Idiopathic herniation of the thoracic spinal cord

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Abstract. Since 1974, when Wortzman et al were the first to describe a case of idiopathic spinal cord herniation (ISCH), the number of reported cases has increased owing to magnetic resonance imaging (MRI) now is routinely available for patients with myelopathy and spinal surgeons are becoming more familiar with this clinical entity. This extremely rare herniation occurs exclusively in the thoracic spine, causing slowly progressive myelopathy. Diagnosis is based on ventral displacement of the spinal cord in the thoracic spine. MRI is the technique of choice to exclude a posterior arachnoid cyst, the most common mistaken diagnosis, and to recognize a spinal cord herniation when an anterior dural defect is present. A case of ISCH is reported and a Literature review of this clinical entity often mis-diagnosed has been obtained. (www.actabiomedica.it)

Key words: Idiopathic Spinal Cord Herniation, Brown-Séquard Syndrome, Magnetic Resonance Imaging

Introduction

Since 1974, when Wortzman et al were the first to describe a case of idiopathic spinal cord herniation (ISCH) (1), the concept of ISCH has gradually been appreciated, and the number of reported cases has increased owing to magnetic resonance imaging (MRI) now is routinely available for patients with myelopathy and spinal surgeons are becoming more familiar with this clinical entity. This extremely rare herniation occurs exclusively in the thoracic spine, causing slowly progressive myelopathy. Diagnosis is based on ventral displacement of the spinal cord observed on sagittal MRI. Surgical reduction of the herniated spinal cord usually improves the myelopathic condition (2). The idiopathic or spontaneous aetiologies are different from documented traumatic or postoperative causes (3). Despite the existence, to our knowledge, at least 133 ISCH cases (Table 1) have been reported in the international Literature, whose 14 in the radiological Literature, misdiagnosis and delayed diagnosis remain a major concern (1-60).

With this article we report another case of ISCH of the thoracic tract and provide a thorough review of the Literature about the clinical-radiological correlation to better recognize and characterize this entity.

Etiopathogenesis

ISCH can be classified on the basis of its aetiology into 4 groups: idiopathic (12, 14, 21, 23, 26, 27, 30, 32, 34, 39, 40), iatrogenic (29, 61, 62), post-traumatic (12, 26, 63, 64), and post-inflammatory (37).

The primitive etiopathogenetic mechanism consists in pre-existing dural defect trough which an arachnoid cyst causing pressure. Actually, the real etiopathogenesis still is unknown (14, 17, 25, 38, 65-67). There have been many hypotheses about the cause of the dural defect as it exists congenitally: a dorsally existing arachnoid cyst causing pressure (23), an unrecognized traumatic event (23, 32, 46, 53, 60), and compression by a thoracic disc prolapse (60).
| No | Author     | Year | Journal                          | Cj/Rj | N of Cases | Age/Sex | Symptoms | Level | mRX | CTm | MRI |
|----|------------|------|----------------------------------|-------|------------|---------|----------|-------|-----|-----|-----|
| 1  | Cobb       | 1973 | J Neurosurg                      | Cj    | NAA        |         |          |       |     |     |     |
| 2  | Hoffman    | 1973 | J Neurosurg                      | Cj    | NAA        |         |          |       |     |     |     |
| 3  | Wortzman   | 1974 | J Neurosurg                      | Cj    | 1          | 63/m    | BS       | D7    | X   |     |     |
| 4  | Masuzawa   | 1981 | J Neurosurg                      | Cj    | 1          | 36/f    | BS       | D4-5  | X   |     |     |
| 5  | Chan*      | 1985 | Neurosurgery                     | Cj    | NR         | NR      | NR       | NR    | NR  | NR  | NR  |
| 6  | Mizuno     | 1986 | No Schinkei Geka                 | Cj    | 1          | 55/m    | SpP      | C6-7  | X   |     |     |
| 7  | Alvisi*    | 1987 | J Neurosurg                      | Cj    | 17         | -       | -        | -     | -   | -   | -   |
| 8  | Andrews*   | 1988 | J Neurosurg                      | Cj    | 5          | NR      | NR       | D     | X   | X   | X   |
| 9  | Oe*        | 1990 | Nippon Seikeigeka Gakkai Zasshi  | Cj    | 1          | 61/m    | SpP      | D4-5  | -   | -   | -   |
| 10 | Isu        | 1991 | Neurosurgery                     | Cj    | 2          | 43/f    | BS       | D5-6  | X   |     |     |
| 11 | Tronnier   | 1991 | Neurosurgery                     | Cj    | 1          | 45/f    | S Def    | D3-4  | X   | ?   |     |
| 12 | Nakazawa   | 1993 | Spine                            | Cj    | 2          | 43/f    | BS       | D2    | X   | X   |     |
| 13 | White      | 1994 | J Neurol Neurosurg Psychiatry    | Cj    | 2          | 61/f    | BS       | D4    | -   | -   | -   |
| 14 | White      | 1994 | J Neurol Neurosurg Psychiatry    | Cj    | 2          | 39/m    | SpP      | D8    | -   | -   | -   |
| 15 | Borges     | 1995 | Neurosurgery                     | Cj    | 3          | 68/f    | BS       | D7-8  | X   | X   |     |
| 16 | Oe*        | 1995 | Neurosurgery                     | Cj    | 2          | 69/m    | BS       | D2-3  | X   | X   |     |
| 17 | Sahl       | 1995 | Rofo                             | Rj    | NAA        | -       | -        | -     | -   | -   | -   |
| 18 | Hausmann*  | 1996 | Neuroradiology                   | Rj    | 4          | 56/f    | BS       | D6    | -   | -   | -   |
| 19 | Batzdorf   | 1995 | Neurosurgery                     | Cj    | 1          | 23/f    | BS       | D6-7  | -   | -   | -   |
| 20 | Kumar      | 1995 | J Neurosurg                      | Cj    | 1          | 38/m    | BS       | D7-8  | X   | X   |     |
| 21 | Matsumura  | 1996 | Rinsho Schinkeigaku              | Cj    | 1          | 63/f    | BS       | D3-4  | X   | X   |     |
| 22 | Miura      | 1996 | Neuroradiology                   | Rj    | 1          | 49/m    | SpP      | D5-6  | X   | X   |     |
| 23 | Siatous    | 1996 | Spine                            | Cj    | 1          | 34/f    | SpP      | D6-7  | X   |     |     |
| 24 | Slavotinek | 1996 | Neuroradiology                   | Rj    | 1          | 22/f    | BS       | D5    |     |     |     |
| 25 | Urbach     | 1996 | Neuroradiology                   | Rj    | 1          | 44/m    | S Def    | D5-6  |     |     |     |
| 26 | Baur       | 1997 | Eur Radiol                       | Rj    | 1          | 66/f    | BS       | D10   | X   | X   | X   |
| 27 | Lee*       | 1997 | British Journal of Neurosurgery  | Cj    | 1          | 19/m    | Paraplegia | -   | -   | -   | -   |
| 28 | Takahashi  | 1997 | Spine Spinal Cord                | Cj    | 3          | 57/m    | BS       | D2-3  | -   | -   | -   |
| 29 | Henry      | 1997 | Arch Phys Med Rehabil            | Cj    | 1          | 30/f    | BS       | D7    | X   |     |     |
| 30 | Uchi      | 1997 | Eur Radiol                       | Rj    | 2          | 71/f    | BS       | D4-5  | X   | X   |     |
| 31 | Dix        | 1998 | AJNR                             | Rj    | 1          | 44/f    | BS       | D7-8  | X   | X   |     |

