Split Spinal Cord Malformations and Associated Spinal Anomalies: Comparison of MRI Findings with Intraoperative Results

Özgür Kılıçkesmez[1], Erol Taşdemiroğlu[2], Ercan İnci[1],
Sibel Bayramoğlu[1], Tan Cimilli[1]

Dr. Sadi Konuk Bakırköy Eğitim ve Araştırma Hastanesi, Radyoloji Servisi[1], İstanbul Eğitim Hastanesi, Nöroşirürji Servisi[2], İstanbul

ÖZET

Yarık spinal kord malformasyonları ve ilişkili spinal anomaliler: MR görüntüleme sonuçları ve operasyon sonuçlarının karşılaştırılması

Amaç: Bu çalışmanın amacı yarık spinal kord malformasyonlarında manyetik rezonans görüntülemenin tanı ve değerlendirmeye olan katkısını araştırmaktır.

Gereç ve Yöntem: Pang sınıflamasını temel alarak hastanemizde tedavi edilen 22 olguyu değerlendirerek bulguları sunduk.

Bulgular: Altı fibröz (%27.3) ve 16 kemik (%72.7) septum saptandı. Olguların 21'inde (%95.4) gergin omurilik sendromu vardı.

Sonuç: Diastematomyeliye, spinal disrafizmin diğer komponentlerinden olan lipomiyelomeningosel, meningosel, intrasakral meningosel, filum terminale lipomu, lipom, dermal sinüs traktı, dural ektazi, siringomiyeli ve kemik anomalileri eşlik etmekteydi.

Anahtar kelimeler: Yarık spinal kord malformasyonları, diastematomyeli, spinal disrafizm, MRG

ABSTRACT

Split spinal cord malformations and associated spinal anomalies: Comparison of MRI findings with intraoperative results

The aim of this study is to assess the contribution of magnetic resonance imaging (MRI) in the diagnosis and evaluation of split spinal cord malformations (SSCM) and associated spinal anomalies.

Material and Methods: According to pang's classification, we have reviewed and analyzed 22 cases of SSCMs treated at our institution and present our findings.

Results: Six fibrous (27.3%) and 16 osseous (72.7%) septums were detected. Twenty one of them (95.4%) had tense cord.

Conclusion: Other components of spinal dysraphism such as lipomyelomeningocele, meningocele, occult intrasacral meningocele, phylum terminale lipoma, lipoma, dermal sinus tract, dural ectasia, syringohydromyelia and bony abnormalities were associated with diastematomyelia.

Key words: Split spinal cord malformations, diastematomyelia, spinal dysraphism, MRI

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INTRODUCTION

Diastematomyelia is an occult spinal dysraphism also known as the split cord malformation or "double-barrelled spine". It was first described in 1837 and refers to an abnormality where the spinal canal is split by a fibrous, cartilaginous or bony septum creating two sleeves each containing a portion of spinal cord which is split sagitally (1-4). Pang and colleagues have defined two types of split cord malformations (SSCMs) (5,6). According to Pang's classification we have reviewed and analyzed 27 cases of SSCMs treated at our institution and present our findings. The aim of this study was to assess the contribution of magnetic resonance imaging (MRI) in the diagnosis and evaluation SSCM and associated spinal anomalies.

Yazışma adresi / *Address reprint requests to*: Özgür Kılıckesmez
Bakirkoy Dr. Sadi Konuk Training and Research Hospital
Radiology Department, Bakirkoy Istanbul 34147 TURKEY

Telefon / *Phone*: +90-532-734-6196

Elektronik posta adresi / *E-mail address*: okilickesmez@yahoo.com

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MATERIAL AND METHODS

The MRI scans and medical records of 27 patients operated for SSCMs were retrospectively rewieved. Twenty two pediatric cases were included in the study. Twenty two patients with SSCMs who were surgically treated in the neurosurgery service of Istanbul Education Hospital between January 1995 and December 2002 were reviewed. Patients' age and gender, symptoms and signs, radiological and operative findings, associated anomalies, outcome, and pathological specimens were

evaluated retrospectively. We routinely performed plain X-rays of the spine in the anteroposterior and lateral projections initially and then MRI scans of the spine on a 1,5 T magnet system were acquired. Imaging protocol was consisting sagittal and axial spin echo (SE) T1, sagittal and axial FSE T2, and in some of the patients coronal SE T1 weighted images were obtained additionally. When diagnosed, all SSCMs were surgically treated, even if the patient was neurologically intact.

