on upper extremities. None of patients had allodynia, but impaired vibratory sensibility was found in all patients with NP. Clinical signs of radiculopathy were present in 6 patients. Hypoesthesia was registered in typical dermatomes in 6/11 (54.5%) of patients, with asymmetric hyporeflexia in 2/11 (18.1%) patients on lower, and none of patients on upper extremities. Despite the fact that almost every patient had symptoms and signs suggesting polyneuropathy, in only four of them demyelinating polyneuropathy was detected by ENG.
Conclusions: NP is common in patients with SSc. Presence of NP is associated with more severe SSc, symptoms of depression and worse quality of life. Almost all SSc patients (90.9%) with NP have typical symptoms and signs for polyneuropathy. However, in only few of them polyneuropathy could be detected by ENG. This finding suggests that pure small-fiber polyneuropathy, which is not detectable by ENG, may be the cause of NP in most of patients.
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FRI0406 MORTALITY PROFILE IN SYSTEMIC SCLEROSIS: A LARGE RETROSPECTIVE POPULATION-BASED STUDY FROM BRAZIL

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Background: Systemic sclerosis (SSc) is an uncommon autoimmune multisystem disease associated with reduced life expectancy compared with the general population.[1] In order to prolong survival of this patient population, clear information on the most important death-related conditions is undoubtedly necessary. No mortality data in SSc, however, are available from Latin America, as well as few large series studies have looked at the mortality profile in SSc.
Objectives: We aimed to describe the causes of death in SSc occurred in the state of Rio de Janeiro, Brazil, from 2006–2015, and also to compare the data gathered with the general population mortality.
Methods: All death certificates issued in the state of Rio de Janeiro, Brazil, from 2006–2015 were screened for the code attributed to SSc according to the tenth revision of the International Classification of Diseases (ICD-10), either as an underlying (UD) cause of death (also referred to as basic cause of death) or a non-underlying (non-UD) cause. In addition to compiling the causes of death in both settings, we calculated the non-adjusted as well as the age bracket-adjusted (age at death $<$50 years and $\geq$50 years) mortality ratio against the general population for each cause of death when SSc was listed as a non-UD cause.
Results: Of 1.294.491 fatalities recorded over the study period, ICD-10 code for SSc was listed on 374 (0.02%) death certificates, being a basic cause of death on 223 occasions and a non-UD cause on 151 occasions. The overall mean (SD) age at death in SSc was 58.7 (15.6) years, with men (n=56) having an earlier mean age at death than women (n=318) [53.5 vs 59.6 years, respectively; p=0.004]. For SSc as a basic cause of death, the main non-UD causes were respiratory system diseases (61.4%), in particular pneumonia, followed by septicemia (37.6%), diseases of the circulatory system (34.9%), and renal failure (9.4%). There were no significant differences between the genders for each cause of death. The mean age at death was significantly lower among men vs women for diseases of the respiratory system, respiratory failure, certain infectious and parasitic diseases, and septicemia. For SSc as a non-UD cause of death, the major conditions leading to death were circulatory system diseases (39%), in particular pulmonary arterial hypertension (PAH; 13.2%), followed by certain infections and parasitic diseases (11.9%), malignant neoplasms (10.5%) and diseases of the digestive system (9.9%). Compared with the overall population, patients with SSc had an excess of death (odds ratio [OR] $>$1) due to PAH (OR 138.94), septicemia (OR 1.92), gastrointestinal hemorrhage (OR 2.40), other systemic connective tissue diseases (OR 24.78) and pulmonary fibrosis (OR 11.05), as well as due to heart failure (OR 6.40) for deaths occurred before age 50.
Conclusions: We have shown large data on the mortality profile of patients with SSc, the first from Latin America. Of note, infections and cardiorespiratory diseases had a strong impact on mortality, as evidenced by previous publications.[1] Taking all into account, these data support an increased vigilance for infections, as well as the need to implement effective measures to control modifiable cardiovascular risk factors, including screening for PAH.
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FRI0407 MACROVASCULAR INVOLVEMENT IN SYSTEMIC SCLEROSIS PATIENTS: IS THERE A RELATIONSHIP WITH MICROVASCULAR PERIPHERAL BLOOD FLOW?