(continued)
| N° | Author   | Year | Journal                                                                 | Cj/Rj | N. of Cases | Age/ Sex | Symptoms | Level | mRX | CTm | MRI |
|----|----------|------|-------------------------------------------------------------------------|-------|-------------|----------|----------|-------|-----|-----|-----|
| 41 | Miyake   | 1998 | J Neursurg                                                              | Cj    | 2           | 45/f     | BS       | D3-4  | X    | X   |     |
| 42 |         |      |                                                                         |       |             | 53/m     | BS       | D2-3  | X    | X   |     |
| 43 | Watters  | 1998 | AJNR                                                                     | Rj    | 1           | 55/f     | BS       | D3-4  | X    | X   |     |
| 44 | Abe      | 1999 | J orthop sci                                                            | Cj    | 1           | 58/     | BS       | D7-8  | X    | X   | X   |
| 45 | Brugieres | 1999 | AJNR                                                                     | Rj    | 2           | 54/f     | BS       | D6    |     |     |     |
| 46 |         |      |                                                                         |       |             | 70/m     | BS       | D5-6  |     |     | X   |
| 47 | Marshman | 1999 | Neurosurgery                                                            | Cj    | 1           | 55/f     | BS-SpP   | D8    |     |     | X   |
| 48 | Vallee   | 1999 | Acta Neurochir (Wien)                                                   | Cj    | 4           | 28/f     | BS       | D3-4  | X    | X   |     |
| 49 |         |      |                                                                         |       |             | 58/f     | BS       | D4-5  | X    | X   |     |
| 50 |         |      |                                                                         |       |             | 40/f     | BS       | D5-6  | X    | X   |     |
| 51 |         |      |                                                                         |       |             | 49/f     | BS       | D4-5  | X    | X   |     |
| 52 | Verny    | 1999 | Neurochirurgie                                                          | Cj    | 2           | 28/f     | SpP      | D3-4  |     |     | X   |
| 53 |         |      |                                                                         |       |             | 58/f     | BS       | D4-5  |     |     | X   |
| 54 | Bartolomei | 2000 | Neurosurgery                                                            | Cj    | 1           | 61/f     | BS       | D3-4  | -    | -   | -   |
| 55 | Ewald    | 2000 | Neurosurgery                                                            | Cj    | 1           | 51/f     | BS       | D5-6  |     |     | X   |
| 56 | Martin   | 2000 | J Clin Neurosci                                                         | Cj    | 1           | 31/f     | BS       | D8    |     |     | X   |
| 57 | Tekkok   | 2000 | Neurosurgery                                                            | Cj    | 1           | 49/f     | BS       | D3-4  | X    | X   |     |
| 58 | Wada     | 2000 | Spine                                                                   | Cj    | 3           | 59/m     | BS       | D4-5  | X    | X   |     |
| 59 |         |      |                                                                         |       |             | 63/f     | BS       | D3-4  | X    | X   |     |
| 60 |         |      |                                                                         |       |             | 48/m     | BS       | D5-6  | X    | X   |     |
| 61 | Adams*   | 2001 | Neuroradiology                                                          | Rj    | 1           | NR       | BS       | NR    | NR   | NR  | NR  |
| 62 | Aizawa   | 2001 | Spine                                                                   | Cj    | 3           | 44/m     | BS       | D8-9  | X    | X   |     |
| 63 |         |      |                                                                         |       |             | 60/f     | BS       | D4-5  | X    | X   |     |
| 64 |         |      |                                                                         |       |             | 59/f     | BS       | D4-5  | X    | X   |     |
| 65 | Berbel   | 2001 | Rev Neurol                                                              | Cj    | 1           | 56/m     | BS       | NA    |     |     | X   |
| 66 | Eguchi   | 2001 | Neurol Med Chir                                                          | Cj    | 1           | 54/f     | SpP     | D4-5  | X    | X   |     |
| 67 | Kawachi  | 2001 | Neurology                                                               | Cj    | 1           | 53/m     | BS       | D10   | X    | X   |     |
| 68 | Miyaguchi| 2001 | Spine                                                                   | Cj    | 1           | 54/f     | BS       | D3-4  | X    | X   |     |
| 69 | Morokoff | 2001 | J Clin Neurosci                                                         | Cj    | 1           | 33/f     | BS       | D8    |     |     | X   |
| 70 | Pereira  | 2001 | Acta Neurochir (Wien)                                                   | Cj    | 1           | 55/m     | BS       | D2-3  |     |     | X   |
| 71 | Watanabe | 2001 | J Neurosurg 95                                                          | Cj    | 9           | 43/f     | BS       | D4    | X    | X   |     |
| 72 |         |      |                                                                         |       |             | 39/f     | BS       | D3    | X    | X   |     |
| 73 |         |      |                                                                         |       |             | 54/f     | BS       | D4    | X    | X   |     |
| 74 |         |      |                                                                         |       |             | 71/f     | SpP     | D4    | X    | X   |     |
| 75 |         |      |                                                                         |       |             | 49/m     | BS       | D4    | X    | X   |     |
| 76 |         |      |                                                                         |       |             | 47/f     | BS       | D5    | X    | X   |     |
| 77 |         |      |                                                                         |       |             | 78/f     | SpP     | D4    | X    | X   |     |
| 78 |         |      |                                                                         |       |             | 56/m     | BS       | D6    | X    | X   |     |
| 79 |         |      |                                                                         |       |             | 47/f     | SpP     | D3    | X    | X   |     |

