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Case Report

Idiopathic Ventral Spinal Cord Herniation: An Illustrative Case and Literature Review

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ABSTRACT

Background and Importance: Idiopathic spinal cord herniation (ISCH), or spontaneous spinal cord herniation, is a rare but serious condition that can cause progressive myelopathy and irreversible neurological deficits if left untreated. The condition is marked by the gradual herniation of the spinal cord through a ventral defect in the dura, leading to compromised blood flow and neurological deficits. Common symptoms include Brown-Séquard syndrome or asymmetrical paraparesis. Treatment options typically focus on reducing the strangulated spinal cord and closing the dural defect with a synthetic patch.

Case Presentation: We present the case of an adult woman with progressive asymmetrical weakness of the lower limbs compatible with spastic paraparesis. Thoracic magnetic resonance imaging (MRI) revealed characteristic features of ISCH at the T3-T4 level. Intraoperative neurophysiological monitoring was used during the surgical intervention, which involved a 3-level laminectomy, dura opening, excision of the dentate ligament, and reduction of the cord across the dural defect. The defect was then filled with an autogenous piece of muscle, followed by the closure of the defect with an artificial dural patch and dural closure. At the 6-month follow-up, the patient showed favorable improvement.

Conclusion: Patients with slowly progressive paraparesis or Brown-Séquard syndrome should consider the possibility of ISCH as a potential cause, despite its rarity. In symptomatic cases, the preferred treatment option often involves reducing the incarcerated spinal cord followed by covering the dural defect.

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Highlights

- ISCH is a rare cause of slowly progressive thoracic myelopathy.
- The clinical presentation of ISCH typically includes slowly progressive paraparesis or Brown-Séquard syndrome.
- ISCH can be diagnosed based on its characteristic MRI features, which include ventral displacement of the spinal cord and an enlarged dorsal subarachnoid space that mimics an intradural arachnoid cyst.
- In symptomatic cases of ISCH with progressive neurological deficits, surgical treatment involves reducing the spinal cord and covering the dural defect with a synthetic dural patch.

Plain Language Summary

Idiopathic spinal cord herniation (ISCH) is a rare condition that happens when the spinal cord and its protective covering protrude through a hole in the protective layer of the spine. This most commonly occurs in the middle of the thoracic spine, between the T2 and T8 levels. Symptoms can include weakness or paralysis of the legs, loss of sensation on one side of the body, or other neurological problems. It is important to diagnose and treat ISCH quickly to avoid further damage to the nervous system. Treatment usually involves surgery to move the herniated spinal cord back into place and repair the hole in the protective layer of the spine.

1. Background and Importance

Idiopathic spinal cord herniation (ISCH) is a rare but potentially treatable cause of thoracic myelopathy that mainly affects middle-aged adults, with a higher incidence in females [1-5]. It typically occurs in the thoracic spine, between the T2 and T8 levels. With the advent of magnetic resonance imaging (MRI) and increased awareness among neurosurgeons and radiologists, there has been a growing recognition and reporting of this rare medical condition [6-9]. Since Wortzman and colleagues first reported a case in 1974, over 250 cases have been published, including two case reports from Iran [10-18]. Taghipour et al. reported the first Iranian case in 2004, and Farrokhi et al. reported a second case in 2023, both from Shiraz [19, 20]. Clinical findings of this condition are typically nonspecific, and patients commonly present with slowly progressive paraparesis or Brown-Séquard syndrome [10-20].

In order to achieve maximum reversal of the neurological deficits, it is mandatory to relocate the herniated cord to its normal intradural position and close the dural defect. We present the third instance of idiopathic spinal cord herniation in an Iranian patient in this report, along with an overview of the literature concerning the diagnosis and treatment of this disorder [11-18].

2. Case Presentation

A 53-year-old woman with a three-year history of predominantly right-sided lower limb weakness and an unsteady gait is presented. These symptoms had become more pronounced in the last three months. Upon examination, apparent weakness of both lower extremities was noted, associated with bilateral lower limb exaggerated reflexes, bilateral Babinski signs, and an American Spinal Injury Association (ASIA) impairment score of C, along with a vague sensory level up to T4. Her MRI showed clear abnormal anterior displacement of the spinal cord at the T3/T4 level (Figures 1A and 1B). A computed tomography (CT) myelogram was utilized to exclude cord displacement caused by an arachnoid cyst, demonstrating the free passage of dye posterior to the displaced spinal cord (Figures 2A and 2B).

