Joint Involvement in Polymyositis/Dermatomyositis

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Summary **The frequency and features of joint involvement were evaluated in 29 patients with polymyositis/dermatomyositis (PM/DM); eight of them (27.5%) presented arthritis. Mean age was 30 years, and mean disease duration was 5.3 years. Oligoarthritis was observed in 5 cases, while 3 presented polyarthritis; in most cases arthritis was concurrent with the musculocutaneous picture. On comparing the 8 arthritis patients with the remaining 21, no significant clinical or serological differences were found. However, males predominated in the arthritis group, 75% vs 19% ($p<0.05$). Arthritis responded favourably to underlying disease treatment and articular sequelae were not observed.**

Key words **Polymyositis, Arthritis, Dermatomyositis, Joint Involvement.**

INTRODUCTION

Polymyositis is an inflammatory muscle disease of unknown aetiology. In 1975 Bohan and Peter proposed their classification and diagnostic criteria in 5 groups (1). In 1979 Schumacher et al., reported 9 patients with polymyositis/dermatomyositis (PM/DM) and joint involvement, without stating its frequency (2). In fact, most of the series described to date have not given attention to the prevalence of joint involvement, nor adequately described their clinical features (3-6). Some classic rheumatology textbooks even contend that the presence of arthritis should suggest the existence of another connective tissue disease (7). In 1976 Bunch described a subluxing arthropathy and more recently, Oddis et al., found it associated to the presence of anti Jo-1 antibodies (8,9). The purpose of our work was to evaluate the frequency and features of joint involvement in PM/DM patients.

MATERIAL AND METHODS

Clinical histories of patients with a diagnosis of PM/DM followed up at our service from 1976 to 1990 were reviewed, and all patients with joint involvement examined. All cases met Bohan and Peter's criteria (1) for definite or probable disease and those without muscle biopsy had been followed up for at least 3 years to rule out alternative diagnoses. Patients were classified in 5 groups, in agreement with the classic scheme (1) and studied as regards age, sex, follow-up time, presenting form, clinical manifestations, laboratory findings, EMG, muscle biopsy, treatment and disease course.

Muscular strength was rated on a 0 to 5 scale, as modified by Gardner-Medwin and Walton (10). EMG was evaluated on the basis of low amplitude and short duration polyphasic potentials, high frequency bizarre discharge and fibrillations (1,11). Throughout, muscle biopsy was obtained by open technique contralateral to the muscle explored by EMG, then studied by light microscopy after staining with hematoxylin-eosin to search for necrosis, regeneration, fascicular and perifascicular infiltrates, vasculitis and atrophy (1,12,13). When required by the overall clinical picture, histochemical and electron microscopy studies were carried out. Laboratory tests specifically included determination of muscle enzyme levels (CPK, SGOT, Aldolase), ANA by immunofluorescence in rat liver, anti RNP antibodies by immunodiffusion and rheumatoid factor using the Singer and Plotz method. In addition, routine conventional tests were performed.

Patients in Group V (Overlap) had to meet ARA criteria for other connective tissue diseases (CTDs), or else present immunological findings and/or anomalies which, without fully complying with CTD criteria, justified their inclusion in this group. On performing the specific analysis of joint involvement, however, Group V cases were excluded from further study on the grounds that arthritis might have been due to other CTDs. Arthritis defined as joint pain and swelling was considered present only on the physician's observation, so that the presence of arthralgias or inflammatory arthritis re-

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Table I: *Clinical features of 43 cases of polymyositis/dermatomyositis. Classified according to Bohan and Peter (1)*

	PM Group I	DM Group II	PM/DM Group III	PM/DM Group IV	PM/DM Group V	Total
Patients (No.)	15	8	2	4	14	43
%	(34.9)	(18.6)	(4.6)	(9.3)	(32.5)	(100)
Female Sex (No.)	9	7	2	1	13	32
(%)	(60.0)	(87.5)	(100)	(25.0)	(92.8)	(74.4)
Mean age in years	39.7	37.1	61.0	8.2	30.7	33
(range)	(20-69)	(17-62)		(5-14)	(15-47)	(5-69)
Muscular weakness %	100	100	100	100	92.7	97.6
Cutaneous rash %	0	100	100	75	35.7	41.8
Dysphagia %	46.6	75	50	25	64.2	55.8
Lung involvement (%)	26.6	25	0	0	21.4	20.9
Heart involvement %	13.3	12.5	50	0	7.9	11.6
Raynaud's phenomenon %	26.6	12.5	0	25	78.5	39.5
Arthritis %	26.6	37.5	0	25	78.5	44

ferred by the patient were not taken into account. All patients underwent chest X-ray, pulmonary function tests (PFT) and EKG. Pulmonary involvement was considered present with the finding of dyspnoea, interstitial lung disease or restrictive pulmonary pattern on PFT. Cardiac involvement was considered when arrhythmia or myocardial failure were present. Statistical analysis was carried out using the chi square test and Student's t-test.