(continued)
| N° | Author       | Year | Journal                          | Cj/ Rj | N. of Cases | Age/ Sex | Symptoms | Level | mRX | CTm | MRI |
|----|--------------|------|----------------------------------|--------|-------------|----------|----------|-------|-----|-----|-----|
| 80 | Barbagallo*  | 2002 | J Neurosurg                      | Cj 2   | 2           | 28/f     | SpP      | D6    | -   | -   | -   |
| 81 |              |      |                                  |        |             | 64/m     | SpP      | D8    | -   | -   | -   |
| 82 | Cellerini    | 2002 | Acta Neurochir (Wien)            | Cj 2   | 2           | 53/m     | BS       | D8    | X   | X   |     |
| 83 |              |      |                                  |        |             | 37/f     | BS       | D4-5  | X   |     |     |
| 84 | Iyer*        | 2002 | Br J Neurosurg                   | Cj 1   | 1           | 63/m     | BS       | D5-6  | X   |     |     |
| 85 |              |      |                                  |        |             | 39/f     | BS       | D6-7  |     |     |     |
| 86 |              |      |                                  |        |             | 50/m     | S Def    | D4    | X   | X   |     |
| 87 | Massicotte   | 2002 | Spine                            | Cj 8   | 8           | 44/f     | SpP      | D5-6  | X   |     |     |
| 88 |              |      |                                  |        |             | 33/f     | BS       | D7-8  |     |     |     |
| 89 |              |      |                                  |        |             | 57/f     | SpP      | D6    |     |     |     |
| 90 |              |      |                                  |        |             | 27/m     | BS       | D9    |     |     |     |
| 91 |              |      |                                  |        |             | 46/f     | BS       | D4    |     |     |     |
| 92 |              |      |                                  |        |             | 21/m     | Headache |       | -   | -   | -   |
| 93 | Inoue*       | 2003 | J Neurosurg                      | Cj 1   | 1           | 77/f     | BS       | D6-7  | X   | X   |     |
| 94 | Nakagawa     | 2003 | J Spinal Disord Tech             | Cj 1   | 1           | 66/f     | Paraplegia | D6-7 |     |     |     |
| 95 | Sagiuchi     | 2003 | Neuro Med Chir (Tokyo)           | Cj 1   | 1           | 48/m     | BS       | D7-8  | X   | X   |     |
| 96 | Sasaoka      | 2003 | J Spinal Disord Tech             | Cj 1   | 1           | 57/m     | BS       | D2-3  | X   | X   |     |
| 97 | Aquellina    | 2004 | Ir Med J                         | Cj 1   | 1           | 37/f     | BS       | D4    | X   |     |     |
| 98 | Najjar       | 2004 | Surg Neurol                      | Cj 1   | 1           | 32/m     | SpP      | D8-9  | X   | X   |     |
| 99 | Rivas        | 2004 | Neurocirugia (Asturias, Spain)   | Cj 1   | 1           | 49/m     | BS       | D6.7  | X   | X   |     |
| 100| Saito        | 2004 | j orthop sci                     | Cj 1   | 1           | 66/f     | Paraplegia | D6-7 |     |     |     |
| 101| Spissu*      | 2004 | Nuero Sci                        | Cj 1   | 1           | -/f      | BS       | -     | -   | -   | -   |
| 102| Srinivasan   | 2004 | Nuerology                        | Cj NAA | -           | -        | -       | -     |     |     |     |
| 103| Maruichi     | 2004 | No Schinkei Geka                 | Cj 1   | 1           | 61/m     | BS       | D4-5  | X   | X   |     |
| 104| White        | 2004 | Br J Neurosurg                   | Cj 3   | 3           | 62/f     | BS       | D7    |     | X   |     |
| 105|              |      |                                  |        |             | 26/f     | BS       | D6-7  |     |     |     |
| 106| Ammar*       | 2005 | Neurosurgery                     | Cj 3   | 3           | -        | -       | D-    | X   |     |     |
| 107|              |      |                                  |        |             | -        | -       | D-    |     |     |     |
| 108| Sugimoto     | 2005 | J Spinal Disord Tech             | Cj 1   | 1           | 48/m     | BS       | D4-5  | X   | X   |     |
| 109| Karadeniz-Bilgili | 2005 | Journal of Clinical Imaging      | Rj 1   | 1           | 36/f     | BS       | D2-3  | X   |     |     |
| 110| Maira*       | 2006 | j Neurosurg Spine                | Cj 5   | 5           | -/f      | -       | -     | -   | -   | -   |
| 111|              |      |                                  |        |             | -/f      | -       | -     | -   | -   | -   |
| 112|              |      |                                  |        |             | -/f      | -       | -     | -   | -   | -   |
| 113|              |      |                                  |        |             | -/f      | -       | -     | -   | -   | -   |
| 114|              |      |                                  |        |             | -/f      | -       | -     | -   | -   | -   |
| 115|              |      |                                  |        |             | -/f      | -       | -     | -   | -   | -   |
| 116|              |      |                                  |        |             | -/m      | -       | -     | -   | -   | -   |