After induction of anesthesia and intraoperative neurophysiological monitoring, the patient was placed on the operating table, and the T2/T3 level was determined by fluoroscopy. This was followed by a 2-level laminectomy and dural opening. The dentate ligaments were then incised, and with subsequent slight rotation of the cord, reduction of the herniated segment of the spinal cord was performed (Figure 3A). Thereafter, the dural defect was filled with an autologous free muscle graft (Figure 3B). The defect was covered with a long

synthetic dural patch, and spinal cord wrapping was the final step of the surgery before dural closure.

The postoperative period was uneventful. At the 6-month follow-up, the patient's neurological condition had significantly improved with an American Spinal Injury Association (ASIA) impairment score of D.

3. Discussion

ISCH is a rare etiology of progressive thoracic myelopathy, with approximately 250 cases documented in the literature [11-20]. It is more common in middle age patients and more common in women than men, with a mean age of 49 years (20-75 years) [11-18]. Ventral dural herniation typically occurs between the T2 and T8 levels of the thoracic spine, with the most common location being the mid-thoracic vertebral column [11-18]. The preponderance of disease manifestation in the thoracic spine can be explained by a combination of factors, including the inherent thoracic kyphosis, which is a normal anatomical curvature of the spine, the natural anterior positioning of the spinal cord within the thoracic cavity, the physiological movement of the thoracic spinal cord as a result of heart pulsations, and the effects of flexion and extension on the spinal cord.

Pathogenesis. Several theories have been proposed to elucidate the pathophysiological basis of this phenomenon, encompassing congenital dural deficiency, post-traumatic dural injury, pressure erosion of the dura mater by osteophytes, as well as adhesions between the anterior thoracic spinal cord and surrounding structures. [21-24]. However, despite these theories, the pathogenesis of spontaneous ventral dural defects and eventual spinal cord herniation has remained controversial.

Clinical Presentation. Most patients typically show signs of progressive myelopathy, which initially presents with spastic paraparesis or Brown-Séquard syndrome [1-6, 11-18, 25]. In a minority, the pathology may present with sensory disorders, such as pain, numbness, or sensory dissociation. Rarely, the condition can be diagnosed incidentally in MRI taken for other reasons.

Imaging. At present, MRI represents the most efficacious modality for precise diagnostic evaluation of ISCH. Notably, a typical hallmark of ISCH is represented by ventral angulation of the thoracic spinal cord, concomitant with an enlargement of the subarachnoid space posterior to the cord, as visualized on MRI scans [6-9, 26-29]. Since the latter imaging feature is similar to an intradural arachnoid cyst, for a definite diagnosis, phase-contrast MRI can be used in which the presence of dorsal pulsatile cerebrospinal flow is in favor of ISCH [30]. Computerized tomography myelography (CTM) represents a potentially useful alternative modality for the diagnostic assessment of ISCH. In computerized tomography myelography (CTM), ventral displacement of the spinal cord and the presence of contrast agent blockage or defects are suggestive of the presence of an intradural arachnoid cyst.

Treatment. The treatment of ISCH has been approached through both conservative and surgical interventions. However, the optimal management strategy for this condition remains unclear, as the natural history of ISCH has yet to be fully elucidated. Therefore, individualized treatment plans should be formulated by physicians to address the unique needs of each patient. Generally, patients presenting with minimal neurological deficits are recommended for conservative management, along with periodic neurological evaluations [17]. In the literature, a conservative treatment strategy has been advocated for patients with minimal and stable neurological symptoms and has been reported in approximately 20 cases [17].

