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ORIGINAL ARTICLE

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The definition and clinical features of the mutilans type of rheumatoid arthritis

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Abstract The mutilans type of rheumatoid arthritis (RA) is refractory to several treatments and involves many types of surgical application. It is difficult to prevent its progress, and it has a poor functional prognosis. However, the definition is not always distinct. In this report, I attempt to clarify the clinical features of the mutilans type of RA and establish an accurate definition of the disease. Previous definitions have been divided into three groups: (1) the mutilans group (Mu-G), which has more than three joints showing severe resorptive bone destruction and joint instability; (2) the suspected mutilans group (sMu-G) which has one to two joints with joint instability or showing no instability but with highly resorptive bone destruction; (3) the Larsen V group (LV-G), which has radiological findings indicating Larsen grade V disease. Patients suffering from rheumatoid arthritis for over 10 years and who do not fall into any of the above groups are referred to as the control group. Among 337 patients who suffered from RA for more than 10 years, 58 were classified as being in the mutilans group, 59 in the suspected Mu-G, 47 in the LV-G, and 173 in the control group. The mutilans group had distinctive features which were different from those of the other groups, which had a radiological finding of Larsen grade V disease or severe resorptive bone destruction without instability. From these data, the definition of the mutilans type of RA should be specified as the Mu-G. Thus, the Mu-G shows a distinctive clinical picture which is unlike that in the other groups. It is important to define the mutilans type of RA clearly because it has a poor prognosis. Early identification will help in the establishment of a treatment plan.

Key words Arthritis mutilans · Larsen grade V · Mutilans type · Mutilating disease (MUD) · Rheumatoid arthritis (RA)

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Introduction

The mutilans type of rheumatoid arthritis (RA) has a poor functional prognosis, it is refractory to several treatments, and involves many types of surgical application. However, a definition has not yet been clearly established. This article investigates how the mutilans type of RA could be appropriately classified. Marie and Leri[1] first described the hand as "la main en lorgnette" because of the telescoping of the fingers in severe destructive osteolytic joint disorder. In 1935, Stursberg[2] used the term polyarthritis mutilans. Since Mather,[3] in 1954, described RA patients with this type of destructive arthritis as having arthritis mutilans, the term arthritis mutilans has become widespread. Larsen et al.[4] described the radiological findings where the original articular surfaces have disappeared as grade V in the classification of the radiographic evaluation of RA, and referred to it as a mutilating abnormality. Ochi et al.[5–7] classified RA in three disease subsets, and described mutilating disease (MUD) as a condition where the number of joints with erosions (NJE) increases rapidly to include the majority of joints within 10 years, but increases little thereafter. Murasawa et al.[8] defined the mutilans type of RA as those patients in whom the radiological changes of Larsen grade V[4] show in more than three joints among the metacarpophalangeal (MP) joints of the fingers and the medium-size and large joints, and also in more than two of the medium-size and large joints. Ryu et al.[9] defined the mutilans type of RA as those patients who have conditions such as the opera-glass deformity of the fingers, or severe resorptive bone destruction in more than three joints. Fukui et al.[10] defined the mutilans type of RA as those patients showing severe resorptive bone destruction and instability such as opera-glass hands in three or more joints. In the case of large joints, two affected joints are sufficient for classification in this group.

This paper divides the previous definitions of the so-called mutilans type of RA into three groups, and justifies this classification through some comparisons.

Patients and methods

Patients

A total of 337 patients at the Department of Orthopaedic Surgery, Showa University Hospital, Tokyo, Japan, and the Kami-itabashi Hospital, Tokyo, Japan, were the subjects of this study. They all met the American Rheumatism Association criteria for the diagnosis of RA,[11] and roentgenograms were taken of 68 joints[12] among patients who had suffered from RA for over 10 years. There were 305 women and 32 men, with a mean age of 62 years (range 23–93 years) at the time of the investigation. These patients were classified into four groups (Table 1). (1) The mutilans group (Mu-G), which included patients showing severe resorptive bone destruction and instability such as opera-glass hands in three or more joints. In the case of large joints, two affected joints are sufficient for classification in this group. There were 58 cases in this group: 54 women and 4 men, with a mean age of 60 years (range 29–90 years). (2) The suspected mutilans group (sMu-G), which included patients showing one or two joints with instability, or with severe resorptive bone destruction without instability. There were 59 cases in this group: 56 women and 3 men, with a mean age of 59 years (range 37–86 years); (3) Larsen V group (LV-G), which included patients showing radiological finding of Larsen grade V[4] but who did not meet the criteria for the first two groups. Radiological Larsen grade V has been defined as a disappearance of the original articular surface. The patients included in LV-G have at least one such joint, but they might not always have severe resorptive bone destruction or obvious instability. There were 47 cases in this group: 42 women and 5 men, with a mean age of 64 years (range 37–85 years). (4) Control group (C-G), which included patients who had suffered from RA for more than 10 years and who did not belong to the other three groups. There were 173 cases in this group: 153 women and 20 men, with a mean age of 61 years (range 23–93 years).

