Adult dermatomyositis: experience of a Brazilian tertiary care center

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ABSTRACT

Objective: To report the results of a retrospective cohort involving 139 patients with dermatomyositis, conducted from 1991 to 2011. **Methods:** All patients met at least four of the five Bohan and Peter criteria (1975). **Results:** The patients' mean age at disease onset was 41.7 ± 14.1 years, and mean disease duration was 7.2 ± 5.2 years. The sample comprised 90.2% white patients and 79.9% female patients. Constitutional symptoms occurred in less than half of the patients. Cutaneous and joint involvements occurred in 95.7% and 41.7% of the patients, respectively. Incipient pneumopathy, ground glass opacities and/or pulmonary fibrosis were present in 48.2% of the patients. All patients received prednisone (1 mg/kg/day) and 51.1% also received intravenous methylprednisolone (1 g/day for three days). Several immunosuppressants were used as corticosteroid sparing agents according to tolerance, side effects and/or refractoriness. Although disease relapse (clinical and/or laboratory) occurred in 53.2% of the patients, 76.3% were in disease remission at the end of the study. The rate of severe infection was 35.3%, and herpes zoster predominated. There were 15 (10.8%) cases of cancer, 12 within one year after the diagnosis. There were 16 deaths (11.5%), and their major causes were sepsis/septic shock (27.5%), pneumopathy attributed to the disease (31.3%), neoplasms (31.3%), and cardiovascular events (12.5%). **Conclusions:** In this study, the clinical and laboratory data were similar to those of other population groups described in the literature, with minimal differences regarding the frequency and characteristics of the extramuscular manifestations.

Keywords: dermatomyositis, myositis, epidemiology.

INTRODUCTION

Idiopathic inflammatory myopathies are a heterogeneous group of systemic autoimmune diseases, such as dermatomyositis (DM), polymyositis (PM), and inclusion body myositis (IBM), which have distinct clinical, epidemiological, histological and pathological characteristics.

Dermatomyositis is characterized by symmetrical and progressive proximal muscle weakness of the limbs, in addition to typical skin changes, such as heliotrope rash and Gottron's papules. Extramuscular manifestations, such as joint, cardiac, pulmonary, and gastrointestinal involvements, can occur in DM.[1,2]

The incidence of DM is 5–10 cases/million people/year. The adult form affects mainly individuals aged 45 to 55 years, while the juvenile form affects individuals aged 5 to 10 years. Women are twice as likely as men to be diagnosed with DM, and no ethnic predilection is observed.[3]

Epidemiological studies on DM are few in the literature.[4–9] So far, only two studies have been published in Brazil. One of them has assessed mortality in DM and PM,[10] while the other has assessed cases of juvenile DM.[11]

The present study aimed at describing the clinical and laboratory characteristics of a large case series of patients with adult DM being followed up at a tertiary care center.

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PATIENTS AND METHODS

This study assessed 139 patients with DM originating from the Service of Myopathies of our tertiary care center. To improve the homogeneity of the population studied, only patients being followed up at our service from January 1991 to August 2011 and meeting at least four of the five Bohan and Peter diagnostic criteria were considered.[12,13] Those diagnosed with amyopathic DM and those under the age of 18 years were not included. This study was approved by the local Ethics Committee [HC 0039/10].

Demographic data, clinical manifestations and laboratory findings were obtained from systematic review of medical records. The laboratory findings considered were those obtained at the time of disease diagnosis, while the clinical manifestations considered were those presented during the patients' follow-up. The following parameters were assessed: constitutional symptoms; skin changes (heliotrope rash, Gottron's papules, ulcers, facial rash, "V" sign, shawl sign, calcinosis, and vasculitis); joint involvement (arthralgia and/or arthritis); dysphagia; dysphonia; dyspnea; and muscle weakness of the limbs (grade 0: no muscle contraction; grade I: signs of mild muscle contraction; grade II: normal range of motion, but inability to move against gravity; grade III: normal range of motion against gravity; grade IV: integral mobility against gravity and against a certain degree of resistance; grade V: complete mobility against marked resistance and against gravity).[14] Complementary tests [electroneuromyography, and muscle biopsy (biceps brachii or vastus lateralis)] were requested routinely in the first medical visit. Creatine kinase (normal range: 24–173 IU/L) and aldolase (normal range: 1.0–7.5 IU/L) levels were measured by using a kinetic automated method. Autoantibodies against cellular components were sought by use of indirect immunofluorescence, with Hep-2 cells as substrate. Erythrocyte sedimentation rate and C-reactive protein were assessed by using Westergren method and nephelometry, respectively.

