Retrospective study of dermatomyositis. Discussion of 40 cases consulting a Dermatology Department

Mercedes Lidia Hassan[1], Ana Capecce[2], María E. Melloni[2], Miriam Saposnik[2], Ivana Ruzzi[3], Josefina Gramajo[4], Juana Lorenzo[5], Roberto Schröh[6], Cristina Corbella[7]

Abstract

Background. Dermatomyositis (DM) is included in idiopathic inflammatory miopathies, together with polymyositis, although clinical variants without muscle involvement are recognized, exclusively showing cutaneous involvement during variable periods of time, or remaining definitely amyopathic. Cutaneous manifestations are so characteristic that a diagnosis may be made even before any muscular involvement occurs. Association to internal malignancy, organ involvement, and evolution possibilities arouse interest of physcicians and dermatologists.
Material and methods. We retrospectively studied 40 patients (33 females) consecutively seen at the Dermatology Department of Hospital Ramos Mejía of Buenos Aires between June 1991, and June 2007.
Objective. To analyse this population composition, cutaneous and systemic involvement characteristics, and disease evolution.
Results. The following was found: 23 percent of amyopathic forms (9 of 40), 16.6 percent associated with internal malignancy (6/36), excluding four cases of childhood/juvenile DM. Malignancies were exclusively gynecological: breast (3), cervix (2), and ovary (1). Lapse between diagnosis of cancer and DM was from 0 to 2 years.Infrequent skin lesions and complications of these patients were described, in addition to currently accepted cutaneous criteria for the diagnosis of dermatomyositis. No "flagellated" erythema lesions or complete antisynthetase syndrome were found. Unsuspected interstitial lung disease with simple chest X-ray was detected in two cases, both non-positive for anti-Jo1 antibodies. One case of this series appeared three years after silicone mammary implant as amyopathic dermatomyositis (ADM). One of 4 deaths was caused by breast cancer metastasis and another by aspiration pneumonia with pharyngeal muscle involvement; in the other two cases the cause was not related to DM.
Conclusions. The analysis of the observed cases enables us to understand data about this population composition (prevalence of ADM, prevalence of internal malignancy, negative results for anti-Jo1 antibodies), to include unusual skin aspects, and to detect unsuspected respiratory diseases requiring special studies for early diagnosis. The lack of previous original reported series of DM in our country is highlighted (Dermatol Argent 2009;15(1):27-36).

Key words: *dermatomyositis, polymiositis.*

Introduction

Dermatomyositis (**DM**) is included in idiopathic inflammatory myopathies, together with **polymyositis** (**PMs**),[1-6] although exclusively cutaneous clinical forms without muscular involvement are recognized for a variable period of time, and even definitely **amyopathic** (**ADM**) forms.[7-15]
It is characterized by the involvement of upper limb proximal striated muscle and neck flexor muscles, with or without dysphagia, and involvement of respiratory muscles. The association with neoplasias and involvement of internal organs arouse interest in clinical physicians and dermatologists; however, the latter rarely see any form of PM.

Bohan and Sontheimer classifications

DM diagnosis criteria in Bohan's classification (**Table 1**) do not include patient subgroups with exclusively or predominantly cutaneous lesions;[8,13] therefore, a broader pathological category has been suggested, designated as "idiopathic inflammatory dermatomyopathies"[14,15] (**Table 2**).
According to Sontheimer, the **adult-onset DM** forms encompass:

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1. Named Professor UBA, and Head of Dermatology Department.
2. Medical Dermatologist of Collagenopathy Sector.
3. Medical Resident, 2nd year, Specialist Course UBA.
4. Attending Physician, 2nd year, Specialist Course UBA.
5. Medical Course Physician, 1st year, Specialist Course UBA.
6. Staff Physician, in charge of Pathology Sector.
7. Staff Physician, Histopathology.

Dermatology Department. Hospital "J. M. Ramos Mejía", Associated to Universidad de Buenos Aires. Autonomous City of Buenos Aires. Argentine Republic.

Correspondence
Mercedes Lidia Hassan. Arcos 2273 1º B, (1428) Autonomous City of Buenos Aires, Argentine Republic. | E-mail: mercedeshassan@yahoo.com.ar

ABBREVIATIONS

LAA: leukocytoclastic allergic angiitis.
ACL: anticardiolipins.
CPK: creatine phosphokinase.
DM: dermatomyositis.
ADM: amyopathic dermatomyositis.
EMG: electromyogram.
FAN: antinuclear antibodies.
TNF: tumor necrosis factor.
IFN: interferon.
ILD: interstitial lung disease.
LDH: lactic dehydrogenase.
MTS: metastasis.
PM: polymyositis.
NMR: nuclear magnetic resonance.
Rx: X-ray.
CAT: computed axial tomography.
HRCT: high resolution computed tomography.
GOT: glutamic-oxalacetic transaminase.

TABLE 1. BOHAN'S CLASSIFICATION OF POLYMYOSITIS/DERMATOMYOSITIS.4

• **Clinical subgroups**
◦ Polymyositis.
◦ Dermatomyositis.
◦ Polymyositis or dermatomyositis associated with malignancy.
◦ Childhood dermatomyositis.
◦ Polymyositis or dermatomyositis with an associated connective tissue disorder.
• **Diagnostic criteria**[5]
◦ Typical skin rash.
◦ Symmetrical proximal muscle weakness with or without dysphagia or respiratory involvement.
◦ EMG compatible with primary muscular involvement.
◦ Alterations in muscle biopsy compatible with primary muscular involvement.
◦ Elevation of muscle enzimes (creatin phosphokinase [CPK], glutamic oxalacetic transaminase [GOT], lactic dehydrogenase [LDH], aldolase).
Confidence limits for diagnosis of dermatomyositis:
Definite DM: rash and three of the four other diagnostic criteria.
Probable DM: rash and two of the four other diagnostic criteria.
Possible DM: rash and one of the four other diagnostic criteria.