Radiological evaluation

Radiological staging was based upon Steinbrocker's criteria.[13] The NJE were counted from routine radiographs of 68 joints[12] according to Ochi et al.[5] Patients found to have radiological Larsen grade V[4] were selected from standard radiographs of rheumatoid arthritis, which were offered from The Department of Radiology, Oslo Sanitetsforeining Rheumatism Hospital. The radiographs were evaluated from the time when the X-rays of 68 overall joints were taken.

Clinical data

Age, onset age of RA, duration of disease, incipient joints, and types and degree of cervical subluxation were examined in each case. The incidence of cases having fused joints was also investigated to find the number of cases in which a change in stage IV of Steinbrocker[12] in more than one joint had fused joints. Age and onset age of RA were evaluated from radiographs from the time when an X-ray photograph of 68 overall joints was taken.

The erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) used were those measured on the occurrence of a mutilating change or at maximum value under follow-up in Mu-G, and at maximum value under follow-up in other groups.

The frequencies of major surgery such as total hip arthroplasty (THA), total knee arthroplasty (TKA), and cervical operations were investigated.

Statistical analysis

Statistical analyses were carried out using Student's *t*-test and the χ^2-test. $P < 0.05$ was considered to be statistically significant.

Results

Stage and class based on Steinbrocker's classification

Among 58 cases of Mu-G, there were: no cases of stages I and II, 22 cases of stage III, and 36 cases of stage IV; no cases of classes 1 and 2, 27 cases of class 3, and 31 cases of class 4. In 59 cases of sMu-G, there were: no cases of stages

Table 1. Classification (grouping) of rheumatoid arthritis

	No. of joints with Larsen grade V radiological change	No. of joints with severe resorptive bone destruction	No. of joints with instability
Mu-G	Not important	More than 3 (more than 2 in large joints)	More than 3 (more than 2 in large joints)
sMu-G	More than 1–5	1–2	None or 1–2
LV-G	More than 1	Not important	Not important
C-G	None	None	None

Mu-G, mutilans group; sMu-G, suspected mutilans group; LV-G, Larsen V group; C-G, control group

I and II, 27 cases of stage III, and 32 cases of stage IV; no cases of class 1, 2 cases of class 2, 36 cases of class 3, and 21 cases of class 4. In 47 cases of LV-G, there were: no cases of stages I and II, 18 cases of stage III, and 29 cases of stage IV; no cases of class 1, 5 cases of class 2, 31 cases of class 3, and 11 cases of class 4. In 173 cases of C-G, there were: 2 cases of stage I, 8 cases of stage II, 97 cases of stage III, and 66 cases of stage IV; 3 cases of class 1, 52 cases of class 2, 99 cases of class 3, and 19 cases of class 4.

In all groups, the majority of cases were stages III and IV, and most were in stage IV with the exception of C-G. In the classification of class, most cases were classes 2 and 3 in C-G, and 3 and 4 in other groups.

Age, onset age of RA, and duration of disease

The mean age of patients was 59.8 ± 12.2 years (range 29–90 years) in Mu-G, 65.3 ± 10.0 years (range 37–86 years) in sMu-G, 64.0 ± 11.4 years (range 37–85 years) in LV-G, and 61.1 ± 12.7 years (range 23–93 years) in C-G. No significant differences were observed among groups except between Mu-G and sMu-G ($P < 0.05$).

The mean onset age of RA was 38.4 ± 13.2 years (range 7–69 years) in Mu-G, 45.2 ± 17.8 years (range 13–72 years) in sMu-G, 42.7 ± 12.7 years (range 20–65 years) in LV-G, and 41.9 ± 12.8 years (range 7–73 years) in C-G. Significant differences were observed between Mu-G and sMu-G ($P < 0.01$). However, no significant differences were observed between Mu-G, LV-G, and C-G.

The mean duration of RA was 22.4 ± 9.5 years (range 3.5–57.8 years) in Mu-G, 19.9 ± 9.2 years (range 2–40 years) in sMu-G, 21.1 ± 10.0 years (range 10–54 years) in LV-G, and 19.4 ± 8.7 years (range 10–54 years) in C-G. No significant differences were observed among groups except between Mu-G and C-G ($P < 0.05$).

Incipient joints of RA

In the search for incipient joints of RA, finger joints were the most frequent with 24 joints (47.1%) out of 51 cases in Mu-G, 11 joints (28.9%) out of 38 cases in LV-G, and 51 joints (38.1%) out of 134 cases in C-G. There were no significant differences among Mu-G, LV-G, and C-G. In sMu-G, finger involvement was 14 joints (26.9%) out of 52 cases, and this was significantly different from Mu-G ($P < 0.05$).

ESR and CRP

The mean ESR at an occurrence of mutilating change in Mu-G was 112.9 ± 26.1 mm/h (range 69–144 mm/h). The mean maximum value under follow-up was 45.2 ± 13.8 mm/h (range 13–72 mm/h) in sMu-G, 42.7 ± 12.3 mm/h (range 20–65 mm/h) in LV-G, and 41.9 ± 12.8 mm/h (range 7–73 mm/h) in C-G. There was a significant difference between Mu-G and all other groups ($P < 0.01$).