The primary aim of surgical intervention for ISCH is to release the entrapped spinal cord and prevent the recurrence of herniation. To this end, various surgical techniques have been described, including the expansion of the dural defect, direct repair of the dural defect through suturing, and the use of autologous muscle packing in conjunction with artificial dural patching, with or without cord wrapping [1-5, 11-16, 18, 19, 31-36]. The optimal surgical approach for ISCH is still a matter of controversy. Nevertheless, the utilization of an artificial dural patch, with or without cord wrapping, has potential benefits, such as reduced spinal cord manipulation during surgery. However, in order to achieve a secure reduction of the herniated spinal cord without any associated neurological complications, it is imperative to possess a thorough understanding of the underlying pathological anatomy and to have extensive experience in the application of microsurgical techniques. [11-16, 18, 19, 31-36].

Intraoperative monitoring (IOM) is essential in the surgical treatment of ISCH, and neuromonitoring, in particular, transcranial motor evoked potentials can help prevent intraoperative gross spinal cord manipulation and inadvertent iatrogenic neurological deficits [37].

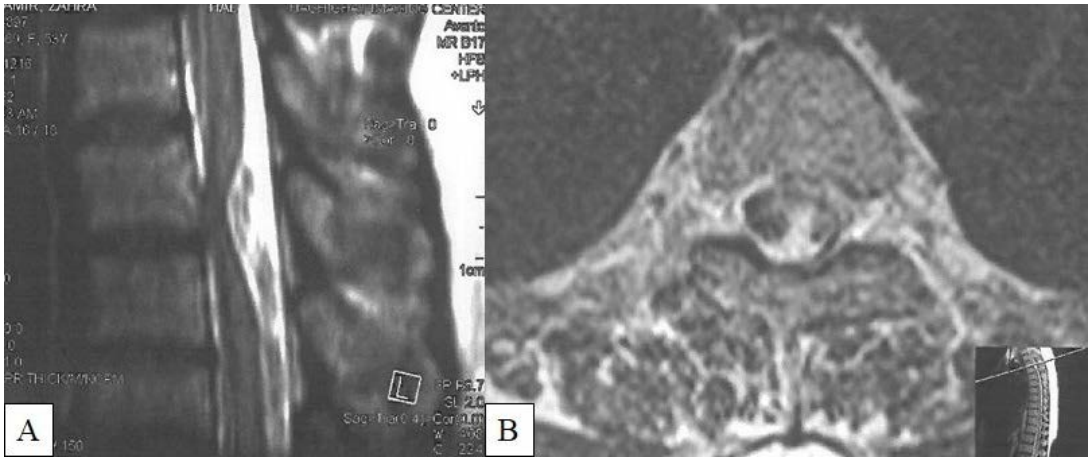

Figure 1. Pre-operative sagittal (A) and axial (B) magnetic resonance imaging (MRI) T2W images showing clear abnormal anterior displacement of the spinal cord at the T3/T4 level, a C-shaped kink, and atrophy

