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ROLE OF MAGNETIC RESONANCE IMAGING (MRI) IN CHARACTERIZATION OF SPINAL DYSRAPHISM

Bandi Arun Kumar[1], Radha Rani K[2], A Ranjith Kumar[3], Duddyala Dilip Kumar[4]

[1]*Assistant Professor, Department of Radiology, Kurnool medical college, Kurnool, Andhra Pradesh, India.*
[2]*Professor, Department of Radiology, Kurnool medical college, Kurnool, Andhra Pradesh, India.*
[3]*Assistant Professor, Department of Radiology, Arunai Medical College and Hospital, Tiruvannamalai, Tamil Nadu, India.*
[4]*Assistant professor, Department of Radiology, Kurnool medical college, Kurnool, Andhra Pradesh, India.*

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Corresponding Author:
Dr. Duddyala Dilip Kumar,
Assistant Professor, Department of Radiology, Kurnool medical college, Kurnool, Andhra Pradesh, India.
Email: ddilipkmc@gmail.com

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ABSTRACT

Background: A spectrum of congenital anomalies known as spinal dysraphism is characterized by an incomplete fusion of the spine's midline mesenchymal, bony, or neural parts. MRI is the preferred imaging technique for identifying and diagnosing spinal dysraphism. **Aim:** To study the role of magnetic resonance imaging for the evaluation of spinal dysraphism.

Materials and Methods: It was a prospective study. This study was performed on patients with clinically suspected spinal dysraphism who referred for MRI spine to the department of Radio-diagnosis and imaging, Kurnool medical college and GGH during the period from February2021-October 2022. In the present study cases of the spine abnormalities and spinal cord malformations are included.

Results: The present prospective study of 30 patients with a clinical suspicion of spinal cord malformations had a female predominance with the age group of 0-30 years accounting for 93% of the cases. Cutaneous markers were seen in 57.14% of cases of CSD with DDS and hypertrichosis being the commonest cutaneous markers. The commonest location for spinal involvement was the lumbar region followed by the thoracolumbar region. In the present study, OSD were more commonly seen than closed spinal dysraphism. The commonest open spinal dysraphism observed was a meningomyelocele, which commonly presented with a vertebral defect and mass. The commonest closed dysraphism observed in this study is intradural lipoma followed by diastematomyelia which again had a female preponderance. The associated abnormalities studied were: Tetheredcord, syringohydromyelia, and Chiari malformation. Tetheredcord was most commonly associated with meningomyelocele and diastematomyelia followed by spinal lipomas. Syringohydromyelia was most commonly associated with myelomeningocele, followed by tetheredcord and diastematomyelia. In our study, all the cases of Chiari malformations are associated with open spinal dysraphism. Of all the vertebral anomalies, spina bifida was the commonest followed by hemivertebra and butterfly vertebra in equal proportions. In 50%(n=8) cases of meningomyeloceles (n=16) was associated with hydrocephalus and these patients were planned for ventriculoperitoneal shunt.

Conclusion: MRI is the investigation of choice for characterizing the anomalies of spinal cord and associated soft tissues in spinal dysraphism. MRI clearly identifies and characterizes the nature of neural tissue protruding through the dysraphic spine in Meningomyelocele, Myelocele and Meningocele.

Keywords: MRI, Dysraphism, Meningomyelocele, Syringohydromyelia, Chiari malformations, Cutaneous markers.

INTRODUCTION

A spectrum of congenital anomalies known as spinal dysraphism is characterized by an incomplete fusion of the spine's midline mesenchymal, bony, or neural parts.[1] Most of these disorders are identified during prenatal, at birth, or in the first few months of life; however, some may be identified in older children or adults.[2] The global incidence of neural tube defects ranges between 1 to 10 per 1,000 births, with nearly half of these cases falling under spinal dysraphism and the other half as anencephaly.[3] Because of antenatal screening and folic acid supplementation, the prevalence of neural tube defects (including anencephaly and spinal dysraphism) has decreased over the last 25 years.[4] Spinal development can be divided into three stages: gastrulation, primary neurulation and then secondary neurulation. Abnormalities in any of these stages can result in spinal cord or spine malformations.[2,5]

