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ORIGINAL ARTICLE

Long-term surgical outcomes of idiopathic spinal cord herniation

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Abstract
Background Because of the lack of long-term postoperative follow-up studies of idiopathic spinal cord herniation (ISCH), there is little information about the long-term effectiveness and complications of the dural defect enlargement in patients with ISCH. The purpose of this study is to determine the long-term effectiveness of this procedure.
Methods Sixteen patients with ISCH were treated surgically by enlargement of the dural defect. The patient's neurological status and surgical outcome were evaluated by the JOA scores for thoracic myelopathy and the recovery rate (mean follow-up period 9.6 years). Correlations between the surgical outcomes and patients' age and duration of disease were assessed retrospectively. The patients were also divided into two groups based on the location of the dural defect: the ventro-lateral (VL) group and the ventral (V) group. The difference in the duration of disease, preoperative JOA score, and the recovery rate were compared between the two groups.
Results There was no recurrence of ISCH after surgery. The mean recovery rate was 42.6%. There was a significant correlation between the patient's age and the recovery rate, and between the duration of disease and the recovery rate. The median recovery rate was significantly lower in the V group than in the VL group. There were no complications related to CSF leakage after surgery.
Conclusions Long-term surgical outcomes of enlargement of the dural defect for ISCH were stable and favorable without recurrences or any complications. This procedure should be considered for patients with ISCH before their neurological deficit worsens, especially for the patients in whom the dural defect is located at the ventral part of the dural canal.

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Introduction

Idiopathic spinal cord herniation (ISCH), first described in 1974 by Wortzman and his colleagues [1], is a rare disease characterized by progressive myelopathy. Surgical restoration of the herniated cord to its normal position usually results in the patient's neurological improvement, although long-term follow-up data are available for only a few cases [2–4]. Previously, we reported that enlargement of the dural defect, which is technically easier and less invasive to the vulnerable spinal cord than direct closure of the defect, resulted in favorable outcomes [5]. However, in a recent study, it was recommended to apply an anterior patch to avoid the cerebral spinal fluid (CSF) circulation disturbance caused by extradural CSF collection [2]. Because of the lack of long-term postoperative follow-up studies of ISCH, it remains unclear which procedure should be selected for ISCH. Here, we report the largest series of ISCH to date, consisting of 16 patients treated surgically at a single center. We evaluated the long-term surgical results of dural defect enlargement to determine the indications and limitations of this surgical procedure for ISCH.

Clinical materials and methods

Our series consisted of 16 patients (7 men and 9 women) with ISCH diagnosed between 1987 and 2005. The patients' mean age was 54 years (range 39–78, Table 1). Enlargement of the dural defect was performed in all cases. The follow-up period ranged from 5 to 23 years (median 9.6 years). Clinical history, radiographic findings, and surgical outcomes were collected retrospectively by reviewing the patient charts and radiological reports. We assessed the correlation between the surgical outcomes and patient age at the time of surgery or the duration of clinical symptoms by non-parametric Spearman test. The patients were also divided into two groups, based on the location of the dural defect determined by MRI: the defect was in the ventro-lateral dural canal in the VL group patients and in the ventral dural canal in the V group patients as previously reported [6]. We compared the duration of clinical symptoms, preoperative neurological status, and neurological recovery after surgery between these two groups using the Mann-Whitneys U test. The patients' neurological status was evaluated by the Japanese Orthopedic Association scores for thoracic myelopathy (JOA score: full score 11), and the recovery rate was calculated by the formula: (postoperative JOA score − preoperative JOA score)/(11 − preoperative JOA score) × 100 (%). This study was approved by the Institutional Review Board of the School of Medicine, Keio University. We obtained signed consent forms for this study from all patients.

