Extrapyramidal type rigidity in rheumatoid arthritis

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Abstract

Objectives. We had noted cogwheel rigidity in a number of patients with rheumatoid arthritis (RA). Based on this finding, we aimed to investigate formally the presence of rigidity and cogwheeling in RA patients. Our secondary aim was to survey the co-existence of RA and Parkinson’s disease (PD).

Methods. A total of 87 consecutive patients with a diagnosis of RA, 78 patients with PD and 67 otherwise healthy patients attending a dedicated headache clinic participated in the study.

Results. Rigidity was observed in 24% of RA, 60% of PD and 2% of headache patients. The frequency among the RA patients was significantly higher compared to that of patients with headache ($\chi^2 = 15.2; P = 0.00009$). The frequency of PD among the RA patients was 2/87 (2.3%), while the frequency of RA among the PD patients was 6/78 (7.7%).

Conclusion. Rigidity can be observed in approximately a quarter of patients with RA.

Key words: Rheumatoid arthritis, Rigidity, Extrapyramidal, Cogwheeling, Parkinson’s disease, Co-existence.

Rigidity is a simultaneous contraction of the agonist and antagonist muscles, leading to constant resistance to passive movement. Being one of the cardinal symptoms of idiopathic Parkinson’s disease (PD), rigidity is also a prominent symptom of many extrapyramidal syndromes. Cogwheel rigidity, a special type of rigidity, is perceived by the examiner as an interrupted and rhythmic muscle resistance during examination of the extremities.

Joint examination in arthritis patients includes passive movement of the extremities by the examiner. One of us (HY) had noted over the years a cogwheel-like phenomenon in many of his patients with rheumatoid arthritis (RA). Based on this finding, we aimed to investigate formally the presence and the level of rigidity and cogwheeling in patients with RA. A secondary aim was to survey the co-existence of RA and PD in a group of hospital-attending patients with the primary diagnoses of RA and PD.

Patients and methods

A total of 87 consecutive patients with a diagnosis of definite RA [1] who were followed by the Division of Rheumatology of the Cerrahpaşa Medical Faculty, and who also consented to a detailed neurological examination, participated in the study. Patients with tremor or central nervous system diseases, those using drugs affecting muscle tone, those patients with RA who had severe pain during physical examination, and those who had had a surgical operation in upper extremity joints or had ankylosed joints were excluded. All patients were examined by one rheumatologist or either of two neurologists. Initially, 49 patients of the RA group were examined by both neurologists to assess interobserver variability. There were only two patients in whom there was a disagreement between the neurologists about the level of rigidity. In one patient, the disagreement was whether he had a rigidity score of 0.5 or 0, and in the other whether the rigidity score was of 1 or 2. Since there was no appreciable interobserver variation in assessing rigidity, the remaining 38 patients were examined by either neurologist, depending on availability.

Seventy-eight patients with definite PD [2] who were followed up by the department of neurology of the same hospital took part in the study as diseased controls. Another diseased control group consisted of 67 age- and sex-matched patients who were attending a dedicated headache outpatient clinic in the same department of neurology. Only around 1% of the attendees in this clinic have any identifiable central nervous system disease. In this control group, similar exclusion criteria as defined for the RA patients were used.

Disease activity in RA patients was assessed by the Steinbrocker scale [3]. In order to survey the frequency of PD among RA patients, a detailed medical history which was specific for PD and drug use was taken.


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Apart from rigidity and cogwheeling, other clinical motor features of PD, such as bradykinesia, bradykinesia and tremor, were also investigated in the upper extremities. Functions such as gait, speech, and foot and finger tapping were also tested. All these positive motor phenomena and functions were rated according to a modified Unified Parkinson’s Disease Rating Scale (UPDRS) [4]. In the original UPDRS scheme, rigidity is measured on a 0–4 scale and a designation of 1+ rigidity implies slight rigidity or that brought on only by the contralateral movement of the opposite hand (mirror movement). Traditionally, our neurology department designates a score of 0.5 to that rigidity only brought on by mirror movement. Our 1+ rigidity, on the other hand, encompasses slight rigidity only. The two scales are otherwise identical.

Disease staging in the PD group was according to the Hoehn–Yahr scale [5]. Also in this group of patients, to determine the co-existence of RA, a detailed medical history which was specific for RA was taken. Clinical positive motor phenomena and motor functions were rated according to UPDRS. In patients whose medical history and clinical findings suggested RA, a panel of laboratory investigations was performed. The erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) levels and the presence of rheumatoid factor (RF) were measured. Roentgenograms of the hands and wrists were also obtained and evaluated by the senior rheumatologist. An ESR over 30 mm/h, a CRP value over 0.5 IU/ml and a positive RF were accepted as supportive for the diagnosis of RA, as well as radiological findings such as a tendency towards symmetrical involvement of the proximal interphalangeal and metacarpophalangeal joints, juxta-articular osteopenia, loss of articular cartilage and bone erosions.