The mean CRP at an occurrence of mutilating change in Mu-G was 5.6 ± 2.9 mg/dl (range 2.8–11.8 mg/dl), and the mean maximum value under follow-up was 3.2 ± 3.9 mg/dl (range 0.4–14.1 mg/dl) in sMu-G, 3.1 ± 1.6 mg/dl (range 0.8–5.6 mg/dl) in LV-G, and 3.0 ± 2.1 mg/dl (range 0.5 ± 11.5 mg/dl) in C-G. The CRP value in Mu-G was significantly higher than those of other groups ($P < 0.05$).

Outbreak period of mutilating changes from the onset of RA

There were many cases in which the duration of the appearance of mutilating changes from the onset of RA was not clarified because not all cases had been observed at the same institution. However, in Mu-G, it was possible to confirm the outbreak periods in 17 out of 57 cases. In Mu-G, it was possible to specify the outbreak period of mutilating change by observing that polyarticular pains and swelling became prominent with a remarkably high value of ESR and CRP, and then destructive and resorptive bone changes were brought about which produced joint instability. This outbreak period was an average of 15.5 years after the onset of RA. The outbreak period could not be specified in sMu-G.

NJE

The mean NJE was 43.4 ± 9.3 joints (range 18–60) in Mu-G, 33.1 ± 9.7 joints (range 7–56) in sMu-G, 28.8 ± 8.7 joints (range 10–42) in LV-G, and 16.2 ± 9.7 joints (range 1–41) in C-G. In Mu-G, a significantly higher NJE was observed than in other groups ($P < 0.01$).

To observe NJE, the joints were divided into large joints such as shoulder, elbow, wrist (radiocarpal, intercarpal, and carpometacarpal joints), hip, knee, and ankle (mortice), and other small joints. In large joints, when patients with fewer than 5 NJE were compared with those having more than 6 NJE, 56 out of 58 cases, or 96.6%, had more than 6 NJE in Mu-G. Thus, the NJE of Mu-G patients was significantly high ($P < 0.01$) compared with other groups. Regarding small joints, when comparing the cases with fewer than 19 NJE with those with more than 20 NJE in Mu-G, 53 out of 58 cases, or 91%, had more than 20 NJE, and NJE in Mu-G was significantly high ($P < 0.01$) compared with other groups (Table 2).

Regarding the distribution of NJE, a C-G peak was observed in 11–15 joints (Fig. 1d), an LV-G peak in 36–40 joints (Fig. 1c), and an sMu-G peak in 31–35 joints (Fig. 1b), whereas in Mu-G the peak was in 46–50 joints (Fig. 1a) showing that NJE was much higher than in the other groups.

Subluxation of atlantoaxial joints of the cervical spine

In the atlantodental interval (ADI), which shows the index of anterior atlantoaxial subluxation of the cervical spine, there were no significant differences between Mu-G and sMu-G. However, in Mu-G, posterior subluxation was observed with a high frequency in 11 cases (19.0%), whereas

Table 2. Number of joints with erosions

		Large joints		Small joints	
		Fewer than 5	More than 6	Fewer than 19	More than 20
Mutilans group	(Mu-G)	2 cases (3.4%)	56 cases (96.6%)	5 cases (8.6%)	53 cases (91.4%)
Suspected mutilans group	(sMu-G)	7 cases (11.9%)	52 cases (88.1%)	14 cases (23.7%)	45 cases (76.3%)
Larsen V group	(LV-G)	11 cases (23.4%)	36 cases (76.6%)	16 cases (34.0%)	31 cases (66.0%)
Control group	(C-G)	105 cases (60.7%)	68 cases (39.3%)	141 cases (81.5%)	32 cases (18.5%)

