Single-centre study comparing surgically and conservatively treated patients with spinal cord herniation and review of the literature

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ABSTRACT

Introduction: Spinal cord herniation (SCH) is a rare cause of progressive myelopathy and Brown-Séguard-Syndrome.
Research question: Evaluation of functional outcome after SCH treatment compared to conservatively treated patients.
Material and methods: We retrospectively analysed functional outcome in SCH patients treated between 2009 and 2020. We conducted a systematic search using PubMed, MEDLINE and EMBASE to perform a pooled analysis in SCH patients.
Results: Our hospital cohort included 17 patients of which 9 were treated surgically. Mean age was 51.9 years, 58.8% of the patients were female. In 4/9 patients (44.4%) the neurological state remained stable after surgery. Four patients improved (44.4%) and one deteriorated after surgery (11.1%). Conservatively treated patients had a higher deterioration rate on follow-up with 3/8 patients deteriorating (37.5%). In our pooled analysis, 109/145 (75.2%) of patients improved, 32/145 (22.1%) remained stable and 4/145 patients deteriorated (2.8%). Among the available data of nine cohorts, mean recovery rate measured by the JOA score was 36.6% (SD 14.4). In our pooled multivariable model lower preoperative JOA score was associated with worse functional outcome (OR 0.86, 95%CI 0.74–0.99, p = .04).
Discussion and conclusion: Our data shows that patients who are treated surgically have a higher improvement rate and acceptable perioperative morbidity compared to conservatively treated patients. Lower preoperative JOA score decreases chances of improved functional outcome on follow-up. We therefore advocate early surgery for symptomatic patients. Wait and see appears outdated due to progressive impairment and decreased chances of recovery. However, it is still an option in asymptomatic incidental SCH patients.

1. Introduction

Spinal cord herniation (SCH) has first been described in 1974 (Wortzman et al., 1974). Even though differentiation of the specific aetiology can be difficult, several have been reported, the most common being idiopathic followed by iatrogenic and posttraumatic. The level of herniation, especially in idiopathic SCH, is frequently located in the thoracic spine; postinterventional and SCH due to trauma can occur anywhere but is commonly located in the cervical spine (Darbar et al., 2006; Borges et al., 1995). The exact location of the dural defect can vary but is most commonly located ventrally or ventrolaterally (Dix et al., 1998). Herniation of the spinal cord leads to compromise of the spinal cord secondary to vascular impairment and adhesions and consecutively symptoms, most commonly Brown-Séguard-Syndrome and myelopathy (Borges et al., 1995). There is still only limited data on the natural course of disease and little is known about functional outcome after surgical treatment (Bartels et al., 2017; Groen et al., 2009; Summers et al., 2013). Another interesting factor to be evaluated is whether extent, duration of symptoms and degree of disability before a surgical intervention influences functional outcome in patients with SCH and how the natural
course of conservatively treated patients is.
Our aim is to retrospectively review our own dataset including both patients who underwent surgery and patients who were treated conservatively. Additionally, we will conduct a review of the literature with the aim of performing a pooled analysis with all available case series assessing functional outcome.

2. Methods

2.1. Data collection

2.1.1. Hospital based cohort study
We retrospectively reviewed a clinical and radiological databases of patients who were treated in our specialist neurosurgical unit between January 2009 and March 2020 for spinal cord herniation as confirmed by MRI and if applicable by intraoperative finding.

We collected information on patient demographics, preoperative symptoms, imaging, history of surgeries or trauma and follow-up data from electronical medical records. Special emphasis was put on functional outcome at discharge and follow-up. Clinical state at the time of first diagnosis and at the last follow up examination was additionally accessed according to the Japanese Orthopaedic Association (JOA) score (Kato et al., 2015).

