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Spine

IDIOPATHIC SPINAL CORD HERNIATION: A NEW THEORY OF PATHOGENESIS

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Najjar MW, Baeesa SS, Lingawi SS. Idiopathic spinal cord herniation: a new theory of pathogenesis. Surg Neurol 2004;62: 161–171.

BACKGOUND

Idiopathic spinal cord herniation is a rare entity that has been described more frequently over the past few years. Its pathophysiology remains obscure, however.

METHODS

We report a case of spinal cord herniation and review the literature extensively. In view of our review, we try to determine the clinical features of the condition and the diagnostic measures used, with emphasis on the role of magnetic resonance (MR) phase-contrast CSF study. The factors affecting the outcome of the condition are also studied including time and type of presentation, as well as the surgical procedure performed. The pathophysiological mechanisms behind spontaneous herniation are discussed, and a new hypothesis is proposed.

RESULTS

Idiopathic spinal cord herniation occurs in the middle-aged adult, with a preponderance of patients being female. Brown-Séquard syndrome is the most common clinical presentation and usually progresses to spastic paraparesis. MRI typically shows a ventral kink in the thoracic cord, with MR phase-contrast imaging proving an important addition to exclude an arachnoid cyst. Better outcomes were noted in the patients treated earlier, and in those with no spasticity. Widening the dural defect seems to afford better results compared to grafting of the defect. The prognosis is favorable after correction, though a vertebral body herniation variant may be associated with worse outcome. In view of the chronology of events and imaging studies in our patient, we hypothesize that herniation occurs as an acquired phenomenon where an inflammatory process results in adherence between the spinal cord and the dura, with erosion, formation of a dural defect, and then later herniation occurring with cerebral spinal fluid (CSF) pulsations.

CONCLUSIONS

Idiopathic herniation of the spinal cord should be recognized and treated early to reach a favorable outcome. It seems to be an acquired condition likely caused by an inflammatory event, the nature of which is to be determined.

KEY WORDS

Spinal cord herniation, Brown-Séquard syndrome, spastic paraparesis, arachnoid cyst, MR phase-contrast imaging.

Idiopathic spinal cord herniation is a rare condition that is being increasingly reported and recognized over the past years. We present one such case that presented with Brown-Séquard syndrome and progressed to parapresis. We found magnetic resonance (MR) phase-contrast imaging of CSF flow to be of special value in the evaluation of these patients. We review the reported cases in the literature and summarize, in the context of this review, the pathophysiological mechanisms behind the condition, as well as diagnostic measures and proposed treatment techniques and their outcome. We propose a new theory on formation of the dural defect other than a congenital cause: an inflammatory reaction leading to cord adhesion and later dural dehiscence.

CASE REPORT

HISTORY AND EXAMINATION

A 32 year-old man had a history dating back to 1991, when he developed pain and hyperesthesia in the upper extremities and the right lower extremity following high-grade fever after septoplasty. He also had weakness in the hand grips bilaterally and diffusely in the right leg. Erythrocyte sedimentation rate was slightly elevated according to the old records. He was treated with nonsteroidal anti-inflammatory agents and physical therapy on the suspicion of postinfective radiculopathy. He improved but presented again in 1994 with weakness in the right leg, apparent to him when he played

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Received March 19, 2003; accepted October 6, 2003.