Classic DM. It shows the two cutaneous and muscular components simultaneously or with a lapse not exceeding 6 months between them. Three presentations are included: exclusive, as part of a collagenopathy, or associated with internal malignancy.

Clínically amyopathic DM. It comprises amyopathic DM (synonym of *without myositis*): distinctive classic DM skin lesions confirmed by biopsy of 6 or more months evolution (*provisional*) to 2 years (*definite*) previous to muscular involvement; and hypomyopathic DM: specific cutaneous involvement, without clinical evidence of muscular involvement, but with some altered supplementary studies (enzymes, electromyogram [EMG] and/or muscle biopsy).

Childhood/juvenile DM forms include **classic DM** and **clinically amyopathic DM** with its two variants (amyopathic and hypomyopathic) defined similarly to the adult onset DM amyopathic and hypomyopathic forms.

TABLE 2. GLOBAL CLASSIFICATION OF IDIOPATHIC INFLAMMATORY DERMATOMYOPATHIES (SONTHEIMER 2002).1

1. Dermatomyositis (DM)		
• ***Adult onset DM:***		
	Classic DM:	exclusive.
		as part of an overlap connective tissue disorder
		associated with internal cancer.
	Clinically amyopathic DM:	amyopathic (provisional or definitive).
		hypomyopathic.
• ***Juvenile onset DM:***		
	Classic DM.	
	Clinically amyopathic DM:	amyopathic.
		hypomyopathic.
2. Polymyositis (PM):		exclusive.
		as part of an overlap connective tissue disorder.
		associated with internal cancer.
3. Inclusion bodies myositis		
4. Other clinical-pathological subgroups of myositis: focal myositis, proliferative myositis, orbital myositis, eosinophilic myositis, and granulomatous myositis.		

Incidence

Incidence in this country is unknown. In the United States, the annual incidence estimation is 1/100,000 inhabitants;[6] for childhood/juvenile DM, 0.06 to 0.32/100,000.[16] According to the Joint Committee of the American Association of Dermatology, in 1998 the annual incidence was 5 cases per million inhabitants.[17]

Etiology

Family-genetic risk of suffering idiopathic inflammatory myopathies (adult and juvenile DM) related to homocygocity for HLADQA1, nucleotide polymorphism in tumor necrosis factor (TNF) promoter (TNF-α 308-A) was found associated with chronicity, calcinosis, and high titers of TNF-α in classic juvenile DM in Caucasian population.[1]Association of DM with lung disease is known, more frequent in Japanese, and occasional genetic deficit of C5 has been reported in DM. Etiology also involves virus (parvovirus B19, Epstein Barr), drugs (such as cholesterol-lowering agents), D-penicillamine, phenytoin, chloroquine, hydroxyurea, interferon (IFN), which may cause myopathy and/or compatible skin lesions.[18-21] Ultraviolet light has been accepted as a trigger, but no photoinduction of the lesion has been attained.

Antibody-associated subgroups

A HLA DR52-related group has been identified, with higher prevalence of Jo-1 antibodies (20-30 percent of PM/DM), characterized by lung fibrosis, arthritis, Raynaud's phenomenon, mechanic's hands, poor prognosis, higher

mortality, and poor response to treatment (*antisynthetase syndrome*); and another DR53-related group, Dw7 with Mi-2 antibodies, present in 15 percent of DM, with florid skin involvement and better response to treatment. PM/Scl present in 25 percent overlaps and in 8 to 12 percent of PM is associated with myopathy plus sclerodermia. Anti-SRP (signal recognition particle) present in 5 percent of PM relates to cardiac involvement without lung fibrosis, resistance to corticoid treatment, and severe myositis.[1,5] DR3, B8, and B14 have been related to juvenile DM; and B14 and B40, to overlap syndrome in adults. Recently, antibodies of 155 kDa (Se) and 140 kDa (US) have been identified in sera of amyopathic DM related to interstitial lung disease or paraneoplasia, respectively.[22]

TABLE 3. CLASSIC CUTANEOUS MANIFESTATIONS OF DM.[14]

Pathognomonic
1. Gottron's papules.
2. Gottron's sign: symmetric confluent macular violaceous erythema on the dorsal aspect of the interphalangeal or metacarpophalangeal joints, oleocranon, patellae and medial malleoli.
Characteristics
1. Heliotrope erithema.
2. Periungual telangiectasia.
3. Symmetric confluent macular violaceous erythema on the dorsal aspect of the hands, extensor aspect of arms and forearms, deltoid areas, posterior part of shoulders and neck (shawl sign), anterior neck and upper chest (V sign), central aspect of the face and the forehead, and scalp.
4. Mechanic's hand.
Compatible with DM
1. Vascular atrophic poikilodermia.
2. Cutaneous calcinosis.

Cutaneous manifestations

Table 3 describes the distinctive cutaneous manifestations of DM.[14]

Signs of poor prognosis[3,9,11,23-25] (acute, progressive, and severe forms) are: early and rapid respiratory, pharingeal, or esophagic involvement, resistance to early corticoid treatment, blisters or necrotic skin lesions in trunk, systemic vasculitis, high erythrosedimentation rate (more than 35 mm in the 1st hour), and old age. **Mortality** varies about 21 percent in adults and 3-31 percent in children, according to diverse case-control studies and authors.[14] **Causes of death** are respiratory failure and those related to associated malignancies in adults (metastasis).[1,3,9,11,25,26] **Incidence of internal malignancies** was found in various case-control studies (between 11 and 43 percent).[34] **Relative risk** for internal malignancy ranges between 1.7 and 7.7, and is maximal in the term of one year.[10,25,26-34]

