Neurological improvement and prognosis after occipito-thoracic fusion in patients with mutilating-type rheumatoid arthritis

Tetsu Tanouchi · Takachika Shimizu ·
Keisuke Fueki · Masatake Ino · Naofumi Toda ·
Yasunori Tatara · Nodoka Manabe

Received: 21 February 2012 / Revised: 28 June 2012 / Accepted: 14 July 2012 / Published online: 27 July 2012
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Abstract

Introduction Mutilating-type rheumatoid arthritis, the most aggressive type of rheumatoid arthritis, is frequently associated with destructive cervical involvement, both at the high-cervical and subaxial levels, causing significant neurological deficit, and their natural course of the disease and the survival are discouraging. For such cases, we have been actively performing occipito-thoracic fusion since 1991. Although medical treatment for rheumatoid patients has represented a marked improvement, it could not treat all of these patients because of several reasons. Therefore, it is still important to evaluate the past treatment results.

Methods We investigated the neurological improvement and prognosis in 51 mutilating-type rheumatoid arthritis patients who underwent occipito-thoracic fusion between 1991 and 2010. The neurological status was evaluated using modified Ranawat classification; class IIIB was subdivided into IIIBa (able to sit upright) and IIIBb (bedridden).

Results The preoperative neurologic status was IIIBa in 19 patients and IIIBb in 17 patients. 15 of the 19 patients with class IIIBa improved to being able to walk (79 %), whereas only 3 of the 17 patients with class IIIBb improved to being able to walk (18 %) after surgery. Of the 51 patients, 28 died during follow-up; the mean age at death was 67.2 years. The postoperative 5- and 10-year survival rates were 60.3 and 26.4 %, respectively.

Conclusion The neurological improvement and prognosis after surgery was poorer in class IIIBb patients than in the

other patient groups. Occipito-thoracic fusion can improve the neurological symptoms and prognosis. However, early surgical intervention is recommended, before a patient becomes bedridden (class IIIBb).

Keywords Rheumatoid arthritis · Mutilating · Cervical spine · Occipito-thoracic fusion · Prognosis

Introduction

The cervical spine is frequently affected in rheumatoid arthritis (RA), and cervical involvement may begin with atlanto-axial subluxation (AAS) and progress to vertical subluxation (VS) and/or subaxial subluxation (SAS). The natural history of rheumatoid cervical disorders is variable.

Ochi et al. [1] subdivided RA patients into three groups using the number of joints with erosion: the subset with least erosive disease (LES), the subset with more erosive disease (MES), and the subset with mutilating disease (MUD). Mutilating-type RA (MUD), the most aggressive form of RA, is frequently associated with severe cervical involvement, both at the cranio-vertebral junction (CVJ) and subaxial levels, causing significant neurological deficit. The natural course of the disease and survival in patients with this type of RA are discouraging [2, 3]. Because of the dynamic change of the underlying rheumatoid arthritis control medicine, we rarely encounter severely affected rheumatoid patients like those affected by mutilating-type. But even now, steroid is only medicine to control in some patients who cannot be treated by biological products due to its side effects or other medico-social reasons. Therefore, it is still important to summarize and re-recognize the past treatment results of destructive cervical disorders in mutilating patients.

T. Tanouchi (✉) · T. Shimizu · K. Fueki · M. Ino · N. Toda ·
Y. Tatara · N. Manabe
Department of Orthopedic Surgery, Gunma Spine Center
(Harunaso Hospital), 878-1 Kamitoyooka, Takasaki,
Gunma 370-0871, Japan
e-mail: teztano@gmail.com

We have been actively performing occipito-thoracic fusion for such cases since 1991. Most of these patients were assessed preoperatively as having Ranawat IIIB myelopathy. Casey et al. [4] reported that they do not recommend surgery for class IIIB patient. However, we believed that class IIIB patients require detailed neurological assessment before determining the indication for surgery. We, therefore, ‘assessed the neurological status according to the modified Ranawat scale [5, 6]. In this scale, the authors subdivided Ranawat IIIB into two categories, IIIBa (unable to walk, but able to sit upright) and IIIBb (unable to sit upright, bedridden) as shown in Table 1. Matsuyama et al. [3] reported a similar classification.

