Conclusions: QMUS with NMD algorithm provides a fair diagnostic value for patients suspected for an IIM and is similar to EMG results. A sizeable NPV indicates a low rate of false negative QMUS results. In addition to the relevant PPV for the presence for NMD, QMUS could serve as a potential screening tool for clinicians to detect possible myopathies and to rule out the presence of IIM.

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AB0777 INFLAMMATORY MYOSITIS ASSOCIATED WITH MYASTHENIA GRAVIS WITH AND WITHOUT THYMIC PATHOLOGY: CASE SERIES AND LITERATURE REVIEW

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Background: Inflammatory myopathies (IM) and Myasthenia gravis (MG) are two well-recognised and distinctive neuromuscular diseases. The association of myasthenia gravis (MG) and inflammatory myositis (IM) is rare and often only one of the diseases is diagnosed. The coexistence of MG and IM might be associated with thymoma. Even less common is the association of IM (polymyositis or dermatomyositis) and myasthenia gravis in the absence of thymoma.
Objectives: Here, we report a case series of 6 patients with concurrent MG and IM who were followed at the Neuromuscular Disease Unit (NMDU) at a tertiary referral centre in Vancouver, British Columbia. We also conducted literature review on clinical characteristics, diagnostic challenge and management of this condition.
Methods: In this study, we retrospectively examined patients seen at NMDU from 2004 to 2017 who had diagnosis of concurrent MG and IM. We reviewed medical records to assess their clinical presentations, laboratory findings, imaging studies and electrophysiological features. The data is presented descriptively.
Results: We identified 6 patients with MG-IM overlap. Three patients had simultaneous onset of MG and IM, 2 of whom presented with myasthenia crisis and fulminant myositis. In the other 3 patients, MG was the initial presentation and IM occurred 3–11 years after MG.
Diagnosis of MG was confirmed with clinical features, electromyography and/or serology. All had symptoms of MG with predominant ocular or bulbar weakness. Among these 6 patients, 3 had underlying thymic pathology including two benign thymoma and one stage IV thymoma; all 3 patients had Acetylcholine Receptor (AChR) Antibody. Of the 3 patients with no thymic pathology by computed tomography (CT) or thymectomy, 1 had high positive AChR antibody and 2 were negative.
Four patients had biopsies confirming the diagnosis of dermatomyositis or polymyositis. The other 2 patients declined biopsy; however, their MRI and EMG findings were consistent with IM. Only one patient had typical dermatomyositis rash.
Among the 3 patients with underlying thymic pathology, thymoma were resected; all 3 were treated with high dose glucocorticoid, IVIG, and methotrexate with complete remission after 2 years. Of the 3 patients with no thymic pathology identified, one patient (AChR+) was in remission on mycophenolate and passed away from pancreatic cancer; two patients (AChR-) had refractory MG and IM, and both responded to rituximab.
Conclusions: In summary, this is one of the largest case series with MG-IM overlap with or without thymic pathology. It is very important to recognise such association and the different pattern of muscle involvement because therapies may be adjusted to treat both conditions. In patients with thymic pathology, conventional disease modifying agents, IVIG and glucocorticoid in addition to thymoma resection appear to be effective. In patients with refractory MG and myositis who were AChR negative, rituximab may be effective.
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AB0778 CLINICAL CHARACTERISTICS AND PROGNOSIS OF POLYMYOSITIS AND DERMATOMYOSITIS ASSOCIATED WITH MALIGNANCY: A 25-YEAR RETROSPECTIVE STUDY

