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Features of polymyositis and dermatomyositis in the elderly: A case-control study

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Polymyositis, dermatomyositis, elderly.

ABSTRACT

Objective

Polymyositis (PM) and dermatomyositis (DM) are uncommon idiopathic inflammatory myopathies (IIM). Little is known about these diseases in the elderly. We attempted to define the characteristics of PM/DM in the elderly by a case-control study involving the retrospective review of medical files of PM/DM patients.

Methods

We drew from among 200 PM/DM patients being followed in our Internal Medicine Department 21 patients (14F / 7M), aged ≥ 65 years at the onset of myositis (17 PM / 4 DM) (mean: 69.9 ± 4.8 yrs.). They were compared with 21 (15F / 6M) randomly selected younger patients with IIM: PM (14) and DM (7) (mean: 46.4 ± 12.4 yrs). Clinical, biological, electrophysiological and pathologic features, treatment regimens and side-effects in the 2 groups were collected.

Results

Clinical features were similar for the 2 groups. Elderly patients tended to have a higher frequency of cancer (24% vs 9.5%, p = 0.06), particularly of rectal adenocarcinoma. The time from disease onset to diagnosis was significantly longer in older patients (26 ± 37 months vs 9 ± 15 months; p = 0.02), normal CK levels were more frequent (40% vs 5%; p = 0.02) and serum CK levels were lower than for the population as the whole (11.5 N vs 22 N, p < 0.03). Electromyography features were more frequently suggestive of a chronic form of the disease in elderly patients. Treatment regimens and short-term side-effects were similar for the 2 groups.

Conclusion

PM and DM are often diagnosed late in the elderly. Biological data and electromyography features argue for a chronic form of the disease in this age group. Clinical and endoscopic rectal examinations should be carried out in elderly patients with PM/DM.

Introduction

Polymyositis (PM) and dermatomyositis (DM) belong to the group of the idiopathic inflammatory myopathies (IIM), which includes sporadic inclusion body myositis (1, 2). There is a specific peak in the prevalence of DM in childhood (2) and a second peak for PM and DM prevalence in mid-life (2). PM/DM seems to be rare in the elderly. Descriptions and features of other autoimmune disorders in the elderly have occasionally been published (3-5). Particular characteristics of the elderly may account for differences in the diagnostic or therapeutic approach in PM/DM: distinctive clinical features (6), the high frequency of iatrogenic complications (7), and of associated conditions (8), especially cancer.

We investigated the characteristics of PM and DM in the elderly by means of a retrospective study in which we compared 21 patients aged 65 years or over at the onset of disease with 21 younger patients.

Patients and methods

Patients

We identified 200 patients discharged from the Internal Medicine Department between 1973 and 1996 with a diagnosis of PM or DM. Patients with inclusion body myositis were excluded. Two groups of patients were defined: Group E consisting of elderly patients (aged 65 years or over at onset) and a control Group Y of younger patients (minimum age 20 years). Medical files were reviewed and a total of 21 consecutive elderly patients were included in the study. A random sample of 21 subjects was selected from among the younger PM/DM patients seen during the study period (group Y). Patients were matched for year of first visit to the department, within 3 year-periods.

Methods

The following features were recorded for each patient.

Clinical features: Age at diagnosis, sex, ethnic group, personal and familial medical history, type of myositis, diagnostic delay (interval between the onset of symptoms and diagnosis), associated autoimmune disease or malignant neoplasm, type of muscular or general symptoms, follow-up duration, associated diseases, diagnosis and therapeutic approach, extramuscular problems (cardiac, esophageal, lung, or renal involve-

ments, arthritis).

Biological features at diagnosis: Serum creatine kinase (CK) and aldolase levels [multiples of normal levels (x N)], ESR, C reactive protein levels (CRP), blood cell count, antinuclear (ANA) and myositis-specific autoantibody concentration.

Electromyographic features at diagnosis: These were of 3 types: no myopathic change, myopathic changes (high insertional activity and duration of motor unit potentials, early recruitment pattern), "acute" myopathic changes (small and fragmented motor unit potentials and fibrillations).

Pathological features of a muscle biopsy specimen: Infiltration of inflammatory mononuclear cells, regenerating muscle fibers, inflammatory lesions of vessels, necrosis. Electron microscopy was performed to exclude inclusion body myositis.

Therapeutic features (initial dose, duration, efficiency, and side-effects of treatments): For each drug, "response" was defined as a clinical improvement of 20% or greater from the initial power (gauged using the British Medical Research Council grading system) and a decrease in biological abnormalities (of more than 30% of initial CK values). "Failure" was defined as a lack of clinical response and/or the need to start another therapy.

