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ORIGINAL ARTICLE

CT and MRI features of patients with diastematomyelia

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Objective: To evaluate computed tomography (CT) and magnetic resonance imaging (MRI) features in patients with diastematomyelia and to investigate clinical characteristics of this lesion.
Study design: A retrospectively study.
Setting: The Second Affiliated Hospital, School of Medicine, Xi'an Jiaotong University.
Methods: A total of 82 diastematomyelia cases were retrospectively studied. All the patients underwent neurological examinations as well as MRI and CT of the spine. A self-established neurological functional grading system was used, and posterior tibial nerve somatosensory cortical-evoked potential (PTNSCEP) was measured to assess the neurological status of the patients. Imaging features of symmetry of splitting, presence of septum, location of lesion and number of split segments were studied. The neurological functional grading, PTNSCEP, and imaging findings were then analyzed and compared, and the difference was considered to be significant if *P*-value was lower than 0.05.
Results: Neurological functional grading and latency of PTNSCEP were significantly different but related in terms of symmetry of splitting, presence of septum and location of lesion. Although no significant differences were present in the number of split segments, the severity of the neurological functional grading and PTNSCEP impairment were not related to the number of split segments.
Conclusion: The imaging features in diastematomyelia are characteristic and relate well with the clinical manifestations according to neurological functional grading and PTNSCEP measurement, except the number of split segments.
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INTRODUCTION

Diastematomyelia is an uncommon congenital malformation of the vertebral axis where the spinal cord is split longitudinally into two. Each hemicord contains a central canal and a set of dorsal and ventral horns and nerve roots. The precise etiology is not known. The clinical manifestations include cutaneous abnormality overlying the spine, neurologic deficits and orthopedic abnormalities.[1–4] Clinically, the symptomatology is not specific and does not differ from that seen in other forms of spinal dysraphism.[5–8]

Despite its low incidence, diastematomyelia results in severe neurological dysfunction. The clinical diagnosis and treatment of the lesion are therefore important. Delayed or improper treatment due to incorrect diagnosis may lead to the worsening of neurological symptoms. Conventionally, diagnosis of diastematomyelia is based on its clinical manifestations and radiological examination. Plain radiography and myelography have traditionally been used. With the improvement in imaging technology, computed tomography (CT) and magnetic resonance imaging (MRI) are established as the primary diagnostic strategies for evaluating the symptoms of diastematomyelia. Although CT and MRI findings of diastematomyelia have been discussed in the literature,[9–11] there are few reports concerning the characteristic imaging and the clinical manifestations.[12,13] The relationship between the clinical severity of diastematomyelia and the morphological changes reflect in imaging findings has never been thoroughly investigated.

We have already demonstrated the clinical characteristics and management strategies of diastematomyelia previously.[14,15] The present study was designed to delineate the imaging (MRI and CT) features of diastematomyelia, and further to investigate the clinical characteristics of this lesion.

PATIENTS AND METHODS

Diastematomyelia cases treated in our hospital during January 1993 to December 2012 were retrieved, and finally 82 patients were included in the retrospective study. The exclusion criteria were age under 3 years, acute trauma, surgical history, combined intracranial lesion, other forms of associated spinal dysraphisim. All the patients underwent clinical examinations including neurological functional grading and orthopedic examination. Data including sex, age and symptoms were recorded. MRI and CT (Figure 1) were performed subsequently to confirm the diagnosis. The interval between imaging and neurological grading was no longer than 7 days.

Neurological functional grading

A neurological grading system was established in our practice based on a previously described clinical scoring.[16] The neurological function in patients of diastematomyelia was graded by scoring gait, urination and defecation (sphincter function), motor, superficial and deep sensation (pinprick, light touch and joint position sensation) and tendon reflex. Decreasing scores indicated the increasing severity of symptoms. Categories of detailed

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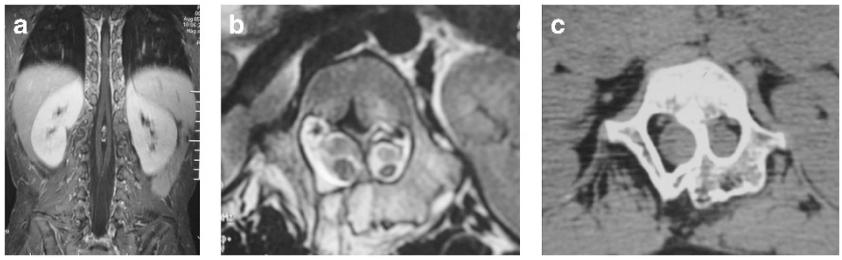

Figure 1 An example of a patient with diastematomyelia. The coronal (**a**) and axial (**b**) planes of MR image, CT image (**c**).