Materials and methods

Patients

Between 1991 and 2010, 51 patients with mutilating-type RA (48 females, 3 males) underwent occipito-thoracic fusion. The mean age of the patients at the time of surgery was 62.5 years (37–78 years). The duration of RA was, on average, 22.5 years (3–51 years). The main clinical finding in these patients was either neurological deficit due to the spinal cord compression or intractable neck-occipital pain. The postoperative follow-up period was 58.0 months (1–187 months). Cervical involvement was diagnosed by plain cervical radiographs, computed tomographic myelogram (CTM), and magnetic resonance imaging (MRI). Eight had AAS + VS, 4 had AAS + SAS, 34 had AAS + VS + SAS, and 4 had SAS + ankylosis at CVJ. One patient previously underwent atlanto-axial fusion because of AAS. Although solid fusion was achieved after atlanto-axial fusion, SAS and VS occurred. The patient characteristics are summarized in Table 2.

Surgical procedure

Preoperatively, the majority of these patients received intermittent halo-traction on the bed for a period of few

Table 2 Patient characteristics ($n = 51$)

Gender	3 males 48 females
Age at surgery	62.5 years (37–78 years)
RA disease duration	22.5 years (3–51 years)
Follow up	58 months (1–187 months)
Cervical lesions	AAS + VS: 8, AAS + SAS: 4, AAS + VS + SAS: 34, SAS + ankylosis at CVJ: 4, Salvage case: 1

AAS atlanto-axial subluxation, VS vertical subluxation, SAS subaxial subluxation, CVJ cranio vertebral junction

days to 2 weeks, to obtain optimal cervical alignment and thereby, pain relief and improvement of the neurological deficit. During the operation, partial posterior decompression by laminectomy and complete resection of inflammatory degenerative ligamentum flava were performed in cases with local stenosis. This procedure was then supplemented with posterior spinal fixation using various instrumentations. Basically, fusion area was from the occiput to T5 in most patients, because we believe that the spinal region around T5 is the most reliable foundation to rebuild up, even in such patients with a fragile spine. Instrumentations and fusion area are shown in Table 3. Additional anterior decompression and fusion was not performed in any of the cases. Autogeneous bone harvested from posterior iliac crest mixed with hydroxylapatite was grafted in whole instrumentation area. Postoperatively, a SOMI brace was applied for 4–6 months.

Clinical evaluation

The neurological status and survival were investigated in the study subjects. Evaluation of the neurological status was performed before the surgery, 6 months after the surgery, and at 2- and 5-year follow-up period. Survival was determined by the Kaplan–Meier method and significance was ascertained by the log-rank test. All values were expressed as means with the 95 % confidence intervals, unless specified otherwise.

Results

Neurological status

The preoperative neurological status in the 51 patients was distributed as follows: class II (1 patient), class IIIA (14 patients), class IIIBa (19 patients), and class IIIBb (17 patients); 36 patients were non-ambulant (IIIB). With regard to the postoperative 6 months, 15 of the 19 IIIBa patients improved to being able to walk (79 %), whereas only 3 of the 17 IIIBb patients improved to being able to

Table 1 Modified Ranawat classification

Class	Neurological status
Class I	No neurologic deficit
Class II	Subjective weakness, hyperreflexia, and dysesthesia
Class IIIA	Objective weakness, long tract sign Ambulatory
Class IIIBa	Objective weakness, long-tract signs Non-ambulatory, but ability to sit upright
Class IIIBb	Objective weakness, long-tract signs Non-ambulatory, inability to sit upright, bedridden

walk (17 %); this difference was highly significant ($P < 0.05$). All of the IIIBb patients could sit upright after surgery, except for two patients who died within a month of the surgery. Changing in neurological status after surgery was shown in Table 4.