RESULTS

There were 17 (77%) female and 5 (23%) male patients with a mean age of 9 year old (Table 1). The presence of neurocutaneous stigmata (hypertrichosis, hemangioma, hyperpigmentation, and subcutaneous lipomas) was the presenting sign in 10 of the 22 patients (45%). Back pain and leg pain were common, being seen in 13 patients (%59). Twelve patients presented with neurological symptoms including leg weakness and/or numbness and dysesthesias (54%). A skeletal deformity was seen in 10 patients (45%). Ten patients presented with scoliosis (45%) and four with leg-length asymmetry (18%). Bowel and bladder symptoms, alone or combined, were present in three patients (13%). After plain X-ray film of the spine revealed widened pedicles, an MRI of the spine was ordered, which led to the diagnosis of SSCM. MRI was capable of detecting septum in all cases. There were six fibrous (27.3%) and 16 osseous (72.7%) septums. Twenty of them (90%) had tense cord. Other abnormalities of spinal dysraphism such as lipomyelomeningocele, meningocele, occult intrasacral meningocele, filum terminale lipoma, lipoma, dermal sinus tract, dural ectasia, syringohydromyelia and bony abnormalities were associated with diastematomyelia. Intraoperative, and MRI findings of these malformations except exact determination of septum type, were similar. We also detected two very rare pathologies accompanying SSCMs. One of them was intrasacral meningocele, and the next was a case with double osseous spurs who had developed lateral sacral pseudomeningocele. To our knowledge lateral sacral pseudomeningocele has not been reported in the literature previously. The goal of surgery was removal of the fibrous or bone septum, resection of any other local spinal cord attachments causing tethering, and

Table 1: The list, radiological and clinical findings of patients with diastematomyelia

Case#	Age/Sex	Level of splitting	Type of septum in MRI versus operation	Termination level of conus medullaris	Other components of spinal dysraphism	Syrinx	Bony abnormalities
1	13/F	L3-5	Fibrous	S1			BV, DE, IDSN, VN
2	17/F	L1-2	Fibrous	L5			BV, DE, IDSN, VN
3	3/F	L1-S3	Double Osseous	S2	LSPM	+	DE, VN,IDSN
4	2/F	T4-8	Osseous	S3	Meningocele	+	HV, DE, BFV, S
5	18/F	T11-L1	Osseous	L2	Intradural lipoma		IDSN, HV, VN
6	3/M	L4	Osseous	S1	Lipomyelomeningocele	+	BV, BFV, DE, IDSN, HV, S, VN
7	1/F	T8-L4	Fibrous	L5		+	BV, HV, S, SH, VN
8	17/F	T12-L1	Osseous	L4	DST		BV, DE, IDSN, HV, S, VN
9	11/F	L1-2	Osseous	L4			DE, IDSN, VN
10	2/M	L3	Osseous	S3		+	DE, VN
11	13/M	L1-2	Osseous	L5		+	DE, IDSN, VN
12	9/M	L5	Osseous	S2	DST		DE, S,
13	5/F	L2-3	Osseous	L3		+	DE, S, VN
14	18/F	T11-L3	Osseous	L3		+	S,
15	10/F	L1-2	Osseous	L3			DE, IDSN, VN
16	6/F	L3	Osseous	L4	Intrasacral meningocele	+	DE, HV, S, VN
17	4/F	L3-4	Fibrous	S1	Lipomyelomeningocele	+	DE, IDSN, VN
18	18/F	L3-4	Fibrous	L5		+	BV, DE, HV, VN
19	3/F	L4-5	Osseous	S1			S
20	18/F	L1-3	Osseous	L3		+	
21	13/M	L3-4	Fibrous	L5	DST	+	DE
22	13/F	L1	Osseous	L3			IDSN, S

Abbreviations: BV: Block vertebra, BFV: Butterfly vertebra, DST: Dermal sinus tract, DE: Dural ectasia, HV: Hemivertebra, IDSN: Intervertebral disc space narrowing, LSPM: Lateral sacral pseudomeningocele, S: Scoliosis, SH: Sacral hypoplasia, VC: Vertebral cleft, VN: Vertebral body anteroposterior diameter narrowing

exploration for associated tethering-related anomalies such as dorsal tethering bands or thick phylum, which can be seen in the majority of patients. Following the detethering procedure, the dura was closed posteriorly with or without placement of a patch graft, whereas anterior dural defects were left open. The patients were kept flat postoperatively for an average period of 72 hours and were then allowed to progressively advanced to full activity levels. We have not experienced any complications in the post operative period in the study group. The patient with lateral sacral pseudomeningocele had undergone sectioning of the phylum terminale in another center. We only resected the double spurs and pseudomeningocele in this patient in whom the symptoms were progressing over the time.