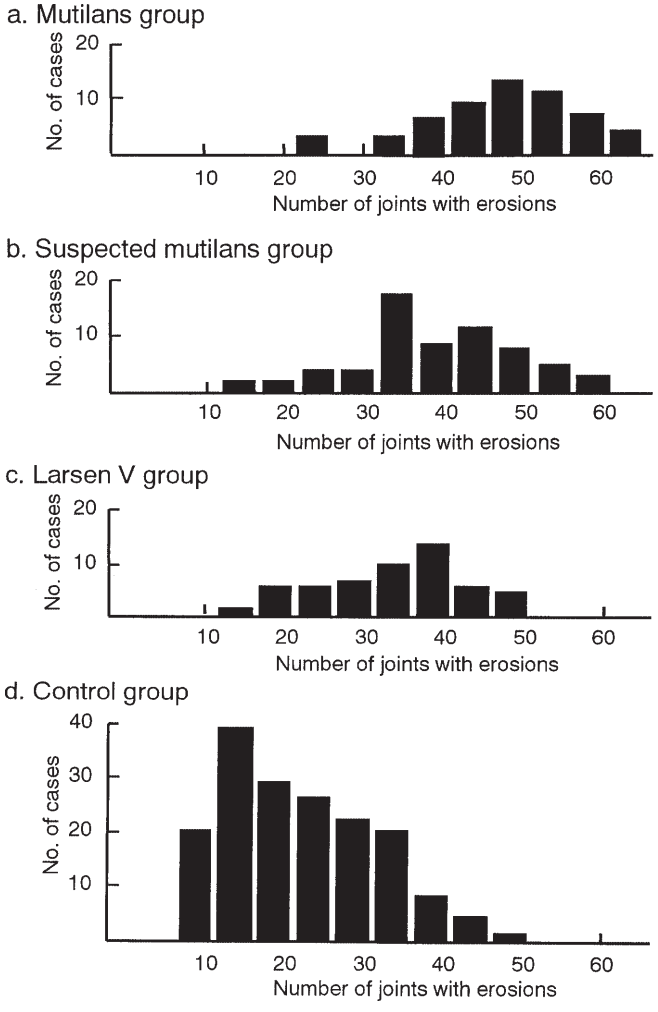

Fig. 1. Distribution of the number of joints with erosions

there were 3 cases (5.1%) in sMu-G, one case (4.2%) in LV-G, and one case (0.6%) in C-G. Thus, in Mu-G, posterior atlantoaxial subluxation was seen to have a significantly higher incidence than in other groups ($P < 0.01$) (Fig. 2a). Ranawat's perpendicular distance (PD), which indicates the grade of vertical atlantoaxial subluxation, was significantly lower, with a mean value of 6.9 mm, in Mu-G than in other groups ($P < 0.01$) (Fig. 2b).

Table 3. Incidence of cases with fused joints

Mu-G	36/58 cases	(62.1%)**
sMu-G	33/59 cases	(55.9%)*
LV-G	30/47 cases	(63.8%)**
C-G	67/173 cases	(38.7%)

χ^2-test in comparison with C-G: * $P < 0.05$; ** $P < 0.01$

Incidence of cases with fused joints

The incidence of cases with fused joints was high in Mu-G, sMu-G, and LV-G, with 62.1%, 55.9%, and 63.8%, respectively, and significantly higher than in C-G ($P < 0.01$). This indicates that the potentiality for bone formation may be preserved in patients with the mutilans type of RA (Table 3).

Incidence of cases that required surgical intervention

The frequency of cases that required surgical intervention was higher in sMu-G and LV-G (56% and 68%, respectively) than in C-G (30%). In Mu-G, it was highest at 84%.

In the itemized surgical interventions, the frequency of cervical operations was almost the same in Mu-G and sMu-G, i.e., 17 out of 58 cases (29.3%) and 14 out of 59 cases (24%), respectively, and was higher than that in other groups. The frequencies of THA and TKA were significantly higher in Mu-G (23 cases, 40%, and 31 cases, 53%, respectively) than in other groups ($P < 0.01$) (Table 4).

Comparison with cases which matched the definition of mutilans type RA according to Murasawa et al.[8]

The cases which matched the definition of mutilans type RA according to Murasawa et al., excluding those which were included in the Mu-G category, were investigated. The distribution of NJE (Fig. 3) showed two peaks at 31–35 and 41–45 joints. This pattern resembled that of sMu-G, and was different from that of Mu-G. Furthermore, NJE was the

Table 4. Operative cases

	Total cases	Cervical operation	(*P* value[a])	THA	(*P* value[a])	TKA	(*P* value[a])
Mu-G	58 cases	17 cases (29.3%)		23 cases (39.7%)		31 cases (53.4%)	
sMu-G	59 cases	14 cases (23.7%)	NS	10 cases (16.9%)	<0.01	16 cases (27.1%)	<0.01
LV-G	47 cases	2 cases (4.3%)	<0.01	4 cases (8.5%)	<0.01	12 cases (25.5%)	<0.01
C-G	173 cases	3 cases (1.7%)	<0.01	4 cases (2.3%)	<0.01	21 cases (12.1%)	<0.01
Total	337 cases	36 cases (10.7%)		41 cases (12.2%)		80 cases (23.7%)	

[a] χ^2-test in comparison with Mu-G; NS, not significant

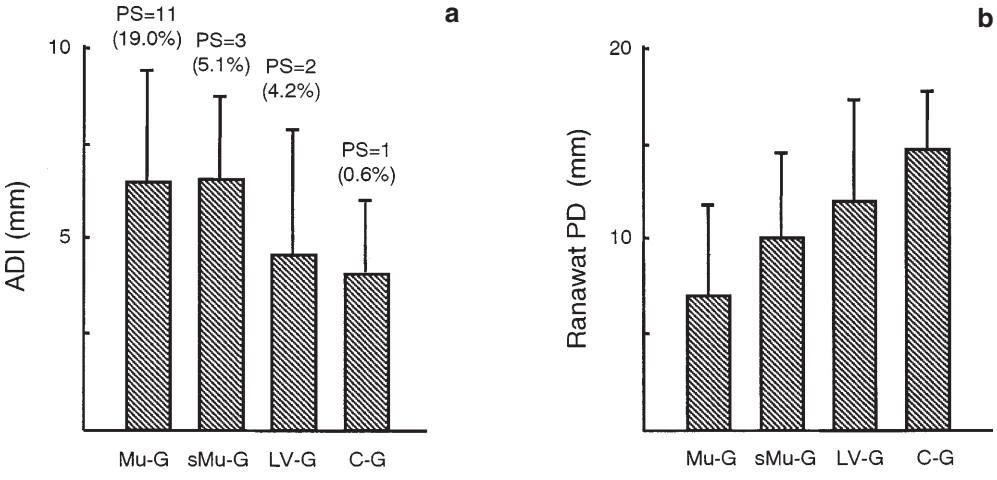

Fig. 2. Cervical (atlantoaxial) lesions. **a** Atlantodental interval (*ADI*). **b** Ranawat perpendicular distance (*PD*). *PS*, number of patients with posterior atlantoaxial subluxation (percentage in parentheses); *Mu-G*, mutilans group; *sMu-G*, suspected mutilans group; *LV-G*, Larsen V group; *C-G*, control group

