Arq Neuropsiquiatr 2000;58(3-B):789-799

DIAGNOSIS OF DERMATOMYOSITIS AND POLYMYOSITIS

A STUDY OF 102 CASES

ROSANA HERMINIA SCOLA, LINEU CESAR WERNECK**, DANIEL MONTE SERRAT PREVEDELLO***, EDIMAR LEANDRO TODERKE****, FÁBIO MASSAITI IWAMOTO******

ABSTRACT - Patients with dermatomyositis (DM) or polymyositis (PM) were studied retrospectively. The patients were divided into four groups: definite PM 24, probable PM 19, definite DM 34 and mild-early DM 25 cases. PM patients complained more often proximal muscle weakness [p <0.01]. DM patients complained more arthralgia [p <0.05], dysphagia [p <0.03] and weight loss [p <0.04]. Five patients had a malignant neoplasm and 9 had other connective-tissue disease. DM presented higher ESR than PM [p <0.002]. PM presented more significant increase in creatine kinase (CK) [p <0.02] and in alanine aminotransferase (ALT) [p <0.001] levels. Electromyography showed myopathic pattern in 76%. Muscle biopsy was the definitive test. Perifascicular atrophy was more frequent in definite DM than in mild-early DM group [p <0.03]. *Conclusion:* A small association with connective-tissue diseases and neoplasms was found. DM and PM are clinically different. DM presents systemic involvement affecting the skin, developing more severe arthralgia, dysphagia and weight loss and presenting higher values of ESR. PM presents a restricted and more significant involvement of muscles generating more weakness complaints and higher levels of serum muscle enzymes.

KEY WORDS: polymyositis, dermatomyositis, inflammatory myopathy, diagnosis.

Diagnóstico de dermatomiosite e polimiosite: estudo de 102 casos

RESUMO - Pacientes com dermatomiosite (DM) ou polimiosite (PM) foram estudados retrospectivamente. Os pacientes foram divididos em quatro grupos: PM definida 24, PM provável 19, DM definida 34 e DM leve-inicial 25 casos. Pacientes com PM queixaram-se mais de fraqueza muscular proximal [p<0,01]. Já os pacientes com DM se queixaram mais de artralgia [p<0,05], disfagia [p<0,03] e perda de peso [p<0,04]. Cinco pacientes tiveram neoplasia e nove tiveram outra colagenose. A DM apresentou VHS mais elevada que a PM [p<0,002]. A PM apresentou um aumento mais importante dos níveis de creatinaquinase [p<0,02] e alanina aminotransferase [p<0,001]. A eletromiografia mostrou padrão miopático em 76%. A biopsia muscular foi o teste definitivo. Atrofia perifascicular foi mais frequente na DM definida que na DM leve-inicial [p<0,03]. *Conclusão:* Foi encontrada baixa frequência de associação com colagenoses e neoplasias. DM e PM são clinicamente diferentes. A DM apresenta envolvimento sistêmico afetando a pele, desenvolvendo quadro mais severo de artralgia, disfagia, perda de peso e gerando valores mais elevados de VHS. A PM apresenta um acometimento restrito e mais importante da musculatura ocasionando maiores queixas de fraqueza e mais altos níveis séricos das enzimas musculares.

PALAVRAS-CHAVE: polimiosite, dermatomiosite, miopatia inflamatória, diagnóstico.

Study accomplished in the Neuromuscular Disorders Service, Neurology Division and Internal Medicine Department, Hospital de Clínicas, Federal University of Paraná, Curitiba, Brazil. Supported in part by a grant from CNPq (Brazil): *Associate Professor of Medicine; **Professor of Medicine and Chief; ***Medical Student CNPq-Research Scholarship Holder; ****Medical Student; *****Resident in Neurology. Aceite: 29-junho-2000.

Dra. Rosana Herminia Scola - Serviço de Doenças Neuromusculares, Hospital de Clínicas da UFPR - Rua General Carneiro 181, 3° andar - 80069-155 Curitiba PR - Brasil. Fax: 55 41 264 3606. E-mail: scola@hc.ufpr.br

The idiopathic inflammatory myopathies (IIM) are a heterogeneous group of acquired muscle diseases, and they can be viewed as comprising three major and discrete groups: polymyositis, dermatomyositis and inclusion-body myositis. Proximal muscle weakness and inflammatory infiltrates within the skeletal muscle characterize polymyositis (PM). When myositis is accompanied by characteristic skin lesions, it is called dermatomyositis (DM)[1-4]. The skin manifestations include a heliotrope rash (blue-purple discoloration) on the upper eyelids with edema, a flat red rash on the face and upper trunk, and erythema of the knuckles accompanied by a raised, violaceous scaly eruption (Gottron's sign). The incidence of PM, DM and inclusion-body myositis is approximately 1 in 100,000[3,5]. The cause of those diseases is still ignored, but the existence of genetic factors and autoimmune mechanisms is known[1,2,4,6]. Viral and bacterial infections as well as some drugs can also be related with the etiology of the inflammatory myopathies, but this type of condition needs to be distinguished from the idiopathic form being discussed[4,6-10].

