CLINICAL FEATURES OF LUPUS MYOSITIS VERSUS IDIOPATHIC MYOSITIS: A REVIEW OF 30 CASES

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SUMMARY

Myositis is a rare but well-recognized complication of systemic lupus erythematosus (SLE). It is reputed to be milder than primary myositis in terms of morbidity and treatment response. This study compares clinical and laboratory features of idiopathic inflammatory myositis in patients with and without evidence of SLE overlap. We performed a case note review of 30 patients with probable or definite polymyositis/dermatomyositis of whom 11 also had definite or probable SLE. Lupus patients were slightly younger at diagnosis than those with primary disease, and more likely to be female. At presentation, quadriceps strength (expressed as a percentage of expected) was significantly reduced in both the lupus (48.9%; 95% CI 29.0–70.4%) and primary (52.0%; 95% CI 43.6–59.4%) myositis groups, and serum creatine phosphokinase (expressed as a multiple of the upper limit of normal) was significantly elevated (11.2; 95% CI 5.3–29.1 vs 10.7; 95% CI 6.1–17.6). During a mean (s.d.) follow-up period of 7.4 (4.1) yr, both groups tended to follow either a relapsing and remitting, or a chronic persistent course, and when last seen quadriceps muscle strength remained significantly depressed. One of the lupus patients and two of the primary myositis patients died due to direct complications of the disease, and one further death was attributable to a complication of therapy. Our results suggest that lupus myositis is often as severe as primary disease and should be treated with equal vigour.

KEY WORDS: Polymyositis, Dermatomyositis, Systemic lupus erythematosus, Myometry.

IDIOPATHIC polymyositis (PM) or dermatomyositis (DM) are reported to complicate 4–16% of cases of systemic lupus erythematosus (SLE) [1–6]. Some [3, 4], but not all [5], authors have suggested that lupus myositis is a relatively mild disease, but might be criticized because they failed to include patients with primary myositis for comparison. Other studies [7–12] have included appropriate controls, but have grouped together patients with myositis who also had SLE and other autoimmune rheumatic diseases, and have sometimes used obsolete or inadequate diagnostic criteria [7, 8]. Unsurprisingly, such heterogeneous groups have been reported to experience myositis which is less severe [10, 11], similar [9, 12] or worse [7, 8] than the primary disease.

This retrospective study compares clinical and laboratory features of patients with PM or DM, with and without associated evidence of SLE, followed up in a single unit during the period 1976–1995. Consistent diagnostic criteria have been used throughout [13], and because clinical assessment can be unreliable, objective measures of muscle strength have been recorded wherever possible.

PATIENTS AND METHODS

Patient records

Hospital records were reviewed of all adult (≥18 yr) patients treated between 1976 and 1995 for suspected PM or DM at the Bloomsbury Rheumatology Unit, University College/Middlesex Hospital. A prospective record of all such patients had been kept by one of us (DAI) during this period. Patients were excluded if they failed to meet the Bohan and Peter [13] criteria for probable or definite PM or DM, or if they had an underlying malignancy (defined as an associated neoplasm diagnosed within a year of the muscle disease). The remaining patients were stratified according to the presence or absence of features suggesting an underlying autoimmune rheumatic disease. Those who met the revised American Rheumatology Association criteria [14] for either probable (three criteria present) or definite (at least four criteria) SLE, and those with apparently primary myositis, formed the study group.

Clinical data

For all patients, demographic data, duration of symptoms prior to diagnosis/treatment, age at onset, and initial and subsequent drug treatment were noted. The presence or absence of a skin rash was noted. The clinical course was assessed and classed as either monophasic (a gradual return to normal without significant relapse), remitting and relapsing characterized by a return to at least 75% of predicted quadriceps muscle strength if measured, or full clinical recovery if myometry not performed, followed by periodic exacerbations and remissions), and chronic persistent (a failure to achieve remission at any point). Functional capacity [15] on last review was assessed by one of us (DAI).