Material and methods

A retrospective observational analysis was carried out in 40 patients (7 males and 33 females) studied in the 1991-2007 period in the Dermatology Department of Hospital "J. M. Ramos Mejía" of Buenos Aires, with confirmed diagnosis of DM. Nine of them were amyopathic (ADM), and 4 were childhood or juvenile cases. Excluded from computation were the cases of difficult differential diagnosis: a mycosis fungoides (photosensitive

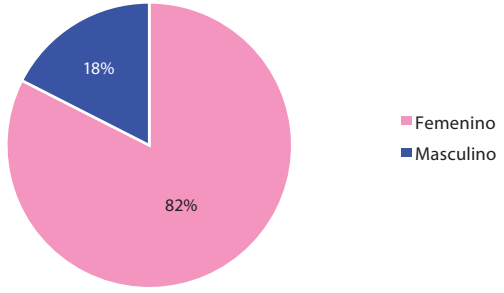

Chart 1. *Dermatomyositis per gender.*

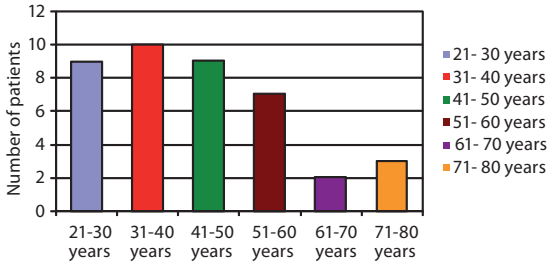

Chart 2. *Dermatomyositis per age at diagnosis.*

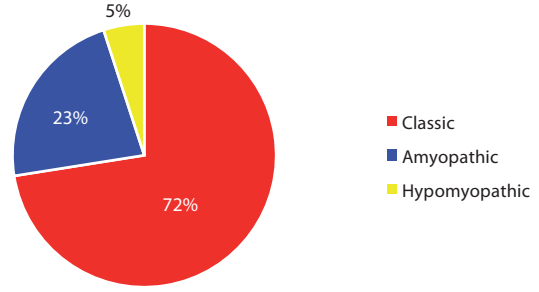

Chart 3. *Dermatomyositis per clinical form.*

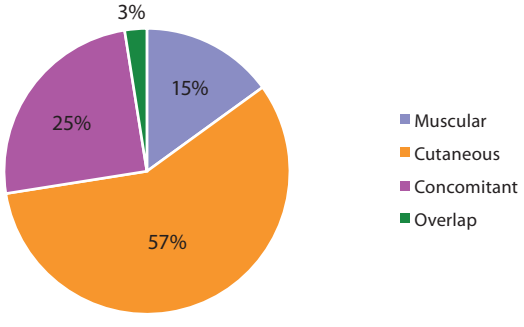

Chart 4. *Dermatomyositis per onset form.*

lesions, facial erythema with eyelid edema, poikiloderma, high reumatoid factor, arthralgia, and Raynaud's phenomenon, together with neurological disease); and 3 female patients in whom the study was not completed. Also not included was a case associated with simvastatin intake, which generated uncertainty about the role of this drug in the appearance of manifestations.

Results

Clincal features are summarized in **Table 4,** and in **Charts 1, 2, 3,** and **4**. Thirty three females and 7 males were studied (F:M ratio = 4.71:1). Four of them had diagnosis of *childhood/juvenile DM*, with age of onset 5, 12, 14, and 15 years, respectively; two were females and two were males. The *amyopathic juvenile forms*, like *calcinosis*, were found in three cases, two females.
As a whole, 9 (23 percent) had diagnosis of ADM, and 31 were classic forms. The period of time lapsing from the cutaneous manifestations to the onset of muscular manifestations lasted up to 11 years; the initial diganosis of these patients was lupus erythematosus (seen at our Department by another observator). In addition to major and minor criteria,[14] in our case-control study we found: ***rare lesions:*** blister lesions (in 2 patients), follicular lesions (2), extended diffuse alopecia, with whitish scales on scalp (1), gingival and lip telangiectasia (1), "milliary" calcinosis (1), "punch-biopsy scar"-like lesions (3), cutaneous vasculitis (2) (**Figures 1, 2, 3, 4, 5,** and **6**).

Complications not previously reported in this pathology: non-tropical pyomyositis (1) (**Figures 7** and **8**), adult tinea (1), and episodic abdominal distension related to peritoneal adherences evidenced by abdominal computed axial tomography (CAT) (1) (**Figure 9**).
No case was found of antisynthetase syndrome (association of Jo-1 antibodies, lung fibrosis o interstitial pneumonitis, Raynaud's phenomenon, and mechanic's hands), or flagellated lesions. Lung involvement was found in two patients with Jo-1, without mechanic's hands in an overlap and a classic DM.
Association with internal malignancy was observed in 16.6 percent (6 of 36), excluding childhood/juvenile cases.
Treatment was carried out based on oral systemic corticosteroids, mostly meprednisone 1-2 mg/kg/day for months, with progressive variable dose reductions to complete 2 years. Not all patients completed this regimen, or complied with it regularly. Occasionally, in special cases due to resistance, irregular intake of corticoids, or concomitant contraindicated conditions, oral methotrexate 7.5 to 12.5 mg/week was used, or low dose intravenous gammaglobulin (0.5 g per kg/day) for two days once a month, with good initial response, but they were discontinued on cost grounds. Mycophenolate mofetil was use in refractary myositis, 2 g/day, starting with 0.5 g/day. Thalidomide, 200 mg/day, and chloroquine phosphate, 200 mg/day, were used in other patients.