The critical tests for establishing and confirming the diagnosis of polymyositis or dermatomyositis are measurement of serum muscle enzymes, electromyography, and muscle biopsy[3,5,8,11].

The most sensitive enzyme is creatine kinase (CK). Levels of aspartate aminotransferase, alanine aminotransferase, and lactate dehydrogenase are often elevated and levels of aldolase, myoglobin, and creatine may also be elevated. However, these enzymes are not specific for PM/DM, and they could be elevated in several other myopathies[3,5,8,11]. The needle electromyography shows myopathic potentials characterized by short-duration, low-amplitude polyphasic units on voluntary activation and increased spontaneous activity with fibrillations, complex repetitive discharges, and positive sharp waves. Besides this electromyographic pattern is not specific for DM/PM, the presence of mixed myopathic and neurogenic potentials may also be found in advanced cases of PM/DM[3,5,8]. Muscle biopsy is the test for establishing the definitive diagnosis of inflammatory myopathies[3,5,8,12,13].

The objective of the present study is to acquire information on clinical signs and symptoms, laboratory features, electromyographies and muscle biopsies from patients with IIM comparing them with the literature with a view to seeking differences between DM and PM. Due to the heterogeneity of presentations, it is essential to have deep knowledge of the ways in which the idiopathic inflammatory myopathies manifest themselves, because had the diagnosis been established, the treatment with steroids and immunosuppressive drugs is effective in the improvement of the symptoms and in the remission of the disease.

METHOD

The records were searched for patients with a diagnosis of polymyositis and dermatomyositis from 1976 to 1998. The patients were appraised in the Neuromuscular Disorders Service of the "Hospital de Clínicas" of the "Universidade Federal do Paraná" in Curitiba-Brazil. All clinical records bearing these diagnoses were then individually revised and the following data were obtained: (1) demographic: age of first hospital diagnosis, sex; (2) dates of disease onset (first symptom); (3) clinical information relevant to PM and DM, neoplasms and other connective-tissue disease including data from clinical history focused on chief complaints, and general/neurological physical examination, which analyzed typical skin rash and proximal and distal muscle strength, tonus and reflexes in upper and lower limbs; (4) laboratory values of erythrocyte sedimentation rate (ESR) and levels of serum muscle enzymes creatine kinase (CK), lactate dehydrogenase (LDH), aldolase (ALD), aspartate aminotransferase (AST) and alanine aminotransferase (ALT), which were analyzed in how many times normal the mean was from normal values; (5) needle electromyographic (EMG) abnormalities and (6) histopathological findings on fresh-frozen muscle biopsy, which was accomplished in all patients included in the study and submitted to the following staining and histochemical reaction: hematoxilin-eosin, modified Gomori trichrome, oil red O, PAS, cresyl violet, sirius red, NADH-tetrazolium reductase, ATPases pH 4.3, 4.6, 9.4, myophosphorylase, non-specific esterase, alkaline phosphatase, acid phosphatase, succinic dehidrogenase and cytochrome c-oxidase[12,13].

A case was included if the diagnosis of PM or DM was accepted after a review of the clinical records. Any feature suggesting another disease was enough to exclude de case. Muscle biopsies showing rimmed vacuoles, whose histology suggests cytoplasm inclusion bodies were excluded intending to exclude the cases of inclusion-bodies myositis[14]. A case was excluded if the nerve conduction study was abnormal. In the circumstance of overlap with other connective-tissue disease, a case was accepted only if the myositis was the primary diagnosis.

Cases were classified as definite or probable polymyositis and definite or mild-early dermatomyositis by using Dalakas established criteria[3]. Each subgroup was subdivided in male and female subgroups with a view to seeking differences of signs and symptoms between them.

Statistical analysis: differences between observed and expected frequencies of clinical presentations among groups and subgroups of PM/DM were tested by using the χ^2 method, with Yates' correction when needed. Differences between mean values such as ages and serum muscle enzyme levels were tested by t-test for independent samples. Statistical differences were considered significant when the probability of α error (p) was smaller than 0.05.

RESULTS

We identified 102 patients, 59 (57.8%) with dermatomyositis and 43 (42.2%) with polymyositis. Among the patients with dermatomyositis, 34 were classified as definite and 25 as mild-early disease. Of the patients diagnosed as having polymyositis, 24 were classified as definite and 19 as probable. As regards the demographic aspects, 61 patients were female and 41 were male (F: M=1.5:1). Considering the patients with dermatomyositis, 36 were female and 23 were male (F: M=1.6:1) and as regards the patients with polymyositis, 25 were female and 18 were male (F: M=1.4:1). The mean age of the patients at diagnosis of the disease was of 21 years old varying from 0,1 to 84 years. The median age of the patients with definite dermatomyositis was 17 years old; with mild-early dermatomyositis, 20 years old; with definite polymyositis, 26 years old; with probable polymyositis, 32 years old. Twelve patients with polymyositis and 27 with dermatomyositis were younger than 14, and there was no significant age difference among groups.