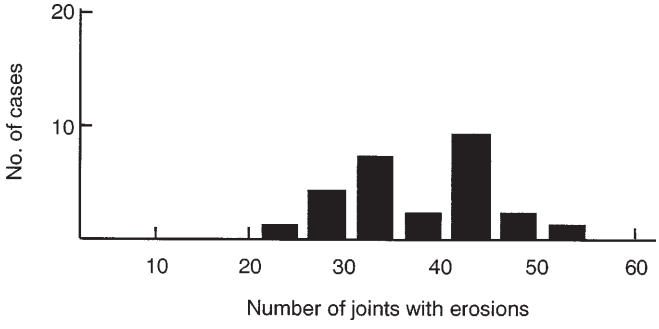

Fig. 3. Distribution of number of joints with erosions (Murasawa definition of mutilans type rheumatoid arthritis)

same as sMu-G and was significantly different from Mu-G ($P < 0.01$) (Table 5). The incidences of posterior atlantoaxial subluxation and the frequency of THA and TKA in the Murasawa definition group were lower than in Mu-G, although this was not significant, and was not significantly different from sMu-G. Many other things, such as the number of fused joints, the grade of anterior atlantoaxial subluxation, the incidence of posterior atlantoaxial subluxation, and the frequency of operative treatments, were almost same as for sMu-G and was not significantly different from sMu-G (Table 5). Therefore, the characteristic features of cases that match the definition of mutilans type RA by Murasawa et al.[8] were considered to be the same as those of sMu-G.

Rheumatoid arthritis patients who have mutilating changes in two large joints only

Cases which had mutilating changes in two large joints only were classified as Mu-G. There were four cases in this category (Table 6).

In case 1, mutilating changes occurred in the bilateral elbows (Fig. 4) in the 9 years after the onset of RA, accompanied by highly inflammatory RA activity before vertical atlantoaxial subluxation occurred. This patient died suddenly 2 years later. The cause of death was not identified, but was assumed to be a heart attack. NJE was fewer than 20, but it was presumed that there would have been many more NJE if the patient had lived longer.

Cases 2 and 3 both had many NJE and several radiological Larsen grade V joints, and also needed THA and TKA. These patients could be considered to have the characteristic features of mutilans type RA (Figs. 5 and 6).

Case 4 does not yet have as many NJE, but does have posterior atlantoaxial subluxation (Fig. 7). As only 3 years have passed since the onset of RA, an increase in NJE could be expected. Therefore this patient was considered to be in the active stage of mutilating type RA.

Incidence of mutilans type RA

Where Mu-G was considered to be mutilans type RA, it occurred in 58 out of 337 cases, or 17.2%, of those who had suffered from RA for more than 10 years. If sMu-G was

Table 5. Comparison of Murasawa's definition of mutilans type rheumatoid arthritis (excluded mutilans group) with mutilans, suspected mutilans, Larsen V, and a control group

	No. of joints with erosions			No. of cases with fused joints	Cervical spine			No. of operative cases		
	Large joints	Small joints	Total joints		ADI	No. of posterior AA subluxation	Ranawat PD	Cervical spine	THA	TKA
Murasawa definition of the mutilans group	9.2 ± 2.4[a]	27.9 ± 5.6[a]	36.6 ± 7.4[a]	17/26 (65.4%)	7.0 ± 4.0[a]	3/26 (11.5%)	10.2 ± 4.4[a]	6/26 (23.1%)	5/26 (19.2%)	10/26 (27.8%)
Mutilans group	10.4 ± 2.1[a] (NS)[b]	32.8 ± 8.6[a] (<0.01)[b]	43.4 ± 9.3 (<0.01)[b]	36/58 (62.1%) (NS)[c]	6.5 ± 3.3[a] (NS)[b]	11/58 (19.0%) (NS)[c]	6.9 ± 5.3[a] (NS)[b]	17/58 (29.3%) (NS)[c]	23/58 (39.7%) (NS)[c]	31/58 (53.8%) (NS)[c]
Suspected mutilans group	8.8 ± 2.7[a] (NS)[b]	24.0 ± 8.0[a] (NS)[b]	33.1 ± 9.7[a] (NS)[b]	33/59 (55.9%) (NS)[c]	6.6 ± 3.3[a] (NS)[b]	3/59 (5.1%) (NS)[c]	10.1 ± 4.7[a] (NS)[b]	14/59 (23.7%) (NS)[c]	10/59 (16.9%) (NS)[c]	16/59 (27.1%) (NS)[c]
Larsen V group	7.6 ± 2.6[a] (NS)[b]	21.4 ± 7.5[a] (<0.01)[b]	28.8 ± 8.7[a] (<0.01)[b]	30/47 (63.8%) (NS)[c]	9.1 ± 5.1[a] (NS)[b]	2/47 (4.2%) (NS)[c]	12.6 ± 3.5[a] (NS)[b]	2/47 (4.3%) (<0.05)[c]	4/47 (8.5%) (NS)[c]	12/47 (25.5%) (NS)[c]
Control group	5.1 ± 2.9[a] (<0.01)[b]	11.7 ± 8.3[a] (<0.01)[b]	16.2 ± 9.7[a] (<0.01)[b]	67/173 (38.7%) (<0.05)[c]	4.1 ± 2.9[a] (<0.01)[b]	1/173 (0.6%) (<0.01)[c]	14.7 ± 3.4[a] (<0.01)[b]	3/173 (1.7%) (<0.01)[c]	4/173 (2.3%) (<0.01)[c]	21/173 (12.1%) (<0.01)[c]

