Objectives: This study aimed to analyze the serum levels of neopterin in patients with dermatomyositis (DM) in association with clinical manifestations, laboratory data and patient prognosis.
Methods: One hundred and eighty-two consecutive DM patients and 30 healthy controls were retrospectively enrolled in the study. Serum levels of neopterin were detected by using the ELISA method. Clinical and laboratory data and patient prognosis were obtained and analyzed in association with serum neopterin.
Results: Serum levels of neopterin were significantly increased in DM patients (median 21.2 nmol/L, IQR 13.9-35.2 nmol/L) compared to healthy controls (median 4.3 nmol/L, IQR 2.9-5.6 nmol/L, P<0.01). High serum neopterin levels were associated with anti-MDA5 antibody, rapidly progressive interstitial lung disease (RP-ILD), and cutaneous involvement including skin ulcer and heliotrope rash. Longitudinal assessment of serum samples revealed that the serum neopterin levels were closely correlated with disease severity. In addition, a significant increase in serum neopterin concentration of non-survivors (median 38.7 nmol/L, IQR 23.5-65.3 nmol/L) was observed when compared to that of survivors (median 19.0 nmol/L, IQR 12.5-29.0 nmol/L) (P<0.01). ROC curves showed that serum neopterin could distinguish non-survivors and survivors at an optimal cut-off level of 22.1 nmol/L with a sensitivity and specificity of 0.804 and 0.625 respectively (P<0.01). Kaplan-Meier survival curves revealed that DM patients with serum neopterin > 22.1 nmol/L had a significantly higher mortality compared to patient group with serum neopterin <22.1 nmol/L (logrank P<0.01). Multivariate Cox regression analysis identified high serum neopterin concentration to be an independent risk factor for poor prognosis in DM (adjusted HR=4.619, 95%CI: 2.092-10.195, P<0.01).
Conclusion: Increased serum levels of neopterin were significantly associated with RP-ILD and reduced survival in DM patients, suggesting it as a promising biomarker in the disease evaluation of DM. These findings highlight the role of cellular immune activation and macrophage activity in the pathogenesis of DM.

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FRI0309 SEVERE ABDOMINAL MANIFESTATIONS IN JUVENILE DERMATOMYOSITIS

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Background: Juvenile dermatomyositis (JDM) is a rare and heterogeneous children-onset idiopathic inflammatory myopathy. It primarily affects skeletal muscle and skin, but also pulmonary, cardiac and gastrointestinal systems in a subset of patients. Gastrointestinal (GI) involvement occurs in 22 to 37% of JDM patients and have been described only in a few case reports.
Objectives: To describe causes of severe abdominal manifestations in JDM patients.
Methods: Retrospective monocentric study. Inclusion criteria were (i) diagnosis of JDM according to the modified Bohan and Peter criteria before 16 years of age, (ii) occurrence of severe involvement of any intra-abdominal organ, (iii) follow-up at Necker Hospital from 2005 to 2018. Severe abdominal manifestation was defined as a potential life-threatening abdominal organ event, need for abdominal surgery and/or parenteral nutrition. Gastrointestinal (GI) involvement was defined by severe abdominal pain, dysphagia, digestive hemorrhage, obstruction, ulceration and/or perforation. Severe hepatitis was defined by an elevation of liver enzymes exceeding five times the upper limit associated with an elevation of γ-glutamyl transferase (γGT) activity and/or liver failure. Abdominal manifestations were classified as certain or probable primary JDM involvement, therapy-related event or infection-related event.
Results: Nine patients with 19 abdominal complications were identified among 110 JDM patients followed during the study period (8,3%). Diagnosis of JDM was made at a median age of 10.7 years-old [range, 3.7 – 14.4], with a median CMAS of 7 [range, 2 – 48], median MMT of 47 [range, 38 – 76], a median creatine kinase level of 3 812 UI/L [range, 170 – 14 054], and a median follow-up of 3 years [range, 0.4 – 6.3]. Seven patients were positive for either anti-NXP2 (n = 4), anti-TIF1-γ (n = 2) or anti-MDA5 (n = 1) antibodies. JDM-related abdominal disease comprised GI involvement (n = 10), acute pancreatitis (n = 4) and hepatitis (n = 3). The revealing symptom for GI involvement and acute pancreatitis was abdominal pain. Five patients with severe abdominal pain, vomiting and/or obstruction, underwent abdominal imaging revealing thickening of colon (n = 2) and/or ileum mucosa (n = 2). Hepatitis was found in 3 patients with an uncommon extensive steatosis with moderate inflammation and perisinusoidal fibrosis (n = 2). Treatment-related complications were considered certain for methotrexate-induced liver toxicity (1 patient) and possible for duodenal perforations (1 patient) 3 days after a methylprednisolone pulse. At last visit, JDM was clinically inactive in four patients. Three patients died from refractory JDM, 2.9 years [range, 2 – 3.6] after the JDM diagnosis, among the 9 patients identified in this study *versus* only 1/92 of the other JDM patients from our cohort.
Conclusion: Primary JDM involvement was the main cause of severe abdominal manifestations and associated to a high mortality rate. We highlight for the first time that pancreatitis should be considered as a main diagnosis in JDM patients who develop severe and/or sustained abdominal pain.
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FRI0310 UNIVERSITY OF CALIFORNIA LOS ANGELES SCLERODERMA CLINICAL TRIALS CONSORTIUM GASTROINTESTINAL TRACT 2.0 REFLUX-SCALE ASSOCIATES WITH IMPAIRED ESOPHAGEAL SCINTIGRAPHY FINDINGS IN SYSTEMIC SCLEROSIS