The patients were initially treated with corticosteroids (oral prednisone, 1 mg/kg/day), with later gradual dose reduction according to clinical and laboratory stability. When disease was severe (progression of dyspnea, dysphagia, significant loss of muscle strength), pulse therapy with methylprednisolone (1 g/day for three consecutive days) was performed. The following corticosteroid sparing agents were used as monotherapy or in combination: azathioprine (2–3 mg/kg/day); methotrexate (20–25 mg/week); cyclosporine (2–4 mg/kg/day); mycophenolate mofetil (2–3 g/day); leflunomide (20 mg/day); cyclophosphamide (0.5–1.0 g/m^2 of body surface); chloroquine diphosphate (< 4 mg/kg/day); and intravenous human immunoglobulin (1 g/kg/day for two consecutive days). Cyclophosphamide was used in the presence of progressive dyspnea associated with pulmonary parenchymal change confirmed on computed tomography ("ground-glass" opacity or honeycombing).

Disease relapse was defined as recurrence of the initial clinical findings and/or increased serum levels of muscle enzymes attributed to disease activity, after ruling out infections and/or neoplasms.

Infections requiring hospitalization or intravenous and/or prolonged antibiotic therapy were considered severe. Cases of herpes zoster and the fatal ones were also assessed.

Continuous variables were expressed as mean ± standard deviation (SD), and categorical variables were expressed as percentages. Student *t* test and chi-square test were used to analyze parametric and nonparametric data, respectively. The program STATA version 7.0 (STATA, College Station, TX, USA) was used for the analysis, and P values < 0.05 were considered statistically significant.

RESULTS

This study assessed 139 patients with DM, 111 of whom were women (4:1). Most patients were white (90.2%), their mean age at disease onset was 41.7 ± 14.1 years (18–84 years), and their mean disease duration was 7.2 ± 5.2 years (0–20 years) (Table 1).

The interval between symptom onset and DM diagnosis was 4.5 ± 5.4 months (1–36 months). Despite that short interval, 41.0% of the patients had dysphagia and 20.9% of the patients were bedridden at the time of diagnosis, evidencing the severity of the disease in that group of patients. In fact, regarding muscle strength in the upper and/or lower limbs, approximately 16.5% of the patients had grade III, most patients had grade IV, and, the minority, grade V.

Constitutional symptoms occurred in more than 40% of the patients. The antinuclear antibody (ANA) was positive

Table 1
Demographic findings in Brazilian patients with dermatomyositis

Age ± SD, mean (years)*	41.7 ± 14.1 (18–84)
Duration of disease ± SD, mean (years)	7.2 ± 5.2 (1–20)
Female sex (%)	111 (79.9)
White color (%)	124 (90.2)
Duration of symptoms prior to diagnosis ± SD, mean (months)	4.5 ± 5.4 (1–36)

SD: standard deviation.

in 62.7% of the patients, with titers ranging from 1:20 to > 1:320, most of them of the nuclear speckled pattern (Table 2). No correlation between ANA positivity and demographic data, clinical and laboratory parameters, and neoplasms was observed in the present study (P > 0.05). The other clinical manifestations and laboratory data are shown in Tables 2 and 3, respectively.

