EFFECTIVENESS OF TOPICAL SODIUM TIOSULFATE 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 needed, additional debridement. TST is compounded at 25% as w/o emulsion, for extensive calcinosis, or as beeler-base or cold-cream ointment, for limited need, additional debridement. TST is compounded at 25% as w/o emulsion, for extensive calcinosis, or as beeler-base or cold-cream ointment, for limited need. Wounds are then covered with a polymeric foam dressing. This cure in control without damaging the periulceral skin, protection against contamination, moisturizing the wound, and creating a moist healing environment shows some advantages over the dry cure (exudate barrier). For minimally involved areas, TST can be used as a poultice. The results are divided into four main sections: clinical improvement, healing, and adverse effects.

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%, as w/o 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 control without damaging the periulceral skin, protection against contamination, moisturizing the wound, and creating a moist healing environment shows some advantages over the dry cure (exudate barrier). For minimally involved areas, TST can be used as a poultice.

Results: Nine patients (7 women) with calcinosis-associated skin ulcers and SSC were included: 2 patients with diffuse SSC (DcSSc), 6 with limited SSC (LcSSc) and 1 with overlap syndrome. Median 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 (diltiazem, colchicine, zolendronic acid, ruxolitinib, and/or aconcomocorol) 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 2.5). They showed clinical improvement (disappearance of the majority of 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). Radiological improvement was also observed in 1 case. No TST related adverse effect has been detected, except for slight maceration of the wound edges due to the ointment preparation, which was resolved by protecting these with zinc oxide cream.

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

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PREDICTORS, LONG TERM CLINICAL AND TREATMENT OUTCOMES IN SOUTH ASIAN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOSITIS: A SINGLE CENTER STUDY

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Background: Idiopathic inflammatory myositis (IM) are a heterogeneous group of immune-mediated disorders with varied presentations and multiple organ involvement. Data on long term outcome among South Asian patients with IM is sparse.

Objectives: To study the long term clinical outcome, treatment responses and factors predicting outcome among adult patients with IM.

Methods: Patients diagnosed as ‘Idiopathic Inflammatory Myositis’ under the department of Clinical Immunology and Rheumatology at CMC, Vellore, India were screened retrospectively. Patients aged 18 years and above, satisfying Bohan and Peter criteria, having follow up of one year or more with at least two outpatient or inpatient visits between January 2010 and April 2019 were included in this study. Those patients with connective tissue disease associated calcinosis were not included. Details on muscle weakness, extramuscular involvement, muscle enzymes and treatment administered were recorded at baseline, 3, 6, 12, 18, 24 months and yearly thereafter. After assessing their cumulative response, categorization of patients into complete and partial responders was done. Complete responders were defined as patients with persistent muscle power of more than 4/5 and/or MMT 8 more than 75/80, complete resolution of skin, articular and lung involvement (if any) as well as muscle enzymes less than twice the upper limit of normal without any documented flares during the entire follow up period. Patients not satisfying the said criteria were grouped as Partial responders. Disease free survival duration was also analyzed.

Results: Out of 310 patients of IM identified, 187 (60.3%) patients satisfied the inclusion criteria. Women were 2.2 times more than men and mean age at symptom onset was 35.7±12.6 years. Dermatomyositis was the predominant myositis subtype seen. All patients were put on steroids with the mean dose being 45.9 ± 18.6 mg/day. At baseline, the key immunosuppressants used were methotrexate in 44.9% and mycophenolate in 37.6% patients. The median follow up duration was 48 (25-80) months. An associated malignancy was diagnosed in 3.2% after a median duration of 24.5 months. Five patients expired after a median duration of 60 months from diagnosis. Normal muscle power was attained in 76.1% patients and 88.6% were vocational by the last follow up visit. Steroids were discontinued in 56.7% patients after a median duration of 24 months (p=0.0002). Discontinuation of the immunosuppressant was feasible in 10.2% patients after a median duration of 44 months. Assessment of

Figure 1: Disease free survival plot of patients with IM
PULMONARY FUNCTION IN PATIENTS DIAGNOSED OF EARLY SYSTEMIC SCLEROSIS: A NEW TOOL FOR SYSTEMIC SCLEROSIS CLASSIFICATION?

