ORIGINIAL ARTICLE

Split-cord malformation and accompanying anomalies

Le syndrome de split-cord malformation et les anomalies associées

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KEYWORDS
Split-cord malformation; Magnetic resonance imaging

Summary
Objective. — To present the magnetic resonance imaging (MRI) appearances of spinal split-cord malformation (SCM) and to investigate the various types of congenital spinal disorders associated with SCM.

Materials and methods. — MR examinations of 23 patients with SCM were carried out in our hospital between June 2002 and May 2007 and retrospectively analysed.

Results. — Nineteen (82.6%) patients were diagnosed as type I SCM, while four (17.4%) were diagnosed as type II SCM. The most commonly involved site of SCM was the dorsolumbar area (47.8%) while cervical involvement was the least common (4.3%). No accompanying congenital spinal disorders were detected in four patients (17.4%). In 19 patients (82.6%), congenital spinal disorders accompanying SCM were detected, the most common of which was a low-lying cord, found in 14 patients (60.9%). Other anomalies included hydromyelia in seven patients (30.4%), lipoma in six (26%), meningomyelocele in four (17.4%), thick filum in three (13%) and dermoid cyst in three (13%).

Conclusion. — In preoperative planning for SCM, its characteristics and those of the accompanying anomalies should be determined. MRI is a valuable tool for making such determinations. © 2007 Elsevier Masson SAS. All rights reserved.

MOTS CLÉS
Split-cord malformation ; Imagerie de résonance magnétique

Résumé
Objectif. — L’objectif de cette étude était de présenter les aspects en imagerie par résonance magnétique (IRM) du syndrome de dédoublement médullaire (split-cord malformation [SCM]) et d’étudier les anomalies congénitales associées.

Matériels et méthodes. — Les examens IRM de 23 patients présentant un SCM et explorés dans notre hôpital entre mai 2002 et juin 2007, étaient rétrospectivement analysés.

Résultats. — Dix-neuf patients (82,6%) présentaient un SCM de type I alors que quatre (17,4%) présentaient un SCM de type II. La localisation la plus fréquente de SCM était la charnière

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Split-cord malformation and accompanying anomalies

Introduction

Spinal dysraphism is a common term used to describe all congenital spinal disorders resulting from anomalous differentiation in the early foetal period and/or incomplete closure of dorsal midline structures. Split-cord malformation (SCM) comprises 3.8% of all spinal dysraphisms. It is a rare entity characterised by symmetrical or asymmetrical separation of the spinal cord into two parts along the sagittal plane [1]. It may be either isolated or accompanied by various spinal or extraspinal anomalies. SCM is usually detected in childhood; however, because it can be asymptomatic in some cases, diagnosis may be delayed until adulthood.

Congenital anomalies in which the spinal cord is split into two cords were previously termed "diastematomyelia" or "diploymelia". In 1992, the confusion in terminology was resolved by a new classification system of the disorder by Pang et al. [2]. In this classification, SCMs are divided into type I and type II, depending on the presence of a septum between the cords. In type I SCM, the two hemicords are housed in separate dural sacs with a rigid septum in between whereas, in type II SCM, both hemicords are housed within the same dural sac, with no rigid septum in between.

SCM may be isolated or accompanied by various spinal or extraspinal anomalies such as scoliosis, kyphoscoliosis, hemivertebra, block vertebra, butterfly vertebra, hypoplastic vertebra, rib abnormalities, low-lying cord, thick filum, meningomyelocele, hydromyelia, lipoma and dermoid cyst. Awareness of these anomalies is important in identifying the clinical symptoms and determining the surgical plan.

This study presents the magnetic resonance imaging (MRI) appearances of spinal SCM and investigates the types and incidence rates of congenital spinal disorders associated with SCM.

Materials and methods

For our study, we retrospectively evaluated MR images of 23 patients (14 women and nine men) who had undergone MRI evaluation between June 2002 and May 2007 in our hospital and been diagnosed as SCM. The ages of the patients ranged from four to 62 years (mean: 30.1 years).

