Spontaneous Spinal Cord Herniation: MR Imaging and Clinical Features in Six Cases

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Abstract
Background and Purpose: Spontaneous spinal cord herniation (SSCH) is a rare but important differential diagnosis of spinal cord disorder. The aim of this study was to evaluate the topography and lesion pattern in six patients with SSCH by magnetic resonance imaging (MRI) and their correlation with clinical symptoms.

Material and Methods: MRI was evaluated according to intramedullary hyperintense signal changes, alterations of the posterior vertebra and dorsal subarachnoid space at the level of the herniation. Neurologic symptoms as well as their progression were analyzed.

Results: SSCH occurred between the level of T3–T7 and were demonstrated best on sagittal and axial T2-weighted images (T2-WI). Hyperintense signals on T2-WI were seen in five patients. Dorsal margins of the vertebra at the level of herniation were scalloped in three patients and one patient showed a sagittal cleft. Mean time from the initial clinical symptoms to diagnosis was 4.2 years. Incomplete Brown-Séquard' syndrome was the most common clinical presentation in our patients (four cases).

Conclusion: SSCH is best diagnosed using multiplanar T2-WI with characteristic imaging findings of herniation and adjacent structures. Coexistent notches or even sagittal clefts in the dorsal vertebra raise the hypothesis that SSCH might originate from malfusion of the notochord.

Key Words: Spinal cord herniation · Brown-Séquard’s syndrome · Dorsal arachnoid cyst · Magnetic resonance imaging · Sagittal cleft

Spontane Myelonherniation: MR-Bildgebung und klinischer Verlauf in sechs Fällen

Zusammenfassung
Hintergrund und Ziel: Die spontane Myelonherniation (SMH) ist eine seltene, aber wichtige Differentialdiagnose spinaler Pathologien. Ziel dieser Studie war es, Topographie und Verteilungsmuster der SMH zu ergründen und mit den klinischen Symptomen zu korrelieren.


Schlüsselwörter: Spinales Myelonherniation · Brown-Séquard-Syndrom · Dorsale Arachnoidalzyste · Magnetresonanztomographie · Sagittaler Spalt

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Introduction

Spontaneous spinal cord herniation (SSCH) is a rare cause of progressive myelopathy, first described by Wortmann et al. in 1974 [1]. Review of the literature yielded SSCH in 89 patients [2].

SSCH is mostly seen in middle-aged patients, predominantly in women. Herniation occurs typically in the upper or middle thoracic vertebra between T2 and T8. To date only one case has been reported with a herniation at lower level at T10 [3]. In most of the patients clinical symptoms are typically slightly progressive over years [4]. Chronic pain and progressive weakness of the legs often lead to incomplete or even complete Brown-Séquard’s syndrome (BSS). In addition, one third of the patients suffer from vegetative disorders, especially from bladder and bowel dysfunction. By contrast, only two cases with sudden symptomatic onset and rapid clinical progression have been described so far [5, 6]. Reports of SSCH show that herniated spinal cord portion measures from 1 up to 3 cm [3, 5, 7–18]. However, one patient with two separated fragments of herniated spinal cord has been reported in the literature up to now [13]. Posttraumatic herniation of spinal cord might be difficult to differentiate from SSCH because the time interval between trauma and symptomatic herniation could amount up to 30 years [8, 18, 19].

The purpose of this study was to evaluate the topography and lesion pattern in six patients with SSCH with special regard to magnetic resonance imaging (MRI) features and clinical symptoms.

Material and Methods

MRI studies of six patients (four women and two men, aged 32–53 years, mean 41 years) with SSCH were evaluated by two experienced neuroradiologists. Routine imaging protocol included sagittal and axial T2-weighted images (T2-WI) with a slice thickness of 3 mm and 2 mm respectively. An additional transverse CISS (Constructive-Interference in Steady State) sequence with 1 mm slice thickness was acquired in one patient and post-myelo-CT was performed in two patients. Inclusion criteria were the absence of remarked history of spinal trauma and degenerative spinal changes at the level of herniation, as well as normal cerebrospinal fluid (CSF) analysis.

Extent of intramedullary hyperintense signal changes was rated for confidence of visualization on a four-point scale with a score of (–) = none, (+) = mild, (+++) = moderate, and (++++) = severe. Alterations of dorsal surface of the vertebra at the level of the herniation were analyzed with regard to scalloping features or sagittal dorsal clefts. Post-myelo-CT and CISS sequences (MRI) were evaluated to define the paramedian aspect of the herniation and its relationship to the dorsal margins of the vertebral body. Dorsal subarachnoid space was screened for associated arachnoid cysts at the level of SSCH. There was total agreement with respect to all findings between the raters. Time interval between onset of clinical symptoms and diagnosis with MRI as well as neurologic history were assessed.

### Table 1: MR features and clinical symptoms in six patients with SSCH.

<table>
<thead>
<tr>
<th>Patient #</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Level</th>
<th>Signal abnormalities T2-WI</th>
<th>Vertebral body</th>
<th>Neurologic symptoms</th>
<th>Time period to MRI</th>
<th>Dorsal subarachnoid space</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>38</td>
<td>F</td>
<td>T4 right</td>
<td>+++</td>
<td>Not scalloped dysfunction</td>
<td>Incomplete BSS, bowel</td>
<td>3 years</td>
<td>No cyst</td>
</tr>
<tr>
<td>2</td>
<td>32</td>
<td>F</td>
<td>T6 right</td>
<td>++</td>
<td>Cleft, Not scalloped</td>
<td>Incomplete BSS</td>
<td>2 years</td>
<td>No cyst</td>
</tr>
<tr>
<td>3</td>
<td>36</td>
<td>M</td>
<td>T7 left</td>
<td>+</td>
<td>Scalloped</td>
<td>Incomplete BSS</td>
<td>4 years</td>
<td>No cyst</td>
</tr>
<tr>
<td>4</td>
<td>35</td>
<td>M</td>
<td>T6 right</td>
<td>++</td>
<td>Scalloped</td>
<td>Bladder and bowel dysfunction</td>
<td>1 month</td>
<td>No cyst</td>
</tr>
<tr>
<td>5</td>
<td>53</td>
<td>F</td>
<td>T3 left</td>
<td>(+)</td>
<td>Not scalloped</td>
<td>Hygalgesia and hemi-dysesthesia below level T6</td>
<td>8 years</td>
<td>No cyst</td>
</tr>
<tr>
<td>6</td>
<td>51</td>
<td>F</td>
<td>T3/4 right</td>
<td>–</td>
<td>Scalloped</td>
<td>Incomplete BSS, no hemiparesis</td>
<td>3–4 years</td>
<td>No cyst</td>
</tr>
</tbody>
</table>