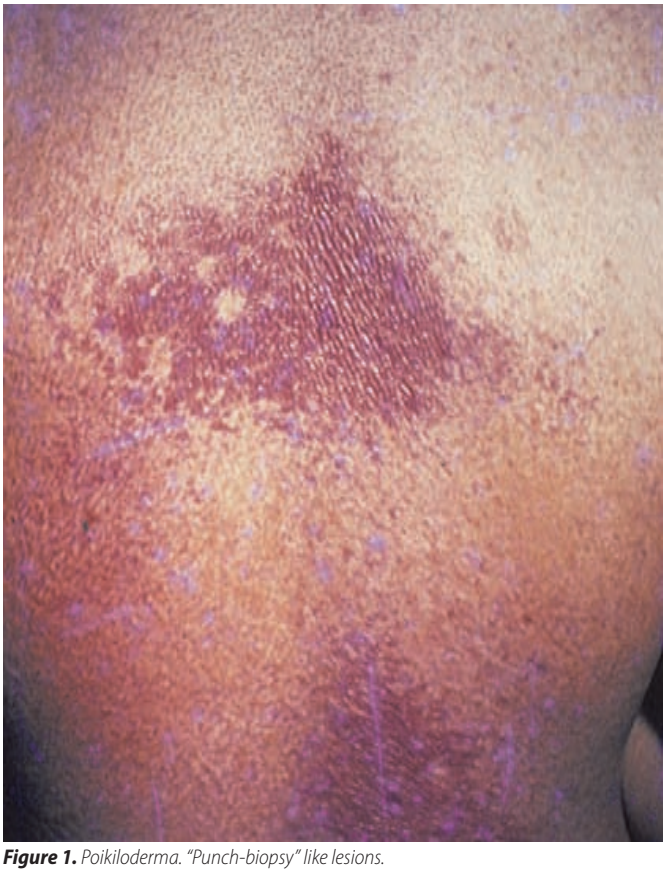

Figure 1. *Poikiloderma. "Punch-biopsy" like lesions.*

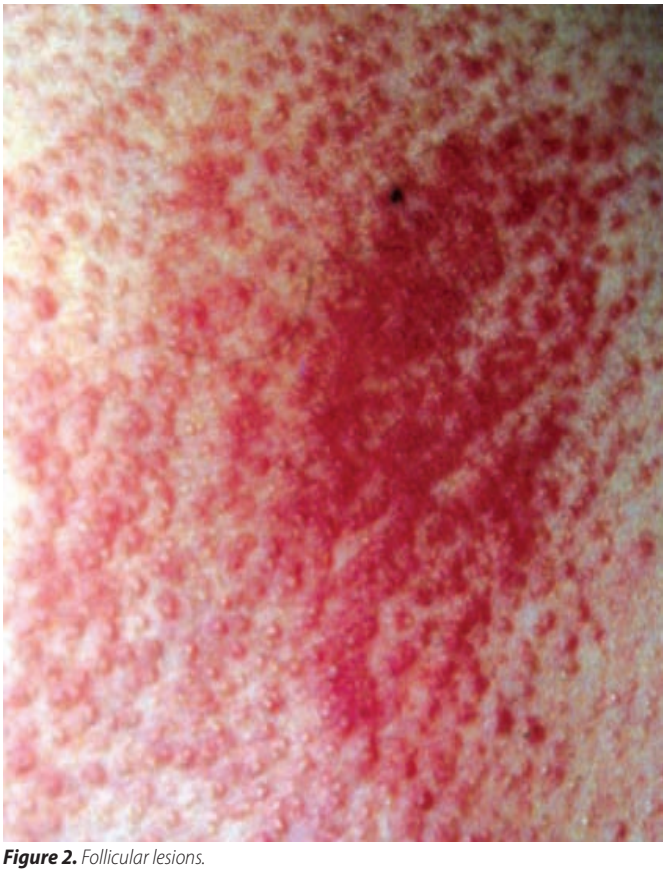

Figure 2. *Follicular lesions.*

TABLE 4. CLINICAL CHARACTERISTICS AND EVOLUTION OF PATIENTS WITH DM DIAGNOSIS IN HOSPITAL "RAMOS MEJÍA". PERIOD 1991-2007.

Pat	Age	Sex	Clinical form	Onset	Antibodies	Neoplasia	Systemic condition or other feature	Evolution
1	50	F	Classic	Muscular	-	Cervix	Loss of weight LAA	Resp. failure
2	60	F	Classic	Cutaneous	-	-	Loss of weight	-
3	57	F	Classic	Concomitant	-	-	-	-
4	37	F	Classic	Cutaneous	-	-	-	Resp. failure
5	35	F	Amyopathic	Cutaneous	ACL	-	Follicular lesions	Remission
6	58	F	Classic	Cutaneous	-	Breast	LAA	-
7	43	F	Classic	Concomitant	-	-	-	-
8	43	F	Classic	Cutaneous	-	-	Blisters	-
9	48	F	Classic	Cutaneous	-	Breast	-	Bone MTS
10	50	M	Classic	Cutaneous	-	-	-	Remission
11	68	F	Classic	Muscular	-	-	-	Resp. failure
12	54	F	Amyopathic	Cutaneous	-	-	-	-
13	12	F	Classic	Muscular	-	-	Loss of weight	Remission
14	36	F	Classic	Muscular	-	-	-	-
15	14	F	Amyopathic	Cutaneous	-	-	Acne due to corticoids	Remission
16	64	F	Classic	Muscular	-	-	-	-
17	15	M	Amyopathic	Cutaneous	-	-	Calcinosis	Irreg. Tt.
18	54	F	Amyopathic	Cutaneous	-	Breast	-	-
19	32	F	Amyopathic	Cutaneous		-	Respiratory Post-silicone	Remisión.
20	56	F	Classic	Concomitant	-	-	Mild resp esoph	Remission
21	15	F	Amyopathic	Cutaneous	FAN+	-	Calcinosis+++	Improvement
22	5	M	Classic	Concomitant	-	-	Loss of weight	-
23	27	F	Classic	Cutaneous	-	-	Abdominal distension Steatosis Dry Sd Lipodystrophy	-
24	46	F	Classic	Cutaneous	FAN 1/2560 Mo	-	-	-
25	33	F	Classic	Concomitant	FAN 1/320 Mo	-	Blisters	Pyomyositis
26	28	M	Classic	Cutaneous	FAN 1/80	-	Dysphagia Follicular lesions	-
27	58	F	Classic	Cutaneous	-	Ovary	Dysglusia	Bone MTS
28	29	F	Classic	Concomitant	-	-	-	-
29	34	F	Classic	Concomitant	FAN 1/80 Mo	-	Tricophytic tinea	-
30	78	M	Amyopathic	Cutaneous	-	-	Fibromyalgia	-
31	27	F	Classic	Muscular	-	-	-	-
32	50	F	Classic	Cutaneous	FAN 1/1600 Mo	-	Erythroderma by chloroquine	-
33	50	F	Classic	Concomitant	FAN+	Cervix	Vasculitis ulcers	-
34	33	M	Classic	Concomitant	FAN 1/400 Mo	-	Lung fibrosis Pharingeal	-
35	75	F	Hypomyopathic	Cutaneous	FAN 1/160 Mo	-	-	Pneumonia.
36	40	F	Classic	Overlap	FAN 1/80 Ro+ La+	-	ILD Cutaneous vasculitis Livedo and acral ulcers	Norwegian scabies. Died due-to neumothorax
37	72	F	Hypomyopathic	Cutaneous	-	-	-	-
38	32	F	Classic	Concomitant	FAN 1/5120	-	ILD	-
39	40	F	Amyopathic	Cutaneous	-	-	-	-
40	50	M	Classic	Cutaneous	-	-	-	-