The definite dermatomyositis patients' most frequent complaint was proximal muscle weakness followed by muscle pain and dysphagia (Table 1). Patients with mild-early dermatomyositis mainly complained proximal muscle weakness, followed by muscle pain, fever and arthralgia. Patients with definite polymyositis mainly complained proximal muscle weakness, followed by muscle pain and dysphagia. Patients with probable polymyositis complained proximal muscle weakness, followed by the muscle pain and distal muscle weakness. The patients with PM differed from the patients

Table 1. Signs and symptoms of 102 patients with dermatomyositis and polymyositis.

General signs and symptoms	Definite Dermatomyositis N=34	Mild-early Dermatomyositis N=25	Definite Polymyositis N= 24	Probable Polymyositis N=19
Proximal weakness	28 (82.4%)	17 (68%)	23 (95.8%)	18 (94.7%)
Distal weakness	6 (17.7%)	2 (8%)	2 (8.3%)	7 (36.8%)
Muscle pains	15 (44.1%)	15 (60%)	11 (45.8%)	10 (52.6%)
Arthralgia	9 (29.5%)	11 (44%)	2 (8.3%)	5 (26.3%)
Arthritis	4 (11.8%)	3 (12%)	1 (4.2%)	1 (5.3%)
Dysphagia	15 (44.1%)	8 (32%)	8 (33.3%)	0
Fever	6 (17.7%)	11 (44%)	3 (12.5%)	4 (21.1%)
Raynaud's phenomenon	8 (23.5%)	3 (12%)	3 (12.5%)	1 (5.3%)
Weight loss	6 (17.7%)	9 (36%)	3 (12.5%)	1 (5.3%)
Erythematous rash-including heliotrope	23 (67.7%)	24 (96%)	0	0
Subcutaneous calcification	3 (8.8%)	6 (24%)	0	0
Gottron's sign	9 (26.5%)	10 (40%)	0	0

There is the possibility that one patient had shown more than one sign or symptom. The weaknesses expressed on this table are the patients' complaints, so they are subjective. The values were analyzed by the χ^2 method, and the statistical differences and the respective probability of α error (p) are in the text.

Table 2. The more frequent neurological signs of 102 patients with dermatomyositis and polymyositis.

Neurological signs	Definite DM n=34	Mild-early DM n=25	Definite PM n=24	Probable PM n=19
Decrease of proximal muscle strength in upper limbs	32 (94.1%)	20 (80%)	24 (100%)	17 (89.5%)
Decrease of proximal muscle strength in lower limbs	**33 (97.1%)**	**16 (64%)**	22 (91.7%)	16 (84.2%)
Decrease of distal muscle strength	11 (32.4%)	4 (16%)	7 (29.2%)	4 (21.1%)
Proximal muscle atrophy	12 (35.3%)	9 (36%)	11 (45.8%)	7 (36.8%)
Distal muscle atrophy	9 (26.5%)	5 (20%)	7 (29.2%)	5 (26.3%)
Hypotonic muscles	5 (14.7%)	2 (8%)	7 (29.2%)	2 (10.5%)
Absence of reflexes	14 (41.2%)	8 (32%)	14 (58.3%)	7 (36.8%)

The same patient can present more than one alteration in the neurological exam.
DM, dermatomyositis; PM, polymyositis. The bold values presented significant statistical difference **(p=0.000825)**.

with DM as they presented more frequently proximal muscle weakness complaint (p=0.0089). On the other hand, patients with DM differed from patients with PM, in that they presented more frequently arthralgia (p=0.046), dysphagia (p=0.027) and weight loss (p=0.038) complaints.

In the two subgroups with PM, distal muscle weakness complaint was more often found among patients with a probable diagnosis (p=0.022).

Between the two subgroups with DM, the fever complaint was found more frequently among patients with mild-early dermatomyositis (p=0.027).

The presence of "mechanic's hands", lateral and palmar areas of the fingers rough and cracked with irregular blackened horizontal lines, was only found in two women, one with definite polymyositis and one with definite dermatomyositis.

The specific signs and symptoms most frequently found in dermatomyositis were skin rash including heliotrope rash, subcutaneous calcifications and Gottron's sign. Skin rash was present in 79.6 % of patients with dermatomyositis, and 9 patients (15.3%) had subcutaneous calcifications. Between the two subgroups with DM, the erythematous rash was found more frequently among patients with mild-early dermatomyositis (p=0.0075).

Each subgroup subdivided into male and female subgroups was analyzed in relation to signs and symptoms. Despite the fact that the frequency of signs and symptoms often differs by as much as 50% between male and female within the same subgroup, there is no statistical difference if the χ^2 method is used with Yates' correction for small samples.

The neurological exam was not statistically able to differentiate DM from PM (Table 2). Among patients with DM, the decrease of proximal muscle strength in inferior members was more often present in definite DM than in mild-early DM (p=0.0008). All patients with definite polymyositis had proximal muscle strength decreased in superior members and 92% had it decreased in inferior members. The patients with probable polymyositis presented proximal muscle weakness mainly in the superior members. It is important to stress that the patients presented, either in superior or inferior members, some level of proximal muscle weakness, except for two men and three women included in the mild-early dermatomyositis, whose strength was normal.

