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ABSTRACT SUPPLEMENT

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Results: NOSVAR includes 381 patients with SSc. 270 SSc patients (71%) fulfil the ACR criteria. The remaining 111 SSc patients meet the modified Medsger and LeRoy criteria for SSc. The mean age at onset is 48 years (SD 0.8), the median disease duration is 10 years (range 1–48 years) and the female to male ratio is estimated to 4:1. The cohort consists of 7 limited (l) SSc patients, 277 limited cutanoeus (lc) SSc patients and 98 diffuse cutaneous (dc) SSc patients. Altogether, 73/381 patients (19%) have bilateral skin thickening extending proximal to the metacarpophalangeal (MCP) joints and are classified as SSc by this criterion alone. Of the remaining 302 patients, 296 gain >9 points by fulfilling other parameters defined in the new ACR/EULAR criteria. Only 6 patients (2%) in the NOSVAR cohort do not meet the new criteria. These 6 patients fulfil only the LeRoy and Medsger's modified SSc criteria and have Raynauds phenomenon, pathological capillaroscopi and a positive antinuclear antibody, but no skin changes.

Conclusion: This study demonstrates the applicability of the new SSc classification criteria in a large, clinical registry cohort. Only 2% of SSc patients in the cohort do not meet the new ACR/EULAR criteria.

References:

1. Van den Hoogen, F *et al*. Classification criteria for Systemic Sclerosis; preliminary results. Annals Rheum Dis 2013;72 (3): 59.

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Descriptive Review Of Morphea Subjects From a Single Cohort Center. Lesley Portugal[1], Muneera Naeem[2], Lakshmi N. Moorthy[3] and Vivien M. Hsu[4]. [1]Robert Wood Johnson Medical School, New Brunswick, NJ, [2]Mountainside Hospital, Montclair, NJ, [3]Robert Wood Johnson Medical School-UMDNJ, New Brunswick, NJ, [4]RWJ Med Schl Scleroderma Prog, New Brunswick, NJ.

Background/Purpose: Morphea, or localized scleroderma, is an idiopathic, rare fibrotic skin disorder that may result in tissue atrophy, pigment changes, and contractures. Disability may range from purely cosmetic, causing great anxiety, to lifelong functional disability.

Methods: We conducted a retrospective chart review of pediatric and adults diagnosed with morphea at UMDNJ, and data on types of morphea, possible triggering events, disease course, complications and therapies. Descriptive analysis was done on all variables.

Results: Table 1 lists the clinical manifestations: there were 26 subjects (2 males), mostly Caucasians females of mean age 45±24 (10–85 years). Five had known associated autoimmune disease. Only 2 patients had a family history of morphea. Thirty-eight percent had a family history of autoimmune disease including SLE, RA, Crohn's and thyroid disease. Types of morphea, and known triggering events are listed in table 1.

Table 1. Clinical manifestations of 26 morphea subjects

Variables	*Frequency* (n = 26)	*% frequency*
Triggering event		
Unknown	23	88
Lovenox injection	1	
Chicken pox	1	
Previous Schamberg's	1	
Morphea type	7	27
Plaque	8	31
Generalized SQ	3	12
Linear	4	12
Coup de Sabre mixed	3	12
Eosinophilic fasciitis	1	< 1

Sizes ranged from small (<3±1.5 cm) to large (16±16 cm) with (85%) associated itching, tightness, dryness, burning, pain, or contracture requiring intervention. More than 90% had more than one lesion involving the trunk or extremity. Generalized SQ and eosinophilic fasciitis were only found in adults. Elevated CPK and inflammatory markers were more common with extensive disease. Extra-cutaneous manifestations presented in 18 subjects, including arthralgias, fatigue, and Raynaud. Two adults had monoclonal gammopathy associated with GSM. Tissue atrophy occurred in 14 subjects and 81% had long-term cosmetic issues (pigmentary changes), pain or depression due to their lesions. One was functionally disabled.