Many patients had objective measurements of muscle strength performed at, or soon after, presentation, and in most cases serial measurements were available. Myometry was performed using the technique
described by Edwards et al. [16]. Maximum voluntary contraction (MVC) of the quadriceps muscle was measured in Newtons (N) force and results expressed as a percentage of expected values based on the patient’s premorbid body weight. Predicted MVC values were estimated using the following equation:

\[
\text{Quadriceps MVC (N)} = 7.91 \times \text{body weight (kg)} - 37.7
\]

Where repeated myometry was available, the lowest and most recent values were recorded. In a subset of patients, spirometry had also been performed; measurements of forced vital capacity (FVC) in litres (l), and carbon monoxide transfer factor corrected for lung volume \( K_{\text{CO}} \) in mmol/min/kPa/l at presentation, were compared to predicted values based on the patient’s age, weight and height.

**Laboratory measurements**

Initial, peak and most recent levels of serum creatine phosphokinase (CPK) were recorded. During the period of study, several laboratories had performed the CPK measurements using different assays. Accordingly, results were expressed as multiples of the upper limit of the appropriate normal adult reference range for each laboratory and assay.

Patient sera were also screened for antinuclear antibodies (ANA) using HEp2 cells [17], and antibodies directed against native double-stranded (ds)-DNA, extractable nuclear antigens [Ro (SSA), La(SSB), RNP, Sm] [18] and Jo-1 antibody [19]. Patients were recorded as positive for anti-ds-DNA antibodies if their titre exceeded twice the upper limit of normal on three occasions. In a subset of patients, serum was assayed for the anti-56 kDa nuclear RNP antibody [20].

**Statistical analysis**

The Mann–Whitney U-test and Wilcoxon matched pairs test were used to compared unpaired and paired continuous data, respectively. Correlations between continuous variables were examined using Spearman’s rank correlation. Proportions were compared using 95% confidence intervals, and categorical data with the \( \chi^2 \) test. All myometric measurements were compared to predicted values by estimating 95% confidence intervals for the median observed values. Confidence intervals were estimated using CIA software [21]. We controlled for potential spurious results by comparing the number of significant associations observed with the number expected due to chance using the Poisson distribution. The Bonferroni correction was not used because it is too conservative when a large number of comparisons are made.

**RESULTS**

Between 1976 and 1995, 34 patients were treated for suspected PM or DM, and all were seen by one of us (DAI). Clinical and laboratory data were obtained from the patients’ hospital medical notes \((n = 28)\), or from alternative sources including microfiche, general practice records and laboratory records \((n = 6)\). One patient did not meet sufficient criteria for myositis, one had an underlying adenocarcinoma, and two others had an undifferentiated autoimmune rheumatic disease, suggesting an overlap of primary Sjögren’s syndrome and scleroderma. Of the remaining 30 patients, 11 had definite \((n = 9)\) or probable \((n = 2)\) SLE, and 19 had myositis alone. The lupus myositis patients belonged to a larger cohort of 165 SLE patients who have attended the Bloomsbury Rheumatology Unit during this same period of time [22], giving a period prevalence of 6.6%. During follow-up, five of the lupus patients exhibited other manifestations of SLE, principally rashes and arthritis \((n = 5)\), pulmonary \((n = 2)\) and neurological \((n = 1)\) involvement. None experienced significant renal disease.

Clinical details of the study patients are shown in

**TABLE I**

<table>
<thead>
<tr>
<th>Clinical and laboratory characteristics of myositis patients at first presentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lupus myositis ((n = 11))</td>
</tr>
<tr>
<td>--------------------------------------------------------------</td>
</tr>
<tr>
<td>Male/female</td>
</tr>
<tr>
<td>PM/DM\‡</td>
</tr>
<tr>
<td>Age at onset (yr)</td>
</tr>
<tr>
<td>Duration of symptoms (months)</td>
</tr>
<tr>
<td>Initial serum CPK*$</td>
</tr>
<tr>
<td>(n = 10)</td>
</tr>
<tr>
<td>Initial quadriceps MVC*$</td>
</tr>
<tr>
<td>(n = 9)</td>
</tr>
<tr>
<td>Duration of SLE (months)</td>
</tr>
</tbody>
</table>

\*Significance value, comparing lupus and primary myositis groups using 95% confidence interval for differences in proportions, and
\†Mann–Whitney U-test.