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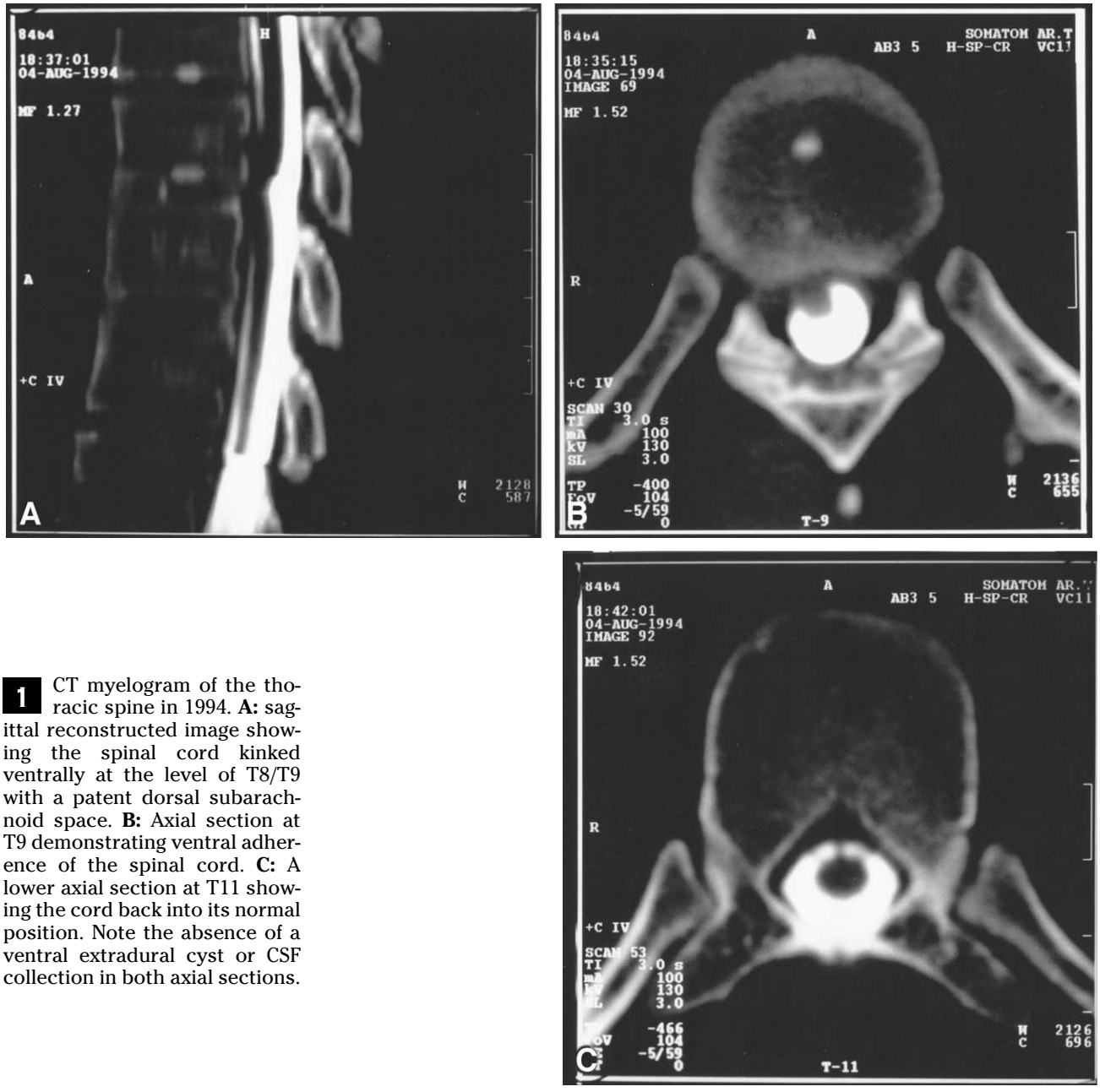
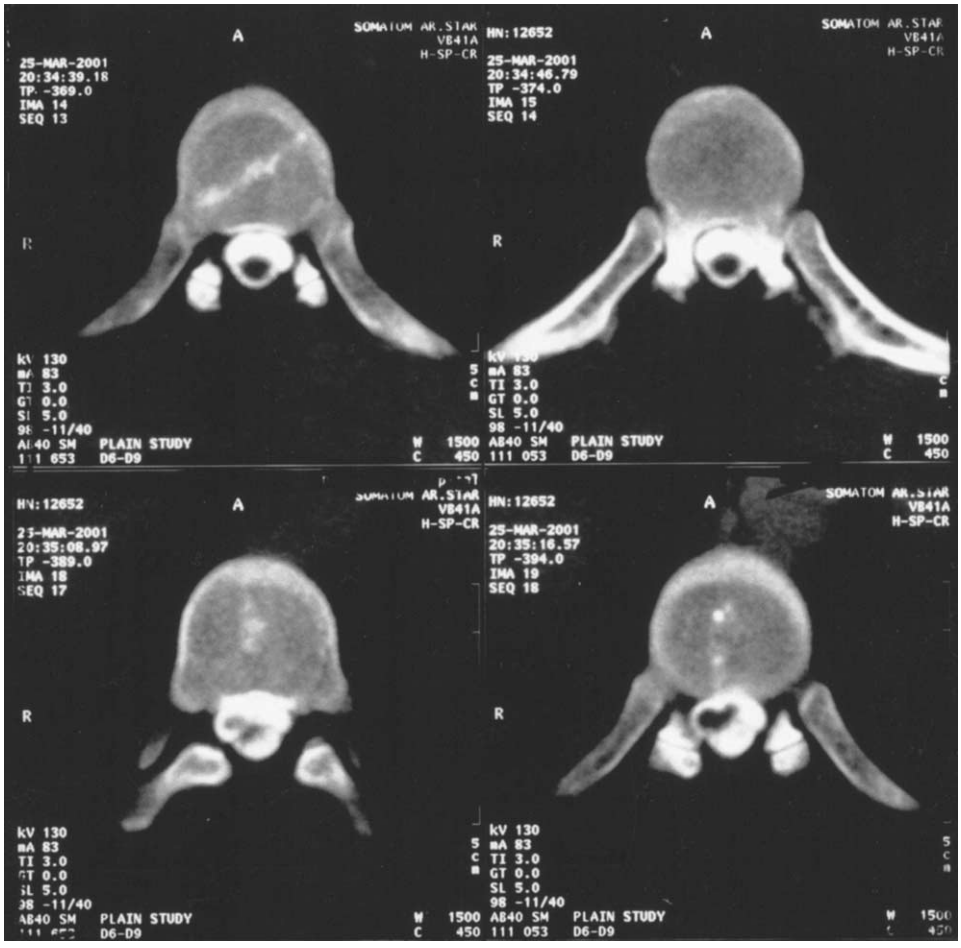
0090-3019/03/$–see front matter
doi:10.1016/j.surneu.2003.10.030