Our clinical protocol required MRI screening of the entire spine in all the patients with SCM for detection of accompanying spinal anomalies. All of these spinal MRI scans were retrospectively reevaluated by two experienced radiologists. The MRI examinations were made using a 1.5-T scanner (Siemens Magnetom Vision, Erlangen, Germany). The evaluation involved determining the level at which the split started and the level to which it extended, whether the hemicords were symmetrical or asymmetrical, whether any spur was present between the hemicords (to differentiate type I from type II) and whether the spur, if present, was complete. Furthermore, any possible associated spinal anomaly was investigated. A conus medullaris terminating below the level of the L2–3 intervertebral disc was considered a low-lying cord. A filum terminale of 2 mm or more was considered a thick filum. Lesions with high-signal intensity on T1-weighted images and homogeneous suppression on fat-saturated images were considered lipomas and those with heterogeneous suppression were considered dermoid cysts. Any differences in the findings between the two radiologists were resolved by their arriving at a common consensus.

Results

SCM was determined to be at the cervical level in one (4.3%) patient, at the cervicodorsal level in two (8.7%) patients, at the dorsal level in three (13%) patients, at the dorsolumbar level in 11 (47.8%) patients and at the lumbar level in six (26.1%) patients. Of all the patients, 19 (82.6%) had type I SCM (Fig. 1) and four (17.4%) had type II SCM (Fig. 2). In 17 (89.5%) patients with type I SCM, the septum extended completely to the opposite side of the canal. In 15 (65.2%) patients, the hemicords were symmetrical (Fig. 3) in size whereas, in eight (34.8%) patients, they were asymmetrical (Fig. 4).

We detected six different congenital spinal disorders accompanying the SCMs. The most common disorder was a low-lying cord, found in 14 (60.9%) of 23 patients. In addition, hydromyelia was detected in seven (30.4%) patients, lipoma in six (26%), meningomyelocele in four (17.4%), thick filum in three (13%) and dermoid cyst in three (13%). In four (17.4%) patients, no congenital spinal disorder accompanying the SCM was detected. The anomalies accompanying the SCM and their incidence are summarized in Table 1. SCM was accompanied by one disorder in five (21.7%) patients, two disorders in nine (39.1%) patients (Fig. 5) and three disorders in four (17.4%) patients. Low-lying cord and hydromyelia were the most common coexisting disorders, seen in five patients (Fig. 6). The meningomyelocele detected in four patients was above SCM level in two of them, at SCM level in one patient and below SCM level in one patient.

The most common symptom was weakness of the extremities, seen in 15 (65.2%) of 23 patients. The other symptoms...
Figure 1  **Type I split-cord malformation.** A: on this sagittal T2-weighted image, a bony septum extending from anterior to posterior within the vertebral canal, in a cranial direction, is seen in the lumbar region. In addition, the spinal cord terminates at the L5 vertebral level (low-lying cord) and, at L1–L2 vertebral levels, the central spinal canal is wider than normal (hydromyelia). B: on this axial T2-weighted image, the bony septum has split the spinal cord into two hemicords.

**Figure 1** SCM ou diastematomyélie de type I. A : coupe IRM sagittale pondérée T2 montrant la cloison osseuse médiane divisant la canal rachidiene et la moelle attachée basse en regard de L5 ainsi qu’un élargissement du canal medullaire (hydromyélie) en regard de L1-2. B : IRM en coupe axiale pondérée T2 montrant la division des hémimoeilles par l’éperon.

Figure 2  **Type II split-cord malformation.** On this axial T2-weighted image, two hemicords are housed within the same dural sac, and the spinal cord is separated by a fibrous septum (black arrow), presenting as a hypodense band.

**Figure 2** SCM ou diastématomyélie de type II. Coupe IRM axiale pondérée T2 montrant les hémimoelles dans un sac unique. La moelle est divisée par la cloison fibreuse (flèche noire) qui apparaît comme une bande hypo-intense.

were pain (n = 12; 52.2%), hypoesthesia (n = 10; 43.5%), muscle atrophy (n = 8; 34.8%), bladder incontinence (n = 4; 17.4%) and bowel incontinence (n = 8.7%). One patient with SCM accompanied by lipoma and two patients with SCM accompanied by no disorders were asymptomatic. In other words, in two (50%) of the four patients with isolated SCM, no symptoms were detected, while only one (5.3%) of the 19 patients with SCM accompanied by at least one disorder was asymptomatic.

