Symptoms of autonomic dysfunction in patients with systemic sclerosis

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Objectives: This study aims to assess prevalence, severity and clinical correlates of symptoms of autonomic dysfunction in patients with SSc.

Methods: Fifty five consecutively recruited SSc patients were included in this study. Thirty seven (67.3%) patients had limited (lcSSc), whilst 18 (32.7%) patients had diffuse cutaneous SSc (dcSSc). Anticentromere antibodies (ACA) were positive in 31 (56.4%) of patients, whilst 28 (26.4%) patients had anti-topoisomerase I antibodies (ATA). All patients completed the Composite Autonomic Symptom Score (COMPASS-31) questionnaire, which consists of 31 items, quantifying six autonomic domains: orthostatic intolerance (OI), vasomotor (VD), secretomotor (SD), gastrointestinal (GD), bladder (BD) and pupillomotor dysfunction (PD). The total score range from 0 to 100, whilst scores for particular domains range as follows: 0-40 for OI, 0-5 for VD, 0-15 for SD, 0-25 for GD, 0-10 BD, and 0-5 for PD. Higher values representing more severe symptoms.

Results: Percentage of SSc with a score >0 in particular domains of the COMPASS-31 were as follows: OI 32/55 (58.2%), VD 49/55 (89.1%), GD 36/55 (65.5%), BD 40/55 (72.7%), PD 30/55 (54.5%). The COMPASS-31 score did not correlate with age or disease duration. There was no relationship between the COMPASS-31 total or subdomain scores and SSc subtype or autoantibody status. Similar values for mean total and subdomain scores were found among patients with different capillaroscopic patterns. Patients with DLCO < 80% had significantly higher mean values of GD, BD and PD scores, compared to patients with DLCO ≥80% (4.42 vs 2.75, 1.51 vs 0.38, 1.93 vs 1.09, respectively). Moreover, the total COMPASS-31 score was significantly higher in patients with decreased DLCO (16.4 vs 11.34, p=0.008).

Conclusion: Symptoms of dysautonomia are common in SSc patients. Patients with a more severe disease, especially decreased lung diffusing capacity, joint pain, and severe gastrointestinal involvement, report more symptoms of autonomic dysfunction.

Disclosure of Interests: None declared

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EVALUATION OF BODY COMPOSITION AND BONE STATUS ACCORDING TO MICROVASCULAR INVOLVEMENT IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Systemic sclerosis (SSc) is a complex autoimmune connective tissue disease, characterized by autoimmunity inflammatory microvascular damage with progressive loss of capillaries, fibrosis and ischemia of skin and internal organs. (1) Nailfold videocapillaroscopy (NVC) is a safe tool for early diagnosis of SSc, it identify morphological changes of vessel that are predictive for clinical disease progression and organ involvement.(2) About clinical complication the loss of bone mass and body composition abnormalities, particularly muscle mass and strength loss (sarcopenia), are recognized in advanced disease.(3) Objectives: To evaluated in SSc patients, the body composition and the bone mass according to the microvascular condition, as assessed and scored by nailfold videocapillaroscopy (NVC, “Early”/“Active”/“Late” patterns).

Methods: Body composition and bone mineral density (BMD) were assessed by DXA in 35 female SSc patients classified according to the 2013 EULAR/ACR criteria and 32 sex-matched healthy subjects. Clinical, laboratory, body composition and bone parameters were analysed according to the different NVC patterns. Means were compared by the Student’s t test or one way analysis of variance; medians were compared by the Kruskall Wallis test; and frequencies by the chi square test.

Results: Higher prevalence of vertebral (26.4% vs 9.3%) and femoral (32.3% vs 9.3%) osteoporosis (OP) was found in SSc. Particularly SSc patients with “Late” NVC pattern showed a significantly higher prevalence of vertebral (p=0.018) and femoral OP (p=0.016). Regional assessment of bone mass (BM) in 7 different body areas showed a significant lower BMD only at the total spine (r=-0.008) and femoral neck (r=0.027) in advanced microvascular damage. Patients with “Late” NVC pattern showed lower whole body lean mass (LM) compared to “Early” and “Active” NVC patterns, particularly at upper limbs. To note, in all body sites, BMD correlate with LM and BMC according to NVC pattern severity.

Conclusion: SSc patients with most severe microvascular damage show a significantly altered body composition and bone status suggesting a strong link between microvascular failure and associated muscle/bone failure.

References:

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CONTRAST-ENHANCED ULTRASOUND IN THE EVALUATION OF MYOSITIS

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Background: contrast-enhanced ultrasonography (CEUS) has been proposed as a tool to assess myositis patients and its accuracy in differentiating myositis from common mimickers.

Methods: 16 patients with myositis (4 polymyositis, 6 dermatomyositis, 2 immune-mediated necrotizing myopathy, 1 inclusion body myositis, 2 overlap Sjogren’s syndrome - myositis and 1 Enterovirus-reactive myositis) and...