AB0421

MANAGEMENT OF ULCERATION IN SYSTEMIC SCLEROSIS BY A SPECIALIST PODIATRY SERVICE; A CASE SERIES

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Background: Systemic sclerosis is a chronic disorder characterised by diffuse fibrosis and vascular abnormalities in the skin and major organs. Digital ulceration is a common complication, often resulting in patient disability. These ulcers can form through a variety of processes and management should be tailored according to aetiology. In Dorset, the specialist podiatry service has created a unique facility for optimal wound care in the setting of digital ulceration.

Objectives: In this case series, we outline podiatric management of 4 common causes of digital ulceration in systemic sclerosis.

Methods: Tissue ischaemia, infection, micro-trauma and calcinosis were identified as four major contributory factors resulting in ulceration or delayed wound healing. Four cases exemplifying management of each different contributory factor were identified and their case notes reviewed in order to extract clinically relevant data and images.

Results: Case 1 illustrates treatment of ischaemic tissue injury to promote earlier healing, using careful wound care, debridement under local anaesthetic and low-level laser therapy.

Case 2 illustrates the importance of identifying repeated micro-trauma as a contributing factor to non-healing ulceration.

Case 3 demonstrates how surgical debridement or removal of the subcutaneous calcinosis in an outpatient setting can be a useful adjunct in encouraging ulcer healing.

Case 4 illustrates the complexity of correctly identifying infection in the setting of ulceration, with prompt management promoting wound healing.

Conclusion: The pathophysiology of ischaemic tissue damage in scleroderma is complex and can reflect a broad range of vascular pathologies, often occurring concurrently¹ including vasospasm, macrovascular disease and microvascular vasculopathy, leading to digital ulceration. Rapid identification of which processes are driving the ischaemia is critical in targeting therapy to the affected individual. A local podiatry service with expertise in management of tissue-related complications of systemic sclerosis is invaluable in promoting wound healing and preventing further complications.

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AB0422

DIAGNOSTIC PERFORMANCE OF THE ACR/EULAR 2013 CLASSIFICATION CRITERIA FOR SYSTEMIC SCLEROSIS IN A ROUTINE CARE SETTING

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Background: An ACR/EULAR task force released new criteria in 2013 to classify patients with systemic sclerosis (SSc).

Objectives: This study evaluates the diagnostic performance of these criteria in a multidisciplinary care setting.

Methods: Patients with an active follow-up in a Systemic Autoimmune Diseases Unit with a clinical diagnosis of SSc were matched by age and gender with consecutive patients referred to a capillaroscopy clinic. The classification criteria were tested on discrimination and diagnostic accuracy between both groups of patients defined as cases-SSc and controls. Receiver operating characteristic (ROC) curve and the area under the curve (AUC) was calculated for the global score to define the best cut off to classify the patients as having SSc.

Results: A total of 130 patients with SSc and 130 matched-controls were included in this analysis, 90% women, with a mean age of 61.5. Main diagnosis for the control groups were primary Raynaud's phenomenon (34.6%), undifferentiated connective tissue disease (13.1%), and mixed connective tissue disease (9.2%). The 92% and 8% of patients in the SSc-cases and control groups met the 2013 ACR/ EULAR SSc classification criteria respectively. Sensitivity and specificity of the criteria were 81.5% and 93.7%, respectively. The best cut offs for the total score were 8 and 9, and the AUC (95%CI) was 0.962 (0.939-0.985). The individual items with a better discriminatory capacity were abnormal capillaroscopy, telangiectasia and antinuclear antibody positivity.

Table 1. Demographic data and ACR/ EULAR SSc classification criteria of SSc patients and controls.

	Cases-SSc, n= 130	Controls, n= 130	p
Age, mean (SD)	62.4 (16.0)	61.3 (14.8)	0.58
Female, %	90.5	90.8	0.92
Disease duration from onset of symptoms, mean (SD)	7.5 (6.4)	7.8 (6.7)	0.73
% Patients with individual ACR/EULAR 2013 Criteria Items			
Skin thickening	15.8	0.7	<0.001
Sclerodactyly	45.5	6.1	<0.001
Puffy fingers	27.7	1.5	<0.001
Digital tip ulcers	30.2	5.3	<0.001
Fingertip pitting scars	18.8	4.6	0.001
Telangiectasia	51.5	6.1	<0.001
Abnormal nailfold capillaries	79.4	0.8	<0.001
Pulmonary Arterial Hypertension	16.3	3.8	0.001
Pulmonary fibrosis	18.8	6.2	0.002
Raynaud's phenomenon	90.7	66.9	<0.001
Anticentromere antibody	60.7	18.9	<0.001
Sci70	12.0	0	<0.001
Total Score	12.5 (4.8)	3.4 (2.9)	<0.0001

Figure 1. ROC curve for global score of the ACR/EULAR2013 SSc classification criteria.

Conclusion: The ACR/EULAR 2013 criteria showed good diagnostic properties in this cohort reflecting daily practice. Individual items showing the highest discriminatory capacity were abnormal capillaroscopy, telangiectasia and antinuclear antibody positivity.