At 2-year follow-up, 14 patients had died and we evaluated 33 patients. 31 of them maintained their neurological improvement and activities of daily life; however, in the remaining two patients, their ability deteriorated because of the aggravation of systemic joint lesions.

At 5-year follow-up, 19 patients had died and we evaluated 23 patients. 16 of them maintained their activities; however, in the remaining seven patients their ability deteriorated because of joint lesions or trauma: 1 with femoral fracture, 1 with pelvic fracture, 1 with multiple spine fracture, 1 with limb amputation with arteriosclerosis obliterans, and 3 with prosthetic knee infection. Limb amputation was performed in two patients with prosthetic knee infection, and three patients underwent other spine surgery. Long-term follow-up of physical activities is shown in Table 5.

Survival analysis

None of the patients died during the operation in this study. However, two patients (3.9 %) died within 1 month after surgery and both patients were class IIIBb. These cases were not related to surgery directly and the causes of death were postoperative extubation trouble and cerebral infarction due to cardiac dysrhythmia in one case each. 28 patients (54.9 %) had died by the time of the last follow-

up. The mean age at death was 67.2 years (49–83 years). The survival rate was 60.3 % at 5 years after the surgery, and 26.4 % at 10 years, as calculated statistically by the Kaplan–Meier method. The mortality was 37 % at 5 years and 53 % at 10 years. The survival curve is shown in Fig. 1.

At the same time, the survival rate was compared for three different preoperative neurological statuses. Among the 28 patients who died by the time of the final follow-up, 6 patients had been classified as IIIA, 6 patients as IIIBa, and 15 patients as IIIBb. The survival rate at 5 years after the surgery of the IIIA patients was 69 % and that of the class IIIBa patients was 74 %, while that of the class IIIBb patients was only 41 %. Thus, the survival rate of the class IIIBb patients was significantly poorer than that of the other classes of patients (log-rank test, $P < 0.05$). The three survival curves are shown in Fig. 2.

The causes of death were respiratory involvement in seven cases, sepsis in four cases, gastrointestinal bleeding in two cases, heart disease in three cases, cerebral infarction in two cases, lung cancer in one case, acute myelogenous leukemia in one case, accident death in one case, and sudden death of unknown cause in seven cases.

Case report (Fig. 3)

A 69-year-old woman had typical mutilating-type rheumatoid joint involvements. She had suffered from RA for 36 years. The patient could not sit upright due to neck-occipital pain and the neurological deficit. A lateral-view

Table 3 Implants and fusion area ($n = 51$)

Instrumentations	Fusion area (from occiput to)								Total
	T1	T2	T3	T4	T5	T6	T7	L1	
Fan shaped rod	2	1	2	1	–	–	–	–	6
ISOLA pre-bend rod	–	–	1	8	6	2	–	–	17
RRS loop spine system	1	2	1	1	19	2	1	1	28
Total	3	3	4	10	25	4	1	1	51

Table 4 Change in neurologic status after surgery (modified Ranawat classification)

Pre-op.	Post-op. 6 months					Total
	Class II	Class IIIA	Class IIIBa	Class IIIBb	Dead	
Class II	1	–	–	–	–	1
Class IIIA	–	14	–	–	–	14
Class IIIBa	1	14	4	–	–	19
Class IIIBb	–	3	10	–	4	17
Post-op. total	2	31	14	0	4	51

Table 5 Long-term follow-up of physical activities

	Pre-op.	Post-op. 6 months	Post-op.2 years	Post-op. 5 years
Class II	1	2	1	–
Class IIIA	14	31	22	11
Class IIIBa	19	14	10	9
Class IIIBb	17	0	0	3
Total	51	47	33	23
Dead	–	4	10	19

Fig. 1 Overall survival rate of the patients who underwent occipito-thoracic fusion. The survival rate was 60.3 % at 5 years after surgery and 26.4 % at 10 years after surgery **Fig. 2** Survival rate by the preoperative neurological status. The white triangle represents the survival rate of the 14 patients with class IIIA myelopathy. The white circle represents the survival rate of the 19 patients with class IIIBa myelopathy. The black circle represents the survival rate of the 17 patients with class IIIBb myelopathy. The survival rate of the class IIIBb patients was significantly poorer than that of the other classes of patients

of the cervical spine X-ray showed anterior subluxation at C2/3 and AAS + VS at CVJ, with spontaneous fusion at C3-7. Cervical cord compression at C2/3 was noted on MRI.