**Discussion**

In terms of embryological development, the spinal cord is formed by the integration of two paramedian notochords along the midline. When this integration is defective, SCM is the result [3]. According to the unified theory of embryogenesis proposed by Pang et al. [2], an error in the formation of an accessory neuroenteric canal between the yolk sac and amnion, which subsequently advances with the mesenchyme to form an endomesenchymal tract that splits the notochord and neural plate, is responsible for all SCMs.

There was confusion with the terminology, including the use of terms such as diastematomyelia and diplomyelia, until Pang et al. came up with the term “double spinal-cord malformations” [2]. However, all double spinal-cord anomalies were termed “SCM”, and these anomalies were classified as either type I or type II. In type I SCM, there are two hemicords, each housed in its own dural sac, but separated by a rigid osseocartilaginous septum, extending from the vertebral body to the neural arch. The entire rigid septum is
Split-cord malformation and accompanying anomalies

Figure 3  Type I split-cord malformation. On these axial (A) and coronal (B) T2-weighted images at thoracic level, the spinal cord is split into two equal parts by an osseocartilaginous septum (white arrows). A dotted appearance of the septum can be seen on the coronal image.

Figure 3  SCM de type I. Coupe IRM axiale (A) et coronale (B) pondérée T2 montrant la moelle épinière divisée en deux parties par une cloison osteocartilyagineuse (flèche blanche). La cloison est visualisée comme un image nodulaire en coupe coronale.

located extradurally. In contrast, in type II SCM, both hemi-
cords are housed within a common dural sac, with no rigid
septum lying between the two hemicords [2].

SCM is known to be common among women [4,5]. In two
separate studies, the female-to-male ratio was reported as
1.3:1 [6,7]. In our study, this ratio was 1.6:1. On the other
hand, some studies have reported higher rates of prevalence
among men [2,8].

SCMs are often located in the lumbar and dorsolumbar
regions [5,6,9]. Sinha et al. [7] reported incidence rates
for cervical, cervicodorsal, dorsal, dorsolumbar, lumbar and
lumbosacral locations as 3, 1, 13, 28, 50 and 5%, respec-
tively. In our study, the most common area for SCM was the
dorsolumbar region, with an incidence rate of 47.8%. The
least common location was the cervical region, with a rate
of 4.3%. The differences in our location rates compared with
the Sinha et al. study could be due to the limited number of
patients in our study.

The incidence rates for type I versus type II SCM are inco-
sistent. Tortori-Donati et al. [10] reported an incidence rate
of 75% for type II SCM; however, in many studies from India,
similar rates have been reported, but for type I SCM. In the
study by Sinha et al. [7], among 203 SCM patients, the type I
incidence rate was 65% and in a study by Kumar et al. [11],
it was 82%, closely similar to the incidence rate found in our
study.

In most type I SCM patients, the osseocartilaginous se-
ptum extends from the vertebral body to the neural arch,
where it unites with the bone on the opposite side [9]. These
types of septum are termed “complete”. However, in some
patients, the septum may be incomplete. In our study, two
(10.5%) of the 19 type I-SCM-patients had an incomplete rigid
septum.

<table>
<thead>
<tr>
<th>Congenital spinal disorder</th>
<th>Number of patients (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low-lying cord</td>
<td>14 (60.9)</td>
</tr>
<tr>
<td>Hydromyelia</td>
<td>7 (30.4)</td>
</tr>
<tr>
<td>Lipoma</td>
<td>6 (26.0)</td>
</tr>
<tr>
<td>Meningomyelocele</td>
<td>4 (17.4)</td>
</tr>
<tr>
<td>Thick filum</td>
<td>3 (13)</td>
</tr>
<tr>
<td>Dermoid cyst</td>
<td>3 (13)</td>
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</tbody>
</table>
Figure 5  
Type I split-cord malformation at the thoracic level. A: sagittal T1-weighted image at the lumbar level showing an intradural-hyperintense lesion (lipoma) with well-defined contours at the L4 vertebral level (white arrow). B: on a sagittal T2-weighted image with fat saturation, the lipoma (white arrow) appears hypointense. In addition, the spinal cord terminates at the L5 vertebral level (low-lying cord).

The hemicords were asymmetrical in size in eight (34.8%) of 23 patients, similar to the reported rate of 30% [12]. The only clinical importance of such asymmetry is the possibility of a thinner lower extremity on the side with the thinner hemicord compared with the lower extremity of the opposite side [13]. Indeed, in six (75%) of the eight patients with asymmetry in our study, the extremity on the side with the thinner hemicord was thinner. In addition to differences in size, the spinal root counts of the two hemicords may also be asymmetrical: an equal number of spinal roots may originate from each hemicord or one root may originate from the thinner side, with three spinal roots originating from the other side. However, in our study, this assessment was not done.