\‡PM/DM, polymyositis/dermatomyositis.

\§Median (range); \*creatin phosphokinase, expressed as a multiple of the appropriate upper limit of the local reference range; \*estimated 95% confidence limits for the median using the Wilcoxon method; \*maximum voluntary contraction expressed as a percentage of the expected value based on the weight of the patient.
Table I. The distribution of PM and DM was similar in the two groups. Patients with SLE were more likely to be female and tended to be slightly younger at presentation. Although not statistically significant, there were differences in the ethnic background of the lupus and primary myositis patients. Both groups were predominantly Caucasian, but patients of Afro-Caribbean extraction were more prevalent in the lupus group, whereas Asian patients were more common in the primary myositis group. The duration of symptoms prior to diagnosis was significantly longer in the primary myositis group, although the absolute difference was quite small. In those patients for whom data were available, initial values of serum CPK and quadriceps strength were very similar in both groups. However, compared to expected reference values, these parameters were significantly elevated and depressed, respectively. Two of the male patients with PM and DM, respectively, had a monoclonal serum paraprotein without bone marrow evidence of an associated myeloma.

Table II shows the immunological characteristics of these patients. As expected, the prevalence of ANA, and antibodies to native DNA and extractable nuclear antigens, was clearly different in the two groups. In those patients with primary myositis who did have a positive antinuclear factor, the median titre was significantly lower compared to the patients with SLE. All patients with a positive antinuclear factor exhibited a speckled pattern of immunofluorescence, while two lupus patients had a mixed homogeneous/speckled staining pattern. Anti-Jo-1 antibodies were rare in both groups, whereas anti-56 kDa antibodies were relatively frequent.

Table III shows the various treatments used for the myositis patients. All received corticosteroid therapy (usually as the initial treatment), and there was no significant difference in the doses used in the two groups.

### Table II

**Immunological features of myositis patients**

<table>
<thead>
<tr>
<th></th>
<th>Lupus myositis (n = 11)</th>
<th>Primary myositis (n = 19)</th>
<th>P*</th>
</tr>
</thead>
<tbody>
<tr>
<td>ANA† positive</td>
<td>11/11</td>
<td>11/19</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Titre</td>
<td>1:5120 (1:40–1:10 240)</td>
<td>1:40 (1:10–1:320)</td>
<td>0.012†</td>
</tr>
<tr>
<td>Staining pattern</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Homogeneous</td>
<td>2</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Speckled</td>
<td>11</td>
<td>11</td>
<td></td>
</tr>
<tr>
<td>Anti-ds-DNA</td>
<td>5/10</td>
<td>0/18</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>ENA§ antibody positive</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Antigen</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ro</td>
<td>4/11</td>
<td>0/18</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>La</td>
<td>6/11</td>
<td>0/18</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Sm</td>
<td>5/11</td>
<td>0/18</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>RNP</td>
<td>5/11</td>
<td>0/18</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>MSA‡ antibody positive</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Antigen</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Jo-1</td>
<td>1/11</td>
<td>1/18</td>
<td>NS</td>
</tr>
<tr>
<td>56 kDa</td>
<td>2/5</td>
<td>9/15</td>
<td>NS</td>
</tr>
</tbody>
</table>

*Significance value, comparing groups using the 95% confidence interval for differences in proportions, or Mann–Whitney U-test.
†Antinuclear antibody.
§Extractable nuclear antigen.
‡Myositis-specific antibodies.