All patients received prednisone (1 mg/kg/day) at the time of diagnosis, and 49.6% underwent pulse therapy with methylprednisolone (1 g/day for three consecutive days) because of the severity of the disease (Table 4). Different immunosuppressant corticosteroid sparing agents were used in monotherapy or in combination, depending on the tolerance, side effects and refractoriness of the disease, as follows: azathioprine in 66.9% of the patients; methotrexate in 50.4%; chloroquine diphosphate in 25.9%; cyclosporine in 23.7%; intravenous human immunoglobulin in 14.4%; intravenous cyclophosphamide in 10.1%; mycophenolate mofetil in 3.6%; and leflunomide in 0.7%. At the end of the present study, 56 of the 80 patients were using one immunosuppressant as a corticosteroid sparing agent, 23 patients were on two, and one patient was on three immunosuppressant corticosteroid sparing agents.

During follow-up, clinical relapse was identified in 74 patients (53.2%), being, in most cases, associated with corticotherapy reduction.

Neoplasm was identified in 15 patients (10.8%), 12 of whom were confirmed within one year from disease diagnosis, as follows: ovary, uterus, lung, and skin cancer (two cases of each); thyroid, prostate, and breast cancer (one case of each); and cancer in two sites (one case). Metastasis was identified in three patients. One patient had ductal neoplasm of the breast 12 years after the diagnosis of DM, and two others had basal cell carcinoma of the face and upper limbs one year before DM. The clinical and laboratory characteristics and extramuscular manifestations were similar in patients with and without neoplasm (P > 0.05).

Severe infection was observed in 49 patients (35.3%) (Table 5). The major cause was herpes zoster (42.9% of the cases), followed by cellulitis (26.5%), abscess in different

Table 2
Clinical findings in Brazilian patients with dermatomyositis

Clinical findings	n (%)
Constitutional symptoms	65 (46.8)
Bedridden	29 (20.9)
Muscle weakness	
Upper limbs	
Grade V	8 (5.8)
Grade IV	108 (77.7)
Grade III	23 (16.5)
Lower limbs	
Grade V	5 (3.6)
Grade IV	107 (78.0)
Grade III	27 (19.4)
Skin lesion	
Gottron's papules	133 (95.7)
Heliotrope rash	117 (84.2)
Ulcers	20 (14.4)
Calcinosis	9 (6.5)
Vasculitis	31 (22.3)
Facial rash	69 (49.6)
"V" sign	21 (15.1)
Shawl sign	14 (14.4)
Articular	58 (41.7)
Respiratory tract	
Dysphonia	24 (17.3)
Dyspnea	43 (30.9)
Gastrointestinal tract	
Dysphagia	57 (41.0)

Table 3
Laboratory findings in Brazilian patients with dermatomyositis

Muscle enzymes	
Creatine kinase ± SD (U/L)	3,677.9 ± 5,463.0
Aldolase ± SD (U/L)	41.4 ± 73.2
ESR (mm/1st hour)	32.5 ± 26.9
CRP (mg/L)	12.0 ± 15.5
Antinuclear antibody (%)	87 (62.6)
Nuclear speckled	51 (37.4)
Nuclear homogeneous	19 (13.7)
Nuclear smooth dense speckled	3 (2.2)
Nucleolar	7 (5.0)
Cytoplasmic	6 (4.3)

SD: standard deviation; ESR: erythrocyte sedimentation rate; CRP: C-reactive protein.

Table 4
Medicamentous therapy used in Brazilian patients with dermatomyositis

Medicamentous therapy	n (%)
Methylprednisolone	70 (49.6)
Prednisone	139 (100.0)
Azathioprine	93 (66.9)
Methotrexate	70 (50.4)
Chloroquine diphosphate	36 (25.9)
Cyclosporine	33 (24.1)
Intravenous human immunoglobulin	20 (14.4)
Cyclophosphamide	14 (10.1)
Mycophenolate mofetil	5 (3.6)
Leflunomide	1 (0.7)

Table 5
Comorbidities in Brazilian patients with adult dermatomyositis

Comorbidities	n (%)
Infections	49 (100)
Herpes zoster	21 (42.9)
Cellulitis	13 (26.5)
Abscess*	10 (20.4)
Bronchopneumonia	7 (14.3)
Sepsis	5 (10.2)
Calcinosis	4 (8.2)
Endocarditis	2 (4.1)
Pyoarthritis	1 (2.0)
Osteomyelitis	1 (2.0)
Deaths	16 (100)
Sepsis/Septic shock	6 (37.5)
Pneumopathy	5 (31.3)
Neoplasya	5 (31.3)
Cardiovascular events	2 (12.5)