1 CT myelogram of the thoracic spine in 1994. **A:** sagittal reconstructed image showing the spinal cord kinked ventrally at the level of T8/T9 with a patent dorsal subarachnoid space. **B:** Axial section at T9 demonstrating ventral adherence of the spinal cord. **C:** A lower axial section at T11 showing the cord back into its normal position. Note the absence of a ventral extradural cyst or CSF collection in both axial sections.

soccer. Examination showed a Brown-Séquard syndrome below T9, with decreased pain and temperature sensation in the left leg and weakness of the right leg. He had a positive Babinski sign on the right. An MRI was reported as showing deformity of the spinal cord at T8-T9, with atrophy and distortion. A myelogram and later a computed tomography (CT) myelogram in 1994 showed the cord adherent ventrally to the dura at the same level (Figure 1), and he was thought to have had transverse myelitis and was treated conservatively. He continued to have progressive deterioration with difficulty in walking, increased deep tendon reflexes, and 4/5 weakness in the left lower extremity, in addition to the 3/5 weakness on the right. MRI of the dorsal spine was done in 1999 and showed an anterior kink of the spinal cord at T8/9 level, typical, though in retrospect, of idiopathic spinal cord herniation. It was interpreted as an arachnoid cyst compressing the cord, and he was thus operated. The surgeon reported several layers of arachnoid dorsally, raising the suspicion of an old inflammatory process but no arachnoid cyst. He dissected the flattened cord ventrally and reported an ante-

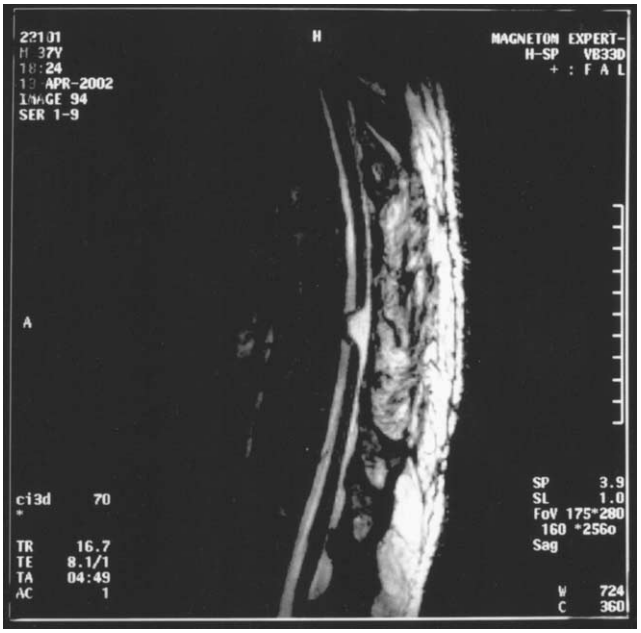


2 CT myelogram of the thoracic spine in 2001. A ventral extradural cystic space has developed. The spinal cord herniates from the intradural compartment into the cystic space through a dural defect. The dorsal subarachnoid space is patent.

rior dural pouch, which was in fact the dural defect, but failed to recognize the pathology and did not reduce the spinal cord. He thought that the patient had an atrophic spinal cord stuck ventrally to the dura. Two postoperative MRIs showed persistence of the same pathology. There was no clinical improvement, but rather gradual deterioration in function of the lower extremities and bladder control. One year later, he had a CT myelogram to rule out a recurrent arachnoid cyst. This showed a patent arachnoid space dorsal to the spinal cord. The defect in the dura is seen, though in retrospect, as well as the formation of an extradural cystic space (Figure 2). The patient was thought to have postinflammatory atrophic changes in the spinal cord and was advised against further surgery. When he came to our attention in 2002, he had become markedly spastic, and barely able to stand on the crutches. He could not walk any more and was bound to the wheelchair. A thin section MRI of the dorsal spine and a phase-contrast study were performed.

NEURORADIOLOGICAL STUDIES

Thin-section MRI of the dorsal spine was performed. It noted anterior deflection of the spinal cord at the T8-T9 level, unchanged from the previous MRIs in 1999 and 2000 (Figure 3). The axial and sagittal 3-D CIS study clearly demonstrated a large defect in the right antero-lateral surface of the thecal sac through which the spinal cord herniated into the anterior epidural space and then went back to the thecal sac. The findings were similar to a CT myelogram done 1 year earlier, showing 2 separate compartments communicating through the defect (Figure 2). Interestingly, these 2 compartments were not existent in the 1994 CT myelogram brought to our attention at a later date. The findings on the older myelogram and their significance as far as pathophysiological mechanisms for formation of



3 Sagittal T2 weighted 3D CIS MR image of the thoracic spine. The spinal cord herniates ventrally in a C shaped fashion at T8/T9 level.

the defect and herniation of the spinal cord will be discussed later.