We detected an association to other diseases in 34 cases (Table 3). There were 20 patients with cardiac abnormalities or pulmonary involvement (arrhythmia, atrioventricular conduction defect, dilated cardiomyopathy, dyspnea and interstitial lung disease), 5 with malignant neoplasms (two

Table 3. Associated clinical findings of 102 patients with dermatomyositis and polymyositis.

Associated clinical findings	Definite DM N=34	Mild-early DM N=25	Definite PM N=24	Probable PM N=19
Cardiac and pulmonary abnormalities	5 (14.7%)	6 (24%)	6 (25%)	3 (15.8%)
Malignant conditions	1 (2.9%)	3 (12%)	1 (4.2%)	0
Overlap syndrome	3 SS (8.8%) 1 RA (2.9%)	4 SS (16%)	0	1 SS (5.3%)

DM, dermatomyositis; PM, polimyositis; SS, systemic sclerosis; RA, rheumatoid arthritis.
There was no statistical difference among groups.

cases of breast adenocarcinoma, an ovary adenocarcinoma, a lung's small cells tumor and a Hodgkin lymphoma), and nine cases with connective-tissue disease (one case of rheumatoid arthritis and eight cases of systemic sclerosis).

Patients with DM had higher ESR than patients with PM (DM=36.9±20.3 mm and PM=16.5±11.6 mm p=0.00112) (Fig 1). Patients with PM presented a more significant increase of CK (DM=781.8±1812 U/L and PM=1770±1866 U/L p=0.015) and of ALT (DM=32.6±27.7 U/L and PM=76.8±61.5 U/L p=0.0005). CK was increased in 67.6% of the patients (1170.5±1886.6 U/L), and it was 15.7 times from normal in definite DM, 5.5 times in mild-early DM, 24.7 times in definite PM and 26.2 times in probable PM. The other enzymes, LDH, AST and ALT, didn't present an increase higher than 3-fold. Aldolase was the second more increased enzyme achieving 12-fold in definite DM and 10-fold in definite PM.

The electromyography in patients with definite DM (n=34) presented a myopathic pattern in 88.2%, mixed potentials in 5.8% and bordering on myopathic in 5.8% of the patients (Fig 2). On the other hand, the EMG showed normal patterns in 35.7%, myopathic in 50%, chronic denervation in 7.1% and active denervation in 7.1% of the patients with mild-early DM (n=14). Patients with

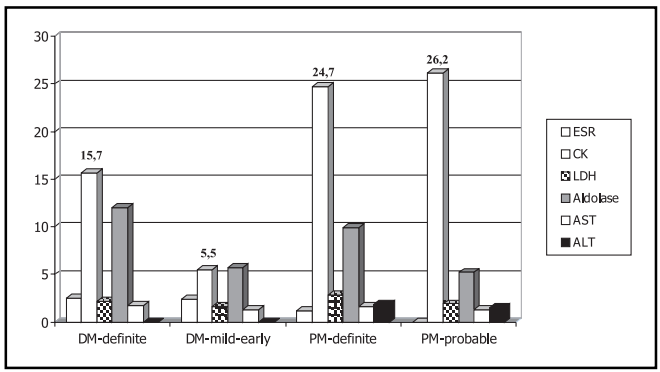

Fig 1. There was no statistical difference when definite DM (dermatomyositis) was compared with mild-early DM or when definite PM (polymyositis) was compared with probable PM by test-t for independent samples. However, the mean value of ESR (erythrocyte sedimentation rate) was higher in DM than in PM (p=0.00112), the mean value of CK (creatine kinase) was higher in PM than in DM (p=0.015) and the mean value of ALT (alanine aminotransferase) was more elevated in PM than in DM (p=0.000472). LDH = lactate dehydrogenase, AST = aspartate aminotransferase.

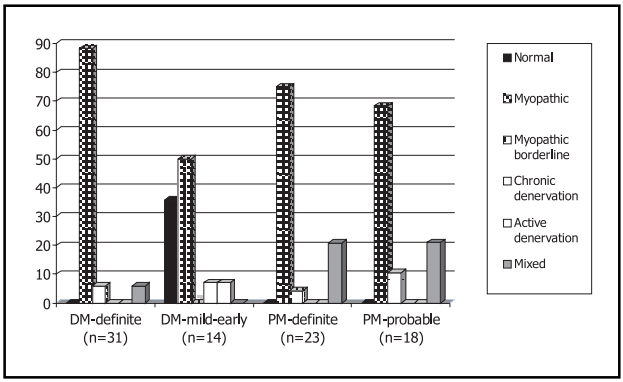

Fig 2. There was no statistical difference among the four groups. DM= dermatomyositis, PM= polymyositis.

definite PM (n=24) presented a myopathic pattern in 75%, mixed potentials in 20.8% and bordering on myopathic in 4.2%. In the patients with probable PM (n=19) the EMG showed a myopathic pattern in 68.4%, mixed in 21% and bordering on myopathic pattern in 10.5% of the patients. The electromyography was not able to differentiate DM from PM due to the fact that the myopathic was the main pattern found among all the patients.

