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Clinical Study

Idiopathic spinal cord herniation: an imaging diagnosis with a significant delay

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Abstract

BACKGROUND CONTEXT: Idiopathic spinal cord herniation (ISCH) is an underrecognized entity that is often underappreciated by the neurosurgery and neuroradiologic communities. This leads to delayed diagnosis, multiple imaging studies, other diagnostic tests, inappropriate surgeries, and repeat office visits.

PURPOSE: To evaluate common associations between ISCH and patient demographics/clinical presentation and to analyze the potential for delayed diagnosis.

PATIENT SAMPLE: Patient sample included those diagnosed with ISCH on imaging at our institution from June 20, 2005 to December 3, 2012.

OUTCOME MEASURES: These were based on the patient improvement/stability/decline based on the patients' most recent clinic/office visit when compared with initial presentation.

METHODS: A retrospective search of radiology reports was performed using Illuminate software from June 20, 2005 to December 3, 2012, using the search term "idiopathic spinal cord herniation." Clinical data were reviewed including patient's age, sex, presenting clinical symptoms, number and type of imaging studies performed as part of the workup, other diagnostic tests, pain procedures, surgeries, and time between original presentation and diagnosis of ISCH on imaging.

RESULTS: A total of 55 patients had the search term "idiopathic spinal cord herniation" included in their radiology report, of which 37 patients were found to meet the imaging and clinical diagnosis of ISCH. The median time from presentation to imaging diagnosis was 20 months in patients younger than 60 years and 5 months in those 60 years or older ($p=.02$). Of the 37 patients evaluated, 27 (73%) had no change in symptoms, 5 patients (14%) experienced worsening of symptoms, and 5 (14%) experienced symptom improvement from original presentation to most recent office visit. Among all patients evaluated, three underwent repair of the ventral dural defect in ISCH, resulting in clinical improvement. There was a median of nine outpatient office visits, three magnetic resonance images (MRIs), and one electromyography (EMG) per patient presenting with ISCH. The most frequent complaints were neck/upper back pain in 70%, upper/lower extremity numbness/paresthesias/weakness in 49%, hyperreflexia in 22%, and burning chest pain in 22%.

CONCLUSIONS: Prolonged time to diagnosis and subsequent treatment of ISCH protracts patient symptoms and is associated with redundant diagnostic tests and patient visits. Earlier use of MRI in younger patients (younger than 60 years) may be warranted in those with a clinical presentation suggestive of Brown-Sequard symptomatology. Increasing recognition of ISCH in imaging and

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surgical communities would lead to improved patient care.

Keywords: Idiopathic spinal cord herniation; ISCH; Overutilization; Dural defect; Health care costs; Delayed diagnosis

Introduction

Idiopathic spinal cord herniation (ISCH) was first described by Wortzman et al. [1] in a 1974 case report involving herniation of the thoracic spinal cord resulting in neurologic symptoms. Idiopathic spinal cord herniation is classically described as occurring in middle-aged adults with a preponderance of women [2]. Most cases describe symptoms of Brown-Sequard syndrome, characterized by ipsilateral paralysis, loss of vibratory and position sense, contralateral loss of pain and temperature sensation, or other myelopathic sequelae [3]. Symptoms are typically unilateral from herniation of the lateral funiculus of the spinal cord in ISCH [3].

Recognition of ISCH in the literature has increased over the past decade, likely because of improvements in technique and utilization of spine magnetic resonance imaging (MRI); however, much of this is based on case reports and small series of patients. As such, the entity remains underrecognized and poorly understood, particularly outside of the surgical spine and neuroradiology communities, leading to a significant delay in clinical diagnosis with few cases undergoing surgical repair. One of the major reasons for delay in diagnosis is a wide spectrum of nonspecific and minor symptoms at patient presentation. These delays, in turn, can lead to redundant diagnostic testing, ineffective therapies, and repeat health care visits because of ongoing symptoms. Currently, there is increased focus on improving the quality of health care delivery, with the goal to improve patient outcome and avoid ineffective medical practices. Therefore, increased awareness and appropriate management of underrecognized entities such as ISCH is an efficient way to improve patient care.

Failure to suspect and recognize ISCH can result in delayed diagnosis and unnecessary use of medical resources. The purpose of this study was to evaluate common associations between ISCH and patient demographics and clinical presentation and to analyze potential causes for delayed diagnosis. The goal of this study was to increase awareness of ISCH as a cause of myelopathy, which can be effectively treated, leading to improvement or at least an arrest of patient symptoms.