(continued)
| N°  | Author     | Year | Journal               | Cj/Rj | N. of Cases | Age/SEX | Symptoms         | Level | mRX | CTm | MRI |
|-----|------------|------|-----------------------|-------|-------------|---------|------------------|-------|-----|-----|-----|
| 117 | Ellger     | 2006 | Clin Neurol Neurosurg  | Cj    | 1           | 59/f    | BS               | D2    | X   | X   |     |
| 118 | Morley     | 2006 | Australas Radiol      | Rj    | 1           | 28/f    | BS               | D5-6  |     |     | X   |
| 119 | Roland     | 2006 | JBR-BTR               | Rj    | -           | -       | -                | -     | -   | -   | -   |
| 120 | Inoue      | 2006 | No Schinkei Geka      | Cj    | 1           | 71/f    | BS               | D2-3  | X   | X   |     |
| 121 | Arts*      | 2006 | Acta Neurochir (Wien) | Cj    | 2           | -       | -                | -     | -   | -   | -   |
| 122 | Barreneche | 2006 | J Neurosurg Spine     | Cj    | 7           | 65/f    | BS               | D4-5  | X   | X   |     |
|     |            |      |                       |       |             | 32/m    | Urinary Dys      | D7-8  |     |     | X   |
| 123 |           |      |                       |       |             | 54/f    | BS               | D2-3  | X   | X   |     |
| 124 |            |      |                       |       |             | 60/f    | BS               | D2-3  | X   | X   |     |
| 125 |            |      |                       |       |             | 59/f    | BS               | D5-6  | X   | X   |     |
| 126 |            |      |                       |       |             | 34/m    | NR               | D7-8  | X   | X   |     |
| 127 |            |      |                       |       |             | 72/m    | BS               | D4-5  | X   | X   |     |
| 130 | Bandai     | 2006 | No To Shinkei        | Cj    | 1           | 63/f    | SpP              | D2-3  | X   | X   |     |
| 131 | Akaza      | 2007 | Internal Medicine (Tokyo) | Cj | 1           | 56/m    | BS               | D2-3  |     |     | X   |
| 132 | Yokota     | 2007 | Neurosurgery         | Cj    | 1           | 33/m    | Horner’s Syndrome | C7-D1 | X   | X   | X   |
| 133 | OUR CASE   | 2007 | Acta Biomed 2021      |       | 1           | 61/m    | BS               | D6-7  |     |     | X   |

Some authors also reported a spinal cord protrusion throughout a defect of the inner layer of a duplicated dura mater (20, 23, 26, 30, 53, 60).

However, to date, there is no radiological or pathological proof to confirm any of this theory.

Tekkok and Coworkers report that it is difficult to define criteria for distinguishing between traumatic and spontaneous cord herniation (51). Many patients, moreover, report a long time, often more than 30 years between the spinal trauma and the onset of symptoms and sometimes it is difficult to understand the relationship between traumatic event and the herniation (1, 54).