DISSCUSSION

The term diastematomyelia, also called the split cord malformation, refers to a sagittal division of the spinal cord into two symmetrical or asymmetrical hemicords, each containing a central canal, one dorsal horn (giving rise to a dorsal nerve root), and one ventral horn (giving rise to a ventral nerve root). Both hemicords are surrounded by a layer of pia. The division may involve the entire thickness of the cord or may only affect the anterior or posterior half of the cord (partial diastematomyelia) (4).

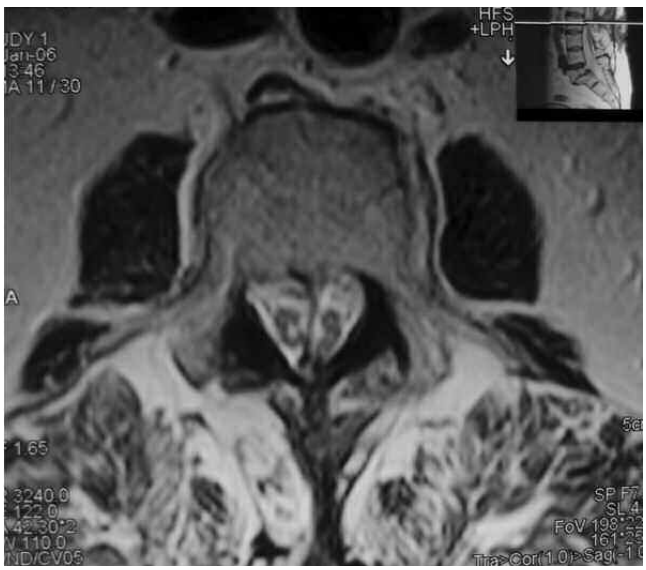

Figure 1a: Diastematomyelia. Axial T2 weighted image of a patient with type I diastematomyelia, the bony septum is easily visible

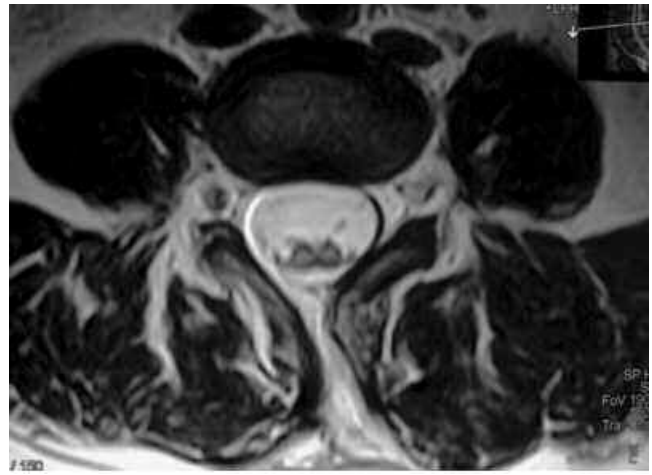

Figure 1b: Diastematomyelia. Axial T2 weighted image of another patient with type II diastematomyelia, No septum may be seen between two hemicords.

Pang and colleagues have defined two types of SSCMs. A Type I SSCM consists of two hemicords, each containing within its own dural tube and separated by a dura-sheathed rigid median septum (Figure 1a). A Type II SSCM consists of two hemicords housed in a single dural tube separated by a non-rigid, fibrous median septum (Figure 1b). SSCMs are seen more often in females with a female to male ratio of 1.5:1. However we detected a higher ratio of 3.29:1 in our study group. The average age at presentation was noticed to be 5.1 year by Andar et al (7). Cutaneous stigmata such as hypertrichhosis, nevi, lipomas, dimples, and hemangiomas overlie the spine in more than half of cases. Half of affected patients manifest orthopedic problems of the feet, particularly clubfoot. A spesific neuroorthopedic syndrome consistitng of weakness and muscle wasting in one leg, often associated with ipsilateral clubfoot, is seen in about half of patients with lumbar diastematomyelia (8,9). Scoliosis is common in older children and adults. Neurological symptoms are nonspecific, indistinguishable from other causes of cord tethering. The majority of septa were localized at the lumbar region, as in other series. Cervical and sacral locations are extremely rare. The conus medullaris is usually located caudal to L-2. The reported spinal lesions in association with SSCMs are a thick phylum terminale, myelomeningocele, meningocele, lipomyelomeningocele, limited dorsal myeloschisis, teratoma, neurenteric cyst, lipoma, dermal sinus tract, dermoid cyst, epidermoid cyst, arteriovenous malformation, epidural venous angioma, and arachnoid