A diagnosis of an inflammatory myopathy was established in 81% of muscle biopsies (Table 4). Among the patients with DM, the inflammatory infiltrates pattern was dermatomyositis specific, with perifascicular involvement, in 68% of the cases. Among the patients with PM, the inflammatory infiltrates pattern was polymyositis specific, with endomysial infiltration (Fig 3), in 39.5% of the cases. The perifascicular atrophy (Fig 4), characteristic of DM, was present more statistically in the group of definite diagnosis (p=0.024). Only one patient with definite polymyositis presented a mild perifascicular atrophy in addition to the characteristic inflammatory infiltrates pattern of polymyositis.

Table 4. The muscle biopsy of 102 patients with dermatomyositis and polymyositis.

Muscle biopsy patterns	DM Definite		DM Mild-Early		PM Definite	PM Probable
	With PFA	Without PFA	With PFA	Without PFA		
Inflammatory myopathy-dermatomyositis	21	7	7	5	0	0
Inflammatory myopathy-polymyositis	0	0	0	0	17	0
Inflammatory myopathy-unspecific	0	4	0	5	7	7
Inflammatory myopathy-borderline	0	1	0	2	0	0
Inflammatory myopathy-absent	1	0	1	1	0	5
Only fibers type 2 atrophy	0	0	0	3	0	5
Normal	0	0	0	1	0	2
Total	22	12	8	17		
	34		25		24	19

PFA, perifascicular atrophy.
The perifascicular atrophy was statistically more frequent in definite dermatomyositis (p=0,024) than in mild-early dermatomyositis.

Table 5. Signs, symptoms and laboratory features found by some studies about idiopathic inflammatory myopathies in general.

	This study Paraná, Brazil	Uthman 1996 Canada	Koh 1993 Singapore	Lundberg 1992 Sweden	Ramirez 1990 UK	Hochberg 1986 USA	Holden 1985 Canada	Tymms 1985 Australia
Mean age at dignosis (yr)	26.6	47.2	50.3	49	37	45.3	48.5	50.4
F/M	1.5:1	2:1	1.9:1	3.8:1	2.5:1	3:1	1.6:1	2.4:1
Number of Patients	102	30	75	29	25	76	36	105
Proximal weakness	95%	90%	87%	100%	100%	93%	_	85%
Dysphagia	30.4%	43%	11%	52%	_	45%	31%	29%
Arthralgia	26.5%	47%	35%	45%	28%	29%	17%	65%
Raynaud's phenomenon	14.7%	23%	7%	59%	36%	26%	_	39%
CK increased	71%	90%	90%	_	84%	96%	_	68%

F, female; M, male; CK, Creatine kinase; yr, year.

DISCUSSION

The primary studies regarding dermatomyositis and polymyositis were frequently conflictant and contradictory due to a lack of clearly defined diagnostic criteria[15,16]. In the present work, muscle pain was the most common symptom after proximal muscle weakness, like in other studies[16-18]. Myalgia and muscle tenderness is a frequent symptom, usually early in the disease, and more often

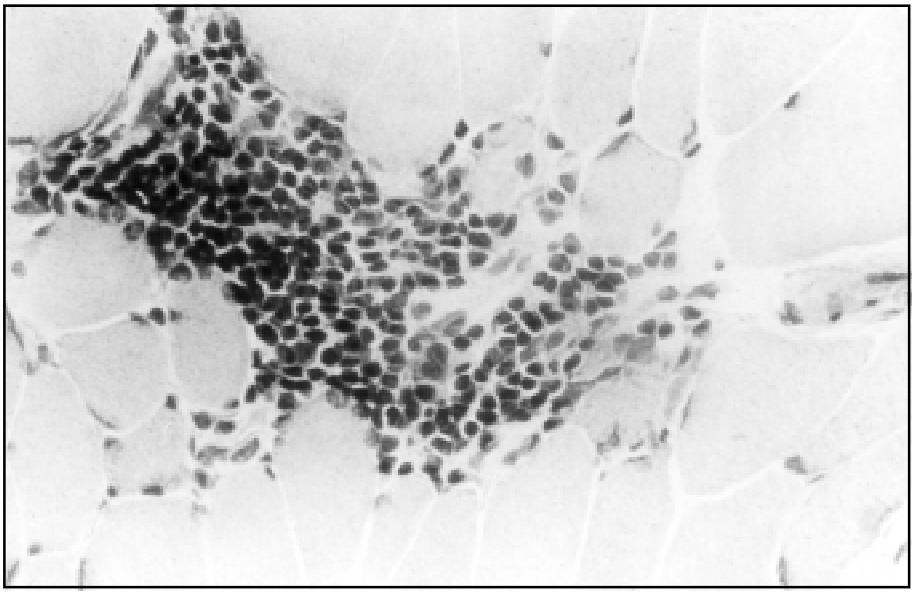

Fig 3. Transverse section of a fresh-frozen muscle biopsy specimen from a patient with polymyositis showing Endomysial mononuclear cells infiltration (hematoxylin and eosin x 348).