In the headache group, only rigidity (including cogwheel rigidity) was assessed.

**Results**

Duration of the disease, age of onset of the disease, sex distribution and mean age of the 85 RA, 72 PD, eight RA and PD, and 67 headache (H) patients are shown in Table 1. There were no patients with RA or PD among the patients in the headache group.

When one considered cogwheel rigidity not brought on by mirror movement (rigidity score 1 or more), the PD patients, with or without co-existing RA, had among them the highest number of patients demonstrating cogwheel rigidity (Table 2). Nearly a quarter (20/85) of patients with RA and only one patient with headache had this clinical sign \( (x^2 = 15.2; \text{d.f.} = 1; P = 0.00009) \) (Table 2). Furthermore, when one considered all levels of rigidity, the patients with RA still had higher levels of rigidity when compared to headache patients \( (x^2 = 22.7; \text{d.f.} = 2; P = 0.00001) \) (Table 2).

Among patients with RA, there were two patients (both female, aged 67 and 65 yr) who had co-existing PD (2.3%), whereas among those with PD, six patients (two males, aged 67 and 56 yr, and four females, aged 51.5, 61, 58 and 67 yr) had co-existing RA (7.7%). Thus, the co-existence rate of the two diseases in the whole group was 4.8% (8/165). In patients with co-existing PD and RA, the onset of RA historically preceded PD by 2–20 yr.

The mean value (±s.d.) of the Steinbrocker scale for the RA patients was 1.6 ± 0.6 and the mean disease stage (±s.d.) according to the Hoehn–Yahr scale for the PD patients was 2.0 ± 0.5. Seventy-eight per cent (66/85) of the patients with RA were RF positive.

Among the RA patients, CRP, ESR values, presence of RF, sex, age, duration of disease, age of onset and Steinbrocker index value were not related to the level of rigidity (data not given).

Table 3 shows the distribution of the various clinical motor features of PD among the groups studied. It is seen that apart from rigidity, the RA patients did not exhibit other features of PD. Finally, none of the RA patients, apart from those with co-existing PD, required specific medication for the control of their rigidity.

**Discussion**

Our study showed that cogwheel rigidity can be seen in patients with RA. Neither the findings in two patients with a primary diagnosis of RA and co-existing PD nor those in six patients with a primary diagnosis of PD and co-existing RA were included in our assessment of rigidity in RA. Inclusion of these patients would obviously have made the rigidity we observed in RA more pronounced.

When we compared the frequency of cogwheel rigidity among the various groups studied, we gave more importance to cogwheeling observed without mirror movement of the opposite extremity for the simple reason that when one examines an arthritic patient one does...

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**Table 1.** The distribution of gender and the mean values of the duration of disease, age at onset of the disease (excluding the headache group) and age of patients with rheumatoid arthritis (RA), Parkinson’s disease (PD), headache (H) and RA and PD

<table>
<thead>
<tr>
<th></th>
<th>RA ( n = 85 )</th>
<th>PD ( n = 72 )</th>
<th>RA and PD ( n = 8 )</th>
<th>H ( n = 67 )</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female/male</td>
<td>71/14</td>
<td>26/46</td>
<td>6/2</td>
<td>50/17</td>
</tr>
<tr>
<td>Duration of disease ( \text{yr} \pm \text{s.d.} )</td>
<td>11.7 ± 8.3</td>
<td>6.3 ± 3.8</td>
<td>6.8 ± 5.5a</td>
<td>–</td>
</tr>
<tr>
<td>Age at onset ( \text{yr} \pm \text{s.d.} )</td>
<td>40.6 ± 11.9</td>
<td>58.2 ± 10.2</td>
<td>58.8 ± 6.2b</td>
<td>–</td>
</tr>
<tr>
<td>Mean age ( \text{yr} \pm \text{s.d.} )</td>
<td>51.9 ± 10.9</td>
<td>64.5 ± 9.3</td>
<td>65.1 ± 5.5</td>
<td>48.6 ± 11.3</td>
</tr>
</tbody>
</table>

*Duration of PD.

*Age of onset of PD.
Table 2. Distribution of rigidity scores in Parkinson’s disease (PD), rheumatoid arthritis (RA), headache (H), and RA and PD groups. \(n\) and \(N\) denote the number of patients exhibiting cogwheel rigidity and the total number of patients in each group, respectively.