cyst. The common bony deformities include scoliosis, block vertebra, hypoplastic vertebra, kyphosis and fused ribs. MRI correctly established the diagnosis in all cases and additional lesions as a cause of tethering were demonstrated in all cases (5,6,10,11). Imaging of diastematomyelia can be difficult because patients have severe scoliosis, often with a rotatory component. The plain radiograph is often the first modality used in the evaluation of these children and may demonstrate a scoliosis or associated vertebral anomalies. The bony spur when present is not often easily visible; however its presence may be suggested by narrowed adjacent intervertebral disc space. Also an increase in the interpeduncular distance may suggest an underlying diplomelia of the cord. Myelography, which in the past would have been advocated for further assessment of this condition, has now been replaced by MRI. At an early age, ultrasound may be used to demonstrate abnormalities of the spinal cord as well as the vertebral bodies and the bony spur. It can also show the presence of an associated syrinx (12,13). Computed tomography is the investigation of choice to optimally demonstrate the bony spur, its extent and relationship to the vertebral bodies as well as showing the vertebral anomalies. Reconstructions can also be obtained, on a helical or multi-slice machine, allowing appreciation of the full extent of the spur and its relationship to adjacent vertebrae (12,13). MRI, with its multi-planar capabilities and superior soft tissue contrast, allows delineation of the extent of the split in the cord and shows the associated anomalies such as lipomas and the syrinx (1,2,4,13). The entire extent of the divided spinal cord in cases of diastematomyelia can be demonstrated consistently on only a few selected coronal MR images. Visualization of associated abnormalities such as tethered cord and other abnormalities is another advantage. T1-weighted images are optimal to visualize the spinal cord and to look for fibrolipomas of the phylum terminale. Bony and cartilaginous spurs are most easily identified on axial T2- or T2*-weighted images or on CT. The bony spur forms from cartilage and has multiple ossification centers. Therefore depending on the age of the patient and the number of ossification centers, the spur can be non-ossified cartilage, cartilaginous with multiple small ossification centers arranged linearly between the two hemicords, a bony strut attached to the wall of the spinal canal by a synchondrosis, or a complete osseous bridge joining the vertebral body with the posterior elements (14). The spur may be isointense or slightly hyperintense compared to CSF on T1-weighted images if nonossified; it will be hyperintense on T1-weighted images if ossified because of the high signal from marrow. Bony, cartilaginous, and fibrous spurs all appear hypointense on T2 weighted spin echo and gradient echo images. The CT attenuation will vary, depending on the stage of ossification. It must be emphasized that even osseous spurs can be missed on T1-weighted spin echo MR images. Therefore, patients with two hemicords should be imaged using T2- or T2*-weighted images (which facilitate identification of bone) in the axial plane or with CT (4). Han et al (15) have stated that MRI was superior except detecting the spur with cortical bone, however we detected the osseous spurs in our study, and certainly the cortical bones were clearly identified as hypointense structures on T2-weighted axial images. Prior to the MRI "era" it was difficult to diagnose patients with occult spinal dysraphism and certainly a tethered cord without utilization of "invasive" diagnostic procedures. Therefore, many infants were untreated and it was possible to observe the "natural history" of the untreated tether. It has been a common observation that the majority of these children experience neurological deterioration during childhood or adolescence. Occasionally, sudden and catastrophic neurological deterioration was associated with normal sports activities or minor injuries. Even the occasional normal child who reaches adulthood is at risk for deterioration in later years. Childbirth may be associated with "precipitous" neurological decline as a result of normal obstetric positioning (16,17). It therefore seems clear that the hazard of deterioration is very significant and surgery must always be considered prior to the evolution of neurological decline. Tethered cord accompanies to the most of the patients with diastematomyelia. So it is obvious that these children should be evaluated also for a probable tethered cord syndrome with MRI (3,10,11,18).

Magnetic resonance imaging was capable of detecting the existence of septum and discriminate the osseous and fibrous spurs each other. However the degree of osteoid matrix was over or underestimated in some cases. On the other hand almost completely all of the patients with SSCMs reviewed in this study had

coexisting spinal abnormalities and multiple causes of tethering associated with their congenital anomaly. These causes should be detected thoroughly before the surgery. With the high quality of MR imaging in the current era, these lesions will almost always be visible on preoperative imaging as it was in our study.

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