2.1.2. Treatment
All of our patients received imaging studies with CT and MRI of the according spine level as per our diagnostic standard. All patients were operated in prone position with fluoroscopic control of the according level. Approach was either via laminoplasty, hemilaminectomy or laminectomy. As per our hospital standard a microscope and intraoperative neuromonitoring via motor evoked potential was used for all patients.
2.2. Statistical analysis

Categorical variables are presented as count and percentage, continuous variables as mean with standard deviation (SD).

2.2.1. Systematic review and pooled analysis

We searched PubMed, MEDLINE and EMBASE for peer reviewed case series of spinal cord herniation using the key words “spinal cord herniation AND myelon herniation NOT disc herniation”, undertaken on the March 28, 2020, in addition to reference lists. We did not limit our search to languages or dates. Per pre-defined analysis plan we only included studies with available abstracts and, if meeting inclusion criteria, available paper and case series of ≥5 patients (Abu-Zidan et al., 2012). Only studies with proven spinal cord herniation independently of aetiology were included. Two authors reviewed all papers independently and independently extracted following information: number of patients, population from which the patients were drawn, mean follow-up time and functional outcome where available. Due to the exact score of the different scoring systems not always being available we dichotomized patients into improved versus not improved versus deteriorated after surgical intervention.

The pooled multivariable model included the prespecified variables age and sex, as well as variables that had a p-value below .2 in the univariable analysis. We performed a pooled multivariable regression analysis for improvement on follow-up using a frailty term to account for differences between study cohorts. Our study is reported according to the STROBE guidelines, and we conducted the pooled-analysis according to the PRISMA guidelines (Moher et al., 2009).

Statistical analysis was performed using STATA 15 (StataCorp. 2011. Stata Statistical Software: Release 15. College Station, TX: StataCorp LP) and biostatistical expertise.

Ethical approval

The data collection was approved by the local Ethics Committee (reference: 185/20 S–KH). A consent form for use of the clinical data in the case illustration has been signed by the patient.

2.2.2. Data availability statement

Anonymized data will be shared by request from any qualified investigator.

Fig. 2. Preoperative MR scan with sagittal and axial T2 sequence.

Fig. 3. A) Exposure of the spinal cord after opening of the dura and visualisation of the asymmetry by anterior dislocation due to the myelon herniation. B) Exposure of the anteriorly located defect upon retraction of the myelon. C) Retraction of the denticulate ligament and repositioning of the myelon. D) Coverage of the herniation defect with TachoSil via inlay/onlay technique.
Characteristics of hospital-based cohort of surgically treated patients with SCH.

| Patient | Age | Sex | Location of herniation | Symptoms at presentation | Duration of symptoms (months) | Surgical approach | JOA preop | JOA postop | JOA on FU | Symptoms on FU |
|---------|-----|-----|-------------------------|--------------------------|-------------------------------|------------------|-----------|-----------|-----------|---------------|
| 1       | 49  | M   | Th7/8                   | BSS                      | 144                           | Laminoplasty      | 12.5      | 12.5      | 12.5      | Stable        |
| 2       | 59  | F   | Th7/8                   | SMP                      | 18                            | Hemilaminectomy   | 12        | 12.5      | 14        | Improved      |
| 3       | 67  | M   | Th1                     | Dysesthesia, unrelated pain | 24                           | Hemilaminectomy   | 16.5      | 16.5      | 16.5      | Stable        |
| 4       | 72  | F   | C6                      | Myelopathy, related pain | 36                            | Hemilaminectomy   | 12        | 14        | 14        | Improved      |
| 5       | 55  | F   | Th7/8                   | Hypoesthesia, ataxia, unrelated pain | 18                           | Hemilaminectomy   | 12.5      | 12.5      | 12.5      | Stable        |
| 6       | 38  | F   | Thb/7                   | BSS, unrelated pain      | 48                            | Laminectomy       | 12.5      | 13        | 13        | Improved      |
| 7       | 79  | F   | L2                      | BSS, related pain        | 20                            | Hemilaminectomy   | 13        | 15.5      | 15.5      | Improved      |
| 8       | 20  | M   | Th5                     | SMP                      | 17                            | Hemilaminectomy   | 16        | 16        | 16        | Stable        |
| 9       | 58  | F   | C3/4                    | Myelopathy               | 4                             | Laminectomy       | 6         | 4         | 4         | Deteriorated  |

BSS = Brown-Séquard-Syndrome; SMP = spastic monoparesis; SPP = spastic paraparesis.