Materials and methods

Institution review board approval was obtained from Henry Ford Hospital for this Health Insurance Portability and Accountability Actcompliant study, and informed consent was waived. A retrospective search of the Henry Ford Health System radiology database was performed using Illuminate software (Softek Solutions, Inc., Prairie Village, KS, USA) from June 20, 2005 to December 3, 2012 with the search term ‘‘idiopathic spinal cord herniation.’’ The diagnosis of ISCH was based on the final report by a Certificate of Added Qualification(CAQ) board certified neuroradiologist. All patient imaging studies were reviewed again by a post graduate year 4 resident to ensure that an alternative diagnosis such as dorsal arachnoid cyst/web was not identified on computed tomography (CT) myelogram, when performed, and that the imaging findings were consistent with this diagnosis on all subsequent imaging. The criteria for diagnosis were based on the reported findings of Parmar et al. [4] and included an acute anterior kink of the thoracic spinal cord with enlargement of the dorsal subarachnoid space, cord deviation limited to one to two vertebral body levels, adherence of the cord to the ventral dura, and focal thinning of the cord. These imaging findings were also correlated to patient symptoms and considered symptomatic if the cord herniation was ipsilateral to the patient’s symptoms and if the symptoms corresponded to the dermatome and/or myotome for that spine level. There also needed to be a lack of an additional competing spine lesion such as multiple sclerosis, spinal/foraminal stenosis, or tumor. Electronic health records were then reviewed to identify patient demographics, presenting clinical symptoms, office visits, and inpatient days directly related to patients’ symptoms, number and type of imaging studies performed as part of the workup for patients’ myelopathic symptoms, EMGs, pain procedures, spine surgeries, and the period of time between the patients’ original presentation and having received the diagnosis of ISCH on imaging.

Statistical analysis

Descriptive statistics of means, standard deviations, medians, and ranges were computed for age at presentation, delay in diagnosis (in months), utilization, and costs. Percentages and sample sizes were computed for the gender and presenting symptoms. Wilcoxon two-sample *t* tests were done to assess the association between patient characteristics and delay in diagnosis. All testing was done at the 0.05 level. Statistical analyses were performed using SAS version 9.2 (SAS Institute Inc, Cary, NC, USA).

Results

The retrospective search yielded a total of 55 reports including the search term ''idiopathic spinal cord herniation,'' all of which were interpreted by board-certified neuroradiologists. Eighteen of the 55 patients were excluded based on the above imaging criteria by Parmar et al. [4] and/or were not considered symptomatic from ISCH on clinical grounds, yielding 37 patients with the imaging diagnosis of ISCH. Three of these were proven at surgery and five others were confirmed on CT myelogram. Of the 37 patients with suspected ISCH, 19 (51%) were women and 18 (49%) were men, with a median age of symptom onset at 57 years (range 25–82 years).

The most frequent presenting symptom was neck/upper back pain (70%), followed by extremity numbness/paresthesias/weakness (49%), hyperreflexia (22%), and burning chest pain (22%). Classic Brown-Sequard was identified in five of the patients (13%), whereas the remainder had varying degrees of partial Brown-Sequard type symptoms.

The median time from clinical presentation to imaging diagnosis was 17 months (range 0–196 months). The median time to diagnosis in patients younger than 60 years was greater than patients aged 60 years or older (median 20 vs. 5 months, $p=.024$). The associations of time to diagnosis with gender and presenting symptoms were not significant (Table 1).

From the time of initial presentation to the most recent office visit, 27 (73%) patients had no change in symptoms, 5 (14%) worsened, and 5 (14%) showed improvement. Three of the five patients showing improvement underwent surgical ventral dural repair. There were seven spine surgeries, other than dural repair, performed in 5 of the 37 patients before an imaging diagnosis of ISCH was suggested (Table 2). The single patient to show improvement with surgery other than ventral dural repair received a syrinx decompression and posterior thoracic laminectomy.

There was a median of nine outpatient office visits (1-67), in which the chief complaint or admission was related to ISCH symptoms. There was also a median of three spine MRIs (0-23), zero CT/CT myelograms (0-7), and one EMG (0-7) per patient presenting with ISCH. These studies/procedures were ordered in the workup of the patient's original chief complaint before ISCH was diagnosed on imaging (Table 3). There was also a delay in obtaining MRI of the thoracic spine with 62% (23/37) receiving cervical spine MRI and 35% (13/37) receiving lumbar spine MRI before imaging of the thoracic spine in these patients.