### Table III

**Treatment of 30 patients with myositis**

<table>
<thead>
<tr>
<th></th>
<th>Lupus myositis (n = 11)</th>
<th>Primary myositis (n = 19)</th>
<th>P*</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. receiving corticosteroids</td>
<td>11</td>
<td>19</td>
<td>NS</td>
</tr>
<tr>
<td>Steroid dose† (mg/day)</td>
<td>50 (15–125)‡</td>
<td>42.5 (25–250)</td>
<td>NS§</td>
</tr>
<tr>
<td>No. receiving other immunosuppressive drugs</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Azathioprine</td>
<td>9</td>
<td>16</td>
<td>NS</td>
</tr>
<tr>
<td>IVMP§</td>
<td>5</td>
<td>2</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Cyclophosphamide</td>
<td>2</td>
<td>5</td>
<td>NS</td>
</tr>
<tr>
<td>Methotrexate</td>
<td>1</td>
<td>4</td>
<td>NS</td>
</tr>
<tr>
<td>Cyclosporin</td>
<td>0</td>
<td>4</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Plasma exchange</td>
<td>2</td>
<td>2</td>
<td>NS</td>
</tr>
<tr>
<td>Immunoglobulin</td>
<td>1</td>
<td>1</td>
<td>NS</td>
</tr>
<tr>
<td>Duration (yr) of treatment</td>
<td>3.5 (1–11)†</td>
<td>6 (2–11)</td>
<td>NS§</td>
</tr>
</tbody>
</table>

*Significance value, comparing lupus and primary myositis groups using 95% confidence intervals for difference, or Mann–Whitney U-test.
†Initial steroid dose.
‡Median (range).
§i.v. methylprednisolone.
### Clinical Course

**Lupus myositis** (n = 11) | **Primary myositis** (n = 19) | 
---|---|---
Monophasic | 1 | 4 | NS†
Relapsing and remitting | 5 | 6 | \( \chi^2 = 0.95 \)
Chronic persistent | 5 | 8 | NS
Duration (yr) of follow-up | 3.5 (1–17)‡ | 7 (2.5–12) | NS§
Serum (CPK)* | | | 
Highest value | 15.7 (2.8–67.7)§ | 13.7 (0.76–41.4) | NS§
95% CL 7.45, 35.7 | 95% CL 8.75, 22.7 |
At last follow-up | 1.48 (0.06–7.24) | 1.0 (0.40–5.6) | NS§
95% CL 0.06, 7.24 | 95% CL 0.8, 1.95 |
**Quadriceps MVC** | | | 
Lowest value | 35.2 (7.0–68.8) | 28.6 (8.07–63.9) | NS§
95% CL 22.0, 41.1 | 95% CL 21.3, 38.0 |
At last follow-up | 55.0 (8.55–93.7) | 68.3 (8.07–63.9) | NS§
95% CL 32.1, 74.4 | 95% CL 8.07, 95.8 |
Functional capacity at last follow-up | II (I–III)† | I (I–III) | NS§
Total no. of deaths | 2 | 4 | NS'
No. of deaths due to disease | 1 | 2 | NS'

*Significance value comparing lupus and primary myositis patients using \( t \) test, †Mann–Whitney U-test and ‡95% confidence interval for difference in proportions.  
§Median (range).  
*Creatine phosphokinase and **maximum voluntary contraction expressed as multiple or proportion of expected values, respectively.  
§P < 0.05, ‡P < 0.01 compared to baseline values using Wilcoxon matched-pairs test.  
†Estimated 95% confidence limits for median using Wilcoxon method.

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**Clinical course**

Various other immunomodulating drugs were used, of which azathioprine was the most frequently prescribed. Patients with lupus myositis were more likely to receive i.v. methylprednisolone (often for a simultaneous flare of the myositis and their SLE), while cyclosporin was only given to those with primary myositis. Treatment duration was similar in the two groups.