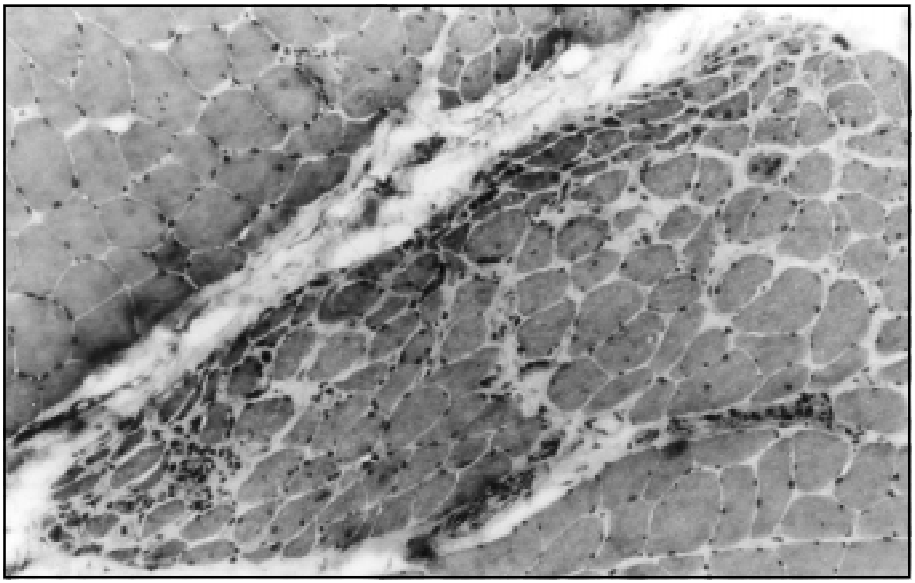

Fig 4. Transverse section of a fresh-frozen muscle biopsy specimen from a patient with dermatomyositis showing Perifascicular atrophy with some mononuclear cells in the endomysium (hematoxylin and eosin x 174).

in dermatomyositis than in polymyositis[3]. Because of this fact, myalgia has already been proposed to be included in the diagnostic criteria[18]. The present study evidences that myalgia is an important symptom in reinforcing the diagnosis, however it is unable to determine it. Table 5 shows the comparison of the demographic data, clinical presentations and laboratory features between our study and others[17,19-24].

Approximately one third of patients with polymyositis and almost a half of the patients with dermatomyositis were under 14 years of age in our study. The important prevalence of children with dermatomyositis resulted in a younger mean age in DM than in PM, but without significant statistical difference. In the same way, the mean diagnosis age was considerably younger in our study than in the consulted ones. This fact corroborates the consulted literature confirming that dermatomyositis occurs more frequently in children[3].

We have found more women with DM and PM forming an index of 1.5:1 (F: M); however there is not an appreciable statistical difference. The frequency of idiopathic inflammatory myopathies between the sexes is classically understood as affecting more women than men[3,8]. In spite of the fact that the whole studied literature is unanimous regarding the fact that women are more often affected with DM and PM, our study presented one of the lowest rates of F: M found, and it is very similar to the one found in the study by Holden at al[24].

The proximal muscle weakness was the most significant aspect found in 95.1% of the patients, in spite of being a complaint of just 85.3%. Almost all patients presented proximal muscle weakness in some studies[21,22]. The patients with polymyositis presented statistically more complaints about muscle weakness than did the patients with dermatomyositis. Because the complaint of distal muscle weakness was more often found in patients with probable polymyositis than in those with definite polymyositis, the cases of probable PM may be a superposition of diseases or even another disease as inclusion-body myositis which classically affects the distal muscles. It is not enough to rule out the diagnosis of inclusion-body myositis on the basis of the muscle biopsy[3,14]. The distal muscle weakness and the lower female to male ratio among the PM patients may be a clue to the fact that some of the patients classified as probable PM are indeed cases of inclusion-body myositis. Such fact reinforces the importance of restrictive criteria in order to have an exact final diagnosis.

Dysphagia happened significantly more in patients with dermatomyositis and it was reported by 30% of all the patients, which is very close to what the studies conducted in Canada and Australia found[17,24]. The patients with dermatomyositis complained more about arthralgia, and other studies are alike[22,23].

Raynaud's phenomenon was not often reported by the patients, and its frequency was not statistically different between DM and PM or between sexes as in other works[17,19]. Fever was statistically more frequently presented by patients with mild-early DM than by those with definite DM, and this aspect corroborates the consulted literature, which refers fever to be one of the commonest initial manifestations of dermatomyositis[3]. We believe that dermatomyositis generates a more important systemic repercussion mainly because the patients with dermatomyositis presented perceptible loss of weight during the disease, which statistically did not happen in the same proportion in the case of patients affected by PM.

The fact that erythematous rash was more often presented by the patients with mild-early DM than by those with definite DM in our study could be explained by the wider contingent of biopsies presenting perifascicular atrophy in the definite DM group. It happens because the presence of perifascicular atrophy in a muscle biopsy is diagnostic of dermatomyositis, if there is clinical and laboratory evidence, even in the absence of inflammation or evident erythematous rash. Perifascicular atrophy is not present in polymyositis or inclusion-body myositis[3].

