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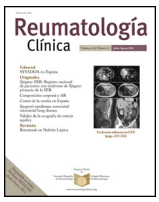

Original Article

Multicenter Registry on Inflammatory Myositis From the Rheumatology Society in Madrid, Spain: Descriptive Analysis☆

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ABSTRACT

Objective: To analyze clinical characteristics, survival and causes of death of patients diagnosed with autoimmune inflammatory myositis in the REMICAM registry from the Society of Rheumatology in the Community of Madrid (SORCOM).
Methods: Multicenter cohort of patients diagnosed with autoimmune inflammatory myopathy with follow-up between January 1980 and December 2014. A total of 313 variables concerning demographic, clinical and morbidity data were collected, and a comparison was performed between clinical subgroups.
Results: A total of 479 patients were recruited from 12 centers with 14% of patients lost to follow-up. Seventy-four percent of cases were women, age at diagnosis of 44 ± 23 years and a mean follow-up period of 10 ± 8 years. The most frequent clinical subgroups were primary myositis (PM 29%, DM 22%), followed by overlap myositis (20.5%), juvenile myositis (18%), myositis associated with cancer (8%), immune-mediated necrotizing myositis (1%) and inclusion body myositis (1%). During the follow-up period, a total of 114 deaths (28%) were registered, the main causes being cancer (24%), infections (23%) and cardiovascular events (21%).
Conclusions: A total of 479 patients were recruited in the REMICAM registry of inflammatory myopathies. Including sociodemographic, clinical and prognostic information, it represents the largest Spanish multicenter registry to date in rheumatology, and constitutes an important source for conducting further substudies.

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Registro de pacientes con miopatía inflamatoria de la Sociedad Madrileña de Reumatología: análisis descriptivo

Palabras clave:
Miopatía inflamatoria idiopática
Registro
Estudio descriptivo

R E S U M E N

Objetivos: Describir las características clínicas, mortalidad y causas de muerte de una serie de pacientes diagnosticados de miositis inflamatoria idiopática del registro REMICAM de la Sociedad de Reumatología de la Comunidad de Madrid (SORCOM).
Métodos: Estudio descriptivo retrospectivo multicéntrico de una cohorte de pacientes con diagnóstico de miositis inflamatoria idiopática en seguimiento en servicios de reumatología de hospitales de la Comunidad de Madrid entre enero de 1980 y diciembre de 2014. Se han recogido hasta un total de 313 variables acerca de aspectos demográficos, clínicos y de morbimortalidad, y se ha realizado una comparación entre subgrupos clínicos.
Resultados: Se han reclutado 479 pacientes procedentes de 12 centros, con un 14% de pérdidas durante el periodo de seguimiento. El 74% de los casos eran mujeres, una edad al diagnóstico de 44 ± 23 años, y una media de seguimiento de 10 ± 8 años. Los subgrupos clínicos más frecuentes fueron las formas primarias (PM 29%, DM 22%), seguidas de síndrome de solapamiento (20,5%), miopatías juveniles (18%), miopatías asociadas a cáncer (8%), miopatías necrosantes inmunomediadas (1%) y miositis por cuerpos de inclusión (1%). Durante el periodo de seguimiento se produjeron un total de 114 fallecimientos (28%), siendo las principales causas el cáncer (24%), las infecciones (23%) y los eventos cardiovasculares (21%).
Conclusiones: En el registro REMICAM de miopatías inflamatorias de la Comunidad de Madrid se han reclutado 479 casos de miositis inflamatoria idiopática con datos sociodemográficos, clínicos y pronósticos, suponiendo el mayor registro multicéntrico español en el ámbito de la Reumatología hasta la fecha, y constituyendo una fuente importante para la realización de posteriores subestudios.

Introduction

Idiopathic inflammatory myopathies (IIM) comprise a heterogenic group of systemic diseases that are characterized by nonsuppurative inflammation of skeletal muscle and progressive muscle weakness, occasionally accompanied by systemic manifestations. It is considered a rare disease with a worldwide mean incidence between 19 cases/million population per year.[1] This varies depending on the geographic region, the research methods utilized and the classification criteria applied. Before the use of steroids and immunosuppressive therapy, the rates of mortality were high.[2,3] The generalized utilization of these treatments, as well as early diagnosis, resulted in a significant improvement in survival.[4–6] The clinical manifestations of IIM, as well as its course and prognosis, are extremely heterogeneous, which together with its low prevalence and the lack of multicenter studies, makes the study of the disease a difficult undertaking.

To date, multicenter studies of IIM in the rheumatology setting have not been performed either in the Community of Madrid or in the rest of Spain. For this reason, and with the support of the Society of Rheumatology of the Community of Madrid (SORCOM), we proposed the creation of a group for the Registry of Inflammatory Myopathies in the autonomous community of Madrid (REMICAM) for the cross-sectional registry of patients, the period of inclusion of which has concluded. The availability of a multicenter observational registry of patients with IIM could enable us to envision the reality of this disease in the Community of Madrid, as well as to determine the rates of morbidity and mortality and compare subgroups of patients, in a disease whose rarity limits the possibility of obtaining significant data.

Patients and Methods

Study Objectives

The main objectives of the REMICAM are to describe and characterize IIM patients, who usually managed in the rheumatology departments of the Community of Madrid, for the purpose of:

1. Describing the sociodemographic and clinical characteristics
2. Studying the rate of comorbidity and the cumulative incidence, and possible differences among distinct types of myopathies
3. Calculating the mortality rate in the overall series and by clinical subgroups

As secondary objectives, the registry proposes a multicenter collaborative study to subsequently examine specific aspects of autoimmune inflammatory myopathies.

Study Design

Retrospective multicenter registry of patients with IIM from units or departments of rheumatology in the Community of Madrid, using a hospital-based cross-sectional registry, recording retrospective information on clinical data, mortality and cause of death.

Patient Selection

Through SORCOM, the invitation to participate in the present study was sent to the different departments or units of rheumatology of public hospitals in the Community of Madrid, as well as the rheumatology department of Hospital Madrid Norte in Sanchinarro, a borough of Madrid. We included unselected consecutive patients with a diagnosis of IIM (dermatomyositis, polymyositis, inclusion body myositis and immune-mediated necrotizing myositis), who had undergone follow-up in the above departments at some time during the period between January 1980 and December 2014, without taking into account their age at onset of the process. The patients ultimately selected met the criteria of Bohan and Peter[7,8] and/or Tanimoto's classification criteria,[9] and we excluded toxic and infectious myopathies and myopathies secondary to neuromuscular disease. The patients were classified into 7 subgroups: idiopathic polymyositis (PM), idiopathic dermatomyositis (DM), juvenile myositis (JM), IIM associated with another connective tissue disease (overlap syndrome), cancer-associated IIM, inclusion body myositis[10] and immune-mediated necrotizing myositis.[11] Patients with overlap syndrome had to meet criteria for IIM and