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Background: Previous studies indicate that malignancies in polymyositis (PM) or dermatomyositis (DM) patients are associated with high mortality. Hence, it becomes important clinically to identify patients who are at high risk of developing malignancy. However, previous studies to characterise such risk factors for malignancy in patients with PM and DM were small size or the findings ware not conclusive.
Objectives: This study investigated factors predictive of malignancy and prognosis in patients with PM and DM.
Methods: We conducted a retrospective study of PM and DM patients who were inpatients at our hospital between January 1992 and September 2016. The diagnosis of PM or DM was made according to Bohan and Peter criteria. The collected data included gender, age at onset, laboratory test results at presentation, clinical features and complications in all progress. Past history of hypertension or diabetes mellitus was determined upon diagnosis of myositis. All patients in this study were followed from the baseline visit until loss of follow-up or death or otherwise until censor date 1 September 2017. Fisher exact test and Mann-Whitney U test were used for group comparisons. Univariate and multivariate analysis of the predictors of malignancy associated with PM or DM were performed by logistic regression to identify independent risk factors. Patient survival was analysed by using Kaplan-Meier curve and log-rank test.
Results: Among 134 patients, 29 had cancer diagnosed between 2 years prior and 3 years following identification of PM or DM. Esophageal cancer (n=5, 14.7%) and lung cancer (n=5, 14.7%) were the most frequent. Univariate analysis showed that male sex (55.2% vs 23.8%, p=0.003), older age (64.6±11.3 vs 53.4±16.5, p<0.001), past history of diabetes mellitus (28.6% vs 2.9%, p<0.001), dysphagia (41.4% vs 17.3%, p=0.01) and absence of interstitial lung disease (37.9% vs 64.8%, p=0.01), arthralgia (17.2% vs 47.1%, p=0.005) and the Raynaud phenomenon (3.4% vs 23.1%, p=0.02) were associated with increased malignancy. Multivariate analysis showed that independent factors included male sex (OR=3.65, p=0.03), older age (OR=1.05, p=0.02), past history of diabetes mellitus (OR=10.4, p=0.005) and absence of interstitial lung disease (OR=0.25, p=0.03) (table 1). Survival was significantly lower in patients with malignancy than in patients without malignancy (p<0.001).

Abstract AB0778 – Table 1. Multivariate analysis of PM/DM with and without cancer

Variables	Adjusted OR	95% CI	*P* value
Male, n(%)	3.65	1.17–11.30	0.03
Onset age, years, mean±SD	1.05	1.01–1.10	0.02
Interstitial lung disease, n(%)	0.25	0.08–0.84	0.03
Diabetes mellitus	10.4	2.00–54.3	0.005
Arthralgia, n(%)	0.55	0.15–2.04	0.37
Dysphagia, n(%)	2.82	0.82–9.71	0.10
Raynaud phenomenon, n(%)	0.25	0.03–2.29	0.22

Abbreviation: OR, odds ratio; CI, confidence intervals.
Conclusions: This study is the first to associate a past history of diabetes mellitus with malignancy; 28.6% of PM or DM patients with malignancy had diabetes mellitus whereas 7.3 percent of cancer patients had diabetes mellitus.

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AB0779 LONG TERM FOLLOW-UP OF A SYSTEMIC SCLEROSIS GROUP TREATED WITH BOSENTAN

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Background: Prospective studies with Bosentan have shown short term efficacy, while it is not clear whether long-term treatment may be effective or whether ulcers may recur once treatment is discontinued.
Objectives: Our objective was to evaluate the long term efficacy and tolerability of bosentan in patients with systemic sclerosis (SSc) who develop digital ulcers (DU).
Methods: In the present prospective, observational, non-controlled study, we followed 26 SSc patients treated with Bosentan from Sept 2014 to Dec 2017 Number of DU, semiquantitative capillaroscopic scoring, VAS (visual analoque scale) for Raynaud, VAS for DU and HAQ were evaluated every 6 month. Results are presented as mean(sd). The difference between efficacy measures at follow-up visits was tested with the Wilcoxon's signed-rank test.
Results: The group included 26 patients, 16 females, 11 diffuse subsets, age was 48.08 (9.8) years, disease duration was 84.35 (76.04) months, number of DU was 4.27 (3.71), most of them had a late scleroderma pattern pattern (16/26).