According to Isu et al (23) an intradural arachnoid cyst, causing pressure and erosion, migrates throughout a dural fissure arising from a congenital defect, a mild trauma, sometimes unknown or an erosion of the dura by a herniated and calcified disc (1, 26, 30). Degenerative disc prolapse with transdural rupture of disc material, often calcified, has been also proposed as a potential cause of the dural defect (20).

Miyaguchi et al (33) reported a case of ISCH with documented intervertebral disc herniated and calcified as the cause of the ventral defect. About the pathophysiological mechanism leading to spinal cord herniation, some authors (17, 29, 34) report the role played by factors as cardiac pulsations, respiratory movements, and the physiological spinal curve. These factors support the contact between spinal cord and dura mater, resulting, over the time, in a total adhesion with generation of a tear of the dura which will be almost totally blocked by the spinal cord.

Even the pulsation of cerebral spinal fluid (CSF) on the dorsal side of the spinal cord, secondary to respiratory movements and cardiac pulsations, can contribute to generate the herniation, pushing the spinal cord into the extradural arachnoid cyst (Fig. 1) (30). Although Masuzawa et al describe this finding as an extradural arachnoid cyst, Sioutos et al suggest that it should be classified as a meningeal diverticulum or an arachnoid pseudocyst (46). This pathogenetical mechanism could...
result in a type IIIB spinal meningeal cyst according to the nomenclature proposed by Kumar et al (26), modifying the classification of Nabors et al (68).

Some authors think that the pressure inside the dorsal arachnoid cyst, as it enlarges, could be enough to produce a progressive thinning of the ventral dura mater until a tear appears and the arachnoid herniates through it (Fig. 1) (21, 23). However, dorsal arachnoid cysts are reported only in 25% of ISCH so other mechanism have to be looked for the most part of the cases (21, 23, 32, 46, 47, 53, 60).

Moreover, as highlighted by White and Firth (60), an erosion of the dura due to the pressure by the arachnoid should be more common in intradural tumor than in arachnoid cysts. To strengthen the unknown trauma hypothesis, Tronnier et al (52), stress the presence of inflammatory changes observed in some of the cases reported by Literature, either in the epidural space (4, 52), or in the arachnoid membrane (60).

Other authors (12, 15, 31, 32, 39, 40, 55), think the herniation of the neural tissue through the dural ventral membrane as consequent to a congenitally duplication of the dura mater.

**Clinical features**

ISCH is an rare clinical entity with almost 90 cases reported in the Literature (66) which typically occurs in middle-aged adults. The median age in all cases reported was 49.9 years with a range from 21 to 78 years and female preponderance (ratio: 2:1) (12, 37, 44, 67).

The most common clinical presentation is a Brown-Séquard syndrome but spastic para-monoparesis, sensory dysfunction or sphincter dysfunction also can be observed (37, 41, 67). The symptoms are slowly progressive suggesting a long course resulting in an arachnoidal adhesions to the nervous tissue with secondary gliosis involving axons. Sudden onset of symptoms has been also reported (30, 60).

The mean duration of the symptoms was 4.25 years (range from 1-12 years) for patients who came in with a Brown-Séquard syndrome and 5.34 years for those presenting with spastic paraparesis (37, 51).

The first symptom is usually a progressive lower-extremity paraesthesia and weakness (54). According to the Literature (20, 34, 60), in the early period, not all of patients become aware of sensorial changes, so the majority of patients arrives at the clinical examination because of increasing difficulty in walking, frequent falls, progressive paraesthesia often preceded by a sensation of warm.

Sphincter dysfunction is reported only in a small percentage of the cases (26, 32, 46, 47, 53, 55, 56).

A increasing impotence is rare but may represent the onset of the symptoms (54). According to the Literature ISCH usually presents in more than 50 % of the cases with symptoms and signs as Brown-Sequard syndrome (3, 14, 26, 30, 34, 39, 41, 46).
The differential diagnosis of ISCH includes Demyelinating Disorders, such as Multiple Sclerosis or a Transverse Myelitis (14, 20, 47).

Case Report

We report a case of ISCH in a 61-year-old man with history of a D7 explorative laminectomy for a suspicious of arachnoid cyst.

He was admitted to our institute with a clinical diagnosis of Brown-Séquard syndrome: weakness and paraplegia of the right lower extremity and paraesthesia of the contralateral lower extremity.

A neurological examination showed bilateral tendon reflexes hyperactive and the Babinski’s sign was also bilaterally present. These data were suggestive of a spinal/medullar suffering below D9-D10.

The MRI of the dorsal spine T1- (performed before and after i.v. infusion of m.d.c.) and T2-weighted (Fig. 2 and 3) showed a focal atrophy with a right ventral displacement of the thoracic spinal cord at the D6-D7 intervertebral level and a straightforward mushroom-shaped herniation of the spinal cord at the D7 level. A dural cyst, cranially to the herniation, was also found.

The diagnosis of ISCH has been established on the basis of the thin-section MRI of the dorsal spine findings. Surgical intervention was performed (reduction of the herniated spinal cord and duroplasty) with a posterior approach. The patient’s postoperative course was uneventful with rapid improvement of the symptoms of the lower extremities within few months.

Imaging

The most part of the ISCH reported have been founded between T2 and T10, with high predominance (79% cfr Brugieres et al) between T4 and T8, and symptoms may appear before than an herniation becomes demonstrable by MRI (15, 66, 67). The MR presentation of ISCH may be characterized by ventral displacement of the thoracic spinal cord.