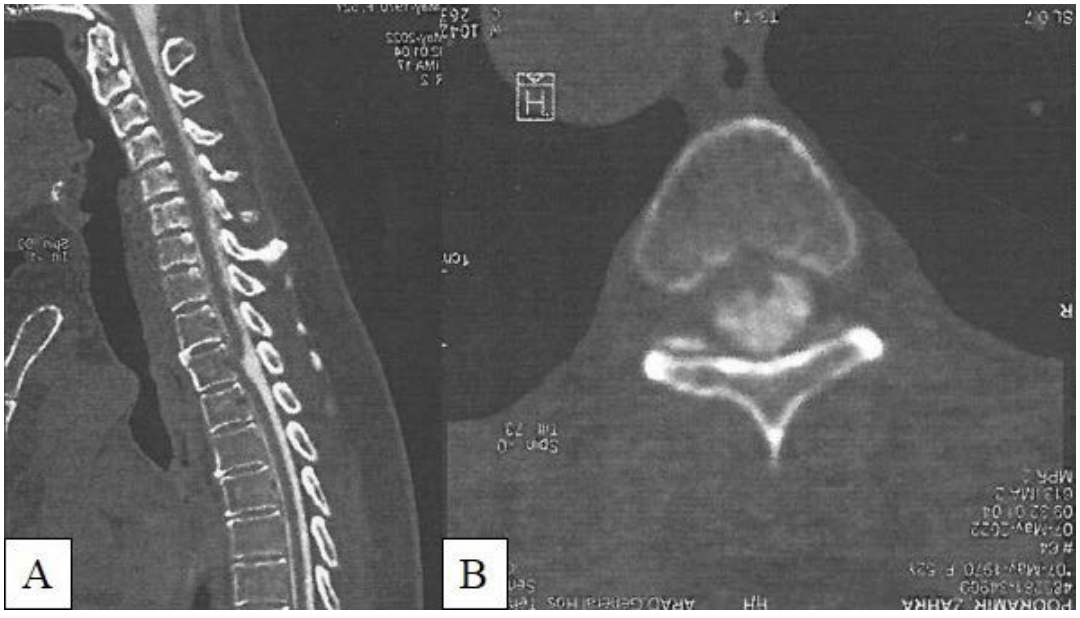

Figure 2. Pre-operative sagittal (A) and axial (B) thoracic CT myelography images utilized to exclude cord displacement caused by an arachnoid cyst, demonstrating free passage of dye posterior to the displaced spinal cord

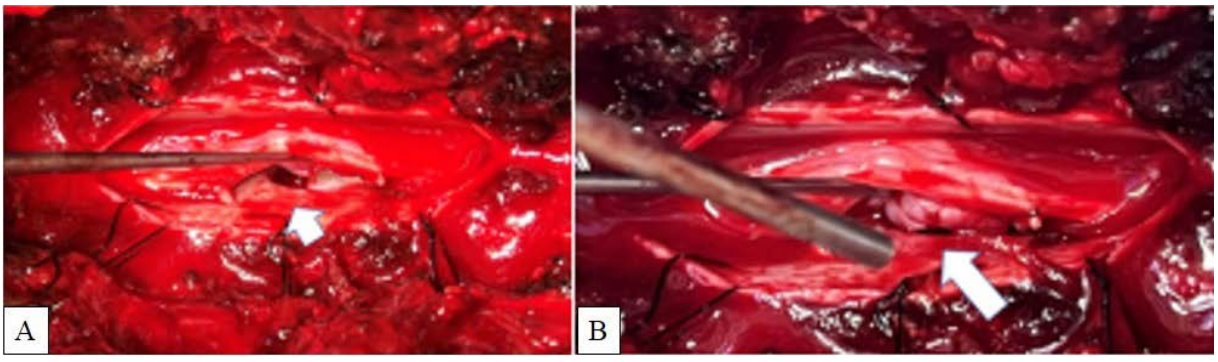

Figure 3. Intraoperative photographs of the spinal cord showing the dural defect exposed by a slight rotation of the cord (A), followed by the reduction of the herniated segment of the spinal cord (B)

Outcome. According to Summers et al., 74% of the surgically treated cases improve postoperatively where 24% remain unchanged and 8% become worse [38]. Surprisingly, the ultimate outcome and neurological improvement are more favorable in the patients presented with Brown Séquard syndrome than those with spastic paraparesis [11-16, 18, 19, 31-40].

4. Conclusion

This case highlights the importance of considering ISCH in patients presenting with slowly progressive myelopathy, such as paraparesis or Brown-Séquard syndrome. The MRI features of ventral displacement of the spinal cord and an enlarged dorsal subarachnoid space are pathognomonic for ISCH. Surgical intervention with reduction of the spinal cord and coverage of the dural defect with a synthetic dural patch is the treatment of choice in symptomatic cases with progressive neurological deficits.

From a medico-legal standpoint, it is essential to inform patients and their relatives that with proper and early surgical intervention, only 75% of patients improve, while in 25% of cases, symptoms may remain unchanged or worsen. Approximately 20% of cases show no change in neurological status and the disease stabilizes. Unfortunately, in 5% of cases, the condition worsens after surgery.

Ethical Considerations

Compliance with ethical guidelines

Ethical approval was waived by the local Ethics Committee in view of the retrospective nature of the study and all the procedures performed were part of the routine care. No information (names, initials, hospital identification numbers, or photographs) exists in the submitted manuscript that can be used to identify patients.