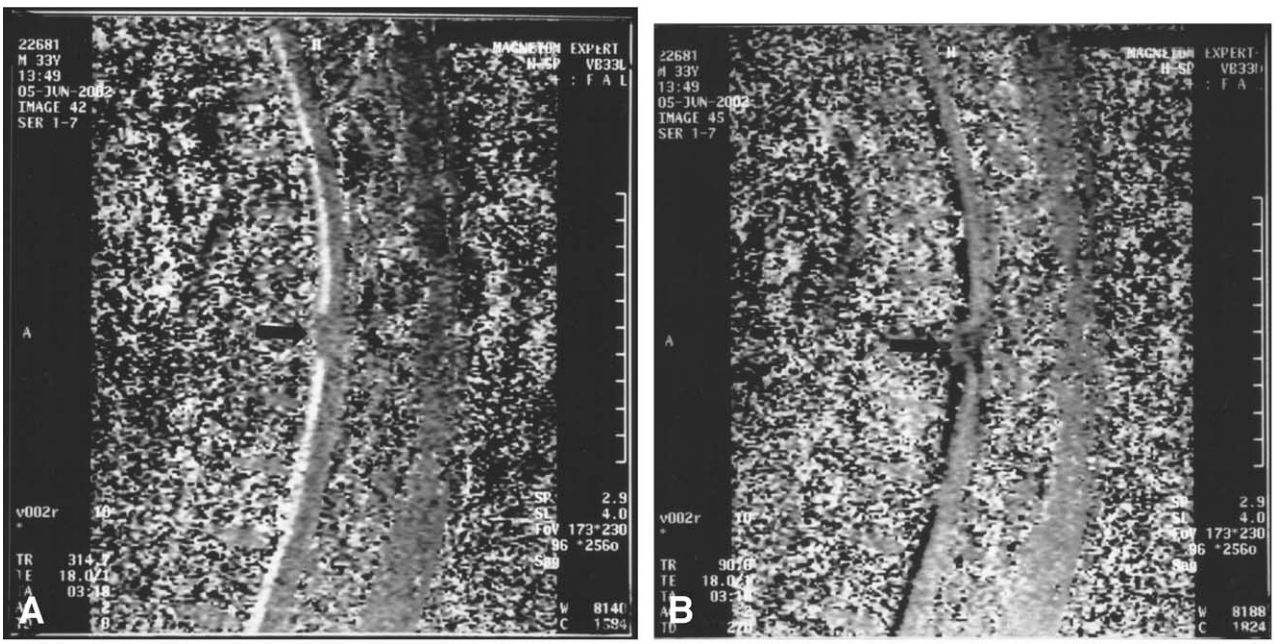
A sagittal and axial phase-contrast cine MR CSF flow study documented interruption of the normal systolic-diastolic flow of the ventral subarachnoid space at the level of the herniation (Figure 4). The CSF flow within the posterior subarachnoid space was continuous with no evidence of any isolated cystic compartment posterior to the cord.

OPERATION

The patient was subjected to revision of his laminectomy at T8-T9 in June 2002 under general anesthesia. The dura and arachnoid were opened again and the spinal cord was seen to be displaced anteriorly and to the right. The spinal cord was turned slightly to reveal the defect. The herniated cord was reduced back into the intradural compartment after dissecting the adhesions between the cord and dural edge sharply. A sheet of polytetrafluroethylene surgical membrane was used to cover the defect and fixed to the dura with 4 stay sutures to prevent recurrence of herniation.

POSTOPERATIVE COURSE

The patient's neurologic condition worsened immediately postoperatively, but then started to improve gradually until he regained his preoperative status after about 4 weeks. He was able to walk with the help of crutches a few weeks later and continues with gradual and slow improvement. Despite the motor improvement, the sensory changes persisted, mostly in the form of numbness in the lower extremities. A postoperative MRI and CSF flow study showed successful reduction of the spinal cord with straightening of the kink at T8-T9, and normal flow pattern ventral and dorsal to the spinal



4 **A** and **B:** Phase-contrast cine MRI of the thoracic spine demonstrating patent flow of CSF in the dorsal subarachnoid space and interrupted flow ventrally at the level of the herniation (arrow).

cord. There was mild increased signal intensity within the cord substance suggestive of myelomalacia.

Discussion

Pathophysiology

Spontaneous ventral herniation of the spinal cord through a previously uninjured dural defect through a dural defect is an exceptional occurrence. A striking similarity in the clinical presentation and radiologic appearance exists among the various cases reported, increasing the index of suspicion among treating physicians and thus the number of cases reported lately. Little is known of the pathophysiology of the condition, however.