[a] Standard deviation
[b] *t*-test with the Murasawa definition of mutilans type
[c] χ^2-test with the Murasawa definition of mutilans type
ADI, atlantodental interval; AA subluxation, atlantoaxial subluxation; Ranawat PD, Ranawat perpendicular distance; THA, total hip arthroplasty; TKA, total knee arthroplasty; NS, not significant

Table 6. Rheumatoid arthritis patients who have mutilating changes in two large joints only

Case	Sex	Age (years)	Duration of disease (years)	Stage	Class	Ankylosed joints	NJE			LV			Mutilating joints	Onset of mutilating change (YA)	Cervical spine			Operation
							Large joints	Small joints	Total	Large joints	Small joints	Total			ADI	PD	PAAS	
1	F	44	11	IV	3	2	4	15	20	2	1	3	Bilateral elbows	9	8	6	−	Synovectomy of right elbow, bilateral wrists
2	F	67	11	III	3	0	12	41	53	5	0	5	Bilateral elbows	9	5	9	−	Bilateral TKA
3	F	72	18	IV	3	5	12	37	49	5	11	16	Bilateral elbows	15	8	10	−	Bilateral THA, bilateral TKA, synovectomy of right wrist
4	F	82	25	III	3	0	7	25	32	4	2	6	Right elbow, left wrist	22	−	12	+	Cervical spine, right FHA

NJE, number of joints with erosions; LV, radiological finding of Larsen grade V; YA, years after the onset of rheumatoid arthritis; ADI, atlantodental interval; PD, perpendicular distance (Ranawat); PAAS, posterior atlantoaxial subluxation; TKA, total knee arthroplasty; THA, total hip arthroplasty; FHA, femoral head arthroplasty

Fig. 4. X-rays of elbows showing mutilating changes in case 1. **a** Right elbow. **b** Left elbow

Fig. 5. X-rays of elbows showing mutilating changes in case 2. **a** Right elbow. **b** Left elbow

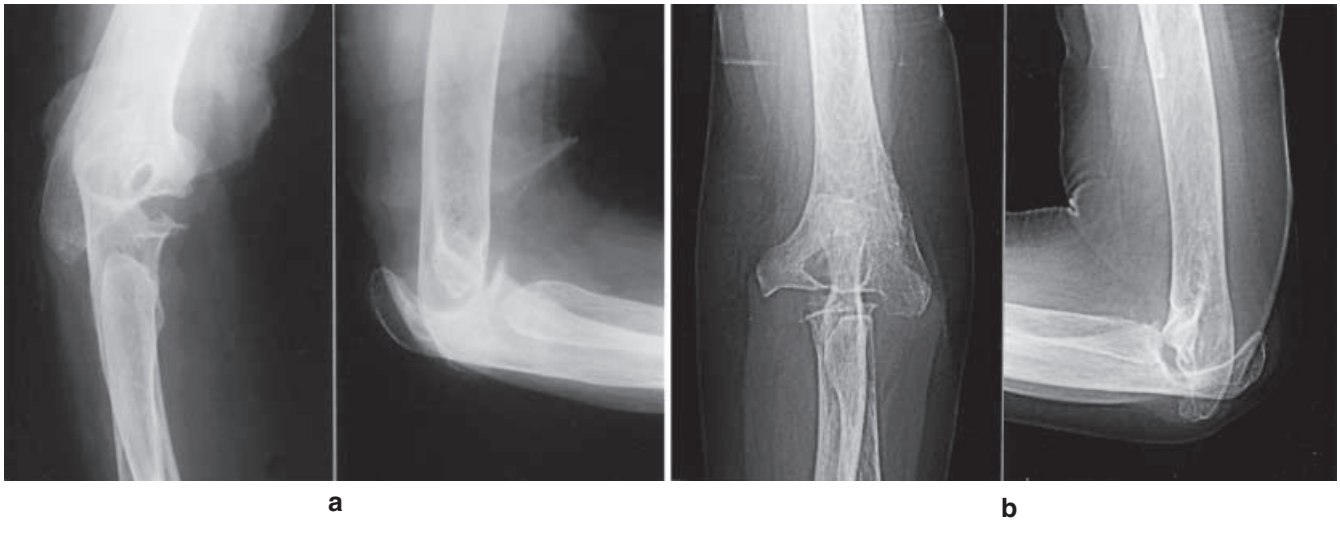

Fig. 6. X-rays of elbows showing mutilating changes in case 3. **a** Right elbow. **b** Left elbow

