EXTENDED REPORT

Interstitial lung disease, a common manifestation of newly diagnosed polymyositis and dermatomyositis

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Objectives: To estimate the prevalence and predictors of interstitial lung disease in newly diagnosed polymyositis and dermatomyositis.
Methods: A prospective study in which consecutive patients with newly diagnosed poly- and dermatomyositis, regardless of clinical symptoms of pulmonary disease, were investigated with chest *x* ray, high resolution computed tomography (HRCT), pulmonary function tests, and biochemical and autoantibody analysis. Patients with inclusion body myositis, malignancy, other defined inflammatory connective tissue diseases (CTDs), or antibody profile indicating other CTDs were excluded.
Results: Between March 1998 and September 2000, 26 new cases of poly- or dermatomyositis were diagnosed; 17 of those patients were included in the study. Interstitial lung disease (ILD), defined as radiological signs on chest *x* ray examination/HRCT or restrictive ventilatory defect, were found in 11 (65%) patients and were more common in men than in women. Arthritis and occurrence of anti-Jo-1 antibodies were found more often in patients with ILD than in those without. There was no statistically significant association between respiratory symptoms, other serological or laboratory variables and ILD.
Conclusions: ILD is a common early manifestation in patients with poly- and dermatomyositis and is not always related to clinical symptoms. Chest *x* ray examination, HRCT, pulmonary function tests, and analysis of anti-Jo-1 antibodies should be included in the initial investigation of patients with myositis regardless of respiratory symptoms.

Polymyositis (PM) and dermatomyositis (DM) are systemic inflammatory disorders with unknown aetiology and pathogenesis. They mainly affect striated muscles, resulting in proximal muscle weakness. However, other organ systems, including the lungs, may be affected, and pulmonary complications are associated with high morbidity and mortality.[1–6]

The reported prevalence of pulmonary involvement in PM/DM varies between 5 and 46% in cross sectional studies depending on whether clinical, radiological, functional, or pathological criteria have been used.[5–10] One serious lung complication of PM/DM is interstitial lung disease (ILD). The presence of ILD in patients with myositis affects the prognosis, and often has an influence on the choice of immunosuppressive treatment. Knowledge of the prevalence as well as predictors of ILD is therefore clinically highly relevant. As far as we know no previous studies have focused on patients with recent onset of PM/DM, and the prevalence of ILD in this group is unknown.

This study was initiated in order to clarify the prevalence and characteristics of ILD in an unselected group of patients with newly diagnosed PM/DM, using chest *x* ray examination, high resolution computed tomography (HRCT), and pulmonary function tests. A second objective was to compare the clinical presentation and biochemical findings in patients with and without ILD in order to evaluate methods for detection of patients at risk of developing ILD in the course of PM/DM.

PATIENTS AND METHODS

Patients

Between March 1998 and September 2000, 26 patients with a recent onset of myositis were identified at the rheumatology unit at the Karolinska Hospital, Stockholm, which has a referral area (northern part of Stockholm County) with a population of about 900 000 inhabitants. Patients with inclusion body myositis (n = 2), malignancy (n = 1), and those with another defined inflammatory connective tissue disease (CTD) or antibody profile indicating another CTD (n = 6) were excluded from this study. Thus 17 patients were included. The diagnosis of PM/DM was based on the criteria suggested by Bohan and Peter.[11 12] Furthermore, the PM diagnosis was confirmed by the presence of non-necrotic fibres invaded by mononuclear inflammatory cells in seven patients. Inclusion body myositis was diagnosed according to diagnostic criteria proposed by Griggs *et al*.[13] The local ethics committee approved the study, and informed consent was obtained from all patients.

Clinical features

Age, sex, ethnicity, smoking habits, and the presence of muscle weakness, myalgia, skin rash, dysphagia, arthritis, arthralgia, Raynaud's phenomenon, swollen hands, cough, and dyspnoea were recorded at the time of diagnosis.

