ROLE OF MRI IN EVALUATION OF SUSPECTED SPINAL DYSRAPHISM

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ABSTRACT

Congenital abnormalities of the spine and spinal cord are referred to as spinal dysraphisms. Spinal dysraphisms can be broadly categorized into open and closed type. This can be grouped as open if the overlying skin is not intact causing leakage of cerebrospinal fluid and occult if the defect is well covered with full thickness skin. The surgical outcome may be better if intervention occurs before the age of 3 years.

Spinal neuroimaging therefore has the important role of determining the presence or absence of an occult spinal dysraphic lesion, so that appropriate surgical treatment can be instituted in a timely manner. MRI is the imaging modality of choice to demonstrate the spectrum of findings in patients with suspected spinal dysraphism, because of its better diagnostic performance, excellent soft tissue characterization and importance in pre-surgical planning.

AIMS

To demonstrate the spectrum of MRI findings in spinal dysraphism.

MATERIALS AND METHODS

Prospective evaluation of 66 patients with suspected spinal dysraphism on 1.5 T MRI.

RESULTS

Age of the patients ranged from 17 days to 13 years. Commonest clinical presentation was swelling in the back. Open spinal dysraphism, in which lumbosacral myelomeningocele was most common presentation. Diastematomyelia was the next most common presentation. Associated abnormalities like hydrocephalus, Arnold–Chiari, syrinx, hydronephrosis were commonly encountered in open defects. We have one case of open spinal dysraphism with osseous hamartoma. MRI with its multiplanar capabilities and superior soft tissue contrast, allows detection of detailed anatomy and characterization of congenital spinal disorders.

CONCLUSION

MRI is an excellent imaging modality for visualizing the spinal cord at all ages and is the imaging modality of choice for defining complex spinal dysraphism.

KEYWORDS

MRI, Spinal Dysraphism, Lumbar Myelomeningocele, Diastematomyelia, Open Spinal Dysraphism with Osseous Hamartoma, Tethered Cord, Dermal Sinus.

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INTRODUCTION

Congenital abnormalities of the spine and spinal cord are referred to as spinal dysraphisms. Spinal dysraphisms can be broadly categorized into open and closed type. This can be grouped as open if the overlying skin is not intact causing leakage of cerebrospinal fluid and occult if the defect is well covered with full thickness skin. The estimated incidence of spinal dysraphism is about 1-3/1000 live birth. About 55-70% of neural tube defects occur in females. Early detection and prompt neurosurgical correction of occult spinal dysraphism may prevent upper urinary tract deterioration, infection of dorsal dermal sinuses or permanent neurologic damage. The surgical outcome may be better if intervention occurs before the age of 3 years.

Financial or Other, Competing Interest: None. Submission 09-01-2016, Peer Review 08-02-2016, Acceptance 15-02-2016, Published 29-02-2016. Corresponding Author: Dr. Raghavendra Y. Junior Resident, Osmania General Hospital, Hyderabad. E-mail: dr.raghavan08@gmail.com DOI: 10.14260/jemds/2016/203 Spinal neuroimaging, therefore, has the important role of determining the presence or absence of an occult spinal dysraphic lesion, so that appropriate surgical treatment can be instituted in a timely manner. Magnetic Resonance Imaging (MRI) is the examination of choice because of its better diagnostic performance, excellent soft tissue characterization and importance in presurgical planning.

MATERIALS AND METHODS

We prospectively evaluated 66 patients with suspected spinal dysraphism and who had undergone MR imaging of the spine from May 2012 to September 2014 were included in the study after informed consent.

Inclusion Criteria

Patients who presented with various neurological problems and skin stigmata suspicious of spinal dysraphism.

Exclusion Criteria

Patients with metallic implants and patients with claustrophobia were excluded from the study. In all patients clinical history and antenatal history were recorded. Sedation was given in required patients.

