</td>
</tr>
<tr>
<td>microhemorrhage</td>
<td>25</td>
<td>23</td>
</tr>
<tr>
<td>ramified</td>
<td>21</td>
<td>17</td>
</tr>
<tr>
<td>avascular areas</td>
<td>19</td>
<td>14</td>
</tr>
</tbody>
</table>

Table 2. NFC abnormalities in SSC patients with ILD

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AB0594 EFFECTIVENESS OF RITUXIMAB IN CSDMARD-RESISTANT ACTIVE MIXED CONNECTIVE TISSUE DISEASE

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Background: Current, most authors accept that mixed connective tissue disease (MCTD) is an independent entity, although there are those who argue that it is actually an overlap syndrome with an undifferentiated early phase of another systemic autoimmune disease (SAD).

Objectives: To analyze the long term evolution of a serie of patients with MCTD.

Methods: Observational, retrospective and multicenter study in patients with MCTD (diagnostic criteria of Alarcón-Segovia et al), followed for a minimum of 2 years.

Results: Fifty-five patients (49 women) with a median age at diagnosis of 38±14 years and with a follow up time (median) of 101 months (range, 24-237 months with a total of 501.2 patients-year) were identified.

At the end of the follow-up period, only 27% (15/55) of the patients kept on fulfilling MCTD criteria. In the remaining 73% (40), 40% (22) had been differentiated to systemic lupus erythematosus (SLE), 13% (7) to systemic sclerosis (SSc) and 20% (11) developed an overlap syndrome [SSc+SLE in 8 cases and SSc+rhematoid arthritis (AR) in 3]. In 8% of these patients, a secondary Sjögren’s syndrome was diagnosed during the follow-up period. The average score in patients who met the EULAR/ACR 2013 criteria for SSc was 11 (minimum 9 - maximum 16) and the average time elapsed from the diagnosis of MCTD to meet SSc criteria was 64.4 months (interquartile range [IQR] 25-75%; 10-127 months).

Applying the 2012 SLICC criteria, only 24 patients of those initially diagnosed as MCTD ended up meeting SLE criteria. The average score in these patients was 5.6 (4-9) and the average time elapsed from the diagnosis of MCTD until fulfilling the SLICC criteria was 59 months (IQR 25-75%; 6-28). When we apply the new ACR/EULAR 2019 criteria, the percentage of patients who meet SLE criteria increased to 30%, with an average score of 17.3 (10-38). The average time elapsed since the diagnosis of MCTD until meeting the new SLE criteria was reduced to 17 months (IQR 25-75; 0-10).

In the multivariate study, the presence of sclerodertry (OR: 2.91; IC 95% 1.90 - 4.1, p = 0.001) and esophageal involvement (OR: 2.05; IC 95% 1.14–3.66, p=0.016) were associated with the evolution to SSc. Any predictor of evolution to SLE was identified.

Conclusion: Only slightly more than a quarter of patients initially diagnosed as MCTD maintain this diagnosis during the follow-up. The majority, ended up evolving towards to another SAD, fundamentally SLE and SSc. The new ACR/EULAR 2019 criteria seems to be more sensitive than the SLICC 2012 criteria for diagnose SLE in these patients.

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**AB0595**

**EFFECTIVENESS OF TOPICAL SODIUM TIOSULFATE PASTE FOR THE TREATMENT OF CALCINOSIS-ASSOCIATED CUTANEOUS ULCERS IN PATIENTS WITH SYSTEMIC SCLEROSIS**

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**Background:** Treatment of calcinosis associated with systemic sclerosis (SSc) mainly involves the use of systemic therapies, which often have limited efficacy. However, little attention has been paid to local treatment, which is especially useful when associated with skin ulcers.

**Methods:** Descriptive analysis of a case series of patients with SSc and calcinosis-associated skin ulcers treated with TST. Wound management procedure: wounds and perilesional skin cleaning and disinfection is performed and, if needed, additional debridement. TST is compounded at 25%, w/v emulsion, for extensive calcinosis, or as beeler-base or cold-cream ointment, for limited calcinosis. Wounds are then covered with a polymeric foam dressing. This cure in moist healing environment shows some advantages over the dry cure (exudate control without damaging the perilesional skin, protection against contamination, and reduction of the needed cures, healing time and pain).

**Results:** Nine patients (7 women) with calcinosis-associated skin ulcers and SSc were included: 2 patients with diffuse SSCs (DsSSc), 6 with limited SSCs (LcSSc) and 1 with overlap syndrome. Mean age was 60 years (IQR 20), 6 patients had localized wounds and 3 had extensive involvement and/or tumoral calcinosis which had been refractory to systemic treatment with diltiazem, colchicine, zoledronate, rituximab, and/or acenocoumarol and had suffered recurrent superinfections. Follow-up results of more than 3 months are available for 8 patients, who have been on TST a median time of 9 months (IQR 8.25). They have shown clinical improvement (disappearing of many calcinosis foci and partial or complete healing of the ulcers together with an improvement in pain, function, quality of life and satisfaction of the patients).

**Conclusion:** In our experience, treatment with TST for calcinosis-associated skin ulcers in patients with SSc is an effective, safe and easily implementable therapeutic alternative in clinical practice.

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