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Background: The University of California Los Angeles Scleroderma Clinical Trials Consortium gastrointestinal tract 2.0 (UCLA SCTC_GIT 2.0) questionnaire is a self-reported tool including 7-multi-item scales and measuring GI quality of life both in clinical trials and day-to-day clinic care of systemic sclerosis (SSc) patients (1). Scarce data are available on the correlation between patient reported GI symptoms and motility dysfunction as assessed by esophageal scintigraphy.
Objectives: To evaluate the UCLA SCTC_GIT 2.0 score in SSc patients undergoing esophageal scintigraphy and correlate their findings.
Methods: Data of SSc patients admitted to our clinic, undergoing esophageal scintigraphy, were reviewed. The score of UCLA SCTC_GIT 2.0, usually administered to all attending SSc patients, was calculated. Data were expressed as percentage (%) and mean ± standard error. Pearson's test was used for Prism 7 correlation analysis. P < 0.05 was considered significant.
Results: Of all the SSc patients admitted to our clinic from 1stSept 2017 to 31st December 2018, twenty underwent esophageal scintigraphy. Seventeen were female, 4 with diffuse subset, mean age was 51 (± 3.4) years, disease duration of 6.8 (± 1.2 years) years. Twelve (60%) patients reported esophageal (reflux, dysphagia) symptoms, 5 (25%) stomach (early satiety, vomiting) and 9 (45%) intestinal (bloating, diarrhea, constipation) symptoms while 3 (15%) reported no symptoms. Mean ± SE scores of UCLA SCTC_GIT 2.0 items were: reflux 0.8 ± 0.2, distention 0.9 ± 0.2, fecal soiling 0.5 ± 0.2, diarrhea 0.5 ± 1, social 0.5 ± 0.1, emotional 0.5 ± 0.2, constipation 0.5 ± 0.1, and total UCLA SCTC_GIT 2.0 0.6 ± 0.1. Nine (45%) patients had no-mild, 4 (20%) moderate and 7 (35%) severe to very-severe reflux score. Esophageal scintigraphy showed hypomotility in 16 (80%) patients who had a higher UCLA SCTC_GIT 2.0 reflux score compared with the remaining 20% patients (1

± 0.2 vs 0.3 ± 0.2). Fifteen/20 (75%) had an abnormal (<90% within 10 sec) esophageal emptying activity. Percentage of esophageal emptying activity negatively correlated with reflux score (r = - 0.52, p = 0.02) while it did not correlate with the other items scales and the total UCLA SCTC_GIT 2.0 score (p > 0.05).

Conclusion: SSc patients with impaired esophageal scintigraphy findings have a higher GIT2 reflux score. The lack of correlation with UCLA SCTC_GIT 2.0 total score may be related to the composite nature of the score capturing overall GI disease aspects. The UCLA SCTC_GIT 2.0 is a complementary tool for objective measurement of esophageal involvement which can be easily administered in day-to-day clinical assessment. A larger number of SSc patients is needed to confirm these preliminary findings.

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FRI0311 PREVALENCE AND CLINICAL MANIFESTATIONS OF ERASMUS SYNDROME IN SYSTEMIC SCLEROSIS: A CROSS-SECTIONAL STUDY

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Background: Erasmus syndrome (ErS) is defined by the association of exposure to silica with the subsequent development of systemic sclerosis (SSc), with or without associated silicosis.