Discussion

Although becoming more prevalent in the literature, ISCH remains an underrecognized entity. In this study, the median time to diagnosis from the time of presentation to recognition on imaging was 17 months, with one patient having a time to diagnosis of 16 years. During this period, patients underwent numerous office visits, imaging/nonimaging diagnostic studies, pain procedures, and unnecessary surgeries without any improvement in symptoms. In this series, most patients (86%) showed no clinical improvement, however three out of the five patients showing clinical improvement actually underwent surgical repair of the ventral dural defect. In fact, all patients who underwent dural repair had improvement in their symptoms. These findings are consistent with those of Massicotte et al. [5] which showed no improvement in four patients without surgical intervention and two of three patients improving after repair of the dural defect. Najjar et al. [2] confirmed the importance of early surgical treatment in improving patient outcome. One patient improved with no treatment and the other patient with clinical improvement received a thoracic laminectomy for syrinx decompression.

As stated earlier, all patients undergoing surgical repair of the dura had significant improvement in their clinical

EVIDENCE & METHODS

Context
The authors seek to document associations between idiopathic spinal cord herniation (ISCH) and presentation/demographic factors. The authors maintain that this is an increasingly misdiagnosed clinical entity with substantial delays in proper recognition.

Contribution
The authors present their experience with 37 instances of ISCH. The authors advocate early use of MRI in young patients with a clinical picture that warrants concern for ISCH, especially those that present with idiopathic Brown-Sequard syndrome.

Implications
As a retrospective review, this study can only deal with the clinical experiences and radiographic features of a small sample of patients with ISCH treated at the authors' center. The reader and the authors cannot have access to the details of patients with ISCH who presented elsewhere for care or those who may have had ISCH but failed to be diagnosed with this clinical entity at the authors' own institution. These potentials for confounding limit the capacity for generalizing the guidelines proposed by the authors. Nonetheless, given the rarity of this condition, the findings presented here may be used to increase sensitivity for ISCH although they likely cannot be considered specific to the condition.

—The Editors

Table 1
Associations between time to diagnosis (in months) and patient characteristics

Patient characteristic	N	Median	p
Gender			.651
Female	19	32	
Male	18	15	
Time to diagnosis			.024
<60 y	23	20	
≥60 y	14	5	
Neck/upper back pain			.393
Yes	26	19	
No	11	9	
Extremity numbness/paresthesias/weakness			.287
Yes	18	20	
No	19	11	
Hyperreflexia			.826
Yes	8	19	
No	29	13	
Burning chest pain			.522
Yes	8	24.5	
No	29	13	
"Classic" Brown-Sequard			.424
Yes	5	20	
No	32	13	

symptoms. The first patient to receive dural repair had pain in the left upper back by the scapula, gait abnormality, left Babinski, and hyperreflexia. Surgery showed the cord significantly herniated through a dural defect. Microsurgical technique was used and the cord was untethered only to identify a 13 to 14 mm dural rent ventrally that connected to a pseudomeningocele (Fig. 1). A 6-0 suture was used to repair the ventral dural tear (Fig. 2). The dura was then sealed ventrally, laterally, and dorsally using fibrin glue. The second patient had midthoracic myelopathy, right Babinski, and weakness/pain. Surgery showed the arachnoid to be extremely thickened with tethering of the cord ventrally. This was dissected with microscope anterior to the spinal cord both superiorly and inferiorly releasing all the arachnoid. The dura was closed with 4-0 nylon sutures and artificial dural matrix. The third patient presented with positive Lhermitte, pain in the neck and right arm, numbness, paresthesias, and weakness in right arm. Surgery demonstrated the cord to be tapered acutely forward at the surgical level. Then over the next couple of hours, the cord was rotated from side to side freeing the adhesion, which was scarred intensely, especially on the right side to the anterior dura. The dura was closed in a watertight fashion with artifical dural matrix and spinal sealant.

Idiopathic spinal cord herniation is typically described as having symptoms of Brown-Sequard syndrome [6]. Most patients in our series (70%) had neck/back pain, which was described by Senturk et al. [7] as possible early presentation of ISCH, even without myelopathy. Approximately half of the patients (49%) had upper/lower extremity weakness/numbness/paresthesias and one-quarter experienced hyperreflexia (22%) and burning chest pain (22%). Only 13% had symptoms of "classic" Brown-Sequard syndrome; however, this is not surprising as this is identified with complete hemisection of the spinal cord, whereas ISCH usually only involves herniation of the ventral/lateral spinal cord except in extreme cases. Therefore, most patients will have partial Brown-Sequard type symptoms.

Several different etiologies for ISCH have been proposed, including duplicated dura mater [8,9]. Herniation of the cord through a preexisting congenital dural defect, followed by adherence of the spinal cord to the edge of the dural defect aided by cerebrospinal fluid (CSF) pulsation and an anterior location of the thoracic spinal cord, with additional herniation caused by negative epidural pressure has also been postulated [10].