Statistical analysis

Results are presented as percentages (%) for discrete variables. The statistical significance of differences was assessed by non-parametric statistical methods for small numbers and non-normal distributions, specifically Fisher's exact test for continuous variables and the Mann-Whitney test for discrete variables. In all tests, $p < 0.05$ was considered to be statistically significant. BMDP statistical software 1.1 (Los Angeles, CA) was used.

Results

Patient characteristics

Except for mean age, the patients characteristics were not significantly different. The mean age of the 21 patients (17 PM / 4 DM) of group E (14F / 7M) was 69.9 ± 4.8 years (range 65 - 80). The mean age of the 21 patients (14 PM / 7 DM) of group Y (15F / 6M) was 46.4 ± 12.4 years (range 23 - 64) ($p < 0.0001$ versus group E).

None of the elderly patients had an associated autoimmune disease, versus 4 of the younger patients (19%) (2 scleroderma, 2 Sjögren's syndrome) ($p = 0.059$). Five (24%) elderly patients had malignant neoplasm: rectum adenocarcinoma (3 patients), endometrial carcinoma (1) and chronic lymphocytic leukemia (1). Two (9.5%) young patients had malignant neoplasm: 1 ovarian cystadenocarcinoma and 1 lymphosarcoma ($p = 0.24$). Associated diseases (systemic hypertension, coronaropathy, hyperuricemia, dyslipidemia, diabetes mellitus, non-alcoholic cirrhosis) were significantly more frequent in group E ($p = 0.001$).

Characteristics of the myositis

The clinical features of the patients are listed in Table I. In elderly patients, myalgias were less frequent (47.5% versus 71.5%, $p > 0.05$), and the mean diagnostic delay was significantly longer (26.3 ± 37.6 months versus 3.9 ± 4 months; $p = 0.02$).

Serum CK levels were recorded in 20 (95%) of the medical files in group E and in 19 (90.5%) for group Y. CK was more frequently normal at diagnosis in the elderly than in group Y (40% versus 5%, $p = 0.02$). Among patients with high CK levels, the mean level was higher in group Y than in group E (22 ± 13.5 N vs 11.5 ± 12.5 N; $p < 0.03$). No significant difference was found in the aldolase level or other biological findings. Electromyographic (EMG) data were available for 17 (81%) patients in each group. "Acute" electromyographic criteria were significantly more frequent in group Y (76.5% versus 12%, $p < 0.001$). No significant difference in pathological features were seen between the 2 groups.

Treatment regimens were studied in 20 of the 21 patients in group E (Table II). The remaining patient recovered from paraneoplastic PM after surgical treatment alone. Glucocorticoid therapy was used for all patients in both groups. The duration of steroid therapy was the only characteristic of steroid treatment which differed between the two groups, i.e. it was longer in group Y ($p = 0.008$).

Side-effects were more frequent in group Y (66.5% versus 50%, $p = 0.35$). Sepsis was reported in 2 (10%) elderly patients and in 8 (38%) patients in group Y. Other corticosteroid complications occurred in 11 (55%) elderly patients and in 8 (38%) patients in the group Y. IVGG was stopped in one elderly patient for hypersensitivity. One elderly patient died from infectious pneumonia during MTX.

Elderly patients had a significant shorter mean follow-up (36 ± 31 vs 66 ± 44 months; $p = 0.03$). Death due to myositis or treatment occurred at statistically similar frequencies in the 2 groups (9.5% in group Y versus 19% in group E).

Discussion

Very few studies have been published about autoimmune diseases in the elderly (3-5). Our study shows that the clinical features of PM/DM in the elderly are similar to those in younger patients. We found a trend towards a higher frequency of malignant neoplasm in the elderly population (24% vs 9.5%, $p > 0.05$). Rectal adenocarcinoma accounted for 50% of cancers in group E. In the literature, colorectal cancer is an uncommon tumor associated with PM/DM (9, 10), but appears to be the second cause of death in these patients (9). Our results suggest that systematic clinical and endoscopic rectal examination of elderly patients with PM/DM would be of value.

The interval between the onset of symptoms and the diagnosis of PM/DM was significantly longer in the elderly. The higher frequency of normal serum CK levels and difficulties in analyzing EMG findings in the elderly may lengthen the delay of diagnosis. It may also be lengthened by the high frequency of comorbidity with possible muscular symptoms (endocrine or rheumatologic diseases, metabolic or iatrogenic disorders) in the elderly (7, 8).