Microangiopathy evolution score was 5.19 (2.04), VAS for DU was 75.52 (16.17), VAS for Raynaud was 67,43 (14.16), HAQ was 1.62 (0.55). 5 patients received Bosentan less then 6 month, so they were excluded from the statistical analysis.
6 month evaluation revelead significant decrease in the number of DU (p<0.01), the VAS for DU (p<0.01), the VAS for Raynaud (p=0.03) and the HAQ (p=0.04), but not of the microangiopathy evolution score. No significant difference was noticed of the above mentioned parameters at the next follow-up evaluations.
Regarding Bosentan safety: 6 patients died during the follow up (3 cases of severe pulmonary arterial hypertension, 1 scleroderma renal crisis, 1 heart failure, 1 post vascular surgery). Bosentan was stopped due to lack of efficacy in 2 case and due to side effects in 3 cases: 2 elevated liver enzymes, 1 severe trombocytopenia and 1 dyspneea agravation.
12 patients had a follow up after a 6 months Bosentan stop. We did not notice any significant increase in the number of DU, the VAS for DU or Raynaud, the capillaroscopic semiquantitative scoring or the HAQ.
Conclusions: We noted a significant decrease in the number of DU, patients perception of Raynaud and of DU after 6 months of treatment and the effect was maintained in the 3 years follow-up, even 6 months after Bosentan was stopped. In this long-term follow-up no new unidentified adverse reactions were found, except for the unexpected severe thrombocytopenia. The present study is limited due to the small sample size, to the observational nature and should be viewed as descriptive. Questions rise about drug costs (6 months or long term treatment), but it also has to be emphasised that most of these lesions were chronic and non-responsive to previous treatments.

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AB0780 PROLONGED PROTON PUMP INHIBITOR EXPOSURE IS ASSOCIATED WITH DEVELOPMENT OF CALCINOSIS IN SYSTEMIC SCLEROSIS

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Background: Long-term use of proton pump inhibitors (PPI) has been associated with some safety concerns, including potential vascular calcification. In a previous retrospective analysis, we noted a possible association between PPI use and calcinosis in scleroderma (SSc).
Objectives: To investigate the association between PPI use and the presence and extent of calcinosis in SSc patients.
Methods: Data from prospectively recruited patients were collected by patient survey, physician assessment and medical records. Calcinosis was graded; size (+=<1 cm, ++=≥1≤3 cm, +++=>3 cm) and number of sites involved (NSI) (I=1 body site, II=2–3, III≥3). A total daily PPI equivalent dose (TDED) was calculated for each patient. We calculated PPI exposure score (PPE) by multiplying the total duration of use by TDED. For analysis, PPE was categorised into four groups; 0=no exposure, 1=up to 5 years, 2=6–10 years, 3≥10 years. Fisher's exact test was used to assess categorical variables. Logistic regression assessed association between calcinosis and independent variables.
Results: 216 patients were recruited, 81.5% females, mean age 57.46 (SD 13.5) years. 56.5% had limited, 31.5% diffuse SSc, 9.7% had overlap features and 2.3% other CTD. Mean disease duration was 10 years (SD 9). ANA subtypes were defined: ACA positive (31.5%), ATA (25.5%), ARA (12.0%), ANA +ENA- (11.6%), U3RNP (5.1%), ANA- (4.2%), PmSCL (3.7%) and 6.5% other antibodies. Gastroesophageal reflux symptoms occurred in 83.3% of patients, most were on PPI (81%) and 14.8% had previously been on PPI. Current calcinosis (CC) was present in 30.1% patients, 9.7% reported past calcinosis. 39.8% had calcinosis at any time (CAT). 60.2% of patients never had calcinosis. Of those with CC, 47.7% had >1 site involved. The most frequent sites affected were; finger (70.8%), elbow (35.4%) and knee (18.5%).
Univariable analysis found an association between disease duration and calcinosis, with odds of CAT increased by 7% per year (OR 1.07, CI 1.04–1.11, p<0.001). Similarly, every year of PPE increased odds of CAT by 3% (OR 1.03, CI 1.01–1.05, p=0.003). Increasing age associated with CAT (odds increasing by 2% per annum, p=0.043). Exposure to a standard dose of PPI for over 10 years increased the odds of calcinosis by 4 times (OR 4.07, CI 1.68–9.85, p=0.002) compared to no exposure. PPE category associated with NSI (p=0.04). 73.3% of patients with large volume calcinosis (>3 cm) had a PPE for >10 years and all with calcinosis >3 cm had exposure to PPI.
Multivariable logistic regression found that disease duration (OR 1.07, CI 1.03–1.11, p=0.001) and antibody specificity strongly associated with calcinosis. Presence of ATA (OR 0.32, CI 0.14–0.75 p=0.008), ANA- (OR 0.13, CI 0.02–0.79, p=0.026), and ANA +ENA- (OR 0.17, CI 0.05–0.52, p=0.002) reduced odds of calcinosis. Although the effect of PPIs on calcinosis was attenuated after adjusting for disease duration and antibodies, higher exposure to PPIs remained a significant predictor of calcinosis, with PPE category (>10) increasing risk of CAT (OR 3.34, CI 1.16–9.17, p=0.025).
Conclusions: Our data support a novel association of PPI exposure with calcinosis and confirm association of disease duration and antibody profile. Given the clinical impact of calcinosis, a potentially modifiable risk factor of PPI exposure warrants further study.
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AB0781 GLUCOCORTICOID DOSE AND CARDIAC INVOLVEMENT MIGHT BE POTENTIAL RISK FACTORS FOR SCLERODERMA RENAL CRISIS