MRI was performed on a 1.5 Tesla electromagnet (General Electrical Medical Systems). The pulse sequences included T1WI, T2WI using spin echo and Short Tau Inversion Recovery sequences. Axial and sagittal T2WI were obtained with TR of 3000ms and TE of 120ms. Sagittal T1WI were obtained with TR of 60ms and TE of 30ms. Sagittal and coronal STIR images were obtained with TR of 3000ms and TE of 40ms. Images were obtained with an interslice gap of 5.2mm, slice thickness of 4mm and a matrix size of 512x512. On MRI, imaging findings in vertebrae, spinal cord and soft tissues were noted.

OBSERVATIONS AND RESULTS

66 patients who presented with various neurological problems and skin stigmata suspicious of spinal dysraphism were referred to our department in Osmania General Hospital and underwent Magnetic Resonance Imaging of the spine in a time period from May 2012 to September 2014 were included in our study. Age of the patients ranged from 17 days to 13 yrs. Most of the children are below 1 yr. of age. Of the 66 patients, 40 were female patients and 26 were male patients. The below table shows the gender distribution of spinal dysraphism.

GENDER	NUMBER	PERCENTAGE			
Males	26	39.4%			
Females	40	60.6%			
Table 1: Gender Distribution					

Of the 66 patients, 38 are open spinal dysraphisms and 28 are closed dysraphisms.

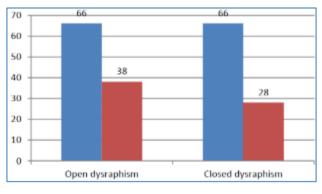


Fig. 1: Types of Spinal Dysraphism

Children with suspected spinal dysraphism can present with various clinical features such as swelling on the back, dimple, hemangioma, dermal sinus, lower limb weakness, bladder and bowel incontinence.

Sl. No.	Clinical Feature	Number	Percentage	
1.	Swelling in the back	51	77.2%	
2.	Hypertrichosis	6	9%	
3.	Dimple	3	4.5%	
4.	Lower	14	21.2%	
5.	Limb weakness	6	9%	
6.	Urinary incontinence	4	6%	
7.	Dermal sinus	2	3%	
	Fecal incontinence			
Table 2: Clinical Presentation of Spinal Dysraphism				

Myelomeningocele followed by diastematomyelia were the commonest presentations in our study. One case was cervical myelomeningocele with osseous hamartoma.

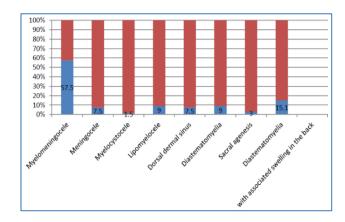


Fig. 2: Different Types of Dysraphism

Location of myelomeningoceles of the 38 patients in our study, 4 were noted in the cervical region, 2 in the dorsal region, 5 in the lumbar region, 7 in the sacral region, 20 in lumbosacral region and 2 in dorsal region.

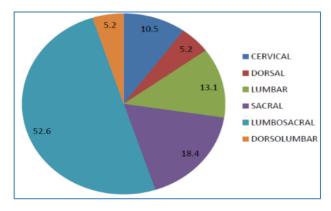


Fig. 2: Location of Myelomeningoceles

Of 66 patients, 16 Diastematomyelia patients were detected. In type 1, two hemicords are encased in respective dural sacs and separated by a bony septum. In type 2, the hemicords are encased in a single sac and separated by a fibrous septum.

Sl. No.	Type of Diastematomyelia	Number	Percentage	
1.	Type-1	4	25%	
2.	Type-2	12	75%	
Table 3: Type of Diastematomyelia				

Open defects were associated with abnormalities such as hydrocephalus, Arnold-Chiari, syrinx.

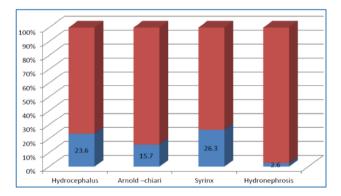


Fig. 4: Open Defects with Associated Abnormalities

Closed defects were associated with abnormalities such as segmentation anomalies, syrinx, scoliosis, tethered cord.