The typical MRI appearance of ISCH shows ventral kinking of the thoracic cord with obliteration of the anterior CSF space and widening of the CSF space posterior to the cord best seen on T2-weighted imaging (Fig. 3). Although a dorsal arachnoid cyst may mimic this appearance, complete effacement of the ventral CSF is not always seen. Idiopathic spinal cord herniation can be confirmed on CT

Table 2
Symptoms, surgeries, and outcomes for patients undergoing surgical treatment

Case	Age (y)	Sex	Presenting symptoms	Previous surgery for same symptoms	Dural repair	Final outcome
1	82	M	LLE numbness/weakness, urinary incontinence	Laminectomy and syrinx decompression	No	Improved
2	65	F	BUE numbness/tingling/burning	Spinal cord stimulator	No	No change
3	66	M	Midthoracic myelopathy, RLE pain/+ Babinski	None	Yes	Improved
4	39	M	LUE loss of temperature/pain/numbness/weakness	(1) Myelotomy/laminectomy (2) Spinal cord stimulator	No	Worsened
5	44	M	Positive Lhermitte, pain in neck, RUE numbness/paresthesias/weakness	None	Yes	Improved
6	49	M	Neck/middle/low back pain	(1) C4–C7 ACDF (2) L4–L5 laminectomy	No	Unchanged
7	53	M	Gait abnormality, hyperreflexia, + Babinski, back pain	C5–C7 ACDF	Yes	Improved

M, male; F, female; LUE, left upper extremity; LLE, left lower extremity; RUE, right upper extremity; RLE, right lower extremity; BUE, bilateral upper extremities; ACDF, anterior cervical discectomy and fusion.

Table 3
Descriptive statistics for types of utilization

Type of utilization	Median	Mean	SD
Office visit	9	14.5	14.6
Spine MRI	3	3.5	3.8
CT/CT myelography	0	1.1	1.6
Electromyography	1	0.9	1.4
Epidural injections/nerve block	0	1.5	3.7
Surgeries other than dural repair	0	0.2	0.5
Inpatient days	0	1.5	3.1

SD, standard deviation; MRI, magnetic resonance imaging; CT, computed tomography.

myelography revealing no contrast anterior to the cord and lack of filling defect posterior to the cord, therefore excluding dorsal arachnoid cyst (Fig. 4). Brugieres et al. [11] showed the value of phase-contrast cine MRI with CSF pulsations posterior to the cord at the level of kinking to exclude a dorsal subarachnoid cyst. However, the presence of a dorsal arachnoid cyst does not exclude ISCH as first reported by Oe et al [9].

One notable difference identified in our study was the increased time to diagnosis in patients younger than 60 years, with a median of 20 months, versus only 5 months in patients aged 60 years or older ($p=.024$). This is likely because of the earlier use of imaging, such as MRI, in older patients with neurologic complaints and more conservative management being reserved for younger patients.

The major limitation of this study is the reliance on typical imaging findings to diagnose ISCH. Eighteen of the original 55 patients given the possible imaging diagnosis of ISCH either did not meet the strict imaging criteria of ISCH and/or were subsequently found to have other pathology causing their symptoms/imaging appearance. Because of the high false positive rate on MRI alone, this diagnosis should be confirmed with CT myelogram before proceeding to surgery. Myelographic studies were only performed on 5 of the 37 patients to exclude dorsal arachnoid cyst. However, studies have found an association of both dorsal arachnoid cyst and ISCH occurring in the same patient in up to 30% during surgery [12].

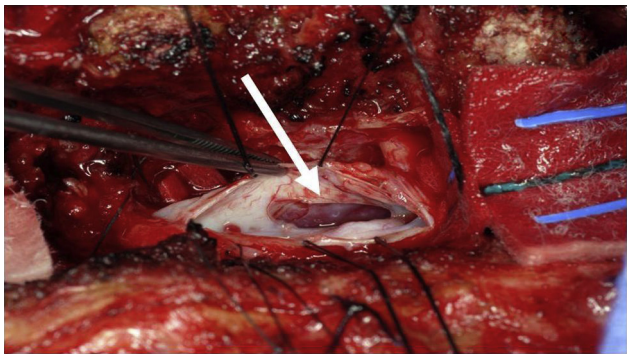

Fig. 1. Intraoperative photograph with arrow pointing to the oval defect in anterior dura mater.