Spinal dysraphism are of two types: open and closed. A defect in the skin covering the neural tissue resulting in an open type spinal dysraphism (OSD), which exposes the neural tissue to the outside environment. Another one is a closed spinal dysraphism, in which skin covers the neural tissue (closed spinal dysraphism-CSD). Based on the presence or absence of a subcutaneous swelling, closed spinal dysraphism is further classified on the basis of with subcutaneous mass and without sub cutaneous mass.[6,7] Clinical manifestations of these patients include cutaneous stigmata, neurological symptoms, bowel, bladder symptoms and orthopaedic symptoms.[8] So, these patients' evaluation and attention are not limited to the primary lesion but also to systems outside the nervous system. Although plain radiographs are helpful in the diagnosis of spinal abnormalities, sometimes patients with significant underlying disease can have normal-appearing spines.[9]

Ultrasonography is used as a screening tool for new-borns and infants and is mostly useful for the antenatal diagnosis of spinal dysraphism.[10] B-mode sonography has significant limitations, including imprecise soft tissue characterization, the inability to perform direct contact scanning on patients with exposed neural tissue, fragile meningeal sacs without running a significant risk, and poor sonographic penetration of dense scar.[11]

As the posterior spinal elements continue toossify, sonography becomes less effective.[12] Due primarily to its multiplanar imaging and tissue characterization capabilities, MRI has improved the prospect of early and case-specific treatment for spinal dysraphism by making the diagnosis simpler, quicker, and more accurate.

A non-invasive method for assessing the spine in children with clinically and/or radiographically suspected spinal dysraphism is MR imaging. With this approach, a conclusive diagnosis can be made without the risks of ionizing radiation or intrathecal contrast injection.[13] MRI is the preferred imaging technique for identifying and diagnosing spinal dysraphism.

Aims & Objectives

1. To study the role of magnetic resonance imaging for the evaluation of spinal dysraphism.
2. To study the magnetic resonance imaging characteristics of different spectrum of lesions in spinal dysraphism.
3. To study usefulness of MRI in pre surgical planning.

MATERIALS AND METHODS

Source of data

This study was performed on patients with clinically suspected spinal dysraphism who referred for MRI spine to the department of Radio-diagnosis and imaging, Kurnool medical college and GGH.

Sample size: Thirty cases (30)

Type of study: Prospective study.

Inclusion Criteria

1. All clinically suspected cases of the spine abnormalities and spinal cord malformations.

Exclusion Criteria

1. All post-operative cases
2. Patients who have contraindications for MRI.
3. Duration of Study:21 months (February2021-October2022)

Method of Collection of Data

The method of study consists of a structured pre-prepared case proforma was used to enter the patient details, antenatal and family history. A thorough general physical examination was done in all cases before the MRI examination.

Equipment and Sequences

All examinations were performed using 1.5 Tesla superconducting magnet, Philips MRI machine, using a Phased array spine coil.

Patient position: Supineposition. Slice thickness – 2 to 3 mm.

Characterization of Spinal Malformations

The various spinal dysraphism were broadly categorized into open or closed types. These were in turn subclassified based on the Clinico-radiological classification. Next the presence and type of associated vertebral anomaly was noted. The following associated findings were noted:

- Syringohydromyelia
- Tethered cord: The classical dorsal displacement of the conus with a large, ventral CSF space was taken as suggestive of tethered cord.
- Arnold Chiarimal formation Type I or II
- Presence of Hydrocephalus.

Diastematomyelia was again subdivided based on the presence or in the absence of bony spur and associated anomalies were noted. [Table 1]

Table 1: The MR parameters were as follows

PARAMETER PLANE	SEQUENCE	FOV (mm)	MATRIX (pixel)	TR (ms)	TE(ms)
SAGITTAL	T1W	320	256x256	450-500	15
	T2W	320	256x256	2500-3500	100
	STIR	320	256x256	2500-3000	75
AXIAL	T2W	200	205X256	2500-3500	110
	STIR	200	205X256	2500-3000	75
CORONAL	STIR	320	256x256	2500-3000	75

RESULTS

In our study spinal dysraphism was most commonly seen in Female [n=17, 56.7%] and male [n=13, 43.3%]. [Table 3]

In our study, total CSD cases were14, in that cutaneous markers were seen in 57.14% [n=8] cases of closed spinal dysraphism. Hypertrichosis constituted 37.50% [n =3], dermal sinus constituted50%[n=4], and portwine stain constituted of 12.50% [n=1]. [Table 4]

In our study of 30 cases, lumbar spine [n=22,73.33%] was most commonly involved followed by lumbosacral spine [n=7,22.5%], thoracolumbar spine [n=4,13.33%], cervicothoracic spine [n=2, 6.67%], cervical spine [n=1,3.33%] and lumbosacral [n=1, 3.34%] in decreasing order. [Table 5]