2.3. Case illustration

A forty-eight-year-old male patient was referred to our outpatient clinic with a 14-year history of increasing gait difficulties, initially mild and first noticed only by his daughter. He was followed-up in a smaller hospital since 2008. Follow-up continued even though neurological symptoms progressed over time. He had history of minor trauma, being in a fight as a young adult but no previous spinal surgeries. On first presentation to our institution, he presented with Brown-Séquard-Syndrome with spasticity of his left leg, weakness of the left leg with hip flexion 4/5, foot flexion 4/5 and foot dorsiflexion 1/5 and positive Babinski on the left side. He did not have any thermal nor sensation to pain on the right side. MRI scan showed evidence of anterior dislocation of the myelon at Th7/8 with an associated cerebro-spinal-fluid (CSF) leak due to the lifted dura, raising suspicion of myelon herniation at this level (Fig. 2). The patient underwent surgery via laminoplasty of Th7/8 and intradural inspection. We found an anteriorly located dural defect, which was primarily sealed by a TachoSil® inlay/onlay (Fig. 3). The lamina were refixed with titanium microplates. Intraoperatively conducted motor evoked potential stayed stable during the entire surgery. Postoperatively the patient experienced transient worsening of the motor function of his left leg (hip flexion 2/5, knee extension 4/5, knee flexion 1/5 and foot dorsiflexion 0/5). The symptoms completely recovered within 24 h to his known baseline, and he was discharged on the third postoperative day. Gait instability when walking improved on clinical follow-up.

3. Results

3.1. Hospital-based cohort study

We identified 17 SCH patients between January 2009 and March 2020. Mean age was 51.9 years, 10 patients were female (58.8%). Median follow-up time was 7 months, 21.7 months for surgically and 5 months for conservatively treated patients.

3.2. Surgically treated patients

We included 9 consecutive patients who were treated surgically in our institution due to SCH. Mean age was 55.2 years, and 66.7% of the patients were female. The demographic, clinical, surgical and functional outcome characteristics are shown in Table 1. Of these 9 patients, SCH was located in the thoracic spine in 6 patients (66.7%), in the cervical spine in 2 (22.2%), and in the lumbar spine in one patient (11.1%). Of all 9 patients, 2 patients had a history of previous surgery for other pathologies on the corresponding level making this a possible cause for the consecutively resulting SMH and one patient had a history of trauma.

Median duration of symptom was 20 months (IQR 18 months). Patients presented with mainly Brown-Séquard-Syndrome (3 patients), spastic monoparesis (2 patients), dysesthesia (2 patients), and myelopathic ataxia (2 patients). Median preoperative JOA score was 12.5 (SD 1). In 4/9 patients (44.4%) the neurological state improved after surgery and in another four patients (44.4%) the neurological state remained stable with no further deterioration. One patient (11.1%) deteriorated after surgery. This patient presented initially with a JOA score of 6 preoperatively and experienced a drop to 4 postoperatively (worsening of arm and leg weakness, sensory disturbances, incontinence). The median postoperative JOA score was 13 (IQR 3). The median JOA score on follow-up was 14 (IQR 2.5).