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Authors' contributions

Conception and design: Abolfazl Rahimizadeh, Saber Zafarshamspour; Data collection: Saber Zafarshamspour, Mahan Amirzadeh, Shaghayegh Rahimizadeh; Drafting the article: Saber Zafarshamspour, Walter L. Williamson; Critically revising the article: Abolfazl Rahimizadeh, Walter L. Williamson; Reviewing the submitted version of the manuscript: All authors; Approving the final version of the manuscript: All authors.

Conflict of interest

The authors declared no conflict of interest.

References

[1] Masuzawa H, Nakayama H, Shitara N, Suzuki T. Spinal cord herniation into a congenital extradural arachnoid cyst causing Brown-Se'quard syndrome. Journal of Neurosurgery. 1981; 55(6):983-6. [DOI:10.3171/jns.1981.55.6.0983] [PMID]

[2] Tronnier VM, Steinmetz A, Albert FK, Scharf J, Kunze S. Hernia of the spinal cord: Case report and review of the literature. Neurosurgery. 1991; 29(6):916-9. [DOI:10.1227/00006123-199112000-00021] [PMID]

[3] Rando TA, Fishman RA. Spontaneous intracranial hypotension: report of two cases and review of the literature. Neurology. 1992; 42(3 Pt 1):481-7. [DOI:10.1212/WNL.42.3.481] [PMID]

[4] Nakazawa H, Toyama Y, Satomi K, Fujimura Y, Hirabayashi K. Idiopathic spinal cord herniation: Report of two cases and review of the literature. Spine. 1993; 18(14):2138-41. [DOI:10.1097/00007632-199310001-00036] [PMID]

[5] Borges LF, Zervas NT, Lehrich JR. Idiopathic spinal cord herniation: A treatable cause of the Brown-Sequard syndrome -case report. Neurosurgery. 1995; 36(5):1028-32; discussion 1032-3. [DOI:10.1097/00006123-199505000-00023] [PMID]

[6] Sioutos P, Arbit E, Tsairis P, Gargan R. Spontaneous thoracic spinal cord herniation: A case report. Spine. 1996; 21(14):1710-3. [DOI:10.1097/00007632-199607150-00019] [PMID]

[7] Slavotinek JP, Sage MR, Brophy BP. An unusual spinal intradural arachnoid cyst. Neuroradiology. 1996; 38(2):152-4. [DOI:10.1007/BF00604803] [PMID]

[8] Watters MR, Stears JC, Osborn AG, Turner GE, Burton BS, Lillehei K, et al. Transdural spinal cord herniation: Imaging and clinical spectra. AJNR. American Journal of Neuroradiology. 1998; 19(7):1337-44. [PMID]

[9] Dix JE, Griffitt W, Yates C, Johnson B. Spontaneous thoracic spinal cord herniation through an anterior dural defect. AJNR. American Journal of Neuroradiology. 1998; 19(7):1345-8. [PMID]

[10] Wortzman G, Tasker RR, Rewcastle NB, Richardson JC, Pearson FG. Spontaneous incarcerated herniation of the spinal cord into a vertebral body: A unique cause of paraplegia. Case report. Journal of Neurosurgery. 1974; 41(5):631-5. [DOI:10.3171/jns.1974.41.5.0631] [PMID]

[11] Wada E, Yonenobu K, Kang J. Idiopathic spinal cord herniation: Report of three cases and review of the literature. Spine (Phila Pa 1976). 2000; 25(15):1984-8. [DOI:10.1097/00007632-200008010-00019] [PMID]

[12] Cho JY, Rhim SC, Jeon SR, Roh SW: Ventral transdural spinal cord herniation: case report and review of the literature. Korean J Spine. 2006; 3(4): 246-249. [Link]

[13] Groen RJ, Middel B, Meilof JF, de Vos-van de Biezenbos JB, Enting RH, Coppes MH, et al. Operative treatment of anterior thoracic spinal cord herniation: Three new cases and an individual patient data meta-analysis of 126 case reports. Neurosurgery. 2009; 64(3 Suppl):ons145-59; discussion ons159-60. [DOI:10.1227/01.NEU.0000327686.99072.E7] [PMID]

