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SINGLE CENTER EXPERIENCE OF CLINICAL PROFILE OF INFLAMMATORY MYOSITIS FROM INDIA

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Background: Clinical profile of myositis differs in respect of the setting. We present a single center experience from a community based referral center

Objectives: STUDY CLINICAL PROFILE OF INFLAMMATORY MYOSITIS (IM)

Methods: We present data from 114 patients of connective tissue disorders (CTD) with dominant Inflammatory Myopathy(IM) evaluated in CRD where we have patient database since 1996. Standard investigations & ELISA, immunoblot and nephelometry to assay autoantibodies (AAb) were done. Data extraction done from 2005-2017

Results: 36 and 28 patients respectively diagnosed as dominant idiopathic dermatomyositis (DM) and polymyositis (PM); remaining 41 patients showed overlap (OCTD). Mean onset age range 33-40 years in each subset with women dominance. Exclusive proximal muscle involvement seen 64% DM, 67% PM and 43% OCTD. 12 of OCTD showed classical DM rash. Raynaud's phenomenon was seen in 38% (25% DM, 10% PM, 65% OCTD). 83% OCTD showed inflammatory polyarthritis; DM 29% and PM 42%. Two patients DM also diagnosed malignancy (ovarian CA). 25% DM, nil PM and 31.7% OCTD showed CT based lung findings. Mean creatinine phosphokinase at diagnosis were DM 1580, PM 2239 & OCTD 830. EMG required in 48 patients confirmed diagnosis (DM 17, PM 16 and OCTD 15). Seven patients with diagnostic dilemma/ poor therapy response required muscle histopathology confirmation. 59% DM, 69% PM and 84% OCTD were seropositive ANA positive in 71% (ENA profile available for most). All Patients received steroids. Methotrexate prescribed in (92%), Azathioprine in (28%), hydroxychloroquine in (88%) with majority showing good response. Mycophenolate prescribed in (6%) for aggressive disease, IVIG in 2 patients for acute IIM with interstitial pneumonitis. Rituximab was prescribed in 4 resistant cases; all responding favourably. Mortality data of 4 patients (severe myositis(1), interstitial pneumonitis(1), ovarian cancer(1), septic shock(1)) was available. Antesyntathase syndrome noted in 9 patients.

Conclusion: Overlap CTD with myositis seems more common profile than DM or PM. Response to therapy was satisfactory with steroids and methotrexate being the mainstay. Rituximab is a promising biological agent in chronic resistant cases.

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AB0587

IMMUNOMODULATORY COMBINATION THERAPIES IN PATIENTS WITH DERMATOMYOSITIS / POLYMYOSITIS: A RETROSPECTIVE CROSS-SECTIONAL STUDY

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Background: Dermatomyositis (DM)/Polymyositis(PM) is an autoimmune disease that typically involve the striated muscle with a variable involvement of the skin and other organs ¹. Lymphocyte subsets disorders may contribute to the pathogenesis of DM/PM. It has been discovered that immunomodulatory drugs such as low-dose interleukin (IL)-2, rapamycin can help to regulate the lymphocyte subsets and control the disease and improve the prognosis ²⁻⁴.

Objectives: To investigate the levels of peripheral lymphocyte and CD4+T subsets of DM/PM patients and further to observe the regulatory effect of modulatory therapy on these cells in PM/DM at a relative large-sample size.

Methods: Total 450 patients with PM/DM and 206 healthy controls (HCs) were enrolled in this study. Among these participations, 320 patients received immunomodulatory combination therapies (immunomodulatory drugs include low-dose interleukin-2, rapamycin, metformin, retinoic acid etc). The absolute numbers of T, B, NK, CD4+T, CD8+T, Th1, Th2, Th17 and Tregs in peripheral blood of these individuals were detected by flow cytometry combined with standard absolute counting beads before and after the treatment.

Results: Patients with DM/PM had lower levels of total T, CD4+T, CD8+T, Th2, Th17, NK, Th1 and Tregs compared with those of HCs (P < 0.05). After immunomodulatory combination treatments, there was a dramatically increases various peripheral lymphocyte subsets such as T, B, CD4+T, CD8+T, NK, Th1, Th17 and Tregs (P < 0.05). Moreover, the increase in Tregs was much more than that in effector T cells (Teffs), resulting a rebalance of immune systems.

Conclusion: The unbalance of lymphocyte cells should contribute to the pathogenesis of DM/PM patients. Immunomodulatory combination therapies could promote the proliferation and functional recovery of Tregs in patients and might help to alleviate disease activity.

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Figure 1: Comparison of lymphocyte subsets in peripheral blood between healthy controls(HC) (n=206) and untreated DM/PM patients (n=450). All the lymphocytes subsets were analyzed by detected by flow cytometry combined with standard absolute counting beads. Data were presented as mean±SD and statistical analysis was determined by two-tailed unpaired t-test. Patients had lower levels of T, NK, CD4+T, CD8+T, Th1 and Tregs, decreased percentages of NK, Th1 and Tregs, and higher ratios of Th1/Th2/Tregs and lower ratios of CD4+T / CD8+T, Th1/Th2 in DM/PM compared with those of healthy controls. *P < 0.05, **P < 0.01, ***P < 0.001.

Figure 2: Changes of peripheral lymphocyte subsets after new immune regulatory combination therapy (n=320). Data were presented as mean±SD and statistical analysis was determined by two-tailed paired t-test. Absolute number of T, B, NK, CD4+T, CD8+T, Th1, Th17 and Tregs were significantly increased. The proportion of B, CD4+T, CD8+T and Th1 were also increased, the ratio of Th1/Th2 and Th1/Tregs were increased while the ratio of Th2/Tregs had a tendency to increase after new immunoregulatory combination therapies. *P < 0.05, **P < 0.01, ***P < 0.001.

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