Twenty-four patients received the following therapy: Topical steroid (n=1), intra-lesional steroid (n=1), UV phototherapy (n=1). Immunosuppression was generally used for extensive disease: Methotrexate (n=17) alone or with systemic corticosteroids, etanercept, penicillamine, hydroxychloroquine, cyclosporine and cyclophosphamide. One patient with GSM required skin grafting. Morphea improved but did not resolve in 19 subjects; only one had resolution after 6 years. One patient had a relapse, two progressed and one child had limb length discrepancy. Of 18 patients with extra-cutaneous manifestations, 5 improved concurrently with their skin lesions.

Conclusion: Many morphea subjects sought therapy for their discomfort or complications. Long-term consequences, including joint pains and cosmetic issues, did not always resolve despite improvement of the morphea. More studies are needed to understand the various types of morphea and its clinical consequences to improve long-term outcomes.

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Characterization Of Lower Limb Cutaneous Ulcers In Systemic Sclerosis: The Analysis of 424 Lesions. Jelena Blagojevic[1], Guya Piemonte[2], Laura Benelli[2], Francesca Braschi[2], Ginevra Fiori[2], Felice Galluccio[2], Francesca Bartoli[2], Lorenza Busco[2], Alberto Pignone[2], Giulia Carnesecchi[2], Gemma Lepri[2], Serena Guiducci[2] and Marco Matucci Cerinic[2]. [1]University of Siena, Siena, Italy, [2]University of Florence, Florence, Italy.

Background/Purpose: Cutaneous ulcers reppresent one of the most frequent complications in course of systemic sclerosis (SSc). They are often disabling and difficult to treat. The upper limb ulcers have been evaluated and characterized extensively, but there are only few studies on lower limb ulcers. SSc is characterized by a microangiopathy that represents the hallmark of disease, but concomitant alterations of arterial, venous and lymphatic circulation may contribute to the pathogenesis of the lower limb cutaneous lesions. The aim of the study is to assess the pathogenesis, characteristics and time to healing of the lower limb cutaneous lesions in course of SSc.

Methods: Fifthy-seven consecutive SSc patients with lower limb cutaneous lesions were followed up for four years. All patients performed an accurate health examination and evaluation of cutaneous lesions, routine blood and urine tests with autoantibodies, lipid and glycemic profile and creatinine clearance, videocapillaroscopy and arterial and venous lower limb Color Doppler Ultrasonography. Arteriography was performed in patients with occlusive peripheral arterial disease.

Results: Four hundred and twenty-four (424) lower limb cutaneous lesions were observed. Lesions were divided into: hyperkeratosis, ulcers (loss of tissue) and gangrene. We observed: 275 (64,9%) hyperkeratosis, 144 (33,9%) ulcers and 5 (1,2%) gangrene. The ulcers were subsetted in: primary ulcers (107 (74,3%)), ulcers secondary to hyperkeratosis (17 (11,8%)) and ulcers secondary to calcinosis (20(13,9%)). The mean time to healing was 152 ± 202 days, and recurrence was observed in 31,6% of lesions. The prevalence of amputations was 1,2%. As regards pathogenesis, 16 (28,1%) patients had a significative peripheral arterial disease, 22 (38,6%) had venous insufficiency and 7 (12,4%) presented lymphedema, besides the microangiopathy. Three patients presented simultaneously a peripheral arterial disease and venous pathology. One patient presented lymphedema and venous insufficiency and two patiens had lymphedema and peripheral arterial disease. Four patients with critical arterial stenosis performed the lower limb angiography which confirmed the presence of stenosis with distal distribution.

Conclusion: Our data indicate that lower limb lesions have often a multifactorial pathogenesis in SSc. This is the first study that characterized extensively a large number of lower limb cutaneous lesions in SSc. The comprehension of characteristics and pathogenesis of these lesions is essential for their correct management.

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