Laboratory studies

Biochemical analyses included serum levels of creatine kinase (CK), alanine and aspartate aminotransferase (ALT, AST), and lactate dehydrogenase (LD). Serological studies included rheumatoid factor (RF) measured by nephelometry, and antinuclear antibodies (ANA) analysed by indirect immunofluorescence using Hep-2 cells. Antiribonucleic protein

Abbreviations: ALT, alanine aminotransferase; ANA, antinuclear antibodies; AST, aspartate aminotransferase; CK, creatine kinase; CT, computed tomography; CTD, connective tissue disease; DM, dermatomyositis; ELISA, enzyme linked immunosorbent assay; HRCT, high resolution computed tomography; ILD, interstitial lung disease; LD, lactate dehydrogenase; PM, polymyositis; RF, rheumatoid factor; TLC, total lung capacity; TLCO, carbon monoxide transfer factor; VC, vital capacity

(RNP), anti-Sm, anti-Ro/SSA, anti-La/SSB, and anticentromere antibodies were detected by enzyme linked immunosorbent assay (ELISA). Antihistidyl-tRNA synthetase (anti-Jo-1) was detected by ELISA or immunodiffusion. Tests were performed at the Departments of Clinical Chemistry and Clinical Immunology, Karolinska Hospital, Stockholm.

Radiology

A chest *x* ray examination was performed by the Fuji digital phosphor-plate technique and included anteroposterior and lateral views. Computed tomography (CT) examinations of the lungs were performed with a Siemens Somatom Plus scanner (Erlangen, Germany) and included both spiral CT and HRCT. All examinations were performed in maximal inspiration without intravenous contrast medium enhancement. The spiral CT was performed with a 10 mm slice thickness and pitch 1.0–1.2, depending on the length of the lungs. The HRCT was performed with a 2 mm slice thickness and 20 mm interspaces covering the entire lung. Both the 2 and 10 mm images were reconstructed with a high spatial frequency algorithm (defined for the actual scanner as AB 7541).

An experienced thoracic radiologist (ER), who was unaware of the clinical details, visually evaluated the radiological findings. The radiologist determined the presence and degree of a reticular pattern caused by a combination of interlobular lines and irregular thickening of interlobular septa as well as a reduction of the lung volumes. An arbitrary scale with a range of 0–3 was created: 0 = absence, 1 = mild, 2 = moderate, and 3 = severe degree of reticular pattern.

Pulmonary function tests

Total lung capacity (TLC) and vital capacity (VC) were determined in a body plethysmograph (Autobox 2800, Gould Electronics, The Netherlands). The single breath transfer factor for carbon monoxide (TLCO) was determined according to a modified Krogh procedure[14] using the above spirometry system, and was corrected for alveolar volume. The results from lung function investigations were expressed as a percentage of the predicted normal using standard reference values[15]; values below 80% of predicted were considered to be abnormal.

Interstitial lung disease

A diagnosis of ILD was defined as the occurrence of radiographic signs of ILD on chest *x* ray examination or HRCT and/or restrictive ventilatory defect (reduction in lung volumes).

Statistics

Statistical analyses of the data were performed using Student's *t* test (age, lung function, and time since onset of symptoms) and maximum likelihood χ^2 test (sex, PM/DM diagnosis, and the prevalence of pathological laboratory tests and symptoms). Significance was established at the $p<0.05$ level.

RESULTS

Clinical features

Of the 17 patients included, nine had PM and eight had DM. Table 1 summarises the main characteristics of the patients at presentation. All patients but one were white. At the time of diagnosis four patients (PM = 2, DM = 2) were current smokers, six were ex-smokers who had stopped smoking 0.5–49 years ago. Pulmonary symptoms, including dyspnoea and/or cough, were present in 71% of the patients. There was a significant difference between PM and DM for the presence of arthritis and Raynaud's phenomenon ($p<0.05$ for both).

Table 1 Classification and clinical and laboratory features of 17 patients with poly- and dermatomyositis

	PM (n = 9)	DM (n = 8)	Total (n = 17)
Diagnostic accuracy (n)			
Definitive	2	7	9
Probable	6	1	7
Possible	1		1
Sex (n), male/female	4/5	2/6	6/11
Age (years), mean (SD)	56.6 (8.4)	59.4 (12.9)	57.9 (10.5)
Initial symptoms			
Musculoskeletal	5		5
Pulmonary	3		3
Musculoskeletal and pulmonary	1		1
Others (rash, Raynaud etc.)		8	8
Symptoms and signs at the time of diagnosis			
Arthralgia/arthritis	6	3	9
Raynaud's phenomenon	3	0	3
Dysphagia	1	4	5
Swollen hands	2	1	3
Cough	5	1	6
Dyspnoea	7	3	10
Crackles on auscultation of lungs	5	1	6
Symptom duration (months), mean (SD)	7.6 (7.3)	3.1 (3.3)	5.5 (6.1)
Laboratory tests (%)			
Raised CK	100	100	100
Raised ALT	75*	75	75†
Raised AST	100*	100	100†
Positive RF	56	25	41
Positive ANA	33	75	53
Positive anti-Ro/SSA	44	25	35
Positive anti-La/SSB	22	0	12
Positive anti-Jo-1	44	0	24

*Data available for eight patients; †data available for 16 patients. PM, polymyositis; DM, dermatomyositis; CK, creatine kinase; ALT, alanine aminotransferase; AST, aspartate aminotransferase; RF, rheumatoid factor; ANA, antinuclear antibodies; Jo-1, histidyl-tRNA synthetase.