Table IV shows the clinical and laboratory outcome in 30 patients with myositis. There was a tendency for more patients with primary myositis to exhibit a monophasic disease course and the median muscle strength at last follow-up was slightly greater in these patients compared to the lupus patients, but the differences were not significant. In both groups, the majority of patients pursued either a remitting and relapsing, or chronic persistent course. Peak levels of serum CPK were significantly higher than those at presentation, while muscle strength frequently fell to levels below those recorded at baseline. With continuing treatment, CPK levels tended to normalize in both groups. In contrast, final muscle strength was significantly higher than the lowest recorded values, but not significantly different to baseline. Patients with primary disease tended to have a slightly higher quadriceps MVC and functional capacity at last follow-up compared to those with SLE, but the differences were not significant. Myometry and CPK measurements were not available for some subjects, and this is indicated in the table. Final functional capacity in these individuals was generally slightly worse than in those in whom measurements were made, although the differences were not significant.

The overall mortality of the two groups was similar; in the lupus group there were two deaths, one due to likely myocardial involvement and one due to multiple complications following corticosteroid-induced osteoporotic fractures. In the primary myositis group, there were four deaths; two were due to incidental pathology (one fatal pneumonia without evidence of underlying respiratory muscle involvement, and one with hypertrophic obstructive cardiomyopathy), and two related to the myositis (one female patient died with a skull fracture having been pushed backwards by her pet dog, and one died as a result of pneumonia complicating poor mobility and respiratory involvement).

Table V shows the correlations between the various measured disease parameters. Serum CPK measurements at onset were not a reliable guide to final functional outcome or muscle strength at any time. Presenting CPK measurements did, however, correlate very well with peak values, whereas neither correlated with final values. Final functional capacity was well correlated with lowest and most recent levels of quadriceps strength (Fig. 1); furthermore, there was a weak but non-significant correlation with baseline strength. A total of 10/11 lupus patients and 7/19 primary myositis patients also had spirometry performed either at presentation (n = 13) or shortly thereafter (n = 4). FVC (expressed as a percentage of expected) was significantly depressed in both the primary [mean 76.7% (95% CI 67.6–85.8%)] and lupus [66.8% (52.6–81.0%)] patient groups. FVC was well correlated with baseline myometry (r = 0.63, P = 0.009), and showed weaker associations with latest myometry (r = 0.60, P = 0.03) and final functional capacity (r = –0.54, P = 0.024). In contrast,
FVC was not related to baseline or peak serum CPK values, but was weakly correlated with the most recent CPK levels ($r = -0.59$, $P = 0.013$). Measurements of $K_{CO}$ were not significantly different from predicted values.

Finally, we estimated the probability that the observed number of associations might have occurred by chance. Of a total of 55 comparisons, 16 achieved statistical significance. The probability that this number of significant results would have occurred by chance is very low ($P < 0.00001$), suggesting that spurious associations are unlikely to have occurred.

**DISCUSSION**

We have directly compared the clinical and laboratory features of lupus myositis with the primary disease in a group of patients followed up in the same centre using uniform methods of assessment over a 20 yr period. Wherever possible, objective measures of muscle strength have been utilized. At presentation, both groups showed similar significant increases in serum CPK, and comparable reductions in muscle strength. There were no substantial differences in morbidity or mortality on prolonged follow-up, and the clinical course was generally similar for both groups. Our findings suggest that myositis occurring in lupus patients is very similar to the primary disease. However, our study is small and potential confounding influences must be considered.

As expected from previous studies [10, 23], our lupus patients were more likely to be female, and more often younger and non-Caucasian compared to the primary myositis patients. The latter group had experienced slightly longer delays before diagnosis and treatment, but the case ratio of PM/DM was identical, and treatment experience and follow-up duration similar. The patients with primary myositis were often referred from other hospitals, whereas the lupus patients were usually already under regular follow-up at the Bloomsbury Rheumatology Unit. Indeed, the latter probably explains the slightly shorter median time to diagnosis experienced by the lupus patients.

