Is extensive cervical laminoplasty an effective treatment for spinal cord sarcoidosis combined with cervical spondylosis?

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Abstract Patients with neurosarcoidosis are usually initially treated with steroid administration even when they have concomitant cord compression on magnetic resonance imaging (MRI). Operative intervention may be indicated in patients with spinal cord sarcoidosis requiring either tissue biopsy for diagnosis or associated with progressive neurologic symptoms. However, there have been no previous reports describing clinical outcomes of laminoplasty for spinal cord sarcoidosis. The objectives of this study are to investigate whether extensive cervical laminoplasty is an effective treatment for spinal cord sarcoidosis combined with spondylotic changes and/or cervical spinal canal stenosis. Open-door laminoplasty was performed in three patients with spinal cord sarcoidosis. All patients received intensive corticosteroid therapy after the operation. MRI imaging was performed in all patients before and after the operation. Operative outcomes were not satisfactory and the clinical courses of the patients fluctuated after corticosteroid therapy. Daily life activities were not significantly

improved after treatments in any of the three patients, and in the long-term follow-up period the clinical course of one patient was one of inexorable deterioration to a state of quadriplegia. The possibility of spinal cord sarcoidosis should be included in the differential diagnosis, when a distinct high signal intensity area is observed within the spinal cord on T2-weighted MR images in patients with spondylotic changes. Laminoplasty is not an effective intervention for the treatment of spinal cord sarcoidosis even when patients have spondylotic changes and/or a constitutionally narrowing cervical spinal canal. Patients with neurosarcoidosis should be treated first with steroid administration even when they have concomitant cord compression on MRI.

Keywords Sarcoidosis · Spinal cord · Cervical spine · Laminoplasty · Spondylosis

Introduction

Sarcoidosis is a multisystem, non-caseous granulomatous disease of unknown cause. Central nervous system involvement occurs in only about 5% of patients [28], but when sarcoidosis affects the nervous system, it most commonly involves the brain and peripheral nerves. About 6–8% of cases with central nervous system involvement show spinal cord lesions [10, 23]. Spinal cord involvement is rare, occurring in only 0.3–0.4% of patients with systemic sarcoidosis [1, 26]. Thus, intramedullary neurosarcoidosis is a rare neurological manifestation, especially as an initial presentation [10]. Neurological symptoms are usually myelopathy and focal weakness. Patients usually present with paraparesis, quadraparesis, hypesthesia, as well as bowel, bladder, and sexual dysfunction [7, 19], symptoms

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which are often also observed in patients with cervical spondylotic myelopathy.

Magnetic resonance imaging (MRI) is the most useful technique for detecting neurosarcoidosis [4, 11, 12, 20, 21]. Commonly, MRI of spinal cord sarcoidosis shows spinal cord swelling on T1-weighted images and similar high intensity areas on T2-weighted images [10, 15, 22, 25]. When spondylotic changes and developmental spinal stenosis are coexistent at the same portion of the spinal cord is sometimes difficult for surgeons to make a differential diagnosis from cervical spondylotic myelopathy [18, 24, 29].

Management of spinal cord sarcoidosis remains controversial but corticosteroid therapy has been shown to improve the neurological status and suppress inflammation and granuloma formation [19]. Operative intervention may be indicated when tissue biopsy is necessary for diagnosis or sarcoidosis is associated with progressive neurological symptoms [11]. When cervical spondylosis is combined with spinal sarcoidosis causing myelopathy, operative decompression by laminoplasty might be indicated. However, there have been no previous reports describing the clinical outcomes of laminoplasty in such cases. This paper presents three patients with spinal cord sarcoidosis combined with cervical spondylosis and describes the clinical outcomes of treatment with open-door laminoplasty combined with steroid therapy.

Case report

Case 1

A 56-year-old woman first noticed numbness in her bilateral hands 6 months before admission to our institution.

She developed a progressive weakness in her lower extremities and a gait disturbance 1 month later followed by urination difficulties. She was admitted to our hospital in January 2003.