3.3. Conservatively treated patients

Eight patients (47.1%) were treated conservatively. We recommend surgery for all of them, but the patients declined. Mean age of conservatively treated patients was 55.1 years, 50% of patients were female. Characteristics of these patients are stated in Table 2. Most patients in the conservatively treated cohort exhibited pain only although pain was unrelated to the lesion in 3 patients (37.5%). Only two patients had neurological deficits as opposed to pain alone, both demonstrating paraesthesia. Two patients (25%) did not have any symptoms. Three patients (37.5%) improved on follow-up, whereas another 3 (37.5%) worsened, one of them however only subjectively keeping the same JOA score, and two remained stable (25%). All 3 patients worsening on follow-up refused surgery, the 2 patients (25%) who presented related...
pain improved during follow-up.

Comparing initial symptoms between surgically and conservatively treated patients, symptoms were less severe in the conservatively treated subgroup, which mainly consisted of pain. Consistent with this, we found a significant difference when assessing the JOA score when first seen (Table 3): patients who were treated surgically had a lower JOA score compared to conservatively treated patients (OR 0.39, 95%CI 0.18–0.87, p = .02). We did not find a significant difference with regards to age, sex, symptom duration and JOA score on follow-up. However, more patients in the conservatively treated group deteriorated on follow-up. Only one of our surgically treated patients (11.1%) deteriorated as opposed to three (37.5%) in the conservatively treated cohort; 4 patients (44.4%) in the surgically treated cohort improved on follow-up compared to 3 (37.5%) in the conservatively treated cohort (Fig. 4). Improvement in the conservatively treated group was limited to pain improvement.

3.4. Systematic review and pooled analysis

Our systematic review yielded 1111 papers overall including case reports (Fig. 1). After reviewing all abstracts, excluding case reports as well as case series with number of patients ≤5, we included 18 studies in addition to our own (Bartels et al., 2017; Ando et al., 2019; Barrenechea et al., 2006; Batzdorf and Holly, 2012; Berg-Boesen et al., 2014; Carter et al., 2015; Gaudino et al., 2016; Hassler et al., 2008; Haraishi et al., 2014; Herring et al., 2019; Imagama et al., 2009; Maira et al., 2006; Massicotte et al., 2002; Menon et al., 2019; Nakamura et al., 2011; Parmar et al., 2008; Prada et al., 2012; Watanabe et al., 2001). All studies were retrospectively conducted studies. We initially included all patients (210 patients). Baseline characteristics of each included cohort is shown in Table 4. Information whether patients improved on follow-up was available for 186 patients (88.6%): 109 patients (58.6%) improved, whereas 44 patients (23.7%) remained stable and 33 patients (17.7%) deteriorated. After excluding patients who were treated conservatively (57 patients, 27.1%), a total of 152 patients remained. In the pooled analysis of 152 available patients with SCH who were treated surgically, information of whether patients improved was missing for one cohort completely (Ando et al., 2019). Of the remaining 145 patients, 109 patients (75.2%) improved after surgery, 32 patients (22.1%) remained stable, and 4 patients deteriorated (2.8%).

We conducted a pooled univariable analysis (see Table 5) and adjusted a pooled multivariable analysis with the pre-specified variables age and sex as well as the preoperative JOA score. In this pooled multivariable model, a lower preoperative JOA score was associated with a lower chance of an improved functional outcome on follow-up (OR 0.84, 95%CI 0.73–0.97, p = .02).

4. Discussion

In our hospital cohort study, patients who were treated surgically did improve or remained stable regarding their neurological state. Symptoms in the conservatively treated cohort were less severe and usually only consisted of pain. In the pooled analysis, 97.1% improved or remained stable, only 4 patients (2.8%) deteriorated. The pooled multivariable analysis demonstrated a lower preoperative JOA score had a lower chance of improved functional outcome on follow-up.