In our study, spinal curvature abnormalities were seen in 23% of cases [n=30]. [Table 6]

In our study, vertebral anomalies were present in 93.33% of cases [n=28], among which spina bifida is the commonest constituting to 89.5% [n=25], hemivertebra and block vertebrae each constituted 3.5% [n=1] and butterfly vertebra constituted 3.5% [n=1]. [Table 7]

In our study, the most common spinal abnormality encountered was myelomeningocele[n=16,54%] followed by syringohydromyelia [n=11,23%], scoliosis [n=7, 14.50%,], intradural lipoma [n=4, 14%], dorsal dermal sinus and diastematomyelia [n= 3, 10%] in decreasing order of frequency. [Table 8]

In our study of30 cases, 53% of cases [n=16] are open dysraphisms whereas 47% [n=14] are the closed type. [Table 9]

In our study of open spinal dysraphism, meningomyeloceles are the commonest ones [n=15, 93%] followed by lipomeningomyelocele [n=1,6%]. We did not report any case myelocele or hemi myelomeningocele. [Table 10]

In our study, out of 14 cases of closed spinal dysraphism, Intradurallipoma was the most common abnormality present in 29% [n=4] followed by diastematomyelia [n=3,21.25%]. [Table 11]

In our study, most common spinal level involved is Lumbar (n=11,68.75%) followed by thoracolumbar (n=3, 18.75%). [Table 12]

In our study, Anomalies associated with myelomeningocele, most common syringohydromyelia and hydrocephalus (n= 8, 27%) and tethered cord and Arnold Chiari malformations (n=7, 23%). [Table 13]

In our study of 13 cases of tethered cord, the most common associated abnormality included MMC [n=8, 61.54% each], followed by DDS and Intradural Lipoma [n=2, 15.38%]. [Table 14]

In our study, Spinal abnormalities associated with syringohydromyelia, most common myelomeningocele (n=9, 60%), Tethered cord (n=5, 33%) and diastematomyelia (n=1,7%). [Table 15]

In our study, Spinal abnormalities associated with hydrocephalus (n=9,30% cases), most commonly myelomeningocele (n=8, 89%) followed by dorsal dermal sinus (n=1, 11%). These patients were planned for ventriculoperitoneal shunt. [Table 16]

Representative cases

Case 1: Cervical Meningomyelocele

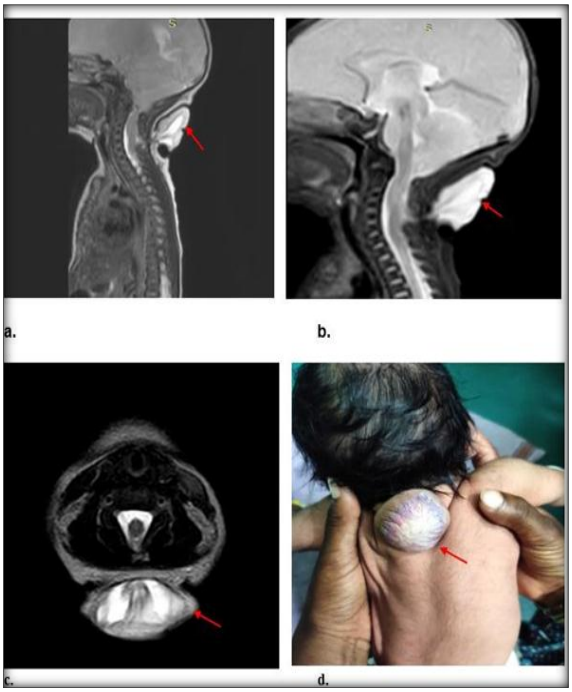

Figure 1: CERVICAL MENINGOMYELOCELE: A 4 Day-old male child with swelling in the cervical region, sagittal T2W image (a) shows cervical meningomyelocele with herniation of the nerve roots over the skin surface, sagittal STIR image(b)shows dilatation of the ventral subarachnoid space, axial T2 weighted image (c) of brain shows cystic lesion with nerve roots and corresponding clinical photo(d) shows swelling in cervical region