Serum CK level was the only biological determination that differed between the two groups. The main reason for this is probably the decrease in muscle mass in aging. Studies have shown a decrease by one-third in muscle mass after the age of 50, an additional decrease of 15% between the ages of 70 and 80 years (11), and a steady reduction in the number of

Table I. Clinical features of the 42 patients in the two groups.

Clinical features	Elderly patients n = 21	Control patients n = 21	P
Age (years)	69.9 ± 4.8	46.4 ± 12.4	< 0.001*
Sex F/M	14 / 7	15 / 6	NS
Diagnostic delay (months)	26.3 ± 37.6	8.7 ± 15.3	0.02*
Muscular			
Proximal weakness	21/21 (100%)	21/21 (100%)	NS
Muscle pain	10/21 (47.5%)	15/21 (71.5%)	NS
General			
Asthenia	5/21 (24%)	5/21 (24%)	NS
Weight loss	7/21 (33%)	4/21 (19%)	NS
Fever	2/21 (9.5%)	3/21 (14%)	NS
Raynaud's phenomenon	0/21	3/21 (14%) [2 DM - 1 PM/SCL]	NS
Extramuscular			
Esophagus	11/21 (52%) [9 PM - 2 DM]	9/21 (43%) [4 PM - 5 DM]	NS
Lungs	1/21 (5%) [1 PM]	6/21 (28.5%) [6 PM]	NS NS
Heart	0/21	3/21 (14%) [2 PM - 1 DM]	NS
Kidneys	0/21	0/21	NS
Arthritis	2/21 (9.5%) [1 PM - 1 DM]	4/21 (19%) [2 PM - 2 DM]	NS

PM: polymyositis; DM: dermatomyositis; SCL: scleroderma; NS: Not significant ($p > 0.05$); *Statistically significant ($p < 0.05$)

Table II. Principal treament regimens for the two groups.

Medication	Elderly patients n = 21 *	Control patients n = 21	P
Glucocorticoid use	20/21 (95%)	21/21 (100%)	NS
First line	16/20 (80%)	19/21 (90.5%)	NS
Response	9/20 (45%)	10/21 (47.5%)	NS
Monotherapy failure	11/20 (55%)	11/21 (52%)	NS
Dependence	7/20 (35%)	10/21 (47.5%)	NS
Mean dose giving dependence	14 ± 2 mg/d	10 ± 2 mg/d	NS
Mean delay to associated therapy	10 ± 5 months	6.4 ± 2 months	NS
Mean duration	23 ± 28 months	43 ± 27 months	0.008
IVGG use	9/21 (43%)	11/21 (52%)	NS
First-line	4/9 (44.5%)	2/11 (18%)	NS
Response	6/9 (66%)	4/11 (36%)	NS
Mean duration	13 ± 15 months	8 ± 9 months	NS
Stopped due to side effects	1/9 (11%)	0/11	NS
Methotrexate use	11/21 (52%)	12/21 (57%)	NS
Mean dose	30 ± 10 mg/week	36 ± 8 mg/week	NS
Response	7/11 (63.5%)	6/12 (50%)	NS
Mean duration	14.5 ± 17 months	11.5 ± 10 months	NS
Stopped due to side effects	2/11 (18%)	2/12 (17%)	NS

NS: not significant ($p > 0.05$); IVGG: intravenous gammaglobulin; * one patient with paraneoplastic syndrome underwent surgical treatment only.

muscle fibers with aging (12). This could result in lower levels of muscle cell lysis and the liberation of enzymes in myositis in the elderly. Lower CK levels could also be explain by less necrosis in PM/DM in the elderly. The pathologic findings were not significantly different between the elderly and younger patients, but our retrospective study did not allow us to quantify the importance of the lesions and the presence of fibrosis and fatty infiltration.

Difficulties is analysing EMG data for the elderly have been described (13). In myositis, myopathic changes could be concealed by the physiological impoverishment of EMG features. In our study, acute changes (spontaneous fibrillations, small and fragmented motor unit potentials) were significantly less frequent in elderly patients, suggesting less necrotic myositis. The results of previous EMG studies of PM/DM in the elderly are consistent with this notion (14).

There was no significant difference in treament between the two groups, taking into account type, dose, duration and side-effects. The efficacy and side effects of steroids were similar in the 2 groups. The similar frequency and impact of these side effects was surprising, as regard to the literature (15), but may be explained by the difference in the mean follow-up period between the 2 groups. The retrospective nature of the data collection does not allow any definite conclusions about some therapeutic features.

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