Table 1 Neurological functional grading

Score	*Gait*	*Urination and defecation*	*Lower limb strength*	*Pinprick sensation*	*Light touch sensation*	*Joint position sensation*	*Deep tendon reflexes*
0	Unable to walk unassisted	Total incontinence	No contraction	Absent	Absent	Absent	− or ++++
1	Severe bilateral deficit	Intermittent incontinence, uncontrolled	Muscle flicker, but no movement	Impaired	Impaired	Impaired	+ or +++
2	Severe unilateral deficit	Intermittent incontinence, controlled	Movement without antigravity	Normal	Normal	Normal	++
3	Mild bilateral or unilateral deficit	Increased frequency	Movement with antigravity, but not against resistance				
4	Walks normally	Total control	Movement against some resistance				
5			Normal function				

assessment and their respective scales are shown in Table 1. Each neurological parameter has a specific measurement.

Gait and sphincter function. The measurement of gait and sphincter function followed Roy's clinical scoring.[16]

Motor. Examination of motor function consisted of testing 14 key muscle groups in each side of the lower limb. The testing was performed on a six-point scale from 0 to 5. The muscle groups tested were the hip joint (flexion, extension, abduction, adduction, internal rotation, external rotation), knee joint (flexion, extension), ankle joint (dorsiflexion, plantar flexion, eversion, inversion) and toes (dorsiflexion, plantar flexion). These muscles were chosen to be evaluated due to availability. Motor scoring was calculated by adding the scores of each key muscle group. Scores for the right and left sides were obtained independently. The total score possible was 70 for each lower extremity.

Sensation. A total number of 10 dermatomes from L1 through S5 were tested separately for pinprick and light touch sensations on both sides of the lower extremity. A three-point scale was used, with the face as the normal control point. The patients' responses to pinprick and light touch were evaluated as being normal (intact), impaired or absent. Sensation scoring was calculated by adding up the scores for each dermatome, for a total score possible of 40 on each side for pinprick and light touch.

Joint position sensation was tested in knee and ankle joints as well as great and little toes. The total score possible was 8 for each lower extremity.

Deep tendon reflex. Knee and ankle responses were assessed for deep tendon reflex. The tendon jerk was graded as '− (absent)', '+ (diminished)', '++ (normal)', '+++ (exaggerated)', or '++++ (clonus)'. The score of each tendon reflex ranged from 0 (absent or clonus) to 2 (normal), and thus a score for one side of the lower extremity ranging from 0 to 4 was derived for each patient.

Reports of all the examinations were reviewed for completeness and internal consistency before inclusion in the master data files. Higher scores reflected better clinical manifestations. The best score, indicative of normal function according to our methods, was 256 (details on score calculation can be seen in the Supplementary File).

Posterior tibial nerve somatosensory cortical-evoked potential

To avoid subjective assessment of disease severity based on the neurological functional grading, posterior tibial nerve somatosensory cortical-evoked potential (PTNSCEP) was measured. In the clinical practice in our country, the central conduction time (N22-P40) is not obtained in children because children do not cooperate well in the testing, while guardians usually will refuse administration of sedatives. For this reason, the central conduction time was not measured in the cohort of patients in this study. Besides, motor-evoked potential measurement was not carried out due to the lack of appropriate equipment in our hospital. As a result, P40 was used as the parameter for PTNSCEP analysis. Recordings were obtained from electrodes at the position Cz′ (= 2 cm behind Cz) with the reference electrode at Fz. Adhesive electrodes attached at the posterior tibial nerve at each ankle. A constant-current square-wave electrical pulse (duration = 300 μs) was delivered to electrodes overlying the posterior tibial nerve at each ankle. The bandpass filters were set at 30 and 250 Hz. The stimulus intensity was increased until there was a visible twitch of the intrinsic muscle. As absolute amplitude values were extremely variable among subjects, latencies were used. All latency values of the cortical SEPs (P40) used for calculations were obtained from each individual separately. Spinal cord lesion was frequently asymmetrical; therefore, results from the more severe leg were used in the data analysis. Waveforms of PTNSCEP were absent in two patients, who were then excluded from the cohort of cases for the convenience of statistical analysis. Each test was repeated at least twice to confirm the reproducibility.

