INFLAMMATORY MYOSITIS ASSOCIATED WITH MYASTHENIA GRAVIS: CASE SERIES AND LITERATURE REVIEW

K. Huang1, M. Mezès2, K. Shoqania1, K. Chapman1, N. Amin1, N. Dehghan1

1Rheumatology; 2Neurology, University of British Columbia, Vancouver, Canada

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 (polyomyositis 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 fulfilling criteria for myasthenia gravis. 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.

Disclosure of Interest: None declared


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

L. Groseclose1,2, F. Berghesi1,2, V. Bojinca1,2, A. Balanescu1,2, A. Borangiu1,2, D. Mazuli1,2, S. Dais-Ilievscu1,2, D. Opris-Belinlik1,2, I. Saulescu1,2, C. Constantinescu3, M. Abbobul3, I. Ionescu1,2, I. Internal Medicine and Rheumatology, Sf Maria Clinical Hospital; 2Carol Davila University of Medicine and Pharmacy, Bucharest, Romania

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 patients...