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CASE REPORT

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Factors predicting relapse in dermatomyositis

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Abstract

Dermatomyositis (DM) is a rare autoimmune disease characterized by classic cutaneous manifestations and proximal muscle inflammation. Clinical symptoms are improved in many patients on treatments, but some experiences relapse. No consensus exists regarding the definition of a DM relapse. In practice, a relapse is diagnosed when the symptoms recur or worsen after first improving under treatment. Published data on both prevalence and risk factors of relapse in DM is limited. The aim of this study was to evaluate the frequency of relapse in DM and identify factors predicting relapse in patients with non-paraneoplastic DM.

Case Report

We analyzed data of patients with DM diagnosed in our department of Dermatology at Ibn Sina Hospital in Rabat between January 1998 and January 2018 collected retrospectively. An inclusion criterion was a diagnosis of DM meeting Bohan and Peter criteria (probable or definite). We did not include patients with paraneoplastic DM.

Initially, 78 patients with DM were identified but only 59 had a non-paraneoplastic DM. The mean age of enrolled patients was 45 years, of whom 62.71% were female. 17 patients (28.8%) experienced relapse at least once (10 showed exacerbation of myositis, 4 showed exacerbations of skin lesions and 2 showed exacerbation of fatigue). The mean period to first relapse after disease stabilization was 8.5 months. The number of relapses per patient ranged from 1 to 6. The time from onset of symptoms to diagnosis of DM tended to be longer in relapsed patients (8 months) than in non-relapsed patients (3 months) and the difference was significant (p 0.04). No statically significant relation was found between relapse and age, gender, educational status, marital status or comorbidities. Patients with alcohol abuse seemed more likely to have a relapse (p 0.02). All patient enrolled had the characteristic cutaneous manifestations of DM but without statistical significance. Proximal muscle weakness was found in 48 (81.3%), distal deficit in 13 patients (22%). Signs of severe muscle involvement consisted of dysphagia (42.3%) and dysphonia (20.33%). A significant relation existed between distal muscle weakness and relapse (p 0.01). All patients received corticosteroid therapy. 6 patients also received methylprednisolone bolus therapy. Methotrexate was used in 8 patients and intravenous immunoglobulin was administered in 1 patient. 17 patients received hydroxychloroquine. Median follow-up was 24 months (1 month to 11 years). Disease stabilization in the enrolled patients was achieved at a mean of 2 months after the initiation of treatment. 40 patients (38%) experienced complications: at least one infection (20 patients), bone complications (5 patients), diabetes mellitus (5 patients) hypertension (2 patients) and 8 deaths. No significant difference was observed for any other factors such as complications, immune status, paraclinic results (myogenic enzymes, myogenic abnormalities in electromyography, histological findings) and treatment status.

Discussion

Several reports have investigated risk factors for mortality, refractoriness to treatment and disease progression in patients with DM but published data on risk factors for relapse is limited. The 28.81% relapse rate in our study is lower than in previous studies (40-65%) [1,2]. Time from diagnosis to first relapse was 8.5 months and most relapses occurred during the corticosteroid taper. Agarwal et al. [2] showed that advanced age and longer time to diagnosis were possible risk factors for relapse in DM without statistical significance. Our study showed that longer time to diagnosis seemed to be a significant factor to have a relapse. We evaluated the relation between sociodemographic variables, comorbidities and relapse, but detected no differences. Patients with alcohol abuse seemed more likely to have a relapse (p 0.02). The presence of autoantibodies in a wide range of tissues in alcohol abusers supports the possibility that other illnesses in the alcoholic are of autoimmune origin like the Alcoholic liver disease and kidney disease [3]. Distal muscle weakness is usually mild in DM patients and it is often severe. Distal deficit at presentation significantly predicted a relapsing course by univariate analysis. Vuong et al. [4] showed that dysphonia and severe skin involvement significantly predicted a relapsing course. Like our study, in 77 adults with polymyositis or DM, dysphonia and dysphagia were not associated with relapse [5]. Tatebe et al. [1] showed that anti-SS-A/Ro positivity was a risk factor for higher relapse-rate.

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