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Background: Scleroderma renal crisis (SRC) is a rare but life-threatening complication of systemic sclerosis (SSc). SRC remains a major risk factor for mortality in SSc. It is important to identify potential risk factors for SRC, and avoid developing overt SRC.
Objectives: To perform a retrospective case series analysis of the characteristics, management and outcomes of SRC in Chinese SSc patients.
Methods: SSc patients hospitalised at Sun Yat-Sen Memorial Hospital from January 1992 to December 2017 were recruited. Clinical data were collected. SRC was defined as new onset, with blood pressure (BP) >140/90 mmHg or a>30 mmHg rise in BP from baseline, rising serum creatinine (Scr) levels and/or oligoanuria. Data were showed as mean ±standard deviation.
Results: ([1] There were 749 SSc patients recruited and 16 patients (2.1%) of them were hospitalised for SRC. Among these 16 patients, 56% were females, age was 54.6±13.6 years, mean duration from SSc onset to SRC occurred was 4 years.
([2] SRC developed in 14 patients (87.5%) with diffuse cutaneous SSc (dcSSc), and in 2 patients (12.5%) with limited cutaneous SSc (lcSSc). Eleven patients (68.8%) were under glucocorticoid treatment before SRC onset: 4 patients received ≥30 mg/d of prednisone, 6 patients received ≥7.5 mg/d prednisone and 1 patient received <7.5 mg/d prednisone. No patient was treated with angiotensin-converting enzyme (ACE) inhibitors before SRC.
([3] All 16 patients manifested progressive renal failure, with Scr levels increase to 420±256 μmol/L. Ten patients manifested new onset hypertension, with systolic BP 175±21 mmHg and diastolic BP108 ±13 mmHg. Five patients who had a history of well-controlled hypertension manifested accelerated increase in BP 178 ±17/108±7 mmHg. One patient was normotensive, but manifested rapidly progressive oliguric renal failure with Scr increase to 969 μmol/L, massive proteinuria and hemolytic anaemia.
([4] Twelve patients (75%) had pulmonary fibrosis, 11 patients (68.8%) had cardiac involvement, 6 patients had pulmonary arterial hypertension (PAH) and 6 patients had gastrointestinal dysfunction. Cardiac involvement was common, manifested pericarditis, myocardial damage and heart failure (n=7, 43.8%, respectively). All 5 dead patients were accompanied by cardiac involvement.
([5] Eleven patients had Raynaud's phenomenon, 8 patients had digital ulcers, 5 patients had arthritis and 2 patients had oliguria. Thirteen patients (81%) manifested anaemia, 8 patients (50%) manifested thrombocytopenia, and 8 patients (50%) manifested microangiopathic haemolytic anaemia (MAHA).
Eleven patients (68.8%) received ACE inhibitor treatment. Fifteen patients were treated with glucocorticoid and 12 patients with immunosuppressant (Cyclophosphamide n=10, Azathioprine n=2). After treatment, renal recovered in 4 patients (25%), kidney function improved and developed to chronic kidney disease (CKD) without dialysis in 5 patients (31%), 2 patients required permanent dialysis (13%). Five patients (31%) died.