The herniation through a dural defect may mimic a epidural tumor either ventral or ventrolateral. There is no contrast enhancement. A secondary enlargement of the dorsal subarachnoid spaces is also present. This sign may mimic a dorsal an arachnoid cyst.

A myelography, which led Wortzmann et al (1) to a surgical treatment of the first case of ISCH, can provide only approximate information on a anterior and/
or lateral displacement of the spinal cord. In the most part of the case, except for the case of Wortzmann et al (1) and White and Firth (60), reported in pre-CT era, a CT-Mielography (CTM) has been performed. The CTM, usually performed before a MRI study, demonstrate no filling defect dorsal to the spinal cord or retention of contrast medium along the ventral aspect of the dural sac. When performed after surgery, it can be useful to exclude the coexistence of a spinal cord herniation with a intradural spinal arachnoid cyst, as reported by Isu et al (2 cases) (23), Oe et al (1 case) (40), Borges et al (1 case) (14). Recently, advances in MRI have reduced the relevance of MCT in the ISCH diagnosis.

MRI typical findings of ISCH show on the sagittal scan an anterior S or C-shaped kink of the spinal cord with secondary enlargement of the dorsal subarachnoid space. On the sagittal plane a decreased spinal cord size (usually atrophic) can be also seen with spinal cord signal changes due to tethering.

The axial MRI images may show the dural defect in addition to the herniation but also arachnoid cyst and associated anomalies including scalloping of the vertebral body, spina bifida or other congenital deformities. Studying CSF dynamics by Phase-contrast cine MRI may be essential to detect a posterior compressing arachnoid cyst replacing MCT.

The most frequent misdiagnosis is: dorsal arachnoid cyst, enlargement of the dorsal subarachnoid space, extradural mass or compression, discal herniation or bulging with secondary spinal cord thinning.

The spinal cord appears typically abruptly deviated to the dorsal parts of the vertebral body at a localized area and the posterior subarachnoid space may be enlarged. These findings and the craniocaudal extent of the displacement are better shown on sagittal MRI scans (12). Hence, radiological techniques are crucial in ISCH diagnosis.

Management

Because of chronic progression of the symptoms, surgery represent the treatment of choice. The aim of surgery is to reduce the herniation, to repair the dural defect and to prevent recurrence.

After the herniation reducing, surgical treatment depends on type of dural defect. There are two main treatment strategies: a) closure of the defect if the nervous tissue is herniated in the epidural space or b) simply widening the aperture when a duplication of the dura mater or a ventral epidural cyst is present (3, 12, 14, 23, 56, 59).

Conclusions

We reported ISCH findings, surgically confirmed, in one case. In addition, we reviewed the Literature of the cases reported of this clinical entity often misdiagnosed. MRI findings are a ventral displacement of the spinal cord in the thoracic spine. MRI is the technique of choice to exclude a posterior arachnoid cyst, the most common mistaken diagnosis, and to recognize a spinal cord herniation when an anterior dural defect is present.

Due to its rare occurrence, its mild, non-specific, and slowly progressive symptoms, it is important to keep in mind this differential diagnosis to achieve an early diagnosis and surgical treatment to prevent major neurological dysfunctions.
Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