RESULTS

Of 53 patients evaluated, 43 met inclusion requirements. Table I lists general clinical features of our population.

Patients in Group V had other associated CTDs, of which 6 were progressive systemic sclerosis (PSS), 3 rheumatoid arthritis (AR), 4 systemic lupus erythematosus (SLE), 2 mixed connective tissue disease (MCTD) and one Sjögren's syndrome.

Thirty-four cases (79%) met criteria for definite disease and 9 (21%) those for probable disease. Muscular weakness was observed in 97.6%, raised enzyme levels in 95.2%, EMG alterations in 95.1% and characteristic muscle biopsy findings in 81.5%. A specific analysis was then performed with data from patients having joint involvement at some time during the course of disease.

Arthritis was present in 4 cases in Group I (26.6%), 3 in Group II (37,5%), one in Group IV (25%) and 11 in Group V (78.5%). None of the patients in Group III presented arthritis. Due to the fact that joint involvement in Group V cases is most likely attributable to associated CTD's, pertinent data were disregarded for statistical purposes in order to achieve greater specificity in the evaluation of such compromise in pure PM/DM cases. Of the remaining 29 patients in Groups I to IV, 8 (27.5%) presented arthritis. Mean age was 30 years (range 7-50) and 6 patients were males (75%). Four cases presented typical DM rash, one of them with juvenile onset, while the remainder belonged to Group I. All of them had proximal muscle weakness, elevated muscular enzymes, typical EMG and muscle biopsy findings, thus meeting criteria for definite disease. Mean follow-up time in this subgroup was 5.3 years (range 2-8). Five patients presented oligoarthritis and 3 polyarthritis. The most commonly affected joints were knees and PIP in 5 cases, MCP in 2 and wrists and elbows in one. Arthritis was associated with muscle involvement in 7 patients. In only one patient, arthritis preceded the remaining symptomatology by 3 months.

None of the patients were taking steroids at the time they developed the articular picture. Arthritis was mild throughout and readily controlled by underlying disease treatment, without requiring specific therapy.

Three patients with joint involvement presented recurrent muscle weakness during the course of the disease, but it should be stressed that arthritis failed to reappear in any of them. In a single case, myositis recurrence was preceded by polyarthralgias by a few days. Two patients presented lung involvement, manifested by dyspnoea and severe restrictive compromise at functional respiratory examination. Radiological lung alterations were absent. The rheumatoid factor test proved transiently positive in one patient, with a 1/640 titer, but several determinations later on were negative. In this patient the duration of arthritis was less than one month and never showed recurrences nor evidence of X-ray erosions during a 6-year follow-up. Positive ANA were observed in 2 cases: in one with a 1/500 titer, and speckled pattern, and in the other with 1/100 and ho-

Table II: *Features of joint involvement in 8 patients with polymyositis/dermatomyositis (PM/DM)*

Case No.	Age (years)	Sex	Diagnosis	Follow-up (years)	Involved joints	RF*	ANA*	RNP*	X-rays
1	49	M	PM	6	3rd-4th right PIP 3rd right DIP	1/640	(-)	(-)	Soft tissue swelling
2	30	M	PM	2	Knees	(-)	(-)	(-)	Normal
3	25	M	PM	8	Left knee	(-)	(-)	(-)	Normal
4	24	M	PM	7	MCPs-PIPs Right elbow	(-)	1/500 Speckled	(-)	Soft tissue swelling
5	32	M	DM	4	Knees-ankles PIPs	(-)	(-)	(-)	Not available
6	23	F	DM	5	PIPs-MCPs wrists- knees	(-)	1/100 Homogeneous	(-)	Soft tissue swelling
7	50	F	DM	5	3rd-4th right PIPs-3rd left PIP	(-)	(-)	ND	Not available
8	7	M	Juvenile DM	5	Knees	(-)	(-)	(-)	Normal

* RF: rheumatoid factor; *ANA: Antinuclear antibodies; *RNP: Antiribonucleoprotein; ND: not done.