Results

Onset and clinical course

In all patients, the initial symptom was numbness of their legs. In spite of conservative treatment with or without the diagnosis of ISCH at the previous hospitals, they were eventually admitted to our hospital because of the progression of myelopathy. The duration of the disease ranged from 1 to 16 years (mean 4.3 years). Preoperatively, gait disturbance and urinary dysfunction were observed in 12 and 8 cases, respectively. The pattern of the neurological deficit was the Brown-Séquard type in 11 cases and the paraparesis type in 5. The mean duration was 3.4 years for the Brown-Séquard type and 6.1 years for the paraparesis type, which was a statistically significant difference between them.

Radiographic findings

The level of the spinal cord herniation was Th3 in 4, Th4 in 7, Th5 in 3, Th6 in 1, and Th7 in 1 case. There were 11 patients in the VL group and 5 in the V group. In all cases, duplication of the ventral dura was observed on MRI and/or CT-myelograms.

Surgical outcomes

The preoperative JOA score of the patients ranged from 2 to 8 (average 5.1). The postoperative JOA scores ranged

Table 1 Clinical characteristics of 16 patients with ISCH

Age	Level	Type of neurological deficit	Location of herniation	Duration of disease (years)	Preop. JOA score	Postop. JOA score	Recovery rate (%)
43	Th4	Brown-Séquard	Ventro-lateral	5	4	8	57
39	Th3	Brown-Séquard	Ventro-lateral	3	4	7	43
54	Th4	Brown-Séquard	Ventro-lateral	4	4	7	43
71	Th4	Paraplegia	Ventral	10	3	0	0
49	Th4	Brown-Séquard	Ventro-lateral	5	5	8	50
47	Th5	Brown-Séquard	Ventro-lateral	5	7	9	50
78	Th4	Paraplegia	Ventral	16	2	4	22
56	Th6	Brown-Séquard	Ventro-lateral	2	5	8	50
47	Th3	Paraplegia	Ventral	3	6	8	40
46	Th4	Paraplegia	Ventral	1	6	8	40
68	Th7	Brown-Séquard	Ventro-lateral	8	5	6	17
67	Th4	Brown-Séquard	Ventro-lateral	3	5	9	67
42	Th3	Brown-Séquard	Ventro-lateral	1	5	9	67
53	Th5	Brown-Séquard	Ventro-lateral	0.5	8	10	67
60	Th5	Brown-Séquard	Ventro-lateral	3	6	9	60
68	Th3	Paraplegia	Ventral	3	7	8	25

from 0 to 10 (average 7.4) at the time of the final examination, and the score increased in 15 of the 16 cases. The average recovery rate was 42.6% at the time of the final examination. There was a significant correlation between age at the time of surgery and the recovery rate ($p = 0.03$) (Fig. 1a), and between the duration of the disease and the recovery rate ($p = 0.02$) (Fig. 1b). While there was no significant difference in the median preoperative JOA score between the VL and V groups (Fig. 2a), the median recovery rate of the V group was significantly lower than that of the VL group (Mann-Whitney test, $p = 0.016$) (Fig. 2b).

Complications

There was no recurrence of spinal cord herniation after surgery. In all cases, the spinal cord was reduced to its normal position, which was confirmed by postoperative MRI (Fig. 3a, b). Although a CSF pooling at the ventral side of the spinal canal was observed in all the patients (Fig. 3a, c), there was no headache or nausea related to a persistent CSF leak after enlargement of the dural defect.