Laboratory findings

Table 1 summarises the laboratory findings. CK was increased in all patients; range 5– >76.8 μkat/l (reference value <2.5 μkat/l for women and <3.3 μkat/l for men). AST was increased in all patients. ALT was increased in all but four, who had a value slightly below the upper reference value. Positive RF was detected in 7 (41%) patients, and ANA with a speckled pattern were present in 9 (53%) patients. Anti-Jo-1 antibodies were present in 4/9 (44%) patients with PM and in none of the patients with DM ($p<0.05$). Anti-Ro/SSA antibodies were detected in 4/9 (44%) patients with PM and 2/8 (25%) with DM. Anti-La/SSB antibodies were detected in 2/9 (22%) patients with PM and in none of the patients with DM. Anti-RNP, anti-Sm, anticentromere, and Scl-70 antibodies were negative in all patients.

Radiographic findings

Radiological signs of ILD on chest *x* ray examination were present in eight patients: five with degree 1 and three with degree 2 abnormalities. HRCT of the lungs was performed in 15 patients, and radiological signs of ILD on HRCT were present in nine (table 2). The most common abnormalities consistent with ILD on HRCT were interstitial thickness (60%) and volume reduction (33%).

Pulmonary function tests

Pulmonary function tests were performed in 11 patients, either before (seven patients) or within 3 weeks after (four patients) starting corticosteroid treatment. VC and TLC were reduced (<80% of the predicted values) for six patients, respectively. The mean VC and TLC were 83% and 86% of

Table 2 Clinical, laboratory data, chest *x* ray, HRCT, and lung function variables at diagnosis of poly- and dermatomyositis

Case	Sex	Age (years)	Diagnosis	Symptoms (months)	CK (μkat/l)	Jo-1	Respiratory symptoms	Smoking	*x* Ray	HRCT	Spirometry (% predicted) VC	TLC	TLCO	ILD
1	M	60	PM	9	>76.8	+	C+D	Never	1	1	77	74	61	+
2	M	49	PM	5	76.8	+	D	Current	1	1	70	75	65	+
3	F	64	PM	1	31.6	+	D	Never	0	0	68	62	NA	+
4	F	65	PM	2	47	+	C+D	Former	2	2	71	66	58	+
5	F	42	PM	10	9.5	−	C+D	Former	2	2	63	67	59	+
6	M	67	PM	3	61.3	−	−	Former	0	1	NA	NA	NA	+
7	M	58	PM	2	>76.8	−	C	Current	1	NA	NA	NA	NA	+
8	F	41	DM	1	6.3	−	C	Current	1	1	NA	NA	NA	+
9	M	64	DM	1	>76.8	−	−	Never	2	2	86	85	76	+
10	M	51	DM	1	37.7	−	−	Current	1	1	53	71	54	+
11	F	71	DM	1	>76.7	−	D	NA	0	1	109	119	64	+
12	F	53	PM	24	15	−	C+D	Never	0	0	92	100	60	−
13	F	51	PM	12	5	−	D	Former	0	0	135	134	48	−
14	F	54	DM	6	6.7	−	D	Never	0	0	NA	NA	NA	−
15	F	65	DM	10	>76.5	−	D	Never	0	NA	91	98	81	−
16	F	80	DM	3	20.7	−	−	Former	0	0	NA	NA	NA	−
17	F	49	DM	2	11.4	−	−	Former	0	0	NA	NA	NA	−

PM, polymyositis; DM, dermatomyositis; CK, creatine kinase; C, Cough; D, dyspnoea; HRCT, high resolution computed tomography; VC, vital capacity; TLC, total lung capacity; TLCO, carbon monoxide transfer factor; ILD, interstitial lung disease; NA, not assessed.

predicted values, respectively. TLCO was reduced (<80% of the predicted values) in all tested patients but one; mean 63% of predicted value (table 2).