ACL: anticardiolipins. **LAA:** leukocytoclastic allergic angiitis. **FAN:** antinuclear antibodies. **Mo:** mottling pattern. **MTS:** metastasis. **ILD:** interstitial lung disease.

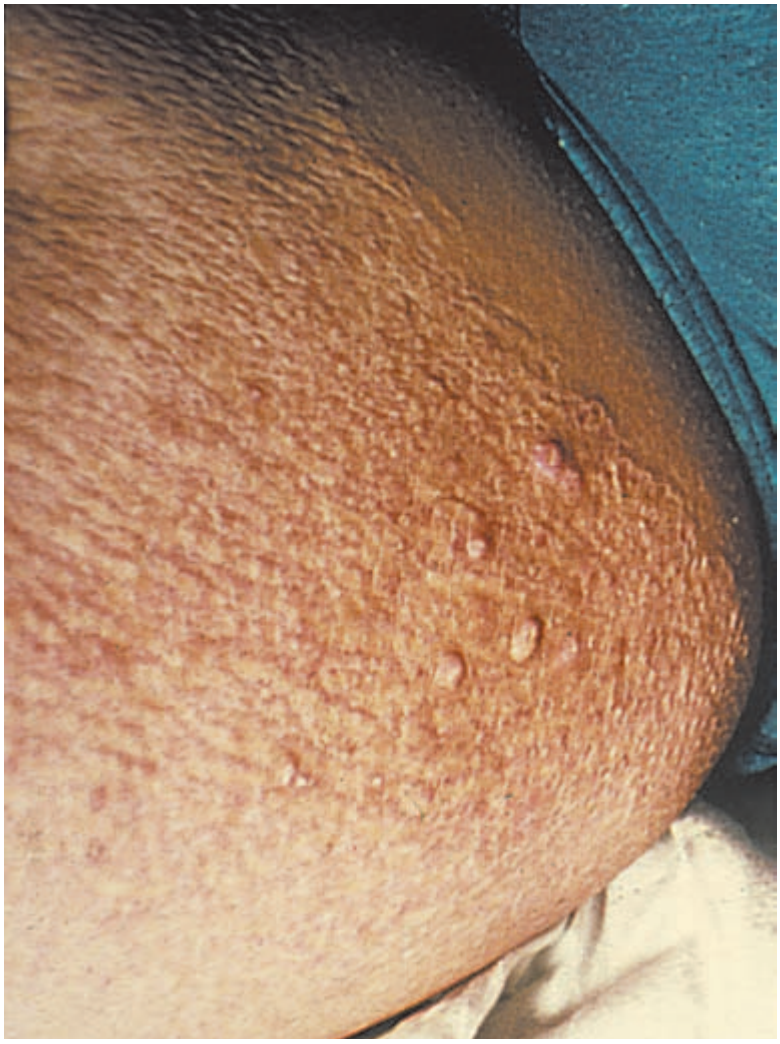

Figure 3. *"Miliary" calcinosis in juvenile DM.*

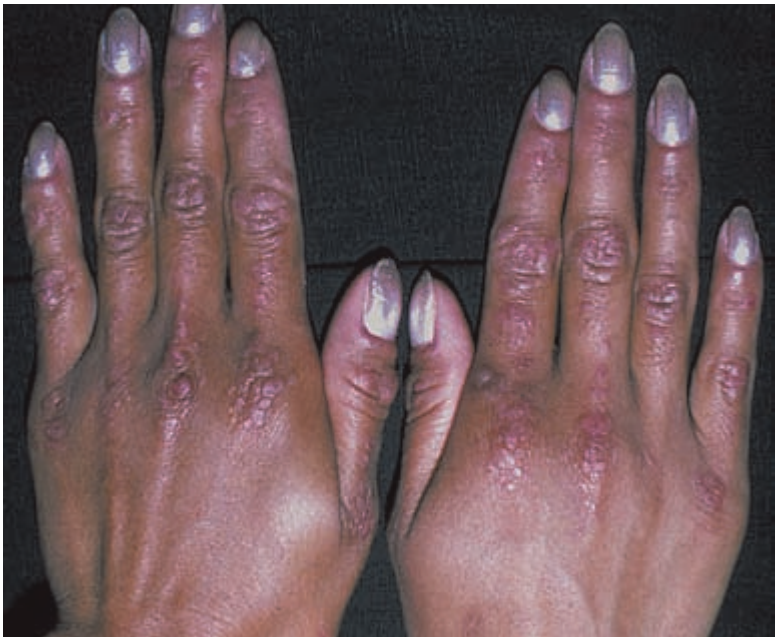

Figure 4. *Gottron's papules.*

No satisfactory results were obtained by corticotherapy or by hydroxychloroquine administration on skin lesions, and these remained in spite of the muscle involvement improvement. In facial lesions, 0.1 percent topical cream tacrolimus, or 1 percent pimecrolimus were indicated in two cases, with partial responses.