Which of these differences might be relevant is controversial. Neither gender [24–27] nor ethnic origin [12, 24] seem important despite early reports to the contrary [28]. Some authors have reported more aggressive myositis in DM vs PM [13, 29, 30], but this has not been confirmed by other studies [23, 25, 27] despite likely differences in pathogenesis [31, 32]. In contrast, older age at onset [8, 12, 24–26, 29, 30, 33] and delayed diagnosis [9, 11, 27, 30, 33, 34] are generally associated with a poorer prognosis, as is the potential adverse effect of an associated autoimmune rheumatic disease [8].

The design of the study may also have introduced bias. Case ascertainment is likely to have been complete because myositis patients were identified prospectively. However, much of the clinical and laboratory data had to be collected retrospectively, and consequently the follow-up data were not always complete. In particular, follow-up myometry was often unavailable, especially for those with primary myositis. Patients with more severe disease may have been more likely to undergo repeat measurements, as

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**TABLE V**

Correlations between quadriceps myometry, serum creatine phosphokinase and final functional capacity in 30 patients with myositis

<table>
<thead>
<tr>
<th>FC* at last follow-up</th>
<th>MVC1†</th>
<th>MVC2</th>
<th>MVC3</th>
<th>CPK1‡</th>
<th>CPK2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Quadriceps MVC†</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>At presentation¹</td>
<td>−0.24</td>
<td>−</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lowest value²</td>
<td>−0.65§</td>
<td>0.47*</td>
<td>−</td>
<td></td>
<td></td>
</tr>
<tr>
<td>At last follow-up³</td>
<td>−0.85#</td>
<td>0.14</td>
<td>0.63*</td>
<td>−</td>
<td></td>
</tr>
<tr>
<td>Serum CPK‡</td>
<td>0.08</td>
<td>0.15</td>
<td>−0.28</td>
<td>0.20</td>
<td>−</td>
</tr>
<tr>
<td>Highest value²</td>
<td>−0.04</td>
<td>0.06</td>
<td>−0.23</td>
<td>0.38</td>
<td>0.90#</td>
</tr>
<tr>
<td>At last follow-up</td>
<td>0.21</td>
<td>−0.24</td>
<td>−0.17</td>
<td>−0.22</td>
<td>0.29</td>
</tr>
</tbody>
</table>

*Steinbrocker functional capacity.
†Maximum voluntary contraction.
‡Creatine phosphokinase.

$r < 0.05$, § 0.001, # 0.0001.
supported by the slightly worse functional outcome in this subgroup.

The overall effect of these various sources of potential bias is difficult to predict. However, the primary myositis patients might have been expected to have had a worse outcome based on their older age at onset, longer delay in diagnosis and likely referral bias in favour of more resistant disease requiring specialist treatment. These factors would have tended to increase any observed difference in severity, an effect likely to be further exaggerated by the differential absence of myometric data in the primary myositis group. However, we found that the two groups were very similar at presentation and on follow-up, thus supporting our conclusion that lupus myositis is just as severe as the primary disease.

It is unclear how lupus myositis gained its benign reputation [32, 35]. Of the three published studies describing the clinical characteristics of myositis in patients with SLE [3–5], only one [5] utilized Bohan and Peter criteria, and none included any controls. The latter study suggested that lupus myositis was very similar to primary disease, in contrast to the milder descriptions of Fessel [3] and Tsokos et al. [4]. However, the diagnostic criteria used by Fessel [3] are not stated, while those used by Tsokos et al. [4] were less than rigorous: all but one of their patients with alleged myositis had a normal serum CPK, and only 5/18 subjects had a confirmatory muscle biopsy. Six others who had a muscle biopsy performed may well have had non-specific changes that can occur in patients with lupus [36]. In contrast to myalgia, which can affect nearly half of patients with SLE [37], true myositis is relatively rare and failure to identify these genuine cases probably explains the ill-deserved reputation of lupus myositis.