The distribution of signs and symptoms with regard to patients' sex showed that the clinical picture is the same for men and women of the same diagnostic group.

The neurological exam could statistically differentiate definite DM from mild-early DM because decrease of proximal muscle strength in inferior members was present in almost all patients with definite DM, while it was present in only 64% of patients with mild-early DM. Studies have demonstrated that the presence of muscle weakness is not necessary for the diagnosis of DM because it is not often found in the early or mild disease[3].

We found 9% of cases in association with connective-tissue diseases (CTD), mainly among patients with DM. Systemic sclerosis was the main disease in association with both DM and PM. Eight had DM while only 1 had PM. The term overlap syndrome is often used to indicate that the characteristics of two different disorders are common to both. Many studies have shown that only dermatomyositis, and not polymyositis, truly overlaps in up to 20 percent of patients, with connective-tissue diseases, and only with systemic sclerosis and mixed connective tissue disease[3,16,19,25]. However, there is a study showing 19 patients with PM and 2 patients with DM in overlap with other CTD among 177 patients[26].

We found only 5% of cases associated with malignancy. It is important to remember that the study of Medsger and collaborators did not find an association of PM and DM with malignancy[27]. Nevertheless, the association of the idiopathic inflammatory myopathies with malignancy was first noticed in 1916 by Stertz and Kankeleit[28]. The prevalence of such an association has been described as varying between 7 to 28 percent of cases and the exact nature of the relationship is not known. A cohort study, conducted in Sweden, showed that cancer risk in the population with PM and DM was significantly increased, as well as a larger mortality rate[28].

The levels of muscle enzymes, mainly CK, were increased in most patients as also shown by other studies (table 7). CK as well as ALT significantly reached higher levels in patients with polymyositis than in those with dermatomyositis. CK is expected to be increased by up to 50-fold its normal value and this happens mainly in PM[3]. The mean value of ESR was significantly higher in patients with dermatomyositis, and it could represent, as previously mentioned, a more intense systemic repercussion in DM than in PM.

Electromyography showed myopathic potentials in most of the patients. However, there were some patients who presented electromyographies showing mixed myopathic and neurogenic

potentials. Furthermore, high amplitude, long duration polyphasic motor unit action potentials (MUAP) were present in 14 percent of the patients with mild-early DM. The neurogenic potentials usually represent a consequence of the regeneration of muscle fibers and chronicity of the disease[29, 30]. The relative high incidence of neurogenic potentials among the patients with mild-early DM can be explained by the fact that the electromyographies had been done during different phases of the disease including the treatment when the regeneration of muscle fibers occurs. It could not be explained by the chronicity of the disease because they are early cases. However, the presence of neurogenic potentials calls for restrictive criteria in order to rule out another disease. The myopathic pattern in needle electromyography is characterized by short duration, low amplitude polyphasic units on voluntary action and increased spontaneous activity with fibrillations, complex repetitive discharges, and positive sharp waves. The electromyographies of both DM and PM are alike. Perhaps the widest use of electromyography is to exclude neurogenic disorders, which reduces the number of axons generating polyphasic units with high amplitude and long duration[3,15,29,30]. Normal electromyographies were found among patients with mild-early DM and are possibly correlated with the beginning of the disease as it is proposed by Dalakas in the diagnostic criteria[3].

It must be stressed that some patients with DM did not present evident erythematous rash or subcutaneous calcifications and the presence of perifascicular atrophy in the biopsy confirmed the diagnosis. There was only one female patient presenting mild perifascicular atrophy in the biopsy assigned to the PM group because she had no signs of skin manifestations and the biopsy showed infiltrates mostly in the fascicles (endomysial inflammation) surrounding and invading individual nonnecrotic muscle fibers besides perifascicular atrophy. The case reported might represent an overlap syndrome that could be called “dermatopolymyositis” or it may represent a simple case of DM without skin manifestation. The presence of perifascicular atrophy occurred mainly in patients with definite DM. The patients with mild-early DM had less perifascicular atrophy, which may indicate an early diagnosis. However, the absence of perifascicular atrophy calls for restrictive criteria in order to rule out another disease.

Finally, we consider DM and PM as being two different diseases. They are clinically different due to the systemic involvement present in DM involving skin, developing more severe arthralgia, dysphagia, weight loss and generating high values of ESR. On the other hand, PM presents a local and more severe involvement of the muscles causing more complaints of weakness and higher levels of muscle enzymes like CK and ALT.