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Background: Raynaud's phenomenon and digital ulcers are common clinical skin manifestations of the microvascular dysfunction in systemic sclerosis (SSc). Although microvascular and macrovascular abnormalities frequently coexist in disease such as diabetes mellitus and other vascular diseases, the possible association between microvascular failure and macrovasculopathy in SSc patients has not been deeply investigated (1).
Objectives: The aim of the study was to estimate in SSc patients the macrovascular function by measuring of the Intima-Media Thickness (IMT) of high-caliber and periferic small-caliber arteries, and to evaluate the possible correlation with microvascular blood perfusion (BP) assessed by Laser Speckle Contrast Analysis (LASCA).
Methods: Twenty-nine female SSc patients (mean age 65.7±12.07 SD years, mean disease duration 97.3±68.01 SD months) were enrolled after informed consent. Carotid IMT was evaluated through B-mode US imaging (Esaote, Genoa) using transducer 18 MHz on both right and left common carotid arteries (CCA) as well as ulnar (UA) and radial arteries (RadA). Therefore, an average IMT score was obtained for any evaluated site. Peripheral BP was assessed by LASCA (Perimed-Sweden) at the level of fingertips, periungueal areas, dorsum and palm of both hands, and face. The BP values were reported as perfusion units (PU) (2). In addition nailfold videocapillaroscopy (NVC) in order to assess the microvascular morphological status was done by nailfold video capillaroscopy (NVC) considering the different progressive SSc patterns and the microangiopathy evolution score (MES) (3–4).
Results: A positive correlation was observed between CCA IMT and age (p=0.004) and disease duration (p=0.02). On the other hand, no significant correlations were observed between IMT of periferic small-caliber arteries (UA and RadA) and age or disease duration. Positive correlations were found between microvascular damage, as assessed by MES and IMT at the level of periferic small-caliber arteries.No significant correlation was observed between macrovascular involvement of high-caliber arteries and peripheral BP as assessed by LASCA. However, at the peripheral site a correlation was observed between RadA IMT and palm (p=0.05), periungueal (p=0.04) and dorsum of hand BP (p=0.0006), as assessed by LASCA. UA IMT showed a correlation limited to the dorsum of the hand (p=0.003).
Conclusions: Significant correlations seem to exist between increased IMT of periferic small-caliber arteries (macrocirculation) and altered peripheral BP (LASCA) at the level of hand microvessels (microcirculation) in SSc patients. These results were found strength by a further correlation with the microvascular damage (MES).
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FRI0408 NEOPTERIN AS A SEROLOGICAL MARKER OF DISEASE ACTIVITY IN PATIENTS WITH ANTI-MELANOMA DIFFERENTIATION-ASSOCIATED GENE 5 ANTIBODY POSITIVE CLINICALLY AMYOPATHIC DERMATOMYOSITIS

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Background: Anti-melanoma differentiation-associated gene 5 (MDA5) antibody in clinically amyopathic dermatomyositis (CADM) is associated with rapidly progressive interstitial lung disease (RPILD). RPILD is a rare disorder with a bad outcome, and therefore the intensive treatment with combinational immunosuppressive drugs in addition to steroid need to be initiated. However until now, good serological markers to evaluate disease activity in CADM have not been established. Activated alveolar macrophage is one of the possible candidates worsening the condition of RPILD.
Objectives: To investigate new serological markers of disease activity in anti-MDA5 antibody positive CADM for therapeutic indication.
Methods: Thirteen anti-MDA5 antibody positive CADM patients were enrolled. We serially measured serum anti-MDA5 antibody, and neopterin and IL-18 as markers of activated macrophage by serum enzyme-linked immunosorbent assay. We tracked them at three points in each patients: before treatment, soon after a series of intravenous cyclophosphamide pulse therapy (about 3months later after onset) and the remission status (about one year later after onset).
Results: Four patients died soon after the initial treatment because of the deterioration of RPILD. At onset of the disease, the levels of serum anti-MDA5 antibody and neopterin were extremely high (169.75±24.3 index and 27.6±24.1 nmol/l) in all patients. However serum IL-18 level was almost normal (479.3±301.4 pg/ml). Among the 9 surviving patients, it took about one year for anti-MDA5 antibody level to decrease to the normal range. On the other hand, neopterin level decreased quickly after the initial treatment. The level of anti-MDA5 antibody transitioned from 169.75±24.3 index to 93.1±50.1 index, and then to 44.8±45.4 index (P=0.003). Neopterin level transitioned from 27.6±24.1 nmol/l to 9.1±6.5 nmol/l, and then to 6.4±5.0 nmol/l (P=0.006). IL-18 level transitioned from 479.3±301.4 pg/ml to 246.0±175.8 pg/ml, and then to 233.3±180.1 pg/ml (P=0.02). The level of anti-MDA5 didn't correlate with the level of ferritin (r=0.28), neopterin (r=0.16), IL-18 (r=0.06) and soluble IL-2 receptor (r=0.27), but the level