Idiopathic cord herniation

TO THE EDITOR: With interest we read the recently published study by Imagama et al. (Imagama S, Matsuyama Y, Sakai Y, et al: Image classification of idiopathic spinal cord herniation based on symptom severity and surgical outcome: a multicenter study. J Neurosurg Spine 11:310–319, 2009), in which they provide “the first evidence for image classification of idiopathic spinal cord herniation (ISCH)” based on their experience in 12 cases, collected retrospectively from the databases of 7 orthopedic institutes. The severity of cord herniation and cord displacement (based on MR images and/or CT myelography) in the sagittal plane was classified as: a kink type (Type K), when showing an obvious spinal kink toward the ventral region; a discontinuous type (Type D), if the spinal cord completely disappears at a herniated site; and a protrusion type (Type P), in which the subarachnoid space of the anterior spinal cord disappears with almost no kink in the posterior spinal cord. On axial images, the location of cord herniation was classified into either central (Type C) or lateral (Type L). Classification of laterality of the herniated spinal cord is based on correspondence (same side as: Type S) or noncorrespondence (opposite side as: Type O) with the hiatus location. In addition, the presence (+) or absence (−) of a bone defect in the vertebral body was scored. This all resulted in a 4-point-construct-score, which represents the image classification for each individual patient. With this classification, the authors started to analyze statistically a number of patient characteristics, preoperative neurological data, and postoperative outcome scores. Dr. Imagama et al. concluded that “Patients with Type P herniation had a good postoperative outcome: a multicenter study.” The authors stated that, “.... the results show the severity of preoperative symptoms and the probable surgical outcome from imaging.”

The authors described a rare and, most probably, underdiagnosed disorder. Therefore, clinical experience is limited, and as a result, treatment strategies are based on individual cases and on small series reported in the literature. We wish to commend Dr. Imagama and colleagues on their attempt to correlate preoperative imaging findings with both the severity of the neurological deficit and postoperative results. However, we feel that a number of limitations in this article warrant further scrutiny and comment.

First, the authors use the abbreviation ISCH (idiopathic spinal cord herniation). However, this denomination seems inaccurate, as the patients the authors report on all meet the criteria of the so-called anterior thoracic spinal cord herniation (ATSCH). In the literature, different terminology is used for the same entity (idiopathic spinal cord herniation, thoracic idiopathic spinal cord herniation, spontaneous spinal cord herniation, spontaneous thoracic spinal cord herniation). Because the cause of cord herniation remains unknown (despite many speculations concerning the etiology), in our opinion, it is more appropriate to use the term ATSCH since this term comprises the radiological features that all these cases have in common (that is, the anterior herniation of the thoracic spinal cord through a ventral dural defect). Second, the image classification designed by the authors is interesting, but the rationale for such a complex classification, composed of 4 different variables, remains obscure to the reader. For the differentiation of the severity of cord herniation and cord displacement in the sagittal plane, the authors distinguish 3 types and employed the following order of severity: 1) Type K (kink), 2) Type D (discontinuous), and 3) Type P (protrusion). In our opinion, it seems more plausible to start this ordinal scale with: 1) Type P (which seems to be similar to the “forme fruste” of ATSCH), followed by 2) kinking of the spinal cord (Type K), and finally ending with 3) complete herniation/protrusion of the spinal cord (Type D). This is in line with a publication by Ewald and Hassler who reported on the evolution of the MR imaging findings in a patient with progressive thoracic myelopathy 6 months, 18 months, and 24 months after onset of the symptoms; finally this patient underwent surgery and ATSCH was confirmed. From a recent review of the literature, we know that the diagnosis of ATSCH is likely to be long delayed, with an average of 5 years from the onset of symptoms. Magnetic resonance imaging abnormalities, in the early stage of the disorder, may be very subtle, showing only slight anterior displacement of the thoracic spinal cord, compatible with the Imagama Type P. Consequently, one may assume that this type of herniation is associated with a mild neurological deficit compared with a more profound cord herniation seen on Type K and Type D images. Unfortunately such a clear theoretical basis for the design of the image classification is missing in their paper.

Third, the authors have not developed a standardized scoring protocol, and apparently both MR imaging and CT myelography can be used, both simultaneously or (in the absence of one of the imaging modalities) separately.
Interrater reliability (do the blinded observers measure what they purport to measure?) was not tested, which makes this classification system invalid.