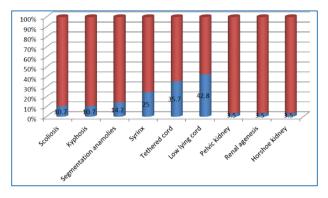
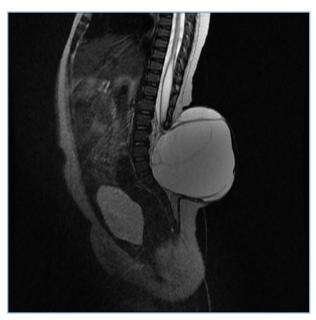


Fig. 5: Closed Defects with Associated Abnormalities



Tethered cord can result from variety of conditions. In our study, 10 cases are associated with closed spinal defects and 4 cases are due to surgery for myelomeningocele.

Sl. No.	Tethered cord	Number	Percentage	
1. 2.	Associated with closed defects Post Surgery	$\begin{array}{c} 10\\ 4 \end{array}$	71.5% 28.5%	
Table 4: Tethered Cord				

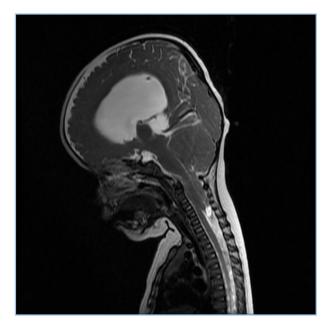
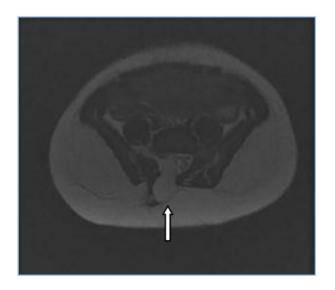


Fig. 6: Sagittal T2WI – Images showing the Herniation of Meninges and Nerve Roots into the Subcutaneous Plane and Tonsillar Herniation - Lumbar Myelomeningocele with Arnold Chiari Type 2



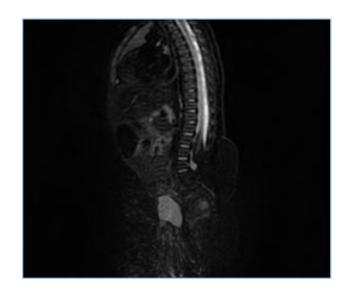


Fig. 7: Axial T2W - Images showing Herniation of Nerve Roots and Altered Signal Intensity Mass Noted in Epidural Space showing Continuity with Subcutaneous Fat. Sagittal STIR - Suppression of the Lesion – Lipomyelocele

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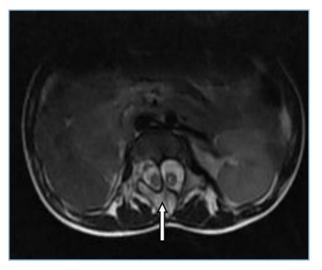
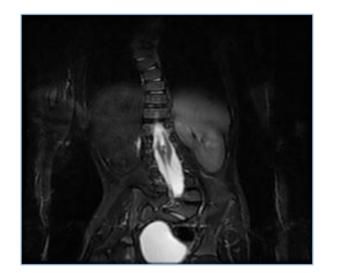


Fig. 8: Coronal STIR and Axial T2W Images showing Two Hemicords with Two Dural Sacs with Bony Septum between them with Absent Right Kidney - Diastematomyelia Type 1