In one case of juvenile DM with calcinosis it was administrated disodium etidronate, 20 mg/day, with very good motor response and X-ray control after three months treatment. Currently, we indicate diltiazem, 10 mg/day, or alendronate.

Patient management was supplemented by kinesiotherapy and muscular rehabilitation, indispensable against contractures and permanent disability, especially in the childhood/juvenile form and to relieve dysphagia.

Causes of death were: respiratory failure due to aspiration pneumonia with pharyngeal muscle involvement (2); metastasis of associated malignancy (1); and two deaths of a cause not related to DM: pancreatitis (1), and traumatic pneumothorax (1).

COMMENT

Dermatomyositis appears with specific skin lesions, taken as diagnostic criteria; in addition, less frequent lesions related to this condition exist.[1,9] These encompass: *follicular lesions* (**Figure 2**), found in two cases located on presternal area and back, having mucin deposits, perifollicular mononuclear infiltrate, follicular hyperkeratosis, and involvement of the arrector muscle of hair; *diffuse alopecia with squamous lesions*,[16,36] which had to be differenciated from adult tinea, psoriasis, anticonvulsant pharmacodermia, and Norwegian scabies. Therefore, direct mycological examination and culture of these lesions are recommended. They have also been referred to as psoriasiform scalp lesions in juvenile DM,[16] but were not exclusive to that age range in our patients. The "*punch-biopsy scar*" appearance is noteworthy in the photosensitive areas of poikilodermia, but is rare, and shows nonspecific histopathology. *Calcinosis* in one case adopted an uncommon "miliary" appearance in the inner aspect of the thighs in a juvenile ADM,[16] and was not disabling. In two patients coexisted morphea-like lesions with compatible histopathology in the root of upper limbs. *Lip and gingival telangiectasia* were found in the physical examination of only one patient; they have recently been described in this disease, and the significance is yet unknown. *Cutaneous vasculitis* (**Figures 5** and **6**) was found

in two cases: one with white atrophy-like lesions in the root of upper limbs, following a linear arrangement with round ulcerations. No anti-phospholipid antibodies were demonstrated. Histopathology showed leukocytoclastic allergic angiitis, which healed with general treatment. Adjacent sclerodermal lesions (clinical and histopathology) with ivory shine and hyperpigmentation were seen. The other case was an overlap DM with petechial fingertip lesions and moderate acral livedo reticularis and ulcerations on the legs; this myositis was refractary to several immunosuppressor treatments and responded to mycophenolate mofetil. The lung instertitial condition remained unaltered, and the patient died of a non-related cause.

Complications observed were the presence of adult tinea caused by *Trichophyton mentagrophytes*,[37,38] with relapses and extension to hairless skin after treating with systemic antifungals, and Norwegian scabies. The first rare condition may be attributed to: (1) immunosuppression inherent to the corticoid treatment, or the disease, (2) a race or cosntitution predisposition (it was recently related to Afro-American women), or (3) insufficient antifungal treatment.

Two unusual conditions posed diagnostic difficulties: (a) episodic abdominal distension caused by peritoneal adherences seen in abdominal CAT, attributed to a sequela of probable prior mesenteric vasculitis in a young 29-year-old female patient with noticeable facial lipoatrophy, and (b) non-tropical pyomyositis.[39,40]

Pyomyositis is a muscular staphylococcal abscess, which may be diagnosed by puncture and culture, biopsy, ultrasonography and/or muscular NMR. It appears in tropical countries favored by parasitosis (filaria), malnutricion, and local trauma. It shows three stages: (1) initial, with mild fever, slight erythema, and local pain, (2) purulent, with fever, erythema, edema and fluctuation, and (3) septic, with hectic fever and poor general condition. Treatment requires systemic antibiotics and surgical drainage of the lesion.

It may pass unnoticed, and myalgia and muscular weakness may be attributed to DM.

One case treated with statins showed association of DM with a fibromyalgia-like picture,[41] with localizad muscular pain on palpation on upper limbs and trunk, without weakness or alteration of muscle enzimes, EMG, or NMR in a DM construed as amyopathic, with normal erythrosedimentation rate.

Not observed were: flagellated dermatitis or antisynthetase syndrome.[42]

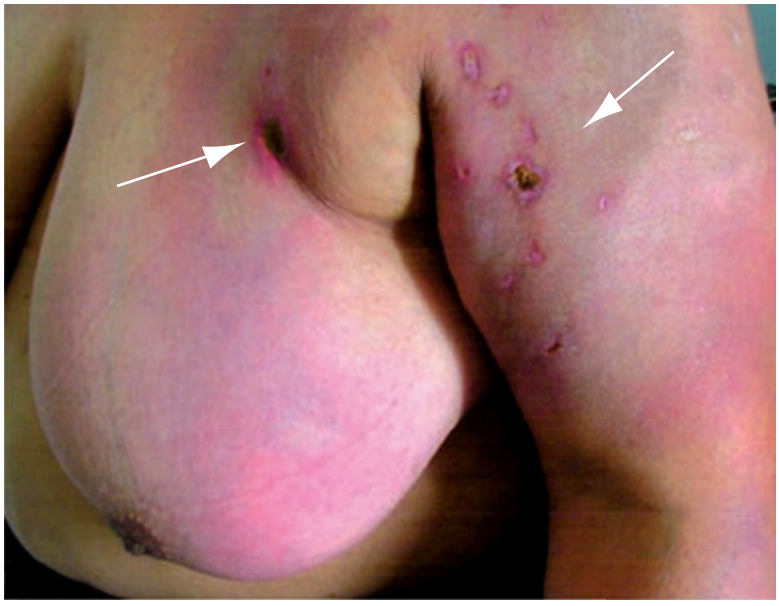

Figure 5. *Cutaneous vasculitis with round ulcerations coexisting with localized sclerodermal lesions* ***(arrows)***.