[14] Ghostine S, Baron EM, Perri B, Jacobson P, Morsette D, Hsu FP. Thoacic cord herniation through a dural defect: Description of a case and review of the literature. Surgical Neurology. 2009; 71(3):362-6, discussion 366-7. [DOI:10.1016/j.surneu.2007.08.022] [PMID]

[15] Prada F, Saladino A, Giombini S, Erbetta A, Saini M, DiMeco F, et al. Spinal cord herniation: Management and outcome in a series of 12 consecutive patients and review of the literature. Acta Neurochirurgica. 2012; 154(4):723-30. [PMID]

[16] Payer M, Zumsteg D, De Tribolet N, Wetzel S. Surgical management of thoracic idiopathic spinal cord herniation. Technical case report and review. Acta Neurochirurgica. 2016; 158(8):1579-82. [DOI:10.1007/s00701-016-2840-2] [PMID]

[17] Randhawa PS, Roark C, Case D, Seinfeld J. Idiopathic spinal cord herniation associated with a thoracic disc herniation: Case report, surgical video, and literature review. Clinical Spine Surgery. 2020; 33(6):222-9. [DOI:10.1097/BSD.0000000000000896] [PMID] [PMCID]

[18] Runza G, Maffei E, Cademartiri F. Idiopathic herniation of the thoracic spinal cord. Acta Bio-Medica: Atenei Parmensis. 2021; 92(Supp 1):e2021143. [PMID]

[19] Taghipour M, Zamanizadeh B, Zare Z, Haghnegahdar A, Javadi S. Herniation of the spinal cord: Case report and review of the literature. Neurosurgery Quarterly. 2004; 14(2)105-107. [DOI:10.1097/01.wnq.0000126696.94879.8c]

[20] Farrokhi MR, Mousavi SR, Rafieossadat R. IIdiopathic spinal cord herniation at the cervicothoracic junction level presenting with unilateral sensory symptoms. Clinical Neurology and Neurosurgery. 2023; ;224:107526. [DOI:10.1016/j.clineuro.2022.107526] [PMID]

[21] Miyaguchi M, Nakamura H, Shakudo M, Inoue Y, Yamano Y. Idiopathic spinal cord herniation associated with intervertebral disc extrusion: A case report and review of the literature. Spine (Phila Pa 1976). 2001; 26(9):1090-4. [DOI:10.1097/00007632-200105010-00022] [PMID]

[22] Najjar MW, Baeesa SS, Lingawi SS. Idiopathic spinal cord herniation: A new theory of pathogenesis. Surgical Neurology. 2004; 62(2):161-70; discussion 170-1. [DOI:10.1016/j.surneu.2003.10.030] [PMID]

[23] Bartels RHMA, Brunner H, Hosman A, van Alfen N, Grotenhuis JA. The pathogenesis of ventral idiopathic herniation of the spinal cord: A hypothesis based on the review of the literature. Frontiers in Neurology. 2017; 8:476 [DOI:10.3389/fneur.2017.00476] [PMID] [PMCID]

[24] Shimizu S, Kobayashi Y, Oka H, Kumabe T. Idiopathic spinal cord herniation: Consideration of its pathogenesis based on the histopathology of the dura mater. European Spine Journal. 2019; 28(2):298-305. [DOI:10.1007/s00586-017-5147-y] [PMID]

[25] Sasani M, Ozer AF, Vural M, Sarioglu AC. Idiopathic spinal cord herniation: Case report and review of the literature. The Journal of Spinal Cord Medicine. 2009; 32(1):86-94. [DOI:10.1080/10790268.2009.11760757] [PMID] [PMCID]

[26] Ferré JC, Carsin-Nicol B, Hamlat A, Carsin M, Morandi X. MR imaging features of idiopathic thoracic spinal cord herniations using combined 3D-fiesta and 2D-PC Cine techniques. Journal of Neuroradiology. 2005; 32(2):125-30. [DOI:10.1016/S0150-9861(05)83127-X] [PMID]

[27] Blasel S, Hattingen E, Baas H, Zanella F, Weidauer S. Spontaneous spinal cord herniation: MR imaging and clinical features in six cases. Clinical Neuroradiology. 2008; 18(4):224-30. [Link]