<table>
<thead>
<tr>
<th>Rigidity score</th>
<th>PD (n = 72)</th>
<th>RA (n = 85)</th>
<th>H (n = 67)</th>
<th>PD and RA (n = 8)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.00</td>
<td>6 (8.3)</td>
<td>65 (76.5)</td>
<td>59 (88.1)</td>
<td>0</td>
</tr>
<tr>
<td>0.50</td>
<td>23 (31.9)</td>
<td>0</td>
<td>7 (10.4)</td>
<td>3 (37.5)</td>
</tr>
<tr>
<td>1.00</td>
<td>29 (40.5)</td>
<td>20 (23.5)</td>
<td>1 (1.5)</td>
<td>4 (50.0)</td>
</tr>
<tr>
<td>2.00</td>
<td>12 (16.7)</td>
<td>0</td>
<td>0</td>
<td>1 (12.5)</td>
</tr>
<tr>
<td>3.00</td>
<td>2 (2.8)</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

Table 3. The median (range) values for the cardinal clinical motor features (bradykinesia, bradydymia, tremor) of Parkinson’s disease (PD) and motor functions (gait and finger tapping) according to UPDRS for rheumatoid arthritis (RA), Parkinson’s disease (PD) and RA and PD patients.

<table>
<thead>
<tr>
<th></th>
<th>RA (n = 85)</th>
<th>PD (n = 72)</th>
<th>PD and PD (n = 8)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bradykinesia</td>
<td>0 (0–1)</td>
<td>1 (0–3)</td>
<td>2 (0–2)</td>
</tr>
<tr>
<td>Bradymenia</td>
<td>0 (0–1)</td>
<td>1 (0–3)</td>
<td>2 (1–2)</td>
</tr>
<tr>
<td>Tremor</td>
<td>0 (0–1)</td>
<td>1 (0–3)</td>
<td>1 (0–2)</td>
</tr>
<tr>
<td>Finger tapping</td>
<td>0 (0–2)</td>
<td>1 (0–3)</td>
<td>1.25 (0–2)</td>
</tr>
<tr>
<td>Gait</td>
<td>0 (0–1)</td>
<td>1</td>
<td>1 (0–2)</td>
</tr>
</tbody>
</table>

not ordinarily use this modality of examination. The frequency of cogwheel rigidity observed among our PD patients, on the other hand, we believe would be quite expected among a group of PD patients under treatment.

During the preparation of our manuscript, we found out that Mumenthaler [6], a German neurologist, in a treatise on neurological diseases, wrote about his impression that a phenomenon similar to cogwheel rigidity could be seen in patients with joint problems, presumably due to changes in muscle tone. No details about the possible presence of RA were given in this treatise. Our results mandate that specificity studies should be conducted among other arthritides as well.

In a hospital-based, retrospective case–control study, it is also hard to derive hard conclusions from co-morbidity rates. The 7.7% frequency of RA observed among our patients with PD is somewhat higher than that found in the general population (0.1–1.96%) [7]. However, it must be emphasized that the prevalence of RA increases with age, being more common among the females in all age groups [8]. This makes our sample of PD patients with a mean age of 65 yr not truly representative of the general population. Among our 30 female and 48 male PD patients, the frequencies of co-existing RA were 13.3% (4/30) and 4.2% (2/48), respectively. On the other hand, the gender-based prevalence rates of RA in a population aged between 65 and 79 yr were reported to be 4.9% for females and 1.8% for males [9]. While these comparisons indeed suggest an increased frequency of RA among the PD patients, it must also be mentioned that reliable reference figures for the prevalence of RA in Turkey are not available. Thus, proper epidemiological studies are needed to clarify further the issue of co-existing RA and PD.

Apart from not including another diseased group with arthritis, another drawback of our study is its non-blinded nature. On the other hand, the difficulty of blinding a clinical study of cogwheel rigidity among patients with RA is obvious. More objective measures of rigidity utilizing electromyography can be tried [10].

We do not have a ready explanation for the existence of cogwheel rigidity among patients with RA. One explanation may be the alterations in joint mechanics and muscle tone, as was historically suggested [6]. Another intriguing possibility concerns the involvement of the dopaminergic system. There is some evidence that the diencephalic–dopaminergic system is involved in the pathogenesis of RA [11]. Some dopamine receptor agonists have an anti-inflammatory effect [12] and in RA and PD patients the past we have shown a marked increase in sebum secretion, a finding very common in PD, among our patients with RA [13]. On the other hand, the paucity of other signs of PD, such as bradykinesia and bradydymia, among our RA patients gives more weight to mechanical problems as the cause of rigidity in our RA patients.

Whatever its cause, or the presence or absence of a pathogenetic link between RA and PD, the clinician should be aware that cogwheel rigidity can be one of the musculoskeletal manifestations of RA.

References