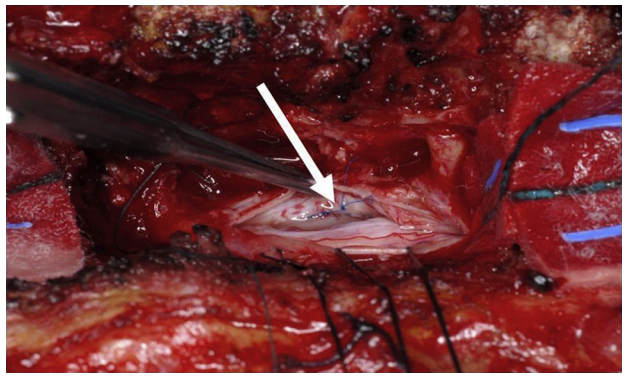

Fig. 2. Intraoperative photograph demonstrating primary closure of dural defect (arrow).

Conclusion

The results of this study confirm the need for increased awareness of ISCH on imaging and as a treatable disease. Delay in diagnosis/treatment prolongs patient symptoms and leads to additional testing and office visits. Earlier use of thoracic spine MRI in younger patients (younger than 60 years) may also be warranted in patients presenting with Brown-Sequard type symptoms. In fact, most of these patients received cervical spine MRI before the thoracic spine was imaged, likely because of nonspecific symptoms. Increasing recognition of this entity among the radiologists and spine surgeons would lead to timely diagnosis and improved patient care.

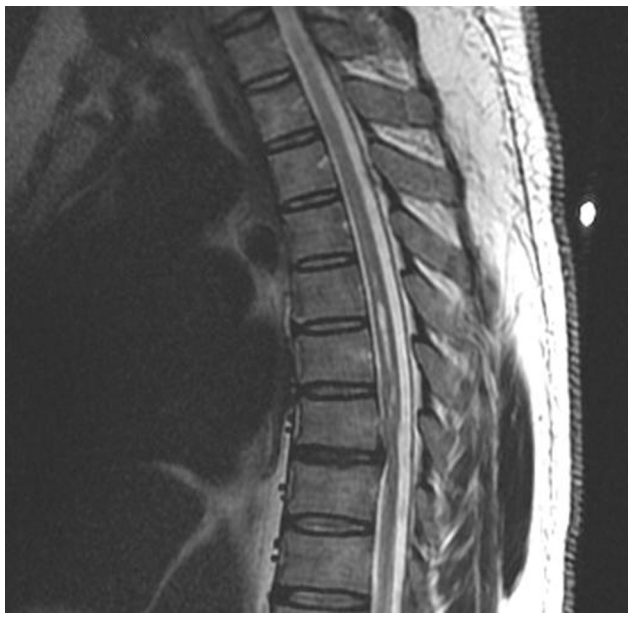

Fig. 3. Sagittal T2 image of the thoracic spine showing anterior kinking of the spinal cord with complete effacement of the ventral cerebrospinal fluid and syrinx formation above this level.

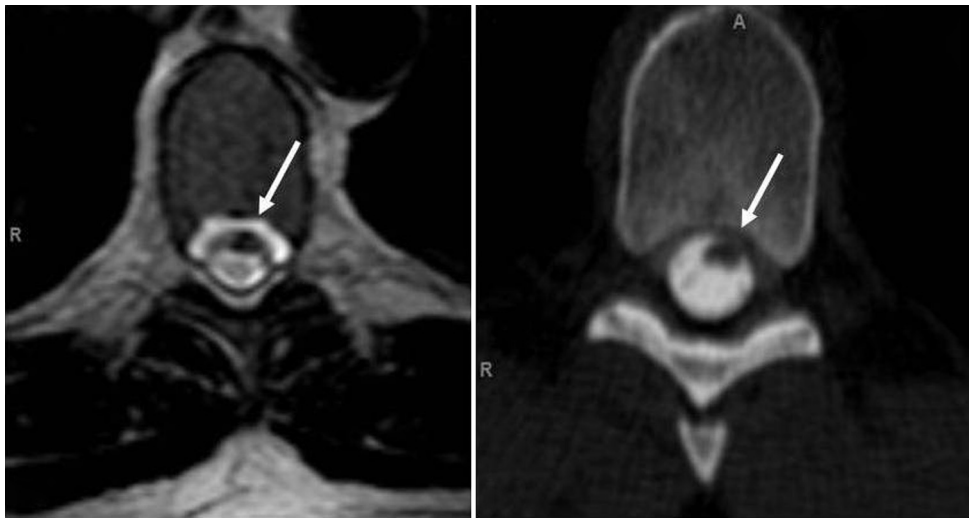

Fig. 4. (Left) Magnetic resonance imaging T2 axial and (right) axial computed tomography myelogram images of the thoracic spine with arrows showing ventrally displaced cord and extradural cerebrospinal fluid/contrast, respectively.

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