The most common clinical course was a relapsing and remitting one, with monophasic illness being relatively rare. It was interesting that although patients often deteriorated despite initiation of therapy, and most subsequently showed some degree of recovery, final myometric measurements were little different to those at onset. It is possible that those with the worst disease tended to have more frequent measurements, but these results are consistent with the clinical assessment of functional outcome, suggesting considerable residual weakness despite apparently adequate treatment and duration of follow-up. Optimal strategies to reduce the long-term morbidity associated with myositis are unclear. Physiotherapy may well be important in the prevention of contractions early on and during remission in the improvement of endurance and strength [38].

As serum muscle enzymes are often used to monitor disease activity [13], it was disappointing to note the poor correlation between CPK levels (at onset or most recent review) and either muscle strength or functional outcome. It may be that serum CPK levels are useful to monitor the active inflammatory component of myositis, but do not accurately reflect accumulated muscle damage or the success of repair processes. However, the situation is probably more complex as myositis can occur in the absence of a rise in the CPK [39]. In addition, CPK inhibitors may be present in the sera of some patients with myositis and in some the amount of inhibitor correlates with the severity of the illness [40].

In contrast to CPK, measured muscle strength correlated well with functional outcome. Myometry may, therefore, provide an integrated measure of the balance of muscle damage and regeneration in patients with myositis. The interrelationship between myometry and spirometry is also of interest, and FVC may be a reasonable, and more accessible surrogate for quadriceps strength. It was unfortunate that follow-up spirometry was not available, as more recent estimates of FVC might have correlated better with final functional capacity than initial measurements. We would agree with others that the use of objective and reproducible measures of muscle strength is to be encouraged [24].

Corticosteroids were used universally among our patients (usually as a first-line agent) and additional immunosuppressive agents were commonly used. Azathioprine was the usual choice, although a wide variety of other drugs were used. There is some evidence that azathioprine used concomitantly with prednisolone reduces the subsequent steroid requirement [41]. The absence of a controlled trial of corticosteroids has been commented on [13], and some older studies have suggested that such treatment may not influence mortality from myositis [8, 42].

The immunological results are of interest. The ANA was less commonly positive (and then in lower titre) in the primary myositis group, as observed in previous studies [12, 24]. Indeed, the presence of a strongly positive ANA may lead to a more diligent search for an associated autoimmune rheumatic disease. However, it is interesting that whenever the ANA was found to be positive, the immunofluorescence pattern was speckled. The significance of this finding is unknown, but it is unlikely to have been due to chance. It has been noted previously by Reichlin and Arnett [17], and by Fudman and Schnitzer [43], and presumably relates to the presence of antibodies to antigens other than DNA/histones. It is interesting to speculate whether those subjects with a positive ANA in the primary myositis group might ultimately develop another autoimmune rheumatic disease, as has been demonstrated in juvenile dermatomyositis [44].

The prevalence of anti-Jo-1 antibody was surprisingly low in this group. Jo-1 antibodies have been reported far more frequently in some studies [24], although Nishikai and Reichlin [19] found the antibody in 30% of patients with primary PM, but only 4% of those with primary DM and <1% of those with an overlap syndrome. In contrast, the anti-56 kDa nuclear RNP antibody was quite common. The latter antibody has been found to be a relatively specific marker for primary myositis, occurring only rarely in patients with SLE without evidence of
muscle disease, or patients with other autoimmune rheumatic diseases [20]. Mortality directly attributable to the myositis was 10% in this study, although one additional death was related to a complication of steroid therapy. This is consistent with recent studies and has been attributed to improvements in general medical care [18]. A wide variety of mortality statistics are quoted in the literature and are well reviewed by Benbassat et al. [26]. It is important to remember that treatment can cause death or disability directly or indirectly, and this has been recognized for many years [9].

In conclusion, we found few differences between lupus and primary myositis patients in terms of measures of disease activity or final outcome. Lupus myositis is not a mild disease and should be treated as aggressively as primary myositis. The importance of measuring muscle strength [24, 33] and function [24], and the superiority of such indices to muscle enzyme changes, have been underlined [23].

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