Occipital fusion to the fifth thoracic vertebra was performed using the RRS loop spine system. The neurologic status improved to IIIBa, with the patient regaining ability to sit upright, and she was discharged from the hospital

without any complication 2 months after the surgery. However, she died 4 months after the surgery because of gastrointestinal tract hemorrhage.

Discussion

Various surgical methods have been reported to manage cervical lesions in RA patients. However, there are few reports of surgical intervention in mutilating-type RA patients who show deteriorating cervical spine lesions, severe neurological deficits, and poor general condition.

In this series, preoperative neurological deficits in the patients were very severe because the neurological status was assessed as Ranawat class IIIB (non-ambulatory patients) in 36 of 51 patients (71 %). After the surgery, half of IIIB patients improved to being able to walk and maintained their ability for a long time before having aggravation of systematic joint lesions or major trauma. It seems that occipito-thoracic fusion gave satisfactory long-term clinical results.

Many researchers have discussed the prognosis of RA patients with myelopathy. Some reports have discussed the prognosis of RA patients with myelopathy treated conservatively [2, 7–10]. Marks and Sharp et al. [7] reported that 12 of the 20 patients died during a 6-month follow-up period, Matsunaga et al. [8] report that all became bed-ridden and died within 8 years, and Omura and Mori [2, 9] reported that the survival rate at 6.2 years was 0 %. These reports suggest that RA patients with myelopathy treated conservatively have a poor prognosis.

Some reports have discussed the prognosis of patients treated with surgical intervention. Grob et al. [11] reported that 36 % patients died within 3.5 years after OC fusion. Hirano et al. [12] reported that 27 % patients treated by OT fusion (for MES and MUD) died during follow-up (36.2 Mo). Matsunaga et al. [8] reported an actual survival rate of 37 % after OC fusion at 10 years, and Mori et al. [9] reported a survival rate of 27 % at 6.2 years; the mean age at death in the two reports was 71 and 68 years, respectively.

Fig. 3 A typical mutilating-type RA patient with cervical myelopathy (IIIBb). Mutilating-type joint involvements (**a** shoulder, **b** elbow, **c** hand). Destructive cervical spine involvements (**d** X-ray, **e** MRI, **f**, **g**

Reconstruction views of CT). She underwent O-T6 fusion. Postoperative X-ray (**h,i**)

It has been well established that the life expectancy of mutilating-type RA patients can be expected to be shorter than that of the general RA population [13–15]. Unlike other previous reports, we evaluated prognosis for limited mutilating-type RA patients (MUD) with the exception of other mild RA patients (MES or LES). Nevertheless, the actual survival rate (60.7 % at 5 years) and the mean age at death (67.4 years) were similar to other reports of surgically treated patients [8, 9, 11–13]. This finding suggests that occipito-thoracic fusion may contribute to better survival of mutilating-type RA patients.

Casey et al. [4] predicted a poor prognosis following surgical interventions in Ranawat IIIB patients (non-ambulatory), because of the limited prospect for functional recovery. However, we found that the neurological improvement and prognosis of Ranawat IIIBa patients were good and similar to those of class IIIA patients. The present study revealed that even among patients with class IIIB myelopathy, those that can still sit upright (IIIBa), surgical treatment may improve the neurological deficit and also the prognosis. We, therefore, recommend early surgical intervention in such patients, before the disease progresses in these patients to modified Ranawat class IIIBb. The study revealed a significant difference in improvement of the neurological status and prognosis between class IIIBa and class IIIBb patients. This finding suggests that the modified Ranawat classification is useful for predicting neurological recovery and prognosis in RA patients with severe destructive cervical lesions.

Conflict of interest None.

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