*Abscess: retroesophageal and gluteal regions, ankles, hands, elbows.

tissues (retroesophageal region, gluteal region, ankles, hands and elbows – 20.4%), and bronchopneumonia (14.3%). Other prevalent causes were calcinosis infection (8.2%), endocarditis (4.1%), pyoarthritis (2.0%), and osteomyelitis (2.0%).

Sixteen patients (11.5%) died, and the major causes were as follows: sepsis/septic shock (37.5% of the patients; three patients had bronchoaspiration); pneumopathy attributed to the disease (31.3% of the patients; pulmonary fibrosis); neoplasm (31.3% of the patients); and cardiovascular events (12.5% of the patients; acute myocardial infarction) (Table 5). The neoplasms were spinocellular carcinoma of the lung, colon neoplasm, and three uterus neoplasms, and with metastases. One hundred and six patients (76.3%) remain on outpatient clinical follow-up.

DISCUSSION

The present study assessed the demographic and therapeutic profiles and outcome of a large case series of patients with DM of a Brazilian tertiary care center, providing, thus, an overview of that population.

The mean age of the patients assessed was 42 years, and most of them were women and white, a demographic profile similar to that of other studies in the literature.[4–6] That distribution is different from those of juvenile DM and DM associated with other connective tissue diseases, whose sex distributions are 1:1 and 10:1, respectively.[5–7,14–16]

Similarly to other systemic autoimmune diseases, almost half of the patients studied had constitutional symptoms. Despite the short interval between symptom onset and the diagnosis of DM (mean of five months), approximately one fifth of the patients were bedridden and almost half of the patients had dysphagia, evidencing the severity of the disease in that group. Early diagnosis is essential not only to improve the quality of life, but also to avoid sequelae, such as muscular atrophy/hypotrophy or dysphagia-related aspiration bronchopneumonia, which increase even more morbidity and mortality.

Although the major symptoms found in DM were muscular, we observed that almost half of the patients also had joint involvement, such as arthralgia and/or arthritides, of neither erosive nor deforming characteristics, affecting their quality of life.

Lungs are involved in 5%–47% of the patients,[17,18] and the following can be found: dyspnea; cough; chest pain; reduced physical exercise tolerance; and respiratory failure. Tomographic images can reveal changes in the pulmonary parenchyma, such as "ground-glass" opacity, linear opacities, pulmonary consolidations and/or micronodules.[17] At the time of clinical investigation, at least half of the patients showed pulmonary parenchymal changes on tomography, and one third had dyspnea on moderate exertion at the time of diagnosis. Regarding clinical outcome and mortality, one third of the patients died due to pulmonary complications.

Dermatomyositis has typical cutaneous manifestations, such as heliotrope rash and Gottron's papules, which can either precede or manifest concomitantly with or after muscle involvement.[19] Among our patients, the most common finding was Gottron's papules, in contrast with studies performed in Senegal[4] and Tunisia,[6] reporting greater frequency of erythematous lesions. Calcinosis, which occurs typically in 40% of juvenile DM, but is not a frequent adult manifestation, was found in only 6% of our patients. The delay in starting treatment and the severity of the disease are risk factors for the development of calcinosis.[20] Other relatively frequent lesions were cutaneous ulcers and vasculitides, which, in addition to being possible signs of the severity of the disease, are secondary infection areas. In fact, in the present study, a high prevalence of cellulitis and abscesses was observed, also contributing to increase morbidity and mortality.

Even without controlled and randomized studies, corticosteroids are considered first-line therapy for DM.[21,22] Of the immunosuppressive agents, the following are second-line treatment: azathioprine; cyclosporine; cyclophosphamide; and methotrexate.[22] There are no protocols defining the best treatment for DM. The medical assessment should be based individually on the severity of the clinical presentation, disease duration, presence of extramuscular manifestations, and contraindication to specific therapeutic agents.