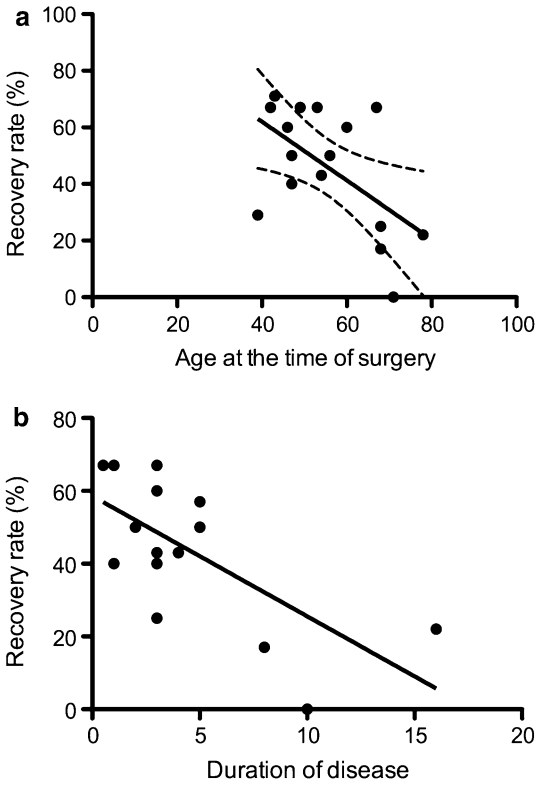

Fig. 1 Correlation between patients' age at the time of surgery and recovery rate (**a**), and between the duration of disease and recovery rate (**b**). There were significant correlations between patients' age at the time of surgery and recovery rate and between the duration of disease and recovery rate

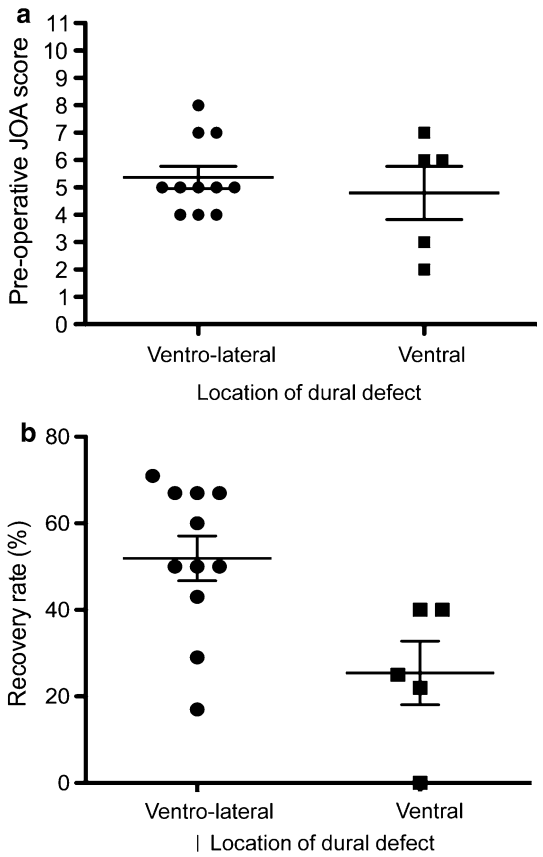

Fig. 2 Comparison of the preoperative JOA scores (**a**) and the recovery rates (**b**) between the VL and V groups. While there was no significant difference in the median preoperative JOA score between the two groups, the median recovery rate of the VL group was significantly higher than that of the V group (Mann-Whitney test, $p = 0.016$)

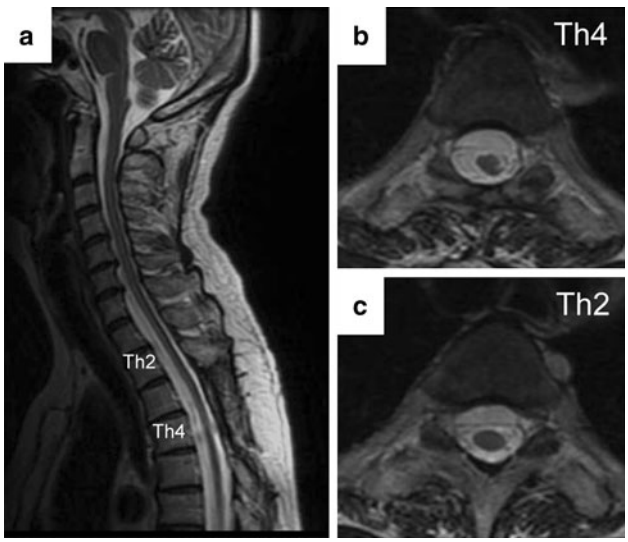

Fig. 3 Representative postoperative sagittal (**a**) and axial (**b**, **c**) T2-weighted MR images of the thoracic spinal cord, demonstrating that the spinal cord was reduced to its normal position (**a**, **b**) and that there was a CSF pooling at the ventral side of the spinal canal (**a**, **c**)