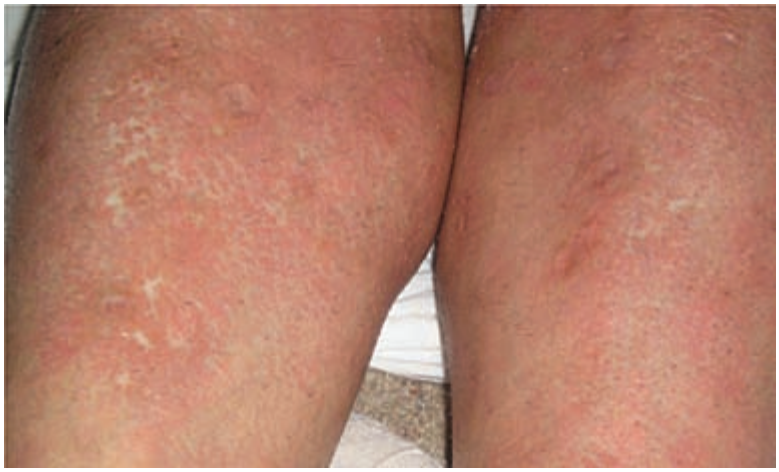

Figure 6. *Cutaneous vasculitis. White atrophy-like lesions on legs.*

Two classic DM cases were diagnosed as interstitial lung disease, with negative Jo-1. Diagnosis was reached by high resolution computed tomography (HRCT), respiratory function testing, and carbon monoxide diffusion, with normal chest X-ray. This indicates that the interstitial lung disease (ILD) must always be searched for by this method in DM/PM patients, even if no clinical manifestations appear, chest X-ray is normal, and anti-Jo-1 antibody is negative, and even in ADM, as mentioned above.[14,15,42-46]

Association with internal malignancy was observed in 16.6 percent of the cases (6/36), three breast cases, two cervix cases, and one ovary case. The periods of time between appearance of malignancy and disease onset were: one year and 4 months (in one case), two years (in two cases), and simultaneous (in two cases), in accordance with previous reports by other authors, based on Scandinavian and European case-control studies, where the higher relative risk (5.9 – 7.7) appears at one year, falls after the second year, but never becomes normal.

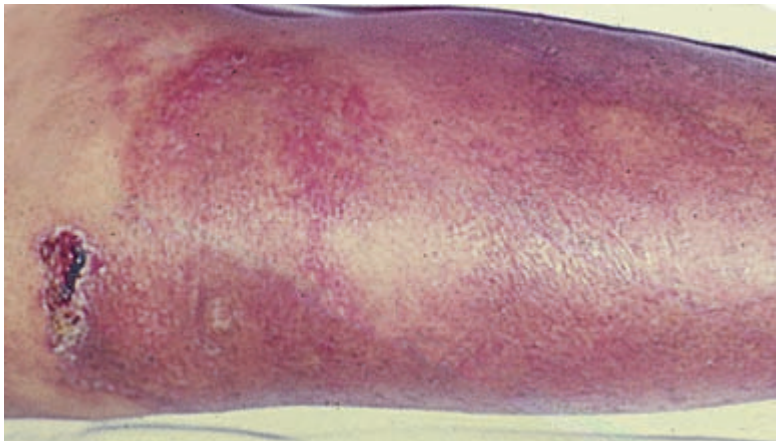

Figure 7. *Non-tropical pyomyositis. Erythema and blister on border of erythematous plate of leg overlying muscle abscess.*

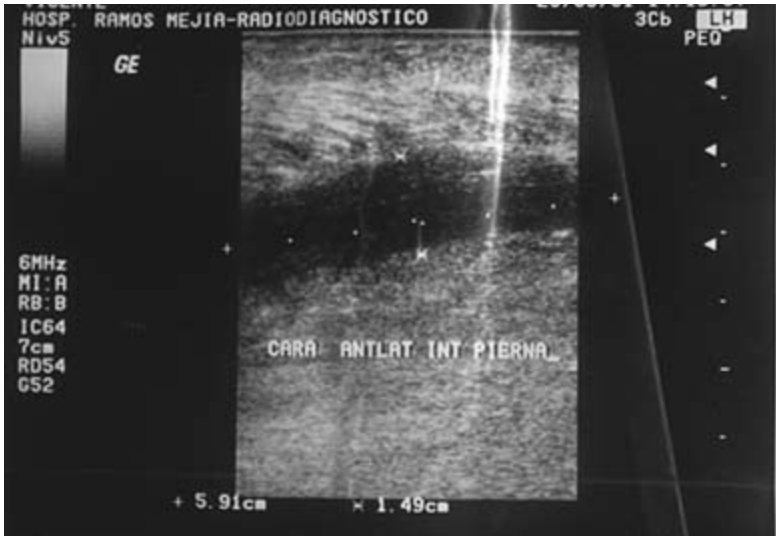

Figure 8. *Muscle ultrasonography. Notice hypoechoic image of intramuscular abscess.*

Therefore, an annual examination is recommended, and upon appearance of any new symptom, within the first three years[24,35] (**Table 5**).

Concomitant onset[34] occurred in an asymptomatic cervical "carcinoma in situ", found in a routine gynecological examination, which was requested at the dermatomyositis diagnosis; therefore, it may be inferred that the delay in diagnosis of a hidden malignancy may be responsible for the extended period lapsing between diagnosis of the disease and appearance of neoplasia.