In this study, all patients received corticotherapy at disease onset and relapses. Due to the severity of the clinical and laboratory manifestations at the time of diagnosis, half of the patients also underwent pulse therapy with methylprednisolone. Regarding immunosuppressive drugs, azathioprine was more frequently used, followed by methotrexate and cyclosporine. The use of chloroquine diphosphate was reserved as adjuvant to cutaneous treatment, while cyclophosphamide was reserved to patients with severe pulmonary involvement defined by the clinical and tomographic findings. Human immunoglobulin was used for refractory cases and those with infections. Mycophenolate mofetil was reserved to the few cases refractory to other immunosuppressive drugs and/or those with contraindications/side effects – leflunomide was predominantly used for joint involvement.

Regarding comorbidities, systemic arterial hypertension and diabetes mellitus were the most often found at the time of diagnosis, and their prevalence increased during the patients' follow-up, which might be related to the medications used. Comparing with the literature, those comorbidities were less frequently reported in our study than in the study by Limaye et al.[23]

Cardiovascular complications are associated with high morbidity and mortality among patients with DM,[24,25] as are neoplasms and pulmonary involvement. In patients with inflammatory myopathies, cardiovascular manifestations are characterized by electrocardiographic abnormalities, valvular heart diseases, coronary vasculitis, ischemic changes, heart failure, and myocarditis.[24,25] However, ischemic cerebrovascular accidents, embolism originating from the heart, and thrombophilia are rare findings in myopathies.[24,25] In the present study, the prevalence of cardiovascular manifestations (acute myocardial infarction and cerebrovascular accident) was low, which can be partially explained by the short follow-up of the patients (mean of eight years). According to the literature, the frequency of cardiovascular involvement in patients with inflammatory myopathies ranges from 6% to 75%.[24,25]

The incidence of neoplasms is approximately ten times greater in inflammatory myopathies as compared to that of the general population, ranging from 3% to 12.6% in patients with DM.[26–29] The risk of cancer is greater in the three first years after the diagnosis, and exists even in patients under the age of 45 years.[26–29] The most commonly affected sites are as follows: gastrointestinal; gynecologic; pulmonary; and hematologic (lymphoma).[26–30]

In this study, the prevalence of neoplasms was around 10%, and most of them were gynecologic (ovary, uterus, and breast). Two thirds of the malignancies were detected right after the diagnosis of DM, within a mean follow-up period of 1–2 years; approximately two thirds are observed within the first year or simultaneously with the myopathy.

Infections are still the major cause of mortality in patients with DM, accounting for 9%–30% of the deaths.[31–33] Several microorganisms have been implicated in opportunistic and pyogenic infections in that subgroup of patients,[31–33] at rates ranging from 14% to 37.3%.[31–33] The most frequent are mycobacteria and fungi (*Pneumocystis jiroveci*, *Candida* sp.).[31–33] In our case series, the infection rate was 14%, and herpes zoster predominated.

In addition to cardiovascular,[24,25] infectious,[31–33] and pulmonary events, the following factors are associated with the increased mortality in DM: thrombocytopenia;[34] diabetes mellitus;[34] neoplasm;[10,34] advanced age;[34] creatine kinase levels;[35] and interstitial pulmonary disease.[35] The percentage of deaths ranges from 7.8% to 28%.[10,34–36] During follow-up, death occurred in 11.5% of the patients, an index lower than that of Chinese[34] (28,6%) and Japanese[35] (26.9%) studies, and slightly greater than the statistics of Taiwan (7.68%).[36] In our case series, sepsis was the major cause of death, followed by neoplasms, and involvement of the respiratory tract and cardiovascular system, as reported in other studies.[24,25,31–33]

Based on the clinical and demographic data shown in this article, the population studied can serve as a reference for the Brazilian population with adult DM due to the following: similarities in the predominance of the female gender and age group; minimal differences in the frequency and characteristics of extramuscular manifestations; mortality rate in accordance with that of the literature; and emphasis on infection as the major cause of death in that group of patients.

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