Neurological examination revealed decrease in light touch and pin prick sensation in both the upper and lower extremities. Deep tendon reflexes were hyperactive in both the upper and lower extremities. A manual muscle strength test of the lower extremities was completely unremarkable, while that of the flexor and abductor of the right fingers decreased to 3/5. Neurological examinations were otherwise normal. The angiotensin-converting enzyme (ACE) level was within the normal range.

Plain radiograph of the cervical spine showed spondylotic changes at C4/5, 5/6 levels. A T2-weighted MR image showed a developmental canal stenosis and a hyperintense intramedullary signal extending from C4 to C6 (Fig. 1a).

The patient was diagnosed with cervical spondylotic myelopathy and open door laminoplasty was performed, because spondylotic changes were observed in the same area as the T2-weighted MR image revealed high signal intensity. After the operation, the patient complained of spasm at the right upper and lower extremities. Subsequent MRI revealed enlargement of the high signal intensity area on the T2-weighted image (Fig. 1b). However, no space occupying lesion such as epidural hematoma was observed. Gadolinium DTPA (Gd)-enhanced T1-weighted MR images showed a spotty enhancement at the C5/6 level (Fig. 2a). Intravenous methylprednisolone (8,480 mg/day) was administered according to the NASCIS (National Acute Spinal Cord Injury Study) regimen, because spinal cord injury was suspected.

Enlarged bilateral hilar lymph nodes were detected by chest radiography and computed tomography (CT). The

Fig. 1 Magnetic resonance images from Patient 1. **a** Sagittal and axial T2-weighted MR image show developmental canal stenosis and hyperintense intramedullary signal extending from C4 to C6. **b** MR image taken after operation reveals enlargement of high signal intensity area. **c** MR image taken after corticosteroid therapy shows marked reduction of high signal intensity area in enlargement of spinal cord

Fig. 2 Gadolinium DTPA (Gd)-enhanced T1-weighted MR images for Patient 1. **a** After operation, spotty enhancement is seen at the C5/6 level. **b** An abnormally enhanced lesion remains 7 months after treatments

pathological diagnosis of sarcoidosis was confirmed upon finding non-caseating granuloma with giant cells of biopsied scalene lymph nodes. A daily dose of methylprednisolone (1,000 mg/day) was given for 3 days. No clinical deterioration in either motor or sensory symptoms appeared, although marked reduction in enlargement of the spinal cord (Fig. 1c) and in the abnormally enhanced lesion (Fig. 2b) was observed 7 months after the operation.

Case 2

A 58-year-old woman noted a slowly progressive weakness in her lower extremities, and both clumsiness and numbness in her bilateral hands 3 months before admission to our institution. She was diagnosed with cervical spondylotic myelopathy and admitted to our hospital in March 2001.

Neurological examination revealed a decrease in light touch and pin prick sensation in both upper and lower extremities. Deep tendon reflexes were hyperactive in triceps tendon reflex (TTR) and in the lower extremities, and Hoffman's, Trömner's and Babinski's signs were positive bilaterally. A manual muscle strength test of both upper and lower extremities decreased to 4/5 except the deltoid muscle. Neurological examinations were otherwise normal. Blood biochemical analyses including ACE were normal, although the cerebrospinal examination revealed a higher protein content.

Plain radiograph of the cervical spine showed spondylotic changes at C4/5 level. A T2-weighted MR image

revealed an area of high signal intensity extending from C2 to Th1 (Fig. 3a). Gd-enhanced T1-weighted MR images showed a focal enhancement at the C4/5 level (Fig. 4a).

The patient was diagnosed with cervical spondylotic myelopathy with sarcoidosis, as confirmed by a lymph node biopsy. Surgical decompression was performed before steroid therapy, because there was a spinal canal stenosis relative to the swollen spinal cord, spondylotic changes were observed at C4/5 level, and the patient had developed progressive weakness in both the upper and lower extremities.

Mild improvement in motor function of the hand appeared after the operation and the patient was discharged 4 weeks after the operation. However, spasticity and numbness of the limbs were exacerbated and gait disturbance worsened 7 weeks after the operation and the patient was admitted to the Department of Neurology at our institution.