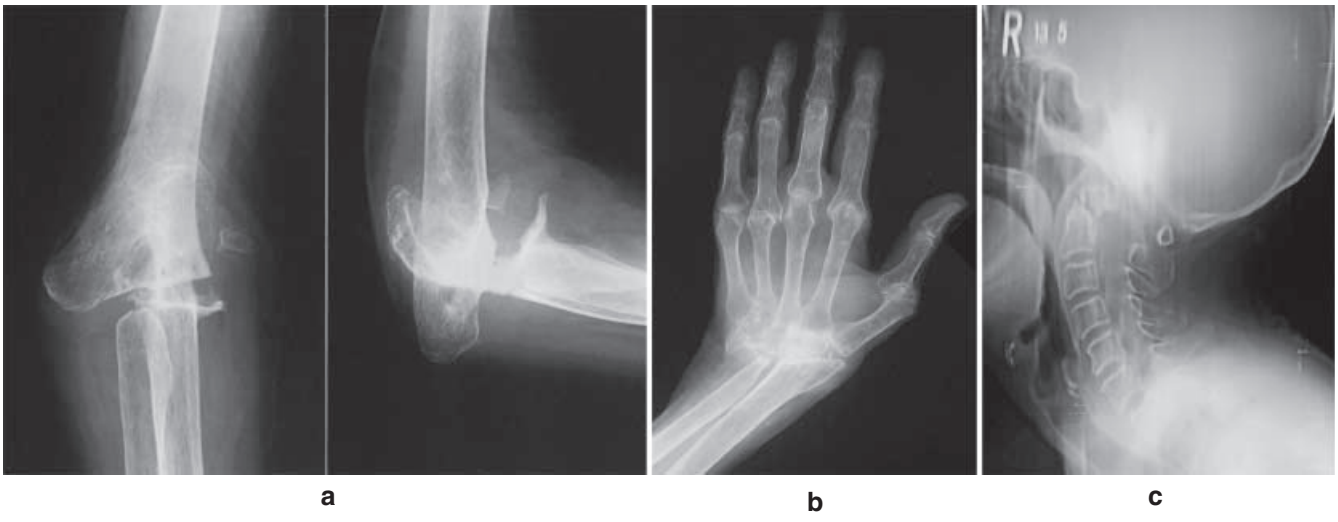

Fig. 7. X-rays of right elbow and left wrist showing mutilating changes, and tomography of the cervical spine showing posterior atlantoaxial subluxation in case 4. **a** Mutilating change of right elbow. **b** Mutilating change of left wrist. **c** Posterior atlantoaxial subluxation of the cervical spine

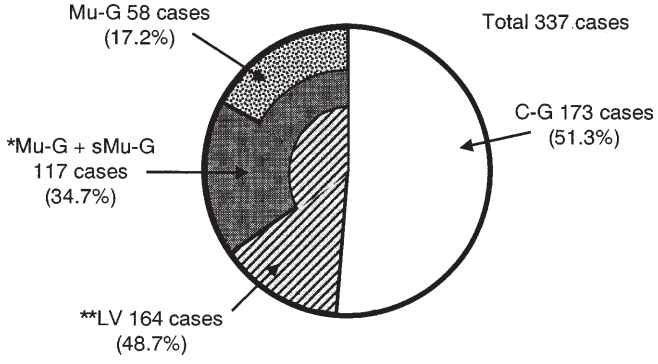

Fig. 8. Incidence of mutilating type RA according to the definition of patients who have suffered from RA for more than 10 years. **Mu-G + sMu-G*, RA patients who had features of both sMu-G and Mu-G. ***LV*, RA patients showing Larsen grade V symptoms (LV-G + sMu-G + Mu-G)

included, there were 117 cases, or 34.7%. Furthermore, if mutilans type RA included LV-G, there were 164 cases, i.e., 48.7%. Thus, the incidence of mutilans type RA varied considerably according to its definition (Fig. 8).

Discussion

Arthritis mutilans[3] that has the characteristic feature of high-grade osteolytic arthritis in fingers and toes has been observed in other rheumatic diseases such as psoriatic arthritis,[14,15] ankylosing spondylitis,[16] progressive systemic sclerosis,[17] and mixed connective tissue disease,[18] infectious diseases such as tuberculosis and osteomyelitis, and chronically lasting arthritis-induced diseases such as neuropathic arthropathy and the unknown cause of multicentric reticulohistiocytosis.[19]

It is not clear when arthritis mutilans in RA patients was first called mutilans type RA, but it was first described by Ishikawa et al.[20] Larsen et al.[4] defined the grade V changes in the radiological classification as a mutilating abnormality. Marie and Leri[1] described a 66-year-old woman with RA who showed marked bone resorption in the wrists, carpals, and metacarpals, which had an opera-glass-like appearance. They also mentioned another patient with a similar deformity which was limited to one finger. Stursberg[2] stressed multiple involvement and called it polyarthritis mutilans. Mather[3] also stressed multiple involvement by proposing the term arthritis mutilans. Ochi et al.[5–7] proposed the name MUD as a disease subset of RA for the type of RA in which the NJE increases rapidly to approximately 50 or more during the first 10 years. However, returning to the starting point, it is now necessary to review the definition of mutilans type RA. As a likely definition, Murasawa et al.[8] described it as cases that have radiological changes of Larsen grade V in more than three joints of the MP and interphalangeal (IP) joints of the fingers and toes. Ryu et al.[9] defined it as RA which results in characteristic deformities and destruction of the joints, such as opera-glass deformity of fingers or severe resorptive bone destruction in more than three joints.