Interstitial lung disease

Eleven patients (65%) demonstrated objective signs of ILD. Table 3 presents the characteristics of the patients with and without ILD. Patients with ILD had significantly lower VC and TLC than those without ($p<0.05$ for both). Three of 11 patients with ILD had neither cough nor dyspnoea, and among the six patients without any objective signs of ILD, four reported respiratory symptoms. There was no statistically significant difference between patients with and without ILD for the presence of cough or dyspnoea. All the men, but only 5 of the 11 women, showed signs of ILD ($p<0.05$). Arthritis was more common in patients with ILD than in those without ($p<0.05$). Anti-Jo-1 antibodies were found only in patients with PM ($p<0.05$), and all had ILD. There was no difference between those patients with and without ILD for the other autoantibodies. ILD was more often present in patients with PM than in those with DM, although this difference was not significant. The average time from onset of myositis symptoms until the diagnosis was shorter in the patients with ILD than in those without ($p<0.05$). There was no significant difference between the groups for the presence of dysphagia, arthralgia, myalgia, Raynaud's phenomenon, or increased levels of CK, LD, ALT, or AST.

Table 3 Characteristics of patients with poly- and dermatomyositis with or without ILD

Characteristics	ILD present (n = 11)	ILD absent (n = 6)	p Value
Sex (male/female)	6/5	0/6	<0.05
Age (years), mean (SD)	57.5 (10.2)	58.7 (11.8)	NS
Time since onset of PM/DM (months) mean (SD)	3.3 (3.3)	9.5 (8.1)	<0.05
PM diagnosis, No (%)	7 (64)	2 (33)	NS
Symptoms, No (%)			
Arthritis	5 (45)	0 (0)	<0.05
Cough	5 (45)	1 (17)	NS
Dyspnoea	6 (55)	4 (67)	NS
Cough or dyspnoea	8 (73)	4 (67)	NS
Raynaud's phenomenon	2 (18)	1 (17)	NS
Dysphagia	3 (27)	2 (33)	NS
Swollen hands	3 (27)	0 (0)	NS
Laboratory tests, No (%)			
RF	6 (55)	1 (17)	NS
ANA	4 (36)	5 (83)	NS
Anti-Ro/SSA	5 (45)	1 (17)	NS
Anti-Jo-1	4 (36)	0 (0)	<0.05
Raised CK	11 (100)	6 (100)	NS
Raised ALT	8 (80)*	4 (67)	NS
Raised AST	11 (100)*	6 (100)	NS
Lung function (% predicted) mean (SD)			
VC	74.6 (16.9)†	106.0 (25.1)‡	<0.05
TLC	77.4 (18.2)†	110.7 (20.2)‡	<0.05
TLCO	62.4 (7.0)†	63.0 (16.7)‡	NS

*Data available for 10 patients; †data available for eight patients; ‡data available for three patients.
ILD, interstitial lung disease; PM, polymyositis; DM, dermatomyositis; RF, rheumatoid factor; ANA, antinuclear antibodies; Jo-1, histidyl-tRNA synthetase; CK, creatine kinase; ALT, alanine aminotransferase; AST, aspartate aminotransferase; VC, vital capacity; TLC, total lung capacity; TLCO, carbon monoxide transfer factor.

DISCUSSION

Polymyositis and dermatomyositis are rare disease entities affecting skeletal muscles and other organs, including the lungs. Interstitial lung disease in PM/DM is increasingly recognised as a serious complication of the disease. To our knowledge this is the first report in which newly diagnosed patients with PM/DM were investigated for the presence of ILD regardless of clinical symptoms that might indicate lung disease. We observed radiological signs or restrictive ventilatory defects compatible with ILD in 65% of the patients, which is a higher prevalence than previously reported.

The reported prevalence of ILD in PM/DM in earlier studies varies widely owing to the lack of uniform diagnostic criteria for ILD, the various stages of the disease in which patients were studied, and the source of patient referral.[5–10] Our study differs from previous studies in that we examined newly diagnosed cases with PM/DM regardless of clinical symptoms indicating pulmonary disease. Thus our patients were not selected on the basis of clinical or laboratory presentation. Our hospital serves as a referral centre for patients with myositis in the northern Stockholm County. The estimated annual incidence of PM/DM during the observation period was approximately 8.5 per million population, an incidence that is in agreement with previous studies.[16–18] This supports our assumption that we have covered most incident patients with PM/DM during the study period, which argues against a selection bias towards more severe cases. Thus we believe that our study cohort constituted unselected PM/DM cases and therefore the result should be representative of the entire

myositis population. The longer duration of myositis symptoms in the patients without ILD compared with those with ILD in this study indicates that lung involvement may be a common early organ manifestation in patients with PM/DM and argues against protracted disease duration before diagnosis as an explanation for the high prevalence of ILD. ILD is not a specific finding for PM/DM, and similar features of ILD have been observed in other CTDs.[19 20] Gabbay *et al* observed abnormalities compatible with ILD in HRCT in a significant proportion (33%) of patients with recent onset rheumatoid arthritis.[21] ILD is also a common manifestation in systemic sclerosis; the prevalence of ILD in early unselected cases before the start of treatment is unknown, however.

