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).

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