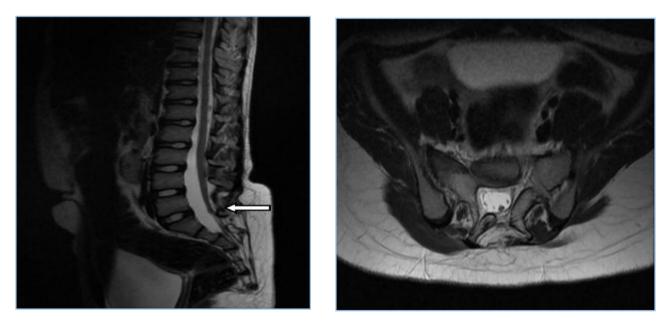


Fig. 9: Sagittal and Axial T2W Images showing Spina Bifida of L5 and S1 Vertebrae with Low Lying and Tethering of Cord - Spina Bifida with Low Lying and Tethered Cord with Thick Filum

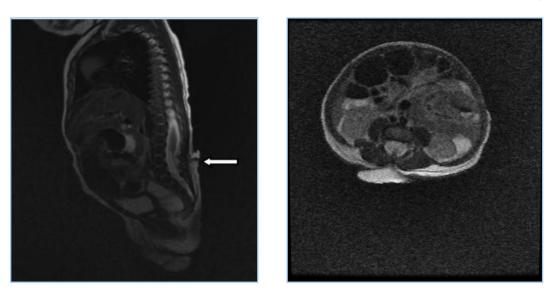


Fig. 10: Sagittal and Axial T2WI showing Hypointense Linear Structure Extending from Skin and Subcutaneous Planes at L4 Level into the Dura s/o Dorsal Dermal Sinus

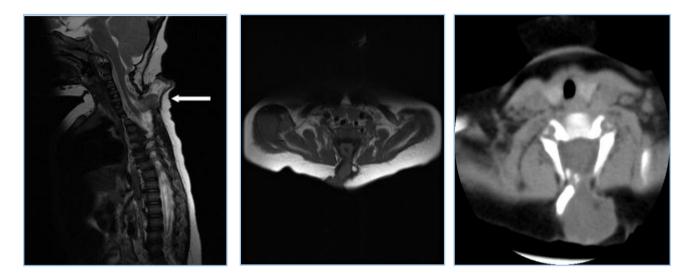


Fig. 11: Sagittal T2WI and Axial T1WI showing Herniation of Spinal Cord and Meninges. Axial CT showing Osseous Component - Cervical Myelomeningocele with Osseous Hamartoma

DISCUSSION

Congenital abnormalities of the spine and spinal cord are referred to as spinal dysraphisms. The purpose of MR imaging in spinal dysraphism is (a) Detection of detailed anatomy (b) For soft tissue characterization of lesion (c) For presurgical planning (d) For early detection and prompt neurosurgical correction of occult spinal dysraphism in order to prevent permanent neurological damage. Our study of role of MRI in evaluation of suspected spinal dysraphism consisted of 66 patients who presented to our hospital with suspected spinal dysraphism and underwent MR imaging of the spine.

The age of the patients in our study ranged from 17 days to 13 yrs. Most of the children are below 1yr of age. In a study by Muhammed Nafees, et al. age of the patients ranged from 16 days old to 37 yrs. old, most of them below 6.4 yrs.¹ In our study, of the 66 patients 40 (66.6%) were female patients and 26 (33.3%) were male patients similar to study by Mohamed Fathy Dawodh et al., in which 18 are female patients and 14 are male patients.²

In our study of the 66 patients, 38 (57.5%) are open spinal dysraphisms and 28 (42.4%) are closed dysraphisms. The results were similar to study by Mohamed Fathy Dawodh et al.; 18 are open dysraphism and 14 are closed dysraphism.²

Children with suspected spinal dysraphism can present with various clinical features such as swelling on the back, dimple, hemangioma, dermal sinus, lower limb weakness, bladder and bowel incontinence. In our study, swelling in the back is the commonest clinical feature which constituted 51 (77.2%). In a study by Mohamed Fathy Dawodh, et al., swelling in the back is the commonest clinical feature which constituted 26 (81.2%). In a study by Kumar R, Singh SN, et al. swelling in the back is the commonest clinical feature which constituted 89 (57%).^{2,3}

Among different types of dysraphism myelomeningocele is the commonest. In our study, myelomeningocele constituted 38 (57.5%). In a study by Mohamed Fathy Dawodh, et al., myelomeningocele is the commonest dysraphism which constituted 15 (46.8%).