We have identified 79 cases (including our case) of idiopathic spinal cord herniation in an extensive review of the literature (Table 1). Most of the cases had been reported over the past 3 years. The first patient was reported by Wortzman et al in 1974, after disclosing the herniation through a thoracic approach and repairing the dural defect [35]. Isu et al, in 1991, postulated that the ventral defect was caused by pressure transmitted from the cord from a dorsal arachnoid cyst and treated both patients reported by excising the arachnoid cyst with some sensory improvement [11]. Since then, various theories to explain the formation of the dural defect have been formulated. The concept of pressure erosion proposed by Isu et al seems unlikely. Arachnoid cysts were reported in association with spinal cord herniation in 14 out of the 79 cases [11,17,18,22,24,31,34]. Most of these cases were early reports. The patient reported by Sioutos et al in 1996 had been previously operated for excision of the arachnoid cyst with no improvement. She improved later after addressing the dural defect and reducing the spinal cord. Thickened arachnoid, raising the suspicion of an arachnoid cyst, has been mentioned in several reports [1,14,26,27,34] and was noted by the neurosurgeon who operated our patient the first time. Releasing the thickened arachnoid and/or resecting the suspected cyst did not cause the spinal cord to move back from its ventral position in both patients reported by Wada et al, and resection of the inner dura to widen the dural defect and treat the herniation was necessary [30]. Although an association with arachnoid cysts is possible, we believe that this does not bear any significance on the pathophysiology of the disease, and the thickened arachnoid seen in many cases may have led to misdiagnosis in some cases [26].

A remote history of trauma is also reported in a few cases [5,28,33,35]. It is difficult to attribute the dural defect and herniation to such remote events, especially that the likelihood of significant trauma to cause a dural tear and no neurologic injury is small [34]. Traumatic and iatrogenic spinal cord herniations are usually posterior rather than anterior and occur in the cervical and dorsal spine [13].

Thoracic disc herniation was thought to be one of the possible causes of herniation by some authors [10,33,35]. The case reported by Miyagushi et al in 2001 is the only case in the literature that is associated with a herniated disc at T3/T4 level visualized at surgery [19]. Several cord herniations occurred at the level of the vertebral body and not the intervertebral disc area [3,5,6,9,10,21,24,34,35]. These were thought to have a worse prognosis in a recent report by Barbagallo et al, where both patients reported had poor clinical outcome after surgery [3].

Another explanation favored by several Japanese authors [2,17,18,21,22] involves the duplication of the ventral dura, with the spinal cord herniating through the inner layer of the duplicated dura. There has been no radiologic or pathologic evidence of such duplication [26]. In the patient reported by Ewald et al, the MRI was normal at initial clinical presentation with a Brown-Séquard syndrome [9]. There was no duplication of the dura nor any ventral cysts, though a defect in the dura cannot be ruled out. One and a half years later, and with progression of the symptomatology, the cord was found adherent to the ventral dura at the level of T6. Six months later, spinal cord herniation was seen on the third MRI. We had a similar experience with the CT myelogram done in 1994 (Figure 1) showing adherence of the spinal cord ventrally but no duplication or ventral cyst. In 1999, 2001, and later 2002, there was a clear ventral CSF filled space into which the cord had herniated through the dural defect (Figure 2). These findings argue against another theory proposed by Wortzman et al and Masuzawa et al, where they postulated that idiopathic spinal cord herniation results from the spinal cord herniating into a preexisting ventral meningocele [16,35]. They also argue against a congenital extradural arachnoid cyst as proposed by Kumar et al [12]. In addition to the late presentation of most of these patients (middle aged adults), there were no spinal deformities such as spina bifida in any of the cases reported in the literature. Therefore, an acquired phenomenon, apparently restricted to one occurring in adult life and invariably associated with de novo symptoms, is favored [3]. Progressive herniation from normal