References

1. Wortzman G, Tasker RR, Rewcastle NB, Richardson JC and Pearson FG (1974) Spontaneous incarcerated herniation of the spinal cord into a vertebral body: a unique cause of paraplegia. Case report. J Neurosurg 41: 631-635
2. Aizawa T, Sato T, Tanaka Y, et al. (2001) Idiopathic herniation of the thoracic spinal cord: report of three cases. Spine 26: E488-491
3. Massicotte EM, Montanera W, Ross Fleming JF, et al. (2002) Idiopathic spinal cord herniation: report of eight cases and review of the literature. Spine 27: E233-241
4. Abe M, Komori H, Yamaura I and Kayano T (1999) Spinal cord herniation into an extensive extradural meningeval cyst: postoperative analysis of intracystic flow by phase-contrast cine MRI. J Orthop Sci 4: 450-456
5. Adams RF and Anslow P (2001) The natural history of transdural herniation of the spinal cord: case report. Neuroradiology 43: 383-387
6. Alvisi C, Cerisoli M, Giulioni M and Guerra L (1987) Long-term results of surgically treated congenital intradural spinal arachnoid cysts. J Neurosurg 67: 333-335
7. Ammar KN, Pritchard PR, Matz PG and Hadley MN (2005) Spontaneous thoracic spinal cord herniation: three cases with long-term follow-up. Neurosurgery 57: E1067; discussion E1067
8. Andrews BT, Weinstein PR, Rosenblum ML and Bararo NM (1988) Intradural arachnoid cysts of the spinal canal associated with intramedullary cysts. J Neurosurg 68: 544-549
9. Aquilina K, Nanra JS and Rawluk D (2004) Idiopathic spinal cord hernia. Ir Med J 97: 115-116
10. Andrews BT, Weinstein PR, Rosenblum ML and Bararo NM (1988) Intradural arachnoid cysts of the spinal canal associated with intramedullary cysts. J Neurosurg 68: 544-549
11. Bartolomei J, Wong J, Awad IA, et al. (2000) Case problems conference: thoracic spinal cord hernia. Neurosurgery 46: 1408-1415
12. Baur A, Stabler A, Pfeffer K, Hamburger C and Reiser M (1997) Imaging findings in patients with ventral dural defects and herniation of neural tissue. Eur Radiol 7: 1259-1263
13. Berbel A, Porta-Etessam J, Martinez-Salio A, et al. (2001) [Idiopathic spinal cord herniation. Presentation of a new case and review of the literature]. Rev Neurol 32: 54-57
14. Borges LF, Zervas NT and Lehrich JR (1995) Idiopathic spinal cord herniation: a treatable cause of the Brown-Sequard syndrome--case report. Neurosurgery 36: 1028-1032; discussion 1032-1023
15. Brugieres P, Malapert D, Adle-Biassette H, et al. (1999) Idiopathic spinal cord herniation: value of MR phase-contrast imaging. AJNR Am J Neuroradiol 20: 935-939
16. Cellerini M, Bayon S, Scaczeri F, et al. (2002) Idiopathic spinal cord herniation: a treatable cause of Brown-Sequard syndrome. Acta Neurochir (Wien) 144: 321-325
17. Dix JE, Griffitt W, Yates C and Johnson B (1998) Spontaneous thoracic spinal cord herniation through an anterior dural defect. AJNR Am J Neuroradiol 19: 1345-1348
18. Eguchi T, Yokota H, Nikiado Y, Nobayashi M and Nishioka T (2001) Spontaneous thoracic spinal cord herniation--case report. Neurosurg 45: 508-512
19. Ewald C, Kuhne D and Hassler WE (2000) Progressive spontaneous herniation of the thoracic spinal cord: case report. Neurosurgery 46: 493-495; discussion 495-496
20. Hausmann ON and Moseley IF (1996) Idiopathic dural herniation of the thoracic spinal cord. Neuroradiology 38: 503-510
21. Henry A, Tunkel R, Arbit E, Ku A and Lachmann E (1997) Tethered thoracic cord resulting from spinal cord herniation. Arch Phys Med Rehabil 78: 530-533
22. Inoue T, Cohen-Gadol AA and Krauss WE (2003) Low-pressure headaches and spinal cord herniation. Case report. J Neurosurg 98: 93-95
23. Miyasaka K, Ito S, Iwasaki Y, et al. (1983) High resolution computed tomography in the diagnosis of cervical disc disease. Neuroradiology 24: 253-257
24. Iyer RV, Coutinho C and Lye RH (2002) Spontaneous spinal cord herniation. Br J Neurosurg 16: 507-510
25. Kumar K, Malik S and Schulte PA (2003) Symptomatic spinal arachnoid cysts: report of two cases with review of the literature. Spine 28: E25-29
26. Kumar R, Taha J and Greiner AL (1995) Herniation of the spinal cord. Case report. J Neurosurg 82: 131-136
27. Maira G, Denaro L, Doglietto F, Mangioli A and Colosimo C (2006) Idiopathic spinal cord herniation: diagnostic, surgical, and follow-up data obtained in five cases. J Neurosurg Spine 4: 10-19
28. Marshman L (2000) Progressive spontaneous spinal cord herniation: case report. Neurosurgery 47: 1469
29. Marshman LA, Hardwidge C, Ford-Dunn SC and Olney JS (1999) Idiopathic spinal cord herniation: case report and review of the literature. Neurosurgery 44: 1129-1133
30. Matsumura T, Takahashi MP, Nozaki S and Kang J (1996) [A case of idiopathic spinal cord herniation]. Rinsho Shinkeigaku 36: 566-570
32. Miura Y, Mimatsu K, Matsuyama Y, Yoneda M and Iwata H (1996) Idiopathic spinal cord herniation. Neuroradiology 38: 155-156

33. Miyaguchi M, Nakamura H, Shakudo M, Inoue Y and Yamano Y (2001) Idiopathic spinal cord herniation associated with intervertebral disc extrusion: a case report and review of the literature. Spine 26: 1090-1094

34. Miyake AP, Tress BM and Kaye AH (2001) Idiopathic spinal cord herniation. J Clin Neurosci 8: 180-183

35. Nakamura H, Kamimura M, Uchiyama S, et al. (2003) Idiopathic spinal cord herniation: a new theory of pathogenesis. Surg Neurol 62: 161-170; discussion 170-171

36. Nakazawa H, Toyama Y, Satomi K, Fujimura Y and Hirabayashi R (1993) Idiopathic spinal cord herniation. No Shinkei Geka 14: 681-685

37. Najjar MW, Baeesa SS and Lingawi SS (2004) Idiopathic spinal cord herniation associated with duplicated dura mater and with an arachnoid cyst. Neuroradiology 38: 155-156

38. Nakagawa H, Kamimura M, Uchiyama S, et al. (2003) Idiopathic spinal cord herniation: report of two cases and review of the literature. Spine 28: 2138-2141

39. Nakagawa H, Kamimura M, Uchiyama S, et al. (2003) Idiopathic spinal cord herniation: case report and review of the literature. Spine 28: 299-305

40. Oe T, Hoshino Y and Kurokawa T (1990) [A case of idiopathic herniation of the spinal cord associated with duplicated dura mater and with an arachnoid cyst]. Nippon Seikeigeka Gakkai Zasshi 64: 43-49