Imaging

Radiological records of the patients were retrospectively analyzed. MRI was performed using a standard imaging protocol in sagittal, coronal and axial planes. No contrast media were administered. The MRI and CT images of each patient were reviewed simultaneously concerning the following

aspects: (a) symmetry of divided cord (symmetry splitting: two hemicords of approximately equal size; asymmetry splitting: one hemicord smaller than its fellow), (b) septum (presence: including bone or cartilaginous septum; absent: including fibrous or fatty septum), (c) lesion location (thoracic, thoracolumbar or lumbar area) and (d) number of split segments (1–3, 4–6 and 7–9).

Data analysis

Data were expressed as mean ± s.d. and were analyzed using SPSS 16.0. A two independent samples Student's *t*-test was performed. One-way analysis of variance was used to compare means of three samples. $P<0.05$ was considered as significant.

RESULTS

Clinical findings

There were 27 male and 65 female patients included in the analysis. The ages of the patients ranged from 3 to 58 years with a median of 6 years. It has been reported that diastematomyelia mostly present in childhood, and its presence in adulthood is extremely rare. As might be expected, most of the patients observed in this study were children at the time of diagnosis. The common symptoms were scoliosis, deformity of the foot and changes of the dorsal skin. The common musculoskeletal anomaly was asymmetry of the legs and weakness of the limb. Patients with urinary incontinence were referred to urologists for the evaluation of bladder function. The lumbar and thoracolumbar lesions in some patients could cause lower motor neuron findings in the legs and feet, such as decreased/absent reflexes, while thoracic lesions might cause upper motor neuron findings, for example, hypermyotonia. Paraesthesia in a dermatomal pattern was compatible with the segmental locations of the diastematomyelia. Approximately 50% of the cases had cutaneous abnormalities on the back, such as a hairy patch, dimple, pigmented nevus, hemangioma and subcutaneous mass.

Neurological functional grading

The neurological signs were variable. Neurological functional grading revealed substantial changes in nearly every case. The scores were significantly different with respect to the symmetry of splitting, presence of septum and location of lesion, while no evident differences were present in number of split segments aspect.

MRI and CT findings

Table 2 shows the MRI and CT findings in this cohort of diastematomyelia patients. The two hemicords appeared symmetrical in 24 cases, while asymmetrical in 58 cases. The septum was present in 66 cases and absent in the rest. Location of lesion was at thoracic level in 21, thoraciclumbar in 29 and lumbar in 32. The number of segments ranged from one to nine: 42 patients had one to three segments, 29 patients had four to six and 11 patients had seven to nine segments.

Comparison of clinical and neuroimaging features

A relation between the neurological functional grading and the imaging features including the asymmetry of splitting, presence of septum and location of lesion was observed ($P<0.05$). The number of split segments was not notably associated with the grading ($P>0.05$).

Meanwhile, PTNSCEP was also related with the parameters. Prolonged latency of PTNSCEP was considered an abnormality. The latency of PTNSCEP was significantly different in the asymmetry of splitting, presence of septum and location of lesion ($P<0.05$), with no difference in the number of split segments ($P>0.05$). The degree of change in PTNSCEP was found to be in accord with the severity of clinical deficits.

Table 2 Imaging and clinical findings in patients with diastematomyelia

Characteristic	*Number*	*Neurological functional grading*	*Latency of PTNSCEP*	P-*value*
Symmetry of divided cord				
Symmetry	24	237.46 ± 8.92	41.46 ± 3.83	<0.05
Asymmetry	58	230.00 ± 8.55	44.22 ± 3.06	
Septum				
Presence	66	230.48 ± 8.67	44.02 ± 3.21	<0.05
Absent	16	239.19 ± 8.49	40.94 ± 3.71	
Lesion location				
Thoracic	21	239.95 ± 8.29	40.67 ± 3.51	<0.05
Thoracolumbar	29	233.21 ± 8.08	42.83 ± 3.26	
Lumbar	32	226.16 ± 6.42	45.75 ± 1.90	
Number of split segments				
1–3	42	232.81 ± 10.40	43.19 ± 3.79	>0.05
4–6	29	232.62 ± 8.57	43.21 ± 3.55	
7–9	11	228.64 ± 5.32	44.82 ± 1.83	

Abbreviation: PTNSCEP, posterior tibial nerve somatosensory cortical-evoked potential.

DISCUSSION

Radiographic studies can provide objective data for determining the severity of diastematomyelia. Prompt and accurate diagnosis can be made based on imaging findings. Understanding the relationship between the clinical severity of diastematomyelia and the morphological characteristics presented in the images often leads to expeditious management, and the imaging findings may also serve as prognostic indicators. Given the paucity of literature in the imaging features of diastematomyelia, the treatment criteria for this lesion are scarcely established. The present study emphasizes the importance of the imaging features of diastematomyelia before making diagnostic and treatment decisions. To the best of our knowledge, this is the first study to address this issue in this disease.