1 Summary of Reported Cases of Spinal Cord Herniation

AUTHORS	AGE	SEX	SYMPTOMS	DURATION	LEVEL	PROCEDURE	OUTCOME
Wortzman et al, 1974	63	M	BS	1.5 yr	T 7	Direct suture	W-IM
Masuzawa et al, 1981	36	M	BS	1 yr	T 4/5	Graft	IM, IS
Oe et al, 1990	61	M	BS	NA	T 4/5	Defect widening	Same
Isu et al, 1991	43	F	BS	1 yr	T 5/6	Arachnoid cyst resection	IS
	45	F	SP	1.5 yr	T 2/3	Arachnoid cyst resection	IS
Tronnier et al, 1991	45	F	BS	3 yr	T 3/4	Patch	W-IS
Nakazawa et al, 1993	43	F	BS	5 yr	T 2	Defect widening	IM+S
	39	F	BS	3 yr	T 4/5	Defect widening	IM
White and Firth, 1994	61	F	BS	1.5 yr	T 4	Graft	Same
	39	M	SP	1.5 yr	T 8	Graft	Same
Kumar et al, 1995	38	M	BS	3 yr	T 7/8	Direct suture	IM+S
Borges et al, 1995	68	F	BS	12 yr	T 7	Direct closure	IM+S
	69	M	BS	8 yr	T 2/3	Direct closure	IM
	48	F	BS	10 yr	T 7	Direct closure	IM
Batzdorf et al, 1995	23	F	BS	2 yr	T 6/8	Patch	IM
Hausmann and Moseley, 1996	56	F	BS	8 yr	T 6	Direct closure	Same
	36	M	SP	10 yr	T 6/7	Resection of hernia	Worse
	51	F	BS	1 yr	T 6/7	Hernia not confirmed surgically	Same
	49	M	SP	3 yr	T 4/5	Hernia not confirmed surgically	Same
Matsumara et al, 1996	63	F	SP	NA	T 3/4	Defect widening	I
Miura et al, 1996	49	M	BS	13 m	T 5/6	Defect widening	IM
Urbach et al, 1996	44	M	Sensory	2 yr	T 5/6	Laminectomy	I
Sioutos at al, 1996	34	F	BS	3 yr	T 7	Graft	W-IM
Slavotinek et al, 1996	22	F	BS	4 yr	T 5	Laminectomy	I
Uchino et al, 1997	71	F	BS	2 yr	T 4/5	Direct closure	Same
	61	F	BS	2 yr	T 6	Direct closure	NS
Baur et al, 1997	66	F	BS	7 yr	T 10	Direct closure	IM+S
Takahashi et al, 1997	57	M	BS	NA	T 2/3	Graft	NA
	56	F	SP	NA	T 3/4	Graft	NA
	68	M	SP	NA	T 7/8	Graft	NA
Miyake et al, 1998	45	F	BS	3 yr	T 3/4	Patch	IM+S
	53	M	BS	6 yr	T 2/3	Patch	IM
Dix et al, 1998	44	F	BS	NS	T 7/8	Graft	IM
Watters et al, 1998	55	F	BS	10 yr	T 3/4	Direct closure	IM
Vallée et al, 1999	28	F	BS	2 yr	T 3/4	Defect widening	IM
	58	F	BS	6 yr	T 4/5	Patch	IM
	40	F	BS	2 yr	T 5/6	Patch	Same
	49	F	BS	4 yr	T 4/5	Patch	Same
Brugières et al, 1999	54	F	BS	5 yrs	T 6	Direct closure+Biopsy	W-I
	70	M	BS	6 m	T 5/6	Direct closure	IS
Marshman et al, 1999	55	F	BS — SP	14 yr	T 7/8	Patch	IM
Abe et al, 1999	58	M	BS	4 yr	T 7/8	Defect widening	IM
Verny et al, 1999	28	F	SP	18 m	T 3/4	NA	I
	58	F	BS	NA	T 4/5	NA	I
Tekkök et al, 2000	49	F	BS	3 yr	T 3/4	Patch	IM
Ewald et al, 2000	51	F	BS	2 yr	T 6	Patch	IS
Wada et al, 2000	59	M	BS	4 yr	T 4/5	Defect widening	IM
	63	F	BS	10 yr	T 3/4	Defect widening	IM
	48	M	BS	2 yr	T 5/6	Defect widening	IM
Pereira et al, 2001	55	M	SP	4 yr	T 2/3	Fill defect w Teflon	IM+S
Miyagushi et al, 2001	54	F	BS	2 yr	T 3/4	Graft	I
Berbel et al, 2001	56	M	BS	NA	T	Could not be reduced	Same

1 continued

AUTHORS	AGE	SEX	SYMPTOMS	DURATION	LEVEL	PROCEDURE	OUTCOME
Morkoff et al, 2001	33	F	BS	NA	NA	NA	I
Egushi et al, 2001	54	F	SP	NA	T 4/5	Laminectomy	Same
Aizawa et al, 2001	44	M	BS	5 yr	T 8/9	Defect widening	IM
	60	F	BS	3 yr	T 4/5	Defect widening	IM
	59	F	BS	20 yr	T 4/5	Defect widening	IM
Watanabe et al, 2001	43	F	BS	5 yr	T 4	Defect widening	I
	39	F	BS	3 yr	T 3	Defect widening	I
	54	F	BS	4 yr	T 4	Defect widening	I
	71	F	SP	5 yr	T 4	Defect widening	Worse
	49	M	BS	5 yr	T 4	Defect widening	I
	47	F	BS	5 yr	T 5	Defect widening	I
	78	F	SP	4 yr	T 4	Defect widening	I
	56	M	BS	2 yr	T 6	Defect widening	I
	47	M	SP	3 yr	T 3	Defect widening	I
Cellerini et al, 2002	53	M	BS	1 yr	T 8/9	Patch	IM
	37	F	BS	6 m	T 4/5	Patch	IM+S
Massicotte et al, 2002	63	M	BS	14 yr	T 5/6	Observation	Same
	39	F	BS	Yrs	T 6/7	Patch	IM+S
	50	M	numbness	6 yr	T 4	Observation	Same
	44	F	SP	11 yr	T 5/6	Patch	Same
	33	F	BS	2 yr	T 7/8	Observation	Same
	57	F	SP	8 yr	T 6	Patch	Same
	27	M	BS	1 yr	T 9	Patch	IM
	46	F	BS	2 yr	T 4	Observation	Same
Giuseppe et al, 2002	28	F	SP	5 yr	T 6	Patch	W-I
	64	M	SP	4 yr	T 8	Patch	Worse
Najjar et al	32	M	SP	8 yr	T 8/9	Patch	W-IM

BS = Brown-Séquard syndrome; SP = spastic paraparesis; I = improved; IM = improved motor function; IS = improved sensation; IM+S = improved motor and sensory; W-I(M or S) = worse then improved (motor or sensory); NS = not specified; NA = not available.

anatomy in Ewald's case and from ventral adherence in our case supports this concept.