Magnetic resonance imaging at the time of admission showed spinal cord swelling from the lower medulla oblongata to T1 on the T1-weighted image and high signal intensity within the spinal cord that was more extensively enlarged than preoperatively on the T2-weighted image (Fig. 3b). Gd-enhanced T1-weighted MR images also showed stronger enhancement at C4/5 level postoperatively than preoperatively (Fig. 4b).

Intravenous methylprednisolone (1,000 mg/day) was given for 3 days. A slight reduction in numbness was observed after the steroid therapy. MRI performed 2 weeks after the steroid therapy showed a marked decrease in the

Fig. 3 Magnetic resonance images from Patient 2. **a** Sagittal and axial T2-weighted MR image reveal area of high signal intensity extending from C2 to Th1. **b** MR image taken after operation shows higher signal intensity within more extensively enlarged spinal cord than preoperatively. **c** MR image performed 2 weeks after steroid therapy shows markedly decreased high signal area

Fig. 4 Gadolinium DTPA (Gd)-enhanced T1-weighted MR images from Patient 2. **a** Preoperative MR image shows focal enhancement at C4/5 level. **b** MR image after operation reveals stronger enhancement of high signal intensity at C4/5 level postoperatively than preoperatively. **c** MR image taken after steroid treatment reveals marked reduction in abnormally enhanced lesion

high signal intensity area on T2-weighted image (Fig. 3c). A marked reduction in the abnormally enhanced lesion was also observed (Fig. 4c). The clinical course of the patient fluctuated, but her ability to participate in daily life activities slowly decreased until she was unable to stand 3 years after the operation.

Case 3

A 59-year-old man noted a slow progressive weakness in his lower extremities, and both clumsiness and numbness of the right hand 2 months before admission. The patient was admitted to another hospital and since Gd-enhanced

T1-weighted MR images demonstrated a spotty intramedullary enhancement, neurosarcoidosis was suspected and the patient was transferred to our hospital.

Neurological examination revealed a decrease in light touch and pin prick sensation in the right upper extremity. Deep tendon reflexes were hyperactive in both upper and lower extremities, and Hoffman's, Trömner's and Babinski's signs were positive bilaterally. A manual muscle strength test of both finger abductors decreased to 4/5. Neurological examinations were otherwise normal. Blood biochemical analyses including ACE were normal.

Plain radiograph of cervical spine showed spondylotic changes at C3/4 and C4/5 levels. A T2-weighted MR image

Fig. 5 Magnetic resonance images from Patient 3. **a** Sagittal T2-weighted MR image reveals area of high signal intensity extending from C3 to C5. **b** MR image taken after operation shows smaller high signal intensity region than preoperatively. **c** MR image performed 6 weeks after steroid therapy shows almost same high signal area as postoperatively

Fig. 6 Gadolinium DTPA (Gd)-enhanced T1-weighted MR images from Patient 3. **a** Preoperative MR image shows focal enhancement at C3/4 level. **b** MR image after the operation shows same enhancement at C3/4 level as preoperatively. **c** MR image taken after the steroid treatment reveals no reduction in abnormally enhanced lesion

revealed an area of high signal intensity extending from C3 to C5 (Fig. 5a). Gd-enhanced T1-weighted MR images showed a focal enhancement at the C3/4 level (Fig. 6a).

The patient was diagnosed with cervical spondylotic myelopathy combined with sarcoidosis as diagnosed by lymph node biopsy. Surgical decompression was performed before steroid therapy, because there was a spinal canal stenosis relative to the swollen spinal cord and spondylotic changes were observed at C3/4 and C4/5 levels.

Mild deterioration in motor function of the hand appeared after the operation, although gait disturbance remained unchanged. The patient was transferred to the Department of Neurology at our institution.

Magnetic resonance imaging at the time of admission showed a smaller high signal intensity region than preoperatively on the T2-weighted image (Fig. 5b). Gd-enhanced T1-weighted MR images showed the same enhancement at C3/4 level as preoperatively (Fig. 6b).