With the technical advancement in the field of radiology, CT and MRI have been widely used in spinal imaging. They are often presented as noninvasive alternatives for the diagnosis and evaluation of spinal anomalies. Before the routine use of CT and MRI, the radiological diagnosis of diastematomyelia was dependent on plain radiographs and myelography. Plain radiographs are generally poor estimates of the disease. Myelography, even though may increase the accuracy of assessment, is an invasive form of investigation.[17] CT and MRI are different diagnostic tools. Both of them are of vital importance in diastematomyelia cases. In our study, all the patients suspected of diastematomyelia underwent CT and MRI. CT best shows the bony spur and the defective vertebrae. Especially, it reveals the type and structure of the spur, that is, whether the spur is bony, cartilaginous or fibrous in nature. In comparison with CT, MRI adequately delineates the presence and extent of the divided spinal cord. In our study, MRI preceded CT for all the patients because of its distinctive advantage over CT in localizing and characterizing the lesions. This imaging technique is considered the best diagnostic imaging modality for identifying diastematomyelia. Most of the diagnostic information in MRI in our practice derived from the

coronal and axial images. Sagittal images served as a supplement. The spinal column findings obtained by MRI were in accordance with those by CT. Compared with the conventional imaging methods, the combination of MRI and CT evidently improves the characterization of lesions. However, MRI or CT alone is inadequate in evaluating diastematomyelia.

Our neurological grading system provides useful information of the severity of neurological deficits in diastematomyelia. On the basis of the clinical symptoms and signs, the severity of neurological deficits of all the patients was scored. The major components of the neurological scoring system were gait, sphincter function, motor function, tendon reflex and sensation. The patients were subclassified according to the imaging outcome in terms of the symmetry of splitting, presence of septum, location of lesion and number of split segments. The clinical manifestations were significantly related to all the imaging features but the number of split segments. Although this feature may reflect the severity of diastematomyelia, it appears less useful in predicting the clinical implications of this lesion.

Somatosensory-evoked potential is a useful neurophysiological study for detecting functional abnormality of the spinal cord. It has been reported that there is a significant relation between clinical manifestations and abnormal somatosensory-evoked potential in patients with diastematomyelia.[18,19] The P40 is generally believed to represent the initial cortical processing of input from large diameter cutaneous and/or muscle afferent fibres, responsible for light touch, vibration and proprioceptive sensation. Abnormal PTNSCEP suggests that impairment of the somatosensory pathways may lead to poorer clinical situations in patients with diastematomyelia, which indicates sensory deficit in the relevant limb. Our study demonstrates that PTNSCEP in diastematomyelia is related to imaging, functional and clinical data. This further validates imaging features and the clinical manifestations in diastematomyelia.

The objective imaging findings of diastematomyelia are associated with the clinical neurological examination findings. Generally, the morphological characteristics reflected on the MRI and CT images are associated with the degree of neurologic deficit. The clinical features could be explained based on the CT and MRI findings of the spinal cord. Notably, neurological functional grading and imaging are two different examination methods for diastematomyelia. The former reflects neurological function; the latter reflects tissue structure. Imaging findings can provide additional evidence to help delineate the extent of the lesion. It is also possible that combining the two methods might produce more accurate assessment of the disease than using either alone. If an accurate assessment is made early, a proper treatment can be instituted and symptoms will be prevented. For example, in patients with asymmetry of splitting, presence of septum, low location of lesion and lower neurological functional grading, surgery should be the first choice of treatment to prevent neurological deficit.

Limitations

This study had the following limitations. First, the retrospective nature of the study performed in a single hospital and a relatively small number of subjects limited the statistical power of the study. Second, only patients over 3 years were included in our study. If patients were under the age of 3 years, detailed sensation examination was difficult and unreliable.

CONCLUSION

The imaging features in diastematomyelia are characteristic; various imaging features in diastematomyelia, except the number of split segments, are associated with the clinical manifestations of the lesion. Imaging findings are useful in defining the clinical status. Clinical and imaging data represent information about different aspects of diastematomyelia, which may be utilized in selecting surgical or nonsurgical therapeutic strategies.

DATA ARCHIVING

There were no data to deposit.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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Supplementary Information accompanies this paper on the Spinal Cord website (http://www.nature.com/sc)