Recently, we reported the results of an individual patient data (IPD) meta-analysis using data derived from 126 case reports with ATSCH. After statistical analysis of the data, it appeared that Brown-Séquard syndrome is a strong independent factor associated with postoperative motor function improvement (p = 0.033). Also, operative results after cord release and subsequent widening of the dural defect were significantly better than those in patients who received an anterior dural patch to cover the dural defect after cord release (p = 0.036). At the time of manuscript preparation, we did not analyze imaging findings.

As a result, associations between imaging characteristics and preoperative neurological deficit or postoperative results were not calculated. To test the imaging classification system described by Imagama et al., for the purpose of this letter, we have used the data of our IPD meta-analysis, supplemented with 29 new cases recently reported by Akaza et al., Chaichana et al., Hassler et al., and others. (It appeared to us that Case 1 and Case 3 have been published before by one of the authors in 2001 [see Ewald and Hassler] and were already included in our IPD meta-analysis). Ishida et al., Sai Kiran et al., Uhl et al., Chaichana et al., Imagama et al., Sasani et al., and Selviaridis et al. For the classification of the severity of herniation and cord displacement, we employed the method of Imagama et al. on sagittal images (Types P, K, and D). In this updated series (now comprising a total of 158 reported cases) no statistically significant associations were found between the severity of herniation/cord displacement as classified by Imagama et al. and preoperative neurological deficits (Brown-Séquard syndrome, paraparesis, sensory deficit, and pain). However, for sensory deficit and pain, these analyses showed a violation of statistical assumptions by empty table cells and frequencies less than 5. To check whether the results were affected by this violation, only the cases with Brown-Séquard syndrome and the cases with paraparesis were used (Table 1). This association also appeared to be not statistically significant (p = 0.17).

The proportions of cases with Brown-Séquard syndrome and paraparesis among individuals in whom no imaging was available did not differ from those classified as Type P, Type K, or Type D (difference of proportions test). Controlling for widening of the dural defect (WDD), Brown-Séquard syndrome and paraparesis remained statistically nonsignificant associated with the extent of herniation (p > 0.05, chi-square). Also we made an attempt to compare imaging type with postoperative results, taking into account only a selected subgroup of cases in which surgery involved a spinal cord release together with widening of the dural defect (all cases in this subgroup were operated in the same way [cord release and WDD] as the patients described by Imagama et al.). There were no statistically significant associations between WDD and Type P (p = 0.09, Fisher exact test), Type K (p = 0.06, Fisher exact test), and Type D (p = 0.22, Fisher exact test) cord herniation.

We are aware that our present analysis also has a number of limitations: 1) data used from publications may have induced information bias because only 1 or 2 images selected by the authors of a particular case report were available; 2) in such cases standardized scoring is not possible; and 3) discrimination between Type K and Type D sometimes is difficult. Nevertheless, the number of cases collected (158 overall) allows for a bivariate analysis to test the hypothesized association between preoperative imaging findings and neurological deficit, and we calculated that such an analysis has sufficient statistical power. This does not apply for the series reported by Imagama et al., which is too small to allow for any statistical analysis at all. Imagama and colleagues have brought up an interesting issue, by focusing the readership on the characteristics of imaging studies in ATSCH. However, as we have shown above, and in contrast to what is stated in their abstract, there is no statistically significant association between imaging characteristics and neurological deficit or operative results.

Clinical experience with ATSCH within a single institution is very limited. Currently, treatment strategies are based on individual cases and on reports in the literature, or they result from ad hoc decisions made during operative exploration of preoperatively unrecognized cases. Standardization of case reports would allow for better comparison of individual cases. Clinicians should be encouraged to report new cases and to describe all the details that are discussed in the recent reviews. Only this will help to better understand the pathophysiology, and the pre- and postoperative findings, and to identify factors that determine outcomes of this rare and very intriguing disorder.

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References
2. Chaichana KL, Scibba DM, Li KW, Gokaslan ZL: Surgical

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<tr>
<th>Neurological Deficit</th>
<th>Imaging Findings (%)</th>
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<tr>
<td></td>
<td>Type D</td>
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<tr>
<td>Brown-Séquard syndrome</td>
<td>15 (68.2)</td>
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<tr>
<td>paraparesis</td>
<td>7 (31.8)</td>
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<tr>
<td>total</td>
<td>22 (100)</td>
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* χ² = 5.05; df = 3; p = 0.17. Type D = discontinuous-type cord herniation; Type K = kinking-type cord herniation; Type P = protrusion-type cord herniation.