Ovary cancer[26] was diagnosed upon bone metastasis in case 27. The finding of gynecological malignancy exclusively confirms that the type of malignancy related to the most frequent according to age, gender and race in each country or geographical region. Of the 6 malignancies, only one was associated to ADM.[33,35]

As regards treatment,[47,54] a noticeable response was detected in a refractary myositis of an overlap DM to mycophenolate mofetil after several immunosuppressor treatments (cyclophosphamide, corticoids, plasmapheresis, gammaglobulin IV, methotrexate). The lung condition remained unaltered, secondary effects were noticeable (gastric intolerance), and death occurred due to a non-related cause. One patient showed an extensive erythrodermal erythematous outbreak due to chloroquine, which subsided with medication discontinuation.

The cause of death was related to respiratory failure (by aspiration pneumonia with pharyngeal muscle involvement in 2 cases), and associated malignancy (recurrence with breast cancer metastasis in one case). Lung involvement was precocious, and has been pointed out as one of the signs of poor prognosis, when it is persistent and refractory to corticotherapy.[1,3,5,14]

CONCLUSIONS

1. In a population of 40 patients who were from 14 to 75 years old at the time of the initial visit between 1991 and 2007, a F:M ratio of 4.71:1 was found.
2. Uncommon skin lesions were observed: blisters (two patients), follicular (2), scaly scalp alopecia (2), miliary calcinosis (1), "punch-biopsy scar" lesions (3), gingival and lip telangiectasia (1), and cutaneous vasculitis (2). Complications during the course of this condition included: adult tinea capitis by *Trichophyton mentagrophytes*, Norwegian scabies, "non-tropical" pyomyositis, and intermittent abdominal distension probably related to intestinal adherences.
3. Two cutaneous vasculitis were found in the studied population with white atrophy-like lesions, and coexistence with adjacent ivory sclerodermal lesions.
4. ADM amounted to 23 percent (9/40) of the study population.
5. ADM did not appear any different from the classic form (DM). One of the 6 cases associated to malignancy appeared in this form, and the remaining 5 were identified among the 36 adult patients of the classic form. Total prevalence of malignancies was 16.66 percent in the adult study population.
6. All malignancies were gynecological, probably due to the female dominance, and since it is the prevailing malignancy in women in our population.
7. Both patients with interstitial lung disease were Jo-1 negative.
8. Although a pathogenic role of silicone implants in this disease has been denied,[6] one case had history of mamary silicone implant. It was an ADM, which evolved after several years of pulmonary fibrosis and remitted after corticotherapy.

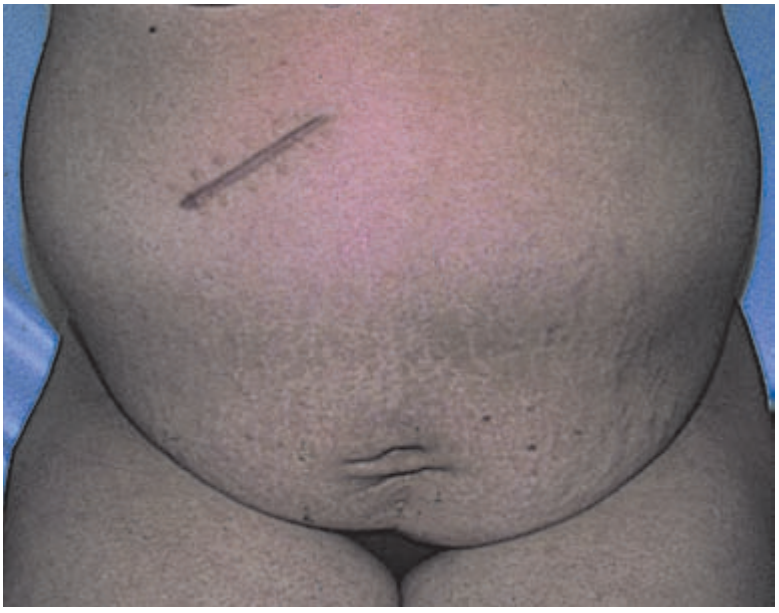

Figure 9. *Intermittent abdominal distension. CAT shows intestinal bridles.*

TABLE 5. DERMATOMYOSITIS. ASSOCIATION WITH MALIGNANCIES. PUBLISHED CASE-CONTROL STUDIES.

Reference quote	No. pats.	Period	RR o percent neoplasia	T. DM-Ca
Sweden 1992[28]	788	20 years	DM:2.4 (M) y 3.4 (F) PM:1.7 (M) y 1.8 (F)	-
Scotland 1996[27]	705	12 years	DM: 7.7; PM: 2.1	0-2 years
Sweden, Denmark, Finland 1997[29]	618 DM 914 PM	10 años	RR: 3 DM: 198 neoplasia PM: 137 neoplasia Ovary, lung, pancreas, stomach, colorectal, lymphoma	-
Singapore, 1997[30-31]	38 DM	6 years	31.6% (38% naso-pharingeal)	-
Denmark, 1997[32]	539	12 years	RR: 5.9. Lung, ovary, lymphoma	0-1 year
Firenze, 2002[10]	1 ADM	-	No malignancy	-
France and Switzerland 2002[34]	33 DM 7 PM	19 años	40%	0-1 año
Buenos Aires 2002 [*]	40	10 years	16.6%. Breast, ovary, cervix	0-2 years

Period: study duration. **RR o percent malignancy:** relative risk or malignancy prevalence.
T. DM-Ca: time between diagnosis of DM and malignancy. **M:** male. **F:** female.
[*]: our case-control study.

9. One patient (overlap syndrome) had, late in the evolution, significant positive titer of anticardiolipin antibodies, and suffered a transient ischemic stroke.
10. The absence of previous work based on own DM patient series in the local references is highlighted.

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