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IDIOPATHIC INFLAMMATORY MYOSITIS JOURNEY THRU' 2019

The Bohan and Peter classification criteria were the most widely used criteria for classifying idiopathic inflammatory myopathies (IIM) and are in use since 1975 [1]. In 2017, the ACR/EULAR criteria based on the score for various features were proposed and have been validated in different ethnicities [2]. All the studies showed that the 2017 criteria performed better than Bohan and Peter criteria in classifying a patient with IIM but performed less satisfactorily in classifying patients as polymyositis. From India, Pinto et al. compared the performance of 2017 ACR/EULAR criteria and Bohan and Peter criteria in 111 IIM patients [3]. Overall both the criteria had a weak agreement between them ($\varkappa$-0.331) but they performed similarly if muscle biopsy findings were not considered. Both the criteria also performed poorly with respect to polymyositis. In their study from Australia, Luu et al. evaluated the addition of MRI and non-Jo1 antibodies to the 2017 ACR/EULAR criteria and concluded that including MRI and non-Jo1 antibodies as covariates may improve the accuracy of 2017 ACR/EULAR classification criteria [4].

In a retrospective analysis from Italy, Cavagna et al. described the clinical course 828 anti-synthetase syndrome (ASS) patients [5]. Patients with anti-Jo1 antibodies had higher rates of arthritis at onset and the majority developed ILD and myositis on follow up. Among patients who did not have ILD at presentation, 63% developed ILD on follow up. Among patients with anti-PL7 antibodies, ILD was the most common finding at presentation and about half of the patients developed myositis and arthritis on follow up. Among patients without myositis at presentation, 61% developed myositis on follow up. Anti-PL12 antibody patients had ILD as the predominant presenting manifestation and only 30% of patients developed complete ASS on follow up. Only 24% of patients without myositis at onset developed myositis on follow up. Majority of the anti-EJ antibody patients had incomplete ASS at onset with ILD being the commonest manifestation. In the follow-up, majority of patients developed myositis. Anti-OJ antibody patients presented with equal frequencies of arthritis, ILD and myositis, and only 22% developed complete ASS on follow up. Survival rates did not differ with the antibody type.

In a retrospective analysis, Melki et al. included 13 anti-MDA5 antibody positive patients and 51 non-MDA5 patients of juvenile IIM [6]. Patients with anti-MDA5 antibodies had more arthritis, skin ulceration, ILD and lupus-like features but milder muscular involvement along with increased levels of IFNα compared to patients without anti-MDA5 antibodies. Also, patients with anti-MDA5 antibodies required more than two immunosuppressant drugs to achieve remission.

In their case-control study, Aggarwal et al. found that 48% of patients out of 48 traditional MSA negative necrotizing myositis patients had anti-HMGCR antibodies while they were found in only 5% in the control group [7]. They also documented statin exposure in 78% of patients with anti-HMGCR antibodies and these patients also had severe muscle weakness, significantly elevated creatinine kinase levels, no other organ involvement and favourable response to immunosuppression. Aussy et al. reported that 78% of

dermatomyositis patients with anti-TIF1γ antibodies had malignancies and 41% of patients died on follow up [8]. The positive predictive value for cancer was 100% if the fluorescence intensity of anti-TIF1γ IgG2 was more than 385.

Gallay et al. reviewed the role of interferons in IIM and concluded that they play a key role in the pathogenesis of IIMs and interferon signature gene transcripts can help refine the IIM subgroups and help to identify patients that can respond to specific therapies like JAK/STAT inhibitors [9].

Challa et al. published histopathology findings of 27 patients with juvenile IIM and showed that juvenile dermatomyositis was characterised by perifascicular atrophy, necrosis, degeneration and regeneration of muscle fibers in all patients and perivascular inflammation in the majority of the patients [10].

The Brazilian Society of Rheumatology recently proposed the guidelines for the treatment of systemic autoimmune myopathies [11]. They recommended oral glucocorticoids as first line therapy (grade C) and methotrexate, azathioprine, and cyclosporine as steroid-sparing drugs (grade B). For glucocorticoid refractory cases, they recommended IVIG, tacrolimus, cyclosporine, cyclophosphamide, azathioprine, methotrexate, abatacept, tocilizumab, and rituximab, either as monotherapy or combination therapy (grade B). They also recommended against the use of anti-TNFα drugs in IIMs.

Both methotrexate and azathioprine were shown to have similar efficacy and adverse event profile in a retrospective analysis of 102 ASS patients by Casal-Dominguez et al. [12]. In another retrospective analysis, Keyber et al. have also demonstrated equal efficacy of methotrexate and azathioprine in the management of adult IIM patients [13]. Azathioprine and mycophenolate mofetil has been shown to be effective in improving FVC and DLCO in patients with myositis associated ILD but adverse events were more frequent with azathioprine compared to MMF [14]. The role of biologics and newer therapies in IIM were reviewed by Khoo et al. and Patwardhan et al. [15, 16].

In their study, Gupta et al. reported a higher risk of abortion (RR= 3.6), obstetric and fetal complications among women who conceived after the onset of myositis compared to pregnancies before the onset of myositis [17]. Infections were the most common cause (63%) of in-hospital mortality among IIM patients as reported by Muhammed et al. in their cohort of 38 IIM patients [18].

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