SCM may be accompanied by many congenital spinal disorders such as low-lying cord, hydromyelia, lipoma, meningomyelocele, thick filum and dermoid cyst. A low-lying cord has been attributed to incomplete retrogressive differentiation with failure of terminal-cord involution [14]. Thick filum is related to abnormal retrogressive differentiation of the secondary neural tube, producing a filum that is thicker than normal. Dermoid cysts result from encystment of part of the dermal sinus tract or iatrogenic implantation of skin cells during back surgery [9]. Hydromyelia is thought to develop after neural-tube closure as a result of abnormal dilatation of the central canal of the closed neural tube [15]. Spinal lipoma arises from premature separation of ectoderm and neuroectoderm during neurulation. This can be found anywhere along the spinal canal and can sometimes be multifocal and huge [9]. Meningomyelocele arises from a lack of carbohydrate molecular expression on the neuroectodermal cell surface, resulting in failed neural-tube closure [14].

The congenital spinal disorders accompanying SCM are clinically and surgically important. Even when the patient has no symptoms related to the SCM, there may be symptoms due to its accompanying disorders. For instance, large lipomas or dermoid cysts may lead to pain and neurological deficits. Furthermore, a low-lying cord can lead to motor and sensory deficits. An awareness of the presence of these additional disorders is surgically important. If the disorders accompanying SCM are overlooked, or the operation is directed solely at the SCM, there may be no improvement in the patient’s clinical symptoms. Moreover, awareness of the accompanying condition(s) is important in determining surgical priorities. For example, if the SCM is accompanied by a tethered cord at the sacral level, the septum should
first be excised to release the cord. Otherwise, the abruptly-released cord will suddenly be pulled upwards, resulting in spinal cord injury at the sacral level.

Since only the MRI findings of our patients were retrospectively evaluated, to avoid any errors in interpretation, skeletal system anomalies were excluded from our study. In 82.6% of the cases, SCM was accompanied by at least one congenital disorder. Six congenital disorders accompanying the SCM were detected. In order of frequency, these disorders were low-lying cord, hydromyelia, lipoma, meningomyelocele, thick filum and dermoid cyst. Sinha et al. [7] reported tethered-cord syndrome as the most common accompanying anomaly. However, all SCM cases could be said to have a tethered cord, as normal cord movements are more or less hindered by an osseocartilaginous or fibrous septum [6,13,16]. Thus, a tethered cord was not included in the list of accompanying anomalies in our study. Our most common anomaly accompanying SCM was a low-lying cord, with an incidence rate of 60.9%. Ersahin et al. [17] reported a rate of 40.5%, while Kumar et al. found a rate of 58.7% for this anomaly [11]. The second most common accompanying anomaly in our study was hydromyelia, with a rate of 30.4%, which is closely similar to the 27.5 and 35.4% rates for concomitant SCM and hydromyelia reported in two separate studies [5,7].

MRI is a highly effective technique for determining SCM and its accompanying anomalies. Particularly on axial and coronal images, SCM is easily detectable whereas, on sagittal images, it is difficult to identify. An osseocartilaginous septum can be detected on all axial images. A fibrous septum, however, can only be detected on high-resolution T2-weighted images. CT is able to display the bony septum well. In addition, on coronal and sagittal-reformatted images, any accompanying vertebral anomalies may be better demonstrated with the new multidetector CT scanning technology. MRI, on the other hand, is relatively more useful in demonstrating accompanying anomalies that are not bony deformities. In addition, MRI can also be used in the evaluation of operation results and follow-up of SCM patients.

Our study has a few limitations. The first is the limited number of patients. Another is the lack of comparison of MRI results with high-resolution, thin-slice, CT myelography, which can clearly show anatomical details and any per operative findings. A third limitation is the exclusion of associated disorders of the skeletal system.

In conclusion, SCM is an extremely rare anomaly of the spinal cord. In the preoperative planning for SCM, the characteristics of the SCM and its accompanying anomalies should be considered. MRI is a highly useful tool for the determination of SCM and its accompanying spinal anomalies.

References