The majority of patients with ILD experienced respiratory symptoms such as cough or dyspnoea, but ILD was also present in four of the six patients without any clinical signs of pulmonary disease. Because a majority of the patients without objective signs of ILD also reported respiratory symptoms, neither cough nor dyspnoea is a valid indicator of pulmonary involvement in myositis and cannot be used for selection of patients who should undergo radiological and lung function assessments. The incidence of subclinical ILD in our study group was 18%. It is unclear if this group of patients will eventually develop clinically significant ILD. Long term prospective follow up studies are needed to evaluate the relevance of subclinical ILD in patients with myositis.

The reported incidence of ILD increases when new methods such as HRCT come into general use for the examination of patients. As expected chest *x* ray examination was less sensitive than HRCT in detecting abnormalities compatible with ILD. With HRCT we could identify parenchymal abnormalities compatible with ILD in two additional cases. HRCT was not carried out in two patients. Patient number 7 had changes on chest *x* ray with interstitial thickening and volume reduction. In patient number 15 HRCT might of course have been positive, which would make the proportion of ILD cases even higher in our study group. In patients with ILD, abnormalities on HRCT were more common than on chest *x* ray examination and lung function test. The abnormalities on HRCT were generally mild, which may suggest detection of an early stage of the disease, and the findings were non-specific, showing the same pattern as seen in patients with other inflammatory CTDs.[22] Restrictive changes on pulmonary function tests and reduced TLCO appeared in almost all patients with radiological evidence of ILD. We believe that small lung volumes and an abnormally low TLCO in the presence of abnormal radiography reflect ILD rather than ventilatory muscle weakness.

An association between anti-Jo-1 antibodies and arthritis with ILD has earlier been reported,[5 10 23–26] and is suggested to constitute a distinct subgroup of myositis, which is named antisynthetase syndrome.[25 27] In the present study all patients with arthritis had evidence of ILD. Our study could also confirm an association between occurrence of anti-Jo-1 antibodies and ILD. A high prevalence of antibodies to Ro/La was seen among our patients with PM/DM, although patients with other CTDs were excluded from our study. An association between anti-Jo-1 antibodies and anti-Ro52 was previously reported, but the mechanism for this association is still unclear.[28–30] As in previous studies there were no statistically significant differences in the serum levels of muscle enzymes and the prevalence of the other autoantibodies between patients with and without ILD.[4 7]

In conclusion, ILD was frequently seen in patients with early onset of PM and DM, and was not related to the presence of respiratory symptoms such as cough or dyspnoea. ILD is a prognostic unfavourable complication that might affect the choice of treatment and monitoring. Our study suggests that chest *x* ray examination, HRCT, pulmonary function tests, and analysis of anti-Jo-1 antibodies should be included in the initial investigations of patients with myositis. The predictive value of subclinical ILD in patients with myositis remains to be determined by a longitudinal study.

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ECHO

Behçet's disease may affect populations of African and Afro-Caribbean origin

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Behçet's disease may occur in ethnic groups that were previously thought to have a negligible risk of it, which raises the possibility that environmental factors contribute to the disease. Consideration needs to be given to this in the differential diagnosis of severe intraocular inflammation, particularly in the presence of mucosal aphthosis.

Behçet's disease is thought to occur mainly in countries bordering the Mediterranean, in Asia, and in the Far East. A case series of eight patients of west African or Afro-Caribbean origin in London showed, however, that seven patients satisfied the criteria of the International Study Group for Behçet's disease, and in the eighth patient, who had ocular and neurological involvement, typical histological changes were seen at necropsy.

The genetic association of the MHC allele HLA-B51 may define risk of the disease, particularly for ocular involvement. All patients had resided in the United Kingdom for several years, and six of them were B51 negative.

The rarity of the disease in populations in central and west Africa may be due to the fact that the HLA-B51 allele is not often found in this region. Another possible explanation may be a bias in case ascertainment, which depends on recognition of the disease and access to medical care.

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