If no clear agreed definition of mutilans type RA exists, then it is not possible to reach a satisfactory diagnosis. Mutilans type RA is the most intractable type, and has the poorest prognosis for joint function in RA. The results of this study showed that ESR and CRP values were considerably elevated, and that there exists a group in which operative frequency and frequency of cervical lesions and their associated problems are characteristic, and that the clinical picture is rather different from the normal definition.

There are clearly several problems about a definition. The concept of MUD proposed by Ochi et al. is an outstanding definition of a subset of the disease. However, as it is likely that many cases do not have mutilating changes as we understand them in this category, it is doubtful whether they can be considered to have the same rank as the mutilating type RA. The classification of Larsen et al.[4] is only based

on radiological findings, and many cases in this group have no instability.

The main factor of mutilans type RA is the deterioration of all the joint tissues, e.g., high-grade resorptive bone destruction and marked instability. Furthermore, it is necessary to consider the systemic factor. How many deteriorated joints should there be for such a diagnosis? Is a single deteriorated joint sufficient to merit the label of mutilans type RA or not? If not, how many of these joints should be involved? How should one manage the cases that have marked resorptive bone destruction without instability? Clearly, there are several questions to be answered. Comparing Mu-G, which has more than three joints (two joints in the case of large joints) having the two main factors, with LV-G, which shows a radiologically high grade of bone destruction, Mu-G showed much more NJE, the ADI of the atlantoaxial joint was larger, Ranawat's PD value was lower, and the incidence of anterior and vertical subluxation was higher. Also, the frequency of major surgery such as cervical operations, THA, and TKA was high in Mu-G. Thus, Mu-G and LV-G clearly show different features.

In the case of large joints only, the reason why just cases with more than two affected joints were included in Mu-G was that in those who were affected in one elbow joint only, it remained localized in that single joint with no marked resorptive bone destruction of fingers or opera-glass hand deformity, and the radiological grade remained at grade V without marked osteolytic changes or instability. However, some of the cases that were affected in two large joints only later began to show opera-glass hand deformity. These cases were compatible with the Mu-G category in other features. Thus, these cases might be very likely to show polyarticular mutilating changes in the future. Cases that had no definite joint instability, even though they had a high level of destructive bone changes such as radiological Larsen grade V changes, were classified as sMu-G. Comparing sMu-G with Mu-G, no significant differences were observed in age, duration of RA affection, stage, class, incidence of cases with ankylosed joints, incidence and degree of anterior subluxation, or frequency of cervical operations. However, in Mu-G, the age of onset of mutilating change was lower, NJE was much greater, the incidence of vertical and posterior atlantoaxial subluxation was higher, and the frequencies of THA and TKA were higher than in sMu-G. These all showed significant differences. The number of joints with radiological Larsen grade V was also significantly much higher in Mu-G than in sMu-G. In the majority of Mu-G cases, the onset of mutilating change could be clearly indicated, but it was difficult to specify the onset time of the mutilating changes in sMu-G.

From the above findings, some discrete, characteristic features of Mu-G and sMu-G were observed. Therefore, Mu-G should be known as mutilans type RA. From these points of view, it could be said that the onset of mutilans type RA was approximately 15 years after the onset of RA. For this reason, RA patients who had been affected for more than 10 years were investigated. This showed that the incidence of mutilating type RA was 58 out of 337 cases, or 17.2%.

Mutilans type RA has many characteristic features, as described by Murasawa et al.[8,21] In general, there are three stages in the disease process of mutilans type RA. In the early stage, a generally high degree of inflammatory symptoms, which are resistant to medical treatment, continue for 2–3 years from approximately 10 years after the onset of RA. In the next active stage, or intermediate stage, a high degree of bone resorptive destruction along with marked joint instability becomes evident, cervical changes progress, and complications increase.[22] After the active stage has continued for 3–4 years, osteolytic changes and instability are complete at the start of the late stage. In the late stage, the inflammation subsides, but the activities of daily living continue to decline. The cases that followed this course were only those with Mu-G. The distinction between the stages was not clear in sMu-G, and the clinical features were quite different from those of Mu-G. However, it could be considered that some of the sMu-G cases might progress to Mu-G.

It has been shown that mutilans type RA, as hitherto defined, has been confused with various other conditions of the RA disease state, and that many cases who had no characteristic features of mutilans type RA had received this diagnosis. As a result of this study, it is strongly suggested that the most intractable cases of RA are Mu-G, and that patients in other groups have a relatively better prognosis. Therefore, an accurate definition of mutilans type RA is essential when establishing a therapeutic plan. The main factors of mutilans type RA are severe resorptive bone destruction and instability in many joints. From this investigation, I arrived at the following definition of mutilans type of RA, i.e., RA patients showing severe resorptive bone destruction and instability such as opera-glass hands in three or more joints, or, if the large joints are affected, two joints are sufficient.

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