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Inflammatory Muscle Diseases.

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Inflammatory Muscle Diseases

TO THE EDITOR: In the review article on inflammatory muscle diseases, Dalakas (April 30 issue)[1] outlines four major subtypes. In 1998, Gherardi et al. described a new but underrecognized clinical variant, called macrophagic myofasciitis,[2] a condition that presents as diffuse myalgias of variable intensity associated with chronic fatigue. Myalgia predominantly affects the lower limbs and is often aggravated by exercise. It is associated with arthralgias in 50 to 60% of patients, as well as fever in 30% of patients.[3] Results on electromyography may be myopathic, and creatine kinase levels may be elevated in up to 50% of patients.[4] This disease is associated with intramuscular injection of aluminum hydroxide, which is used as an adjuvant in several vaccines (hepatitis A and B).[3] Muscle biopsy, including the fascia, at the site of the immunization reveals a pathognomonic focal macrophage infiltration with intracytoplasmic crystalline inclusions corresponding to aluminum deposits.[2] This picture is part of the inflammatory autoimmune syndrome induced by adjuvants.[5]

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No potential conflict of interest relevant to this letter was reported.

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TO THE EDITOR: We are concerned about the statement in the review article by Dalakas that the long-term outcome of inflammatory myopathies has improved, with "a 10-year survival rate of more than 90%." In 2006, Airio et al. reported a 10-year survival rate of 55% among patients with polymyositis and 53% among those with dermatomyositis.[1] In 2012, Schiopu et al. reported a 10-year survival rate of 62% in inflammatory myopathies.[2] In 2014, Aggarwal et el. reported a 10-year survival rate of 70% among patients with a positive autoantibody response to histidyl-transfer RNA synthetase (Jo-1) and 50% among patients with a negative response.[3] Taborda et al. reported a 10-year survival rate of more than 90% in the United Kingdom,[4] as cited by Dalakas. In general, the mean 10-year survival rate among patients with inflammatory myopathies from the cited four studies is approximately 69%, a rate that might be affected by ethnic group, serologic presentation, and disease severity. Moreover, in patients in whom the myopathy is associated with cancer, the prognosis is even worse. Though diagnostic methods and therapies have improved, we should avoid being overly optimistic about the prognosis.

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No potential conflict of interest relevant to this letter was reported.

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THE AUTHOR REPLIES: Young et al. refer to a macrophage-mediated inflammatory disorder that affects muscle fascia but not myofibers that are

remote from the macrophagic infiltrates.[1] In contrast to inflammatory muscle diseases, macrophagic myofasciitis presents with myalgia and chronic fatigue but not muscle weakness, has focal histopathological features that are limited to the fascia and occasionally perifascicular fibers of deltoid muscles in sites of previous vaccinations, and is predominantly, if not exclusively, seen in France, where the deltoid muscles (used for vaccinations) are routinely preferred for diagnostic biopsies.[1] Macrophagic myofasciitis is a seemingly toxic fasciitis, not a primary inflammatory muscle disease, that is linked to aluminum-containing substrates in some vaccines, similar to the eosinophilia–myalgia syndrome caused by adulterated L-tryptophan.[2] Whether these focal fascial macrophagic collections are incidental reactions to the injected vaccines or trigger the reported systemic symptoms of fatigue, myalgia, and cognitive dysfunction remains unclear. The proposed hypothesis that there is an autoimmune or inflammatory syndrome induced by adjuvants is beyond the scope of my review.

Chen et al. refer to survival rates in four different studies with heterogeneous diagnostic criteria that have not been adjusted for systemic conditions or systematically applied immunotherapies. Thus, lumping the rates into one group to obtain a mean survival rate is not appropriate. The study they cite by Airio et al. refers to patients who were evaluated 30 to 46 years ago, whereas the study they cite by Aggarwal et al. was limited to patients with Jo-1 antibodies who often have interstitial lung disease. The reference that I cited (in spite of its limitations) is more representative because it includes patients from one center in which practitioners were experienced in modern immunotherapies. Improved supportive care and the application of new therapies, such as intravenous immune globulin and rituximab, probably account for improved rates of survival. A life expectancy of 81 years was reportedly normal even for patients with inclusion-body myositis, the most disabling of the myositis subtypes.[3] The numbers of patients who were included in these studies are arguably small, and we need better survival studies that take into consideration various factors including correct diagnosis (ensuring specific rates for each disease subtype), the patient's age at onset, the presence of cancer or pulmonary involvement, the time from the initiation of therapy, disease severity, and the judicious use of immunotherapies and good supportive care. I hope that the data in my review article will lead to increased diagnostic accuracy, early initiation of more effective immunotherapies, and higher survival rates.

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Since publication of his article, Dr. Dalakas reports having received consulting fees from Hoffmann–La Roche. No further potential conflict of interest relevant to this letter was reported.

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