41. Pereira P, Duarte F, Lamas R and Vaz R (2001) Idiopathic spinal cord herniation: case report and literature review. Acta Neurochir (Wien) 143: 401-406

42. Sagiuchi T, Iida H, Tachibana S, et al. (2003) Idiopathic spinal cord herniation associated with calcified thoracic disc extrusion--case report. Neurol Med Chir (Tokyo) 43: 364-368

43. Sahl H, Forsting M and Sartor K (1995) [Post-traumatic herniation of the spinal cord: a rare cause of slowly progressing transverse spinal cord syndrome]. Rofo Fortsch Geb Rontgenstr Neuen Bildgeb Verfahr 162: 350-352

44. Saito T, Anamizu Y, Nakamura K and Seichi A (2004) Case of idiopathic thoracic spinal cord herniation with a chronic history: a case report and review of the literature. J Orthop Sci 9: 94-98

45. Sasaoka R, Nakamura H and Yamano Y (2003) Idiopathic spinal cord herniation in the thoracic spine as a cause of intractable leg pain: case report and review of the literature. J Spinal Disord Tech 16: 288-294

46. Sioutos P, Arbit E, Tsairis P and Gargan R (1996) Spontaneous thoracic spinal cord herniation. A case report. Spine 21: 1710-1713

47. Slavotinek JP, Sage MR and Brophy BP (1996) An unusual spinal intradural arachnoid cyst. Neuroradiology 38: 152-154

48. Spissu A, Peltz MT, Matta G and Cannas A (2004) Traumatic transdural spinal cord herniation and the nuclear trait sign: case report. Neurol Sci 25: 151-153

49. Srinivasan A, Bourque P and Goyal M (2004) Spontaneous thoracic spinal cord herniation. Neurology 63: 2187

50. Sugimoto T, Kasai Y, Takegami K, et al. (2005) A case of idiopathic spinal cord herniation with duplicated dura mater. J Spinal Disord Tech 18: 106-111

51. Tekkok IH (2000) Spontaneous spinal cord herniation: case report and review of the literature. Neurosurgery 46: 485-491; discussion 491-482

52. Tronnier VM, Steinmetz A, Albert FK, Scharf J and Kunze S (1991) Hernia of the spinal cord: case report and review of the literature. Neurosurgery 29: 916-919

53. Uchino A, Kato A, Momozaki N, Yukiike M and Kudo S (1997) Spinal cord herniation: report of two cases and review of the literature. Eur Radiol 7: 289-292

54. Uehara H, Kaden B, Peckstein U and Solymosi L (1996) Herniation of the spinal cord 38 years after childhood trauma. Neuroradiology 38: 157-158

55. Vallee B, Mercier P, Menei P, et al. (1999) Ventral transdural herniation of the thoracic spinal cord: surgical treatment in four cases and review of literature. Acta Neurochir (Wien) 141: 907-913

56. Verna C, Mercier P, Hayek G, et al. (1999) [Spontaneous spinal cord herniation: a little-known cause of Brown-Sequard syndrome. Report of two cases and review of the literature]. Neurochirurgie 45: 225-231

57. Wada E, Yonenobu K and Kang J (2000) Idiopathic spinal cord herniation: report of three cases and review of the literature. Spine 25: 1984-1988

58. Watanabe M, Chiba K, Matsumoto M, et al. (2001) Surgical management of idiopathic spinal cord herniation: a review of nine cases treated by the enlargement of the dural defect. J Neurosurg 95: 169-172

59. Watters MR, Stears JC, Osborn AG, et al. (1998) Transdural spinal cord herniation: imaging and clinical spectra. AJNR Am J Neuroradiol 19: 1337-1344

60. White BD and Firth JL (1994) Anterior spinal hernia: an increasingly recognised cause of thoracic cord dysfunction. J Neurol Neurosurg Psychiatry 57: 1433-1435

61. Winters JP and Conley FK (1978) Progressive neurological dysfunction secondary to postoperative cervical pseudomeningocele in a C-4 quadriplegic. Case report. J Neurosurg 48: 289-291

62. Hosono N, Yonenobu K and Ono K (1995) Postoperative cervical pseudomeningocele with herniation of the spinal cord. Spine 20: 2147-2150

63. Cheng MH (1996) Intraspinal extradural arachnoid cyst with spinal cord herniation. J Formos Med Assoc 95: 712-714

64. Lee ST, Lui TN and Jeng CM (1997) Spinal cord herniation after stabbing injury. Br J Neurosurg 11: 84-86
65. Karadeniz-Bilgili MY, Castillo M and Bernard E (2005) Transdural spinal cord herniation: pre- and postoperative MRI findings. Clin Imaging 29: 288-290

66. Barrenechea IJ, Lesser JB, Gidekel AL, Turjanski L and Perin NI (2006) Diagnosis and treatment of spinal cord herniation: a combined experience. J Neurosurg Spine 5: 294-302

67. Ellger T, Schul C, Heindel W, Evers S and Ringelstein EB (2006) Idiopathic spinal cord herniation causing progressive Brown-Sequard syndrome. Clin Neurol Neurosurg 108: 388-391

68. Nabors MW, Pait TG, Byrd EB, et al. (1988) Updated assessment and current classification of spinal meningeal cysts. J Neurosurg 68: 366-377

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