In a study by Muhammed Nafees, et al. myelomeningocele is the commonest dysraphism which constituted 29 (39.2%).^{2,1}

Location of myelomeningoceles of the 38 patients in our study, 4 were noted in the cervical region, 2 in the dorsal region, 5 in the lumbar region, 7 in the sacral region, 20 in lumbosacral region and 2 in dorsal region. Among these lumbosacral region is the commonest location, which constituted 20 (52.6%). In a study by Muhammed Nafees et al., lumbosacral region is the commonest location which constituted 38 (51.4%). In a study by Mohamed Fathy Dawodh et al., lumbosacral region is the commonest location which constituted 11 (73.3%).^{1,2}

In our study of the 66 patients, 16 (24.2%) Diastematomyelia patients were detected. Of which type 2 constituted 75% and type 1 constituted 25%. In a study by Taahira Nishtar et al., of the 53 patients 2 (4%) Diastematomyelia patients were detected.⁴

Open defects are associated with abnormalities such as hydrocephalus, Arnold-Chiari, syrinx. In our study hydrocephalus constituted 9 (23.6%), Arnold-Chiari type 2 constituted 6 (15.7%). In a study by Kumar R, Singh SN, et al., hydrocephalus constituted 71 (49%), Arnold-Chiari type 2 constituted 62 (45%).³

We had a rare case of cervical myelomeningocele with osseous dysraphic hamartoma. Zuppani HB, et al. reported a case of lipomyelocele with osseous dysraphic hamartoma in a child.⁵

A prospective study at NUR Research Centre Institute of Nuclear Medicine and Allied Sciences, Lucknow Marg, Delhi, Dept. of Neurosurgery, GB Pant Hospital New Delhi, was carried out in 100 cases of suspected occult spinal dysraphic anomalies with MRI in order to determine its diagnostic efficacy as the initial imaging modality. MR imaging provided accurate preoperative information in 91 out of 92 cases (98.9%). It is concluded that MRI is an excellent primary diagnostic tool, together with a plain radiograph for complete preoperative evaluation of mid-line spinal anomalies.

Comparing with the above mentioned studies, our study revealed that MRI is imaging modality of choice to demonstrate the spectrum of findings in patients with suspected spinal dysraphism.

CONCLUSION

MRI features of 65 patients with suspected spinal dysraphism were studied and imaging findings were interpreted.

 The age of the patients in our study ranged from 17 days to 13 yrs. Most of the children are below 1 yr. of age.

- 2. Female predominance was noted in our study, which constituted 66.6%.
- 3. Open defects (57.5%) predominated in our study than closed defects (42.4%).
- 4. Swelling in the back is the commonest clinical feature in our study, which constituted 51 (77.2%).
- 5. Myelomeningocele is the commonest type of dysraphism, which constituted 38 (57.5%).
- 6. In myelomeningoceles, lumbosacral region is the commonest location which constituted 20 (52.6%).
- In our study, 16 (24.2%) Diastematomyelia patients were detected. Of which type 2 constituted 75% and type 1 constituted 25%.
- 8. Open defects are associated with abnormalities such as hydrocephalus, Arnold-Chiari syrinx. Other spinal cord abnormalities such as tethered cord, low lying cord, segmentation anomalies, scoliosis, kyphosis were well delineated in our study.
- 9. Most of the myelomeningoceles in our study are associated with abnormalities.

Hence, MRI with its multiplanar capabilities and superior soft tissue contrast, allows detection of detailed anatomy and characterization of congenital spinal disorders.

MRI is an excellent imaging modality for visualizing the spinal cord at all ages and is the imaging modality of choice for defining complex spinal dysraphism.

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