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AB0423

SAFETY AND TOLERABILITY OF RITUXIMAB IN THE TREATMENT OF SYSTEMIC SCLEROSIS: LONG-TERM FOLLOW-UP

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Background: Rituximab (RTX) has been used for the treatment of systemic sclerosis (SSc) for a long time, but data on tolerance and long-term adverse events (AE) are insufficient.

Objectives: To assess the tolerability and safety of RTX in the patients (pts) with SSc in long-term prospective follow-up.

Methods: Data on the safety and tolerability of RTX were evaluated in 149 SSc pts who received at least one RTX infusion in a long-term open-label prospective observational study. The mean age was 48±13.5 years (17-74), women - 122 (82%), diffuse cutaneous subset of the disease had 52%, limited-37% and overlap-11%.

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±4.8mg/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 (CI) 8-11). 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 2.22/100 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 (n=53), with no serious opportunistic infections reported. The overall incidence of all infections was 6.4/100 PY (95% CI 4.9-8.3), serious infections – 1.32/100 PY (95% CI 0.7-2.4). The level of immunoglobulin G during follow-up period decreased from 12.9±4.9 to 10.1±3.4g/l ($p=0.0001$), but remained within normal limits. Infusion reactions occurred in 15 pts (1.8/100 PY, 95% CI 1-3). Other AE were observed in 9 pts (6%) (1.1/100 PY, 95% CI 0.53-2.12). Sixteen deaths were recorded – 10.7% or 1.91/100 PY, 95% CI 1.2-3.1. In most cases, pts died from the progression of the major organ failure. The causes of death were: progression of the interstitial lung disease (ILD) in 4 pts, heart failure associated with SSc cardiomyopathy (2), renal crisis (4), pulmonary arterial hypertension and ILD (2), pneumonia (2), sepsis after tooth extraction (1), acute pulmonary embolism (1).

Conclusion: In our study, we considered the overall safety profile of RTX in SSc as favorable. It was similar to the AE profile in other autoimmune diseases treated with RTX. With an increase of the cumulative dose of RTX, there was no increase in AE. RTX could be considered as a relatively safe drug for the complex therapy of SSc when standard therapy is ineffective or impossible.

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AB0424

NUTRIENT DEFICIENCIES IN SYSTEMIC SCLEROSIS: A SYSTEMATIC REVIEW

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Background: Malnutrition is a critical concern in patients with systemic sclerosis (SSc).^{1,2} However, the extent and types of nutrient deficiencies in SSc remain unclear.

Objectives: 1) To identify the nutrient deficiencies commonly reported in SSc; 2) To evaluate associations between specific nutrient deficiencies and SSc subtype and clinical manifestations.

Methods: We conducted a systematic review of all published reports on SSc and nutrition in PubMed from its inception to August 2020. Clinical trials, observational studies, and case series (with ≥20 cases) containing data on nutritional deficiency and SSc were included. We followed the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) for reporting our findings. Two reviewers (ADN and ERV) studied the titles and abstracts of all search results with pre-specified inclusion and exclusion criteria (Figure 1).

Results: Among 790 retrieved publications, 35 full-length articles and 3 abstracts met the inclusion criteria. Included studies took place across multiple geographic locations and included patients with both diffuse and limited cutaneous SSc. Vitamin D deficiency was the most commonly reported deficiency described in SSc, followed by vitamin B12, folate, selenium, zinc, and iron (Table 1). In addition, some small studies found deficiencies in thiamine, pyridoxine, alpha-tocopherol, and carotene. Interstitial lung disease was associated with vitamin D deficiency and elevated homocysteine (Hcy), while pulmonary hypertension was associated with elevated Hcy. Vitamin D deficiency was also associated with increased modified Rodnan skin score (mRSS) and increased risk of SSc-related organ involvement.

Conclusion: Nutrient deficiencies are common in SSc and are associated with specific SSc features. Routine screening for nutrient deficiencies may lead to early detection of malnutrition. Future studies are needed to understand how nutritional interventions affect patient outcomes in SSc.

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Figure 1. Article selection for systematic review.

Table 1. Nutrient deficiencies and their clinical correlates in SSc.

	Number of studies that found deficiency ^a	Number of studies that did not find deficiency	Clinical correlates of nutrient deficiency
Vitamin D	23	0	■Increased risk of SSc-related organ involvement ■ILD ^c
Homocysteine ^b	5	4	■Higher mRSS ^c ■ILD ^c ■PH
Vitamin B12	3	4	■None identified
Folate	3	2	■None identified
Vitamin C	2	0	■None identified
Selenium	5	0	■Increased risk of SSc-related organ involvement
Zinc	3	1	■None identified
Iron	1	1	■None identified
Copper	0	2	■None identified
Other nutrients			■None identified for all nutrients
Thiamine	1	0	
Pyridoxine	1	0	
Alpha-tocopherol	1	0	
Carotene	1	0	

^a Deficiency defined as below normal levels based on study reference ranges^b Studies looked for elevation versus no elevation of homocysteine in SSc^c Some studies found significant correlation while others did not