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Background: Interstitial lung disease (ILD) is a frequent complication of systemic sclerosis (SSc) and is often progressive and has a poor prognosis. A restrictive ventilatory defect could suggest ILD either alone or in combination with pulmonary arterial hypertension. Nowadays, Early-SSc is well defined as preliminary stage of SSc. Patients who meet criteria for Early-SSc could benefit from an early diagnosis of pulmonary involvement.

Objectives: Our aim was to assess the pulmonary function in patients diagnosed of Early-SSc.

Methods: Retrospective observational study of a wide and unselected series of patients diagnosed as Early-SSc from a single university hospital from 2012 to 2019. Patients were classified as Early-SSc following Le Roy criteria. Despite this, patients already did not meet 2013 ACR/EULAR classification criteria for SSc. We reviewed pulmonary function by conventional spirometry and diffusing capacity of lung for carbon monoxide (DLCO).

Results: We included 56 patients with a mean age of 52.3±12.1 years (96.4% women; 3.6% men). At the diagnosis of Early-SSc, no one of our patients evidenced a restrictive ventilatory pattern. DLCO was below normal limits in 18 patients (32.1%). Small airway obstruction expressed according decreased maximal (mid-) expiratory flow (MMEF) 25-75 was present in 24 patients (42.8%). After a mean follow-up period of 38.3±2.4 months, 29 (51.8%) patients fulfilled 2013 ACR/EULAR criteria. The average time between diagnosis of Early-SSC and achieve SSSC classification was 24.4±1.8 months. The remaining 27 patients continued classified as Early-SSC.

An analysis of the subgroup of patients which progressed to SSc showed that DLCO was decreased in 15 of those 29 patients (51.7%) and 18 of 29 patients (62.1%) presented decreased MMEF 25-75. Comparing with the subgroup of patients which not progressed to SSSC were significant differences (Decreased DLCO: 51.7% vs 11.1%; p=0.02 and decreased MMEF 25-75: 42.8% vs 22.2%; p=0.05).

The analysis of pulmonary function of the subgroup of patients continued classified as Early-SSC after follow-up period did not show significant changes after follow-up.

Conclusion: In our study, a third of the patients classified as Early-SSC presented at diagnosis abnormal values of DLCO and/or signs of small airway obstruction without the presence of a restrictive ventilatory pattern. Moreover, this pulmonary dysfunction was significantly more frequent in patients who progressed to definitive SSc. Patients which remains classified as Early-SSC did not experience significant changes.

Our results support the concept that pulmonary function was impaired in Early-SSC and that I should probably be considered for future Early-SSC classification criteria.

Table 1. Proportion of patients receiving immunosuppressive treatment at each year after disease onset in SSc diagnosed before and after 2007.

<table>
<thead>
<tr>
<th>Years after the first non-RP symptom</th>
<th>Total N of pts seen at each year</th>
<th>% receiving immunosuppressives</th>
<th>Total N of pts seen at each year</th>
<th>% receiving immunosuppressives</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Before 2007</td>
<td></td>
<td></td>
<td>After 2007</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>13</td>
<td>15</td>
<td>47</td>
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<td>21</td>
<td>82</td>
<td>18</td>
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<tr>
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<td>49</td>
<td>14</td>
<td>107</td>
<td>14</td>
<td>&gt;0.9</td>
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<tr>
<td>lcSSc</td>
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<td>dcSSc</td>
<td></td>
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<tr>
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</tbody>
</table>

IS administration was associated with male gender, ILD, a-Scl-70 positivity, ACA-negativity and U disease in lcSSc, and with ACA-negativity and a higher mRSS in dcSSc. Multivariate logistic regression analysis showed that IS treatment could be predicted by ACA-negativity in lcSSc patients (Exp(B) 0.317, p=0.012) and younger age in dcSSc patients (Exp(B) 0.974, p=0.002).

Conclusion: Over the past decade, there has been a trend to prescribe IS more often, especially MTX, and earlier in dcSSc patients. MMF has gained favour over CYC. Autoantibody status was the most consistent predictor whether a patient is likely to take IS over the course of the disease.

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