When evaluating differences between surgically and conservatively treated patients in our series, patients with conservative management demonstrated a statistically higher JOA score when first seen. More patients in the conservatively treated group deteriorated (37.5% versus 11.1%) whereas more patients in the surgically treated group improved on follow-up (44.4% versus 37.5%). The conservatively treated group is clearly biased towards patients with a less severe disease, which worsens over time as demonstrated by the patient described in our case illustration. In our experience, improvement is clearly associated with surgical treatment of the SCH. Although patients seen in our institution

| Table 3 | Difference between surgically and conservatively treated patients. |
|---------|---------------------------------------------------------------|
|         | Univariable analysis |                                           |
|         | OR     | 95%CI        | p-value |
| Age     | 1.03   | 0.96–1.1     | .38     |
| Sex     | 0.5    | 0.07–3.55    | .49     |
| Symptom duration | 1.02 | 0.98–1.06 | .31     |
| JOA score first seen | 0.39 | 0.18–0.87 | .02     |
| JOA score on follow-up | 0.78 | 0.48–1.26 | .3      |

Fig. 4. Comparison of symptom improvement, deterioration and stability of surgically and conservatively treated patients.
become symptomatic further down the line. we advise close follow-up for the patient to verify that they do not experience they are unlikely to improve and more likely to deteriorate. However, in true incidental SCH without any underlying symptoms, undergoing conservative treatment had a higher JOA score, in our experience they are unlikely to improve and more likely to deteriorate. However, in true incidental SCH without any underlying symptoms, including negative electrophysiology (sensory and motor evoked potentials), there might be a rational for conservative treatment. In these cases, we advise close follow-up for the patient to verify that they do not become symptomatic further down the line.

We confirm previous findings of a female preponderance (Groen et al., 2009). As previously reported, the most common level of disease pathology was the thoracic spine (Menon et al., 2019). In contrast to previous publications, Brown-Séquard-Syndrome was present in only 33.3% of patients in our cohort (Watanabe et al., 2001). However, this may be due to relatively low number of patients in our and previously published series.

Several theories exist on how the defect in SCH develops: some believe that it is a congenital defect of the dura mater into which the spinal cord herniates (Borges et al., 1995). The patient in our case illustration reported a traumatic event as a teenager when he was kicked in the back. We consider this as a minor trauma and unlikely being the cause of the herniation. Another way of causing trauma to the anterior dura might be through a calcified disk (Parmar et al., 2008). A probably pathophysiological more plausible aetiology might be the underlying motion of the kyphotic thoracic spine (Tekkok, 2000): here the dura can be fixed anteriorly predisposing it to even minor trauma (Ewald et al., 2009).
4.1. Limitations

This was a retrospective evaluation of all patients presenting to our institution with SCH. Although all available means (medical reports, outpatient clinic letters, discharge summaries and follow-up via telephone) were used, data was not collected prospectively potentially leading to recall bias. Some of our patients undergoing surgery with a suspicion of SCH preoperatively did not have SCH intraoperatively and were excluded from this study. This issue cannot fully be excluded for conservatively treated patients. Additionally, in our pooled analysis the neurological impairment could not always be deduced specifically for the surgically treated patients. Although we report a relatively large number of patients with SCH, including a pooled analysis, it still is a small cohort and results should be interpreted with caution. Therefore, our findings should be verified by a prospectively collected cohort, ideally as a multicentre study due to the rare nature of this pathology.

Due to the relative rarity of this disease we continue to make decisions on an individual basis. Based on our own data and pooled analysis, we do emphasize surgical treatment in patients with symptomatic SCH as per our clinic’s standard and the findings of our case series demonstrating a significantly higher deterioration rate in conservatively compared to surgically treated patients.

5. Conclusion

Our data shows that surgically treated patients have a higher improvement rate with acceptable perioperative morbidity compared to conservatively treated patients. Patients with a lower preoperative JOA score have lower chances of improved functional outcome on follow-up. We therefore advocate early surgery for symptomatic patients. Wait and see appears outdated due to progressive impairment and decreased chances of recovery.

Disclosure

The authors have no personal, financial, or institutional interest in any of the drugs, materials, or devices described in this article.

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Conflict of interest

None.

Ethics

The data collection was approved by the local Ethics Committee (reference: 185/20 S–KH). A consent form for use of the clinical data in the case illustration has been signed by the patient.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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