Discussion

Although ISCH is rare, almost 150 patients have now been described in the English literature, because this condition has been increasingly recognized in recent years with the increased use of MRI and awareness of the disease among physicians. Although long-term postoperative follow-up reports are sparse, retethering or syrinx development has been documented in some cases [7]. Therefore, a longer follow-up study is needed to determine the effectiveness of surgical interventions for ISCH.

The aim of the surgical treatments for ISCH is to reduce the herniated spinal cord, either by closing the dural defect, with or without a patch, or by widening the defect to untether the spinal cord. The direct closure of the dural defect through an anterior approach was first described by Wortzman et al. [1]. Borges et al. [8] performed primary suturing of the defect without complications; however, as emphasized by Tronnier et al. [9], there are risks of damaging the spinal cord during the suturing of the dural defect in the limited ventral subarachnoid space. To eliminate these risks, several materials, including fascial flaps or patches [10, 11], free fat grafts [12], lyophilized dura [13], and Gore-Tex membranes [14], have been used to repair the defect. In any case, intradural fixation is important to minimize the risk of recurrence. Batzdorf [13] reported a case of recurrence after lyophilized dura graft, but did not clearly indicate whether the graft was placed between the herniated cord and the dura (intradurally) or on the dura (extradurally), or whether it was fixed or not.

We previously reported another technique, enlarging the dural defect to prevent spinal cord incarceration [5], in nine cases where duplication of the dura mater was the cause of ISCH. Although this procedure is technically easier and safer compared to the other procedures such as primary suture and repair of the dural defect, it still remains unclear whether the long-term surgical outcomes is favorable or not in terms of the recurrence of spinal cord herniation and complications. In the present study, we examined retrospectively the long-term surgical outcomes of our 16 ISCH patients, who had all been treated by enlargement of the dural defect, and found that the long-term surgical outcomes were favorable and stable without recurrence of ISCH. Since there were significant correlations between age at the time of surgery and the recovery rate, and between the duration of disease and the recovery rate, an early diagnosis of ISCH is essential to obtain better surgical outcomes, and the surgical treatment should be considered before the progression of myelopathy. Based on the clinical course of our 16 patients, we do not recommend the conservative treatments for ISCH, which could cause irreversible changes in the spinal cord and result in poor outcomes. In addition, the surgical outcome was more favorable in the patients with Brown-Séquard syndrome and less favorable in patients with spastic paraparesis, which was consistent with the previous studies [7, 8, 15–20], suggesting that spastic paraparesis at presentation may indicate a poor prognosis.

Another known but rare symptom of ISCH is cerebrospinal fluid hypotension-related headache in the upright position [21, 22]. However, none of our 16 patients had such a cerebrospinal fluid hypotension-related headache, probably because their ISCH was caused by dural duplication and the residual outer layer of the dura mater would prevent the leakage of cerebrospinal fluid. Previous studies have demonstrated that a congenital duplication of the ventral dura leads to herniation of the spinal cord through the inner dura [3, 15], which was considered to be the pathology of our cases. Enlargement of the dural defect should be considered for patients with ISCH caused by dural duplication.

In conclusion, long-term surgical outcomes of enlargement of the dural defect for ISCH were stable and favorable without recurrences or any complications. This procedure should be considered for patients with ISCH, before their neurological deficit worsens, especially for the patients in whom the dural defect is located at the ventral part of dural canal.

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