REFERENCES

1. Mastaglia FL, Ojeda VJ. Inflammatory myopathies: part 1. Ann Neurol 1985;17:215-227.
2. Mastaglia FL, Ojeda VJ. Inflammatory myopathies: part 2. Ann Neurol 1985;17:317-323.
3. Dalakas MC. Polymyositis, dermatomyositis, and inclusion-body myositis. N Engl J Med 1991;325:1487-1498.
4. Miller FW. Myositis-specific antibodies: touchstones for understanding the inflammatory myopathies. JAMA 1993;270:1846-1849.
5. Plotz PH, Dalakas M, Leff RL, Love LA, Miller FW, Cronin ME. Current concepts in the idiopathic inflammatory myopathies: polymyositis, dermatomyositis, and related disorders. Ann Intern Med 1989;111:143-157.
6. Garlepp MJ. Genetics of the idiopathic inflammatory myopathies. Curr Opin Rheumatol 1996;8:514-520.
7. Illa I, Nath A, Dalakas M. Immunocytochemical and virological characteristics of HIV-associated inflammatory myopathies: similarities with seronegative polymyositis. Ann Neurol 1991;29:474-481.
8. Dalakas MC. Immunopathogenesis of inflammatory myopathies. Ann Neurol 1995;37:S74-S86.
9. Smadja D, Bellance R, Cabre P, Arfi S, Vernant JC. Clinical characteristics of HTLV-1 associated dermato-polymiositis. Acta Neurol Scand 1995;92:206-212.
10. Higuchi I, Hashimoto K, Kashio N, et al. Detection of HTLV-I provirus by in situ polymerase chain reaction in mononuclear inflammatory cells in skeletal muscle of viral carriers with polymyositis. Muscle Nerve 1995;18:854-858.
11. Plotz PH, Rider LG, Targoff IN, Raben N, O’Hanlon TP, Miller FW. Myositis: immunologic contributions to understanding cause, pathogenesis, and therapy. Ann Intern Med 1995;122:715-724.
12. Werneck LC. O valor da biópsia muscular em neurologia. Análise de 290 exames a fresco e pela histoquímica. Rev Bras Clin Terap 1981;10S:2-22.
13. Werneck LC. Estudo da biópsia muscular e sua relação com enzimas séricas e eletromiografias nas doenças musculares. Tese de Professor Titular, Universidade Federal do Paraná. Curitiba, 1991.
14. Scola RH, Werneck LC, Franco CR. Corpos de inclusão citoplasmática: estudo em diversas doenças e revisão da literatura. Arq Neuropsiquiatr 1996;54:245-259.

15. Bohan A, Peter JB. Polymyositis and dermatomyositis. N Engl J Med 1975;292:344- 347.
16. Bohan A, Peter JB, Bowman RL, Pearson CM. A computer assisted analysis of 153 patients with polymyositis and dermatomyositis. Medicine 1977;56:255-286.
17. Tymms KE, Webb J. Dermatopolymyositis and other connective tissue diseases: a review of 105 cases. J Rheumatol 1985;12:1140–1148.
18. De Vere R, Bradley WG. Polimyositis: its presentation, morbidity and mortality. Brain 1975;98:637-666.
19. Uthman I, Vázquez-Abad D, Senécal JL. Distinctive features of idiopathic inflammatory myopathies in French Canadians. Semin Arthritis Rheum 1996;26:447-458.
20. Koh ET, Seow A, Ong B, Rathagopal P, Tjia H, Chung HH. Adult onset polymyositis/dermatomyositis: clinical and laboratory features and treatment response in 75 patients. Ann Rheum Dis 1993;52:857-861.
21. Lundberg I, Nennesmo I, Hedfors E. A clinical serological, and histopathological study of myositis patients with and without anti-RNP antibodies. Semin Arthritis Rheum 1992;22:127–138.
22. Ramirez G, Asherson RA, Khamashta MA, Cervera R, D'Cruz D, Hughes GR. Adult onset polymyositis/dermatomyositis: description of 25 patients with emphasis on treatment. Semin Arthrits Rheum 1990;20:114–120.
23. Hochberg MC, Feldman D, Stevens MB. Adult onset polymyositis/dermatomyositis: an analysis of clinical and laboratory features and survival in 76 patients with review of the literature. Semin Arthritis Rheum 1986;15:168–178.
24. Holden DJ, Brownell AKW, Fritzler MJ. Clinical and serologic features of patients with polymyositis or dermatomyositis. Can Med Assoc J 1985;132:649–653.
25. Targoff IN. Immune manifestations of inflammatory muscle disease. Rheum Dis Clin N Am 1994;20:857-880.
26. Oddis CV, Conte CG, Steen VD, Medsger TA Jr. Incidence of polymyositis-dermatomyositis: a 20-year study of hospital diagnosed cases in Allegheny County, PA 1963-1982. J Rheumatol 1990;17:1329-1334.
27. Medsger TA Jr, Dawson WN Jr, Masi AT. The epidemiology of polymyositis. Am J Med 1970;48:715–723.
28. Sigurgeirsson B, Lindelöf B, Edhag O, Allander E. Risk of cancer in patients with polymyositis and dermatomyositis. N Engl J Med 1992;326:363-367.
29. Barkhaus PE, Nandedkar SD, Sandersm DB. Quantitative EMG in inflammatory myopathy. Muscle Nerve 1990;13:247-253.
30. Robinson LR. AAEM case report#22 Polymyositis. Muscle Nerve 1991;14:310-315.