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Rheumatology: Current Research

Perspective

Identifying and Treating of Inflammatory Myopathy

Caldeira Ferreira*

Department of Rheumatology, University College London, London, UK

ABOUT THE STUDY

Inflammatory myopathy is a condition marked by muscle weakening, inflammation, and (in certain kinds) discomfort. Many cases of inflammatory myopathy are idiopathic, meaning they have no known etiology. These cases are categorized based on their symptoms, signs, electromyography, MRI, and lab results. Moreover, it may be linked to underlying malignancy. Polymyositis (PM), Dermatomyositis (DM), and inclusion-body myositis are the three primary subtypes of Idiopathic Inflammatory Myopathy (IIM).

Diagnosis

A physician will only identify an Idiopathic Inflammatory Myopathy (IIM) condition in a case after all other known causes of myopathy have been ruled out. Weakness in the muscles of the head, neck, trunk, upper arms, or upper legs, elevated blood serum concentrations of various muscle enzymes, such as creatine kinase, abnormal muscle alterations on electromyography, and biopsy findings of PM are the typical criteria for a diagnosis of PM. Muscle cell regrowth and ageing as well as inflammatory infiltrates that are chronic in nature. The diagnosis is DM if heliotrope (purple) rash or Gottron's papules are also present. Myositis in DM may not be clinically obvious, but it can be found with biopsy or MRI.

Muscle cell biopsy results showing:

- Basophilic granules surround the cytoplasmic vacuoles in the cell
- Inflammatory infiltrate consisting primarily of CD8 T lymphocytes and macrophages, along with filamentous inclusions in both the cytoplasm and nucleus, should raise suspicion of IIM if the criteria for PM are satisfied, but muscle weakness spreads to the hands and feet or does not involve pain.

Treatment

Because of the very low prevalence of inflammatory myopathies, there haven't been many randomized therapy trials. Enhancing daily life skills and muscle strength is the aim of treatment. Immunosuppression, a form of treatment, involves reducing immune system function. Nearly all PM or DM patients respond to treatment in some way, at least initially, and many make a full recovery with maintenance therapy. (If treatment for PM or DM does not result in an immediate improvement, the diagnosis should be carefully reexamined.) The majority of IIM patients will require assistive aids like a cane, a walking frame, or a wheelchair because there is no known effective treatment for the condition.

Polymyositis and dermatomyositis

However, often, treatment starts with a single daily, high dose of oral corticosteroid taken after breakfast. In severe cases of PM and DM with systemic symptoms, an initial three to five days on intravenous corticosteroid (methylprednisolone) may be utilized (prednisone). To lessen the side effects of the prednisone, the strength of the dose given every other day is gradually decreased after about a month over the course of three to four months. Prednisone may be combined with "steroid sparing" oral immunosuppressants including azathioprine, mycophenolate mofetil, methotrexate, and cyclosporine when a high dose cannot be reduced without compromising muscle strength or when prednisone is working but causing serious side effects.

Some of these steroid-sparing medications may take several months to show results. Patients on corticosteroids should adhere to a rigorous high-protein, low-carb, low-salt diet to reduce side effects. Long-term corticosteroid treatment should also be supplemented with a daily calcium supplement, a weekly vitamin D supplement, and a weekly dosage of fosamax for postmenopausal women. Little evidence supports the use of intravenous immunoglobulin, cyclosporine, tacrolimus, mycophenolate mofetil, and other medicines in individuals who do not react to this treatment strategy. Rituximab trials have also suggested a potential therapeutic benefit.

Inclusion-body myositis

There is no established effective therapy for IIM, despite the fact that it has a very similar clinical presentation to PM. IIM does not respond to the medications that successfully treat PM. Intravenous immunoglobulin may help with dysphagia (difficulty

Correspondence to: Caldeira Ferreira, Department of Rheumatology, University College London, London, UK, Email: Ferreira89@yahoo.com

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swallowing), while additional research is required. Exercise that builds strength steadily and without fatigue has proven to be beneficial. Occupational and rehabilitation therapists can recommend safe walking techniques and help with fine motor activities. They can also recommend the right wheelchairs, braces, and canes. Speech therapists can offer suggestions on avoiding choking incidents and lowering anxiety associated with a potential aspiration in both patients and careers.

Epidemiology

Between 2.18 and 7.7 persons per million are given a PM or DM diagnosis each year. Juvenile dermatomyositis, also known as DM, is diagnosed in roughly 3.2 children per million each year, with an average age of onset of seven years.

Adult DM diagnosis often arises between the ages of 30 and 50. The onset of PM, an adult condition, often occurs after the age of twenty. The prevalence of PM and DM is higher in women, higher in Caucasians, and lower in Asians.

IIM appears after the age of 30 (typically after 50), and may be more prevalent in males. At any given time, 35.5 individuals per million have it.