We would like to propose a new mechanism behind the formation of the dural defect and thus later spinal cord herniation. We have to take into consideration the fact that the spinal cord lies ventrally in the thoracic canal, and that all cases of spontaneous spinal cord herniation were ventral or ventrolateral. It seems likely that an inflammatory process involving the spinal cord and/or the meninges is the initial event that leads to ventral adhesion of the cord to the dura (because of its normal position). The initial event may be asymptomatic clinically or cause mild clinical symptomatology, but symptoms gradually worsen as the spinal cord is tethered and later starts to herniate through a dural defect probably caused by the inflammation, adhesion and the pulsatility of the now fixed spinal cord. Adhesions are invariably encountered at the dural edge of the dural defect and prevented reduction of the herniated cord in the case reported in 2001 by Berbel et al [4]. Dorsally thickened arachnoid has been cited by many authors [1,14,26,27,34]. It was thought that this thickening was secondary to inflammation resulting from incarceration of the cord [34]. We have demonstrated that ventral adherence of the cord occurred before herniation, and thus think that the inflammatory reaction causing thickening of the arachnoid takes place early in the pathogenesis of the disease. It seems likely that pulsations of the now stuck spinal cord may cause erosion of the dura and the formation of a dural tear or defect. The cord then progressively herniates through the defect, and pulsating CSF seeping around the herniating cord causes the formation and gradual enlargement of an anterior extradural cyst. The growth of the extradural cyst is demonstrated comparing the MRI done in 1999 to that done in 2002 in our patient. The cyst was not present in the 1994 CT myelogram. Intraoperatively, the ventral wall of the cyst was covered with a whitish membrane. Several authors [12,35] have reported a similar finding of pseudocapsular tissue rather than dura, in further support of our hypothesis. Histologic diagnosis of the ventral cyst wall is needed, however, for verification. Histologic assessment of the dural edge, on the other hand, had been performed by several authors and showed normal dura in all the cases [2,9,20,31].

CLINICAL PRESENTATION

Idiopathic spinal cord herniation typically occurs in middle-aged adults. The median age in all 79 cases reported was 49.88 years or 50 years, with the youngest patient being 22 years of age and the oldest 78 years. There is a female preponderance with 50 female patients compared to 29 males, and a ratio of slightly less than 2:1. The most common clinical presentation was that of a Brown-Séquard syndrome (75% of the patients), which slowly progresses, as seen in our patient and in other reports [9,14], into a spastic mono- or paraparesis, suggesting a predominant involvement of the anterolateral funiculus. The herniated funiculus is incarcerated in the dural defect, and may appear as a tumor-like nodule resulting in biopsy [9,10,16,24, 28], or even frank resection [12] in earlier cases. This is to be avoided to prevent postoperative neurologic deterioration. Histologic sections had shown degenerated spinal cord tissue in all the cases. Patients usually presented within few and up to 20 years in the 3rd case reported by Aizawa et al. The mean duration of symptomatology was 4.25 years for patients who came in with a Brown-Séquard syndrome and 5.34 years for those presenting with spastic paraparesis, an expected finding. Posterior column involvement as presenting symptomatology has been recorded [12,24], and Barbagello et al reported a patient with bilateral sensory level affecting all modalities and associated with paraparesis [3]. Sphincter dysfunction is also frequently cited in idiopathic spinal cord herniation [12,18,24,28,35].

RADIOLOGIC EVALUATION

Idiopathic or spontaneous herniation of the spinal cord occurred in the thoracic spine in all the cases reported. The herniation is either ventral or ventrolateral through an oval dural defect. MRI findings are typical showing on the sagittal scan an anterior C-shaped kink of the spinal cord, together with secondary enlargement of the dorsal subarachnoid spaces. The spinal cord may be thinned because of atrophy or may show signal changes. The axial MRI scans may show the dural defect in addition to the herniation. A “double cord” sign may be a clue to herniation of the cord rather than presence of an arachnoid cyst. Phase-contrast cine MRI is crucial to exclude a posterior compressing arachnoid cyst and establish the diagnosis [1,6,7,20,23,29]. A normal CSF pulsatile pattern is observed within the dorsal subarachnoid spaces, whereas this pattern is absent on the ventral side, at the level of the herniation. Together with an MRI scan, phase-contrast cine MRI provides the diagnosis without the need for an invasive diagnostic measure such as CT myelography.

MANAGEMENT

There are two main treatment strategies. One is closure of the defect after reducing the spinal cord, and the other is simply widening the aperture to prevent strangulation of the cord. Isu et al treated the suspected arachnoid cyst and reported some improvement [11]. This has not been the case in other cases [24,25] where the suspected arachnoid cyst was treated including our patient who continued to deteriorate until the spinal cord herniation was addressed. Tekkök believes that the idea of an association between arachnoid cysts and spinal cord herniation is mistaken and that the cases reported, actually involved enlarged CSF spaces with loosened arachnoid membranes [26].