Table III: *Joint involvement in polymyositis/dermatomyositis (PM/DM) Comparison of patients with and without arthritis*

	With arthritis	Without arthritis
No. of patients	8	21
Mean age (years)	30	36.8
Male sex (%)	75*	19
Dysphagia (%)	37.5	47.6
Raynaud's phenomenon	25	19
Lung involvement (%)	25	19
ESR > 30 mm/h (%)	87.5	76
ANA postive (%)	25	14.2
RF positive (%)	12.5	4.8
Citotoxic drug treatment (%)	25	14.2

*: $p < 0.05$

mogeneous pattern. In these two patients arthritis duration was also shorter than a month and none of them showed clinical or serological evidence of other CTDs during five and seven years follow-up, respectively. No anti RNP antibodies were observed in any of the 7 patients studied. Increased erythrosedimentation (ERS) rate was found in 7 patients; all patients with arthritis presented raised muscular enzyme levels. Remaining laboratory tests failed to show significant alterations. In one case a knee arthrocentesis was carried out rendering 15 cc of amber synovial fluid, with preserved viscosity. White blood cell count was 1500 cells/mm^3, mainly lymphocytes. A search for crystals with polarized light proved negative.

Radiological studies were available in 6 patients, which were normal in 3 and presented soft tissue swelling in hand X-rays in the remainder. Erosive changes and/or periarticular calcifications were lacking throughout.

The main features of patients with joint involvement are listed in Table II. A comparative analysis was then made of patients with or without arthritis in Groups I to IV (Table III): 75% of arthritis cases were males vs 19% in the arthritis-free group ($p<0.05$). The remainder of studied variables failed to show significant differences.

DISCUSSION

Arthritis is a poorly recognized manifestation during the course of PM/DM not associated to other CTDs. Some series prior to the subdivision into the 5 classic groups report the presence of arthritis in 25-50% of cases (3,14,15); however, most of such patients would nowadays be included in Group V and the arthritis attributed to another CTD. In 1977, Bohan et al., reviewed 153 patients with PM/DM and described the presence of arthralgias in 25% and 20% of Groups I and II respectively, emphasizing the absence of overt arthritis (16). Kagen expressly states that overt synovitis is sufficiently rare, that its presence suggests other connective tissue syndromes (7). In 1979, Schumacher et al. reported 9 patients with PM/DM and joint involvement describing clinical, radiological and anatomopathological features but without mentioning frequency (2). In 1976 Bunch and more recently Oddis et al., describe a subset of PM/DM patients with a subluxing arthropathy associated with interstitial lung disease and anti Jo-1 antibodies (8,9). Love et al., found arthritis in 47% of patients with idiopathic inflamma-

tory myositis, usually associated with the presence of anti-synthetase antibodies (17).

In our series of 43 patients with PM/DM, 14 belonging to Group V were disregarded for analysis of joint involvement. Of the 29 remaining cases, 8 (27.5%) presented arthritis concurrently with the onset of the muscular picture; throughout, we were extremely cautious in differentiating frank arthritis as opposed to arthralgias. Except for articular compromise, the general features of our population are not dissimilar to those reported for other series (5,6,14,16).

No correlation was observed between the presence of arthritis and lung compromise, which was a constant finding in the description of Schumacher et al. (2). Neither did the progress of the clinical picture, the number of recurrences nor the need for specific drug therapy differ for the 2 groups. Although a greater frequency of males was observed in the arthritic group, no explanation could be found for this predominance. None of the other variables compared allowed the patients with arthritis to be included in a different PM/DM subset. The presence of joint involvement in the course of an inflammatory myopathy could have distinct interpretations. Firstly, it was regarded as an overlap syndrome, in whose development the most commonly observed diseases are PSS and SLE. In both conditions, arthritis may be found in 60-90% of cases according to the various series (18,21), so it would be quite tempting to attribute its presence to the symptomatic concomitance of these two entities. Secondly, a distinct clinical entity termed rheumatoid myositis has been described (22), which features muscular necrosis showing electromyographic and anatomopathological expression in the course of classic RA. It is often accompanied by other systemic manifestations, a disproportionate increase in ESR with respect to the degree of synovitis and high CPK levels. Thirdly, in patients receiving high steroid doses, a common condition in PM/DM cases, joint effusion has been observed, mostly in the knees, as a result of changes in synovial vasculature. Such fluid is colorless and displays a cell count lower than 500/mm^3 (23,24). Finally, the arthritis could be considered as another feature of PM/DM by itself. For reasons already stated, none of our patients may be included in any of the first 3 situations.

Although the presence of arthritis in the course of classic PM is hardly a relevant finding, its frequency and features are indeed noteworthy. It is quite likely that its mild and transient character superimposed on a clinical picture involving the patient's overall status leads to its being grossly underdiagnosed.

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