Objectives: The objectives of this study were, on one hand, to evaluate the prevalence of ErS in a population of SSc patients and to characterize the cases and, on the other hand, to evaluate the clinical and laboratory characteristics of SSc patients with or without exposure to silica.

Methods: We performed a cross-sectional study of the patients with SSc in our department. Demographics, clinical and laboratory data were collected from all patients with SSc diagnosed according to ACR/EULAR criteria. Moreover, a telephone call was made in order to detail the professional activity and possible exposure to silica.

Results: The prevalence of ErS in this population was 15.3% (9/59). All cases identified were male, corresponding to 75% of men with SSc followed at our department. There was a statistically significant association between ErS and male gender (p<0.001), initial pulmonary manifestation (p=0.025), history of digital ulcers (p=0.014) and smoking (p=0.047). On the other hand, a lower risk of gastrointestinal involvement was found in ErS cases (p=0.008). All patients with ErS had positive autoantibodies (mainly anti-Scl70 and anti-centromere) with titters tending to be higher than SSc without ErS, although without statistically significant differences. In addition, although with no statistical significance, we found that pulmonary artery systolic pressure (PASP) estimated by echocardiogram was higher in patients with ErS.

Conclusion: In our study, prevalence of ErS was higher than data from previously published literature. For a more accurate ErS diagnosis it is necessary to be aware of and investigate less intense silica exposures, which may have occurred many years before diagnosis.

Statistically significant differences were found between ErS and SSc without exposure to silica; this fact may have impact in diagnosis, treatment and prognosis.

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FRI0312 HYDROXYCHLOROQUINE SIGNIFICANTLY REDUCES SERUM MARKERS OF ENDOTHELIAL INJURY AND NEMO VIDEOCAPILLAROSCOPY SCORE IN SYSTEMIC SCLEROSIS: A 3-MONTHS PROSPECTIVE OBSERVATIONAL STUDY

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Background: The most-commonly postulated model of disease progression in Systemic sclerosis (SSc) is based on the interplay between endothelial damage and tissue repair. Hydroxychloroquine (HCQ) is commonly used in systemic sclerosis (SSc) for the management of joint involvement [1]. In addition, HCQ showed to exert a protective role on endothelial dysfunction in several pathologic conditions [2] but no data exist in SSc.

Objectives: aim of our study was to assess the effect of 3-months HCQ administration on serum levels of markers of endothelial injury and microvascular changes, assessed with NEMO score.

Methods: 20 SSc patients considered for treatment with HCQ 400 mg/day and 16 SSc patients not treated with HCQ were consecutively recruited, having clinical assessment, blood samples and nailfold capillaroscopy (NVC) at recruitment and after 3 months. Median age was 51.5 (19.5). Median onset of Raynaud's phenomenon was 83 (66) months. 29 patients had lSSc and 7 patients dSSc. Both groups had comparable vasoactive drugs, disease activity, duration and subtype. Any kind of overt vasculopathy was an exclusion criteria. The NEMO score was calculated as the total number of micro-haemorrhages (MHE) and microthrombosis (MT) and the giant capillaries (GC) score was calculated as the total number of GC [3]. Serum E-selectin, Endothelin-1 (ET-1), Vascular cell adhesion molecule-1 (VCAM-1), vascular endothelial growth factor (VEGF-A) levels were assayed by Magnetic Luminex Assay, Human Premixed Multi-Analyte Assay (R&D Systems). Plasma dimethyl arginine (ADMA) levels were determined by competitive ELISA (DLD Diagnosticka GMBH). Data were assessed with SPSS software version 25.0 and expressed as median and interquartile range. Wilcoxon rank-sum test, Mann-Whitney U test and Fisher's exact test were used for statistical evaluation. P values < 0.05 were considered significant.

Results: in the group treated with HCQ serum levels of E-Selectin (35.28 (16.88) ng/ml vs 30.84 (12.60) p = 0.004), VCAM-1 (847.30 (344.85) ng/ml vs 761.55 (396.87) p = 0.014) and ET-1 (12.25 (5.53) pg/ml vs 10.94 (4.35) pg/ml (p = 0.022) decreased significantly, as well as NEMO Score (7.5 (11.25) vs 6.5 (8) p = 0.001), MHEs (6 (7) to 5 (5.75) p = 0.003) and MTs (1 (4.5) to 0.5 (2) (p = 0.003). VEGF-A (p = 0.68), ADMA (p = 0.91) and GCs (p = 0.30) were unaffected. No significant improvements were observed among patients not treated with HCQ.