Direct closure of the dural defect was first performed by Wortzman et al through an anterior approach. It was later done via a laminectomy exposure with acceptable results. To avoid excessive manipulation of the cord, grafting the dura with fascia or synthetic patch was recommended by several authors [3,7–9,12,14,15,19,24,26,27,34]. Duraplasty to accommodate the mass-like swollen cord after reducing it has also been performed. Another surgical method involves widening of the dural defect through a laminectomy exposure to relieve the strangulated cord. It was first described by Nakazawa et al who thought that resecting the inner layer of the duplicated dura frees the cord and reverses the symptomatolgy [21]. Watanabe et al argue that this procedure is easier and safer than repairing the dural defect with sutures [32]. They recommend resection of the dural ring to be done after release of the spinal cord to avoid neurologic deterioration. Pereira et al describe a unique technique of filling the defect with a piece of Teflon® and then fixing it with fibrin glue with a favorable result [23].

Conservative observant management is another option in some patients with mild symptomatology. Massicotte et al report on 8 cases of idiopathic spinal cord herniation where 4 of them were stable on observant treatment, and they recommend microsurgical repair in patients with progressive neurologic deterioration [15]. We feel that the presence of long tract signs, even if seemingly stable, warrant surgical intervention. All 4 patients who underwent observant treatment had sensory disturbances only, with no other signs.

OUTCOME

The outcome is favorable in most cases with Brown-Séquard symptoms (41 out of 46 patients, i.e., 90%). Most of the patients had improvement in motor symptoms with residual sensory deficit or numbness. Patients who presented with spastic paraparesis had a smaller chance of improvement (69%), with the spasticity benefiting the least. There was initial worsening of neurologic function followed by improvement in several cases, including our patient [3,6,24,27,35]. We arbitrarily divided the patients into those presenting with 3 years or less of symptomatology, and those with more than 3 years of symptoms. The chance of having a good outcome was 86% and 77%, respectively, implying that the earlier we treat the herniation, the better the outcome. Widening the dural aperture was the surgical technique associated with the best outcome in the series reported in the literature (90% improved compared with 70% in grafting). Less manipulation of the already compromised spinal cord is one possible explanation. A retrospective or prospective analysis with other variables being accounted for is needed to determine the best surgical technique. Finally, a recent report by Barbagallo et al described 2 cases with spinal cord herniation at the vertebral body level, where the outcome was less favorable [3]. Our analysis supports their hypothesis; these patients may present a distinct subgroup whose prognosis is less satisfactory. One of their patients, in addition to 2 other patients reported in the literature, had herniation into a vertebral body cavity [10,35]. This was associated with an atypical presentation and poor prognosis. It may also support our hypothesis of an inflammatory process causing erosion through the dura and later the vertebral body.

CONCLUSION

In summary, idiopathic or spontaneous spinal cord herniation is a rare entity that is being increasingly recognized and reported. It presents in the middle aged adult as Brown-Séquard syndrome and usually progresses into spastic paraparesis. MRI findings are typical, showing a ventral or ventrolateral kink of the spinal cord in the thoracic spine. Phase-contrast cine MR is very useful in excluding an arachnoid cyst, the most common mistaken diagnosis. The treatment is usually surgical by either grafting the dural defect through which the spinal cord herniates or simply widening it. The pathophysiology behind formation of the defect is not understood. We report a case of spinal cord herniation and review the literature extensively. In view of the chronology of events in our patient and other patients reviewed, we think that idiopathic spinal cord herniation is an acquired phenomenon and propose a new hypothesis whereby an inflammatory event or process may cause the spinal cord to adhere to the dura ventrally. Together with CSF pulsations, this event may cause erosion of the dura and formation of a dural defect, with progressive herniation of the cord thereafter. The nature of the initial inflammatory insult is to be determined.

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COMMENTARIES

The authors present a case of idiopathic spontaneous spinal cord herniation, an entity that is being diagnosed with increasing frequency. I was interested in the fact that their patient was originally diagnosed as having "transverse myelitis," even though the imaging studies were clearly abnormal. This misdiagnosis occurred in 1994, but we have recently seen a patient with identical studies who was similarly misdiagnosed last year at a major teaching hospital. I agree that patients with a progressive deficit should have surgery, but ours has only nonprogressive sensory symptoms and a normal examination, so we are treating her nonoperatively at present. Whether the authors' theory regarding pathogenesis is correct is unclear, but it is an interesting idea.

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Najjar et al report on the clinical details of a patient with spinal cord herniation at T8–9. They are to be commended for their thorough reinvestigation of this unfortunate patient in 2002 and their recognition that he still had an untreated spinal cord herniation, despite prior surgery, that changed the imaging findings on the CT-myelogram. Their careful investigation and reoperation led to clinical improvement in the patient's neurological function.

The authors also review the literature on this topic and help to increase the overall awareness of this clinical condition, which often goes undiagnosed, even by experienced neurologists and neurosurgeons.

The authors propose a new hypothesis regarding the pathogenesis of this problem. They propose that an unknown inflammatory condition develops at the level of the spine at which the herniation will develop. This inflammation causes the ventral spi-