[28] Ishida M, Maeda M, Kasai Y, Uchida A, Takeda K. Idiopathic spinal cord herniation through the inner layer of duplicated anterior dura: Evaluation with high-resolution 3D MRI.Journal of Clinical Neuroscience. 2008; 15(8):933-7. [DOI:10.1016/j.jocn.2006.10.024] [PMID]

[29] Carter BJ, Griffith BD, Schultz LR, Abdulhak MM, Newman DS, Jain R. Idiopathic spinal cord herniation: An imaging diagnosis with a significant delay. The Spine Journal. 2015; 15(9):1943-8. [DOI:10.1016/j.spinee.2015.04.013] [PMID]

[30] Brugières P, Malapert D, Adle-Biassette H, Fuerxer F, Djindjian M, Gaston A. Idiopathic spinal cord herniation: Value of MR phase-contrast imaging. AJNR. American Journal of Neuroradiology. 1999; 20(5):935-9. [PMID]

[31] Miura Y, Mimatsu K, Matsuyama Y, Yoneda M, Iwata H. Idiopathic spinal cord herniation. Neuroradiology. 1996; 38(2):155-6. [DOI:10.1007/BF00604805] [PMID]

[32] Watanabe M, Chiba K, Matsumoto M, Maruiwa H, Fujimura Y, Toyama Y. Surgical management of idiopathic spinal cord herniation: A review of nine cases treated by the enlargement of the dural defect. Journal of Neurosurgery. 2001; 95(2 Suppl):169-72. [DOI:10.3171/spi.2001.95.2.0169] [PMID]

[33] Barrenechea IJ, Lesser JB, Gidekel AL, Turjanski L, Perin NI. Diagnosis and treatment of spinal cord herniation: A combined experience. Journal of Neurosurgery. Spine. 2006; 5(4):294-302. [DOI:10.3171/spi.2006.5.4.294] [PMID]

[34] Chaichana KL, Sciubba DM, Li KW, Gokaslan ZL. Surgical management of thoracic spinal cord herniation: Technical consideration. Journal of Spinal Disorders & Techniques. 2009; 22(1):67-72. [DOI:10.1097/BSD.0b013e318165fe2a] [PMID]

[35] Batzdorf U, Holly LT. Idiopathic thoracic spinal cord herniation: Report of 10 patients and description of surgical approach. Journal of Spinal Disorders & Techniques. 2012; 25(3):157-62. [DOI:10.1097/BSD.0b013e31823d79f6] [PMID]

[36] Hawasli AH, Ray WZ, Wright NM. Symptomatic thoracic spinal cord herniation: Case series and technical report. Neurosurgery. 2014; 10 Suppl 3(0 3):E498-504; discussion E504. [DOI:10.1227/NEU.0000000000000437] [PMID] [PMCID]

[37] Novak K, Widhalm G, de Camargo AB, Perin N, Jallo G, Knosp E, et al. The value of intraoperative motor evoked potential monitoring during surgical intervention for thoracic idiopathic spinal cord herniation. Journal of Neurosurgery. Spine. 2012; 16(2):114-26. [DOI:10.3171/2011.10.SPINE11109] [PMID]

[38] Summers JC, Balasubramani YV, Chan PC, Rosenfeld JV. Idiopathic spinal cord herniation: Clinical review and report of three cases. Asian Journal of Neurosurgery. 2013;8(2):97-105. [DOI:10.4103/1793-5482.116386] [PMID] [PMCID]

[39] Maira G, Denaro L, Doglietto F, Mangiola A, Colosimo C. Idiopathic spinal cord herniation: Diagnostic, surgical, and follow-up data obtained in five cases. Journal of Neurosurgery. Spine. 2006; 4(1):10-9. [DOI:10.3171/spi.2006.4.1.10] [PMID]

[40] Hassler W, Al-Kahlout E, Schick U. Spontaneous herniation of the spinal cord: Operative technique and follow-up in 10 cases. Journal of Neurosurgery. Spine. 2008; 9(5):438-43. [DOI:10.3171/SPI.2008.9.11.438] [PMID]