Intravenous methylprednisolone (1,000 mg/day) was given for 3 days. Few significant changes were observed after the steroid therapy. MRI performed 6 weeks after the steroid therapy showed almost the same high signal intensity area as postoperatively on T2-weighted image (Fig. 5c). No reduction in the abnormally enhanced lesion was observed (Fig. 6c). Activities of daily life were not significantly improved after the treatments.

Discussion

Several reports have described MRI findings of spinal cord sarcoidosis [3, 6, 9–11, 13, 25]. Typical findings reveal both focal and diffuse hyperintensity on T2-weighted image, sometimes misdiagnosed as myelomalacia when patients have spondylotic changes and/or a narrowing of the spinal canal.

There is no known cure for neurosarcoidosis. Treatment is implemented to reduce symptoms and can possibly lead to remission. A literature review of histologically proven intramedullary sarcoidosis showed that 50% of patients who underwent a more extensive resection suffered a postoperative deterioration in neurological function [16]. In contrast, the deterioration was only seen in 28% of limited biopsy patients. As a result of these findings, a limited laminectomy and biopsy or subtotal resection are usually performed, when tissue diagnosis is required in patients with no history of sarcoidosis in order to differentiate an inflammatory from a neoplastic lesion [11]. However, there is no consensus on the pharmacological treatment of neurosarcoidosis. High doses of corticosteroid therapy are recommended for initial treatments, but the most appropriate points for tapering off and discontinuing treatment after response have not been standardized [14]. Immunosuppressive agents are occasionally used to supplement corticosteroids, although a clinical response cannot be guaranteed [27]. Laminoplasty has also been used to manage the decreased space occupied by the spinal cord caused by spondylotic bars, intervertebral disc hernia, ossification of the posterior longitudinal ligament, and/or a constitutionally narrowing cervical spinal canal [2, 5]. When dynamic factors further reduce the functional diameter of the spinal canal, this contributes to the development of cervical myelopathy [8, 17]. Laminoplasty becomes a feasible posterior decompression procedure for the cervical spinal cord as an attempt to manage multi-segmental cord compression. However, this procedure is sometimes inadequately performed in patients with a spinal canal stenosis relative to the swollen spinal cord such as that observed in intramedullary spinal cord diseases such as neurosarcoidosis, multiple sclerosis, or intramedullary tumor.

In patient 1, laminoplasty was performed before neurosarcoidosis was diagnosed, however, symptoms deteriorated soon after the operation. With corticosteroid therapy, the progression of symptoms was halted and then slightly improved. Patient 2 and 3 showed cervical canal stenosis caused by spondylotic changes and surgical decompression before steroid therapy was selected for treatment because we believed enlarging the cross-sectional area of the spinal canal would be a more effective treatment than administering only steroid therapy due to spinal canal stenosis relative to the swollen spinal cord. The symptoms of patient 2 significantly improved soon after the operation; however, the clinical course fluctuated after corticosteroid therapy. In patient 3, the symptoms improved slightly after the operation and then progressively deteriorated even after corticosteroid therapy. Daily life activities were not significantly improved in any of the three patients after the treatment, and in the long-term follow-up period, the clinical course of patient 2 was one of inexorable deterioration to a state of quadriplegia.

Decompression surgery including laminoplasty has been considered safe and effective for enlarging the cross-sectional area of the spinal canal. However, as shown in this study, the effects of laminoplasty are not predictable in the treatment of spinal cord sarcoidosis even when patients have spondylotic changes and/or a constitutionally narrowing cervical spinal canal. It has been reported that a long interval from the onset of disease to the initiation of corticosteroid therapy contributes to poor functional recovery [1]; however, poor results were achieved in all our three patients even with the rapid administration of corticosteroid therapy. Thus, laminoplasty is not an effective intervention for the treatment of spinal cord sarcoidosis with or without spondylotic changes. The possibility of spinal cord sarcoidosis should be included in the differential diagnosis, when a distinct high signal intensity area within the spinal cord on T2-weighted MR images is observed in patients with spondylotic changes. Patients with neurosarcoidosis should be treated first with steroid administration even if they have concomitant cord compression on MRI.

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