Case 2: Dorsal Dermal Sinus

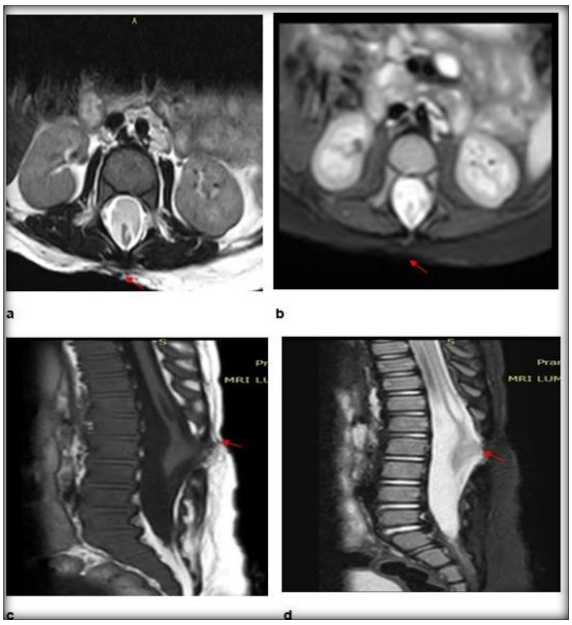

Figure 2: DORSALDERMALSINUS: A18-Month-old male child presented with skin sinus over the lumbar region and Axial T2 (a) andSTIR (b) weighted image shows spina bifida with dermal sinus and sagittalT1(c), T2(d)weighted images shows dilated terminal ventricle and tethered cord.

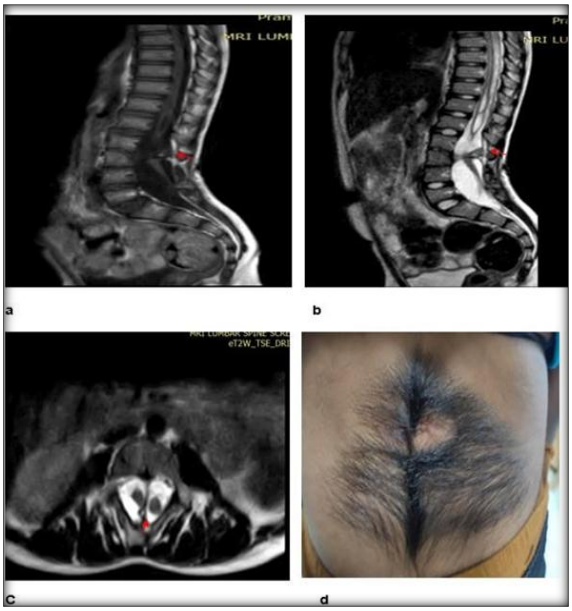

Figure 3: DIASTEMATOMYELIA: A18-Month-old male child presented with hypertrichosis over the lumbar region and sagittalT1(a)andT2(b) andaxialT2 (c) images shows spina bifida with images shows dilated terminal ventricle and tethered cord and clinical photo(d) shows hypertrichosis in lumbar region

Case 3: Diastematomyelia

Table 2: Age distribution of spinal dysraphism

AGE (IN YEARS)	NO.OFCASES (n=30)	PERCENTAGE(%)
<1	13	43.3
1-3	4	13.3
3-10	3	10
11-20	3	10
21-30	5	16.7
31-40	2	6.7
TOTAL	30	100

Table 3: Sex distribution of spinal dysraphism

SEX	NO.OF CASES (n=30)	PERCENTAGE(%)
Male	13	43.3
Female	17	56.7
Total	30	100.0

Table 4: Distribution of cutaneous markers in closed spinal dysraphisms

CUTANEOUSMARKERS	NO. OF. PATIENTS (N=30)	PERCENTAGE (%)
Hypertrichosis	3	37.5
Dorsaldermalsinus	4	50
Portwine stain	1	12.5
Total	8	100

Table 5: Distribution of patients on the basis of level of spine involvement

LEVEL	NO. OF. PATIENTS	PERCENTAGE(%)
CERVICAL	1	3.33
CERVICOTHORACIC	2	6.67
THORACOLUMBAR	4	13.33
LUMBAR	22	73.33
LUMBOSACRAL	1	3.34

Table 6: Distribution of spinal curvature abnormalities

CURVATURE ABNORMALITY	NO. OF. PATIENTS	PERCENTAGE(%)
Present	7	23
Absent	23	77
Total	30	100

Table 7: Distribution of various vertebral anomalies associated with spinal dysraphism

VERTEBRAL ANOMALIES	NO.OFCASES (n=30)	PERCENTAGE(%)
Spinabifida	**25**	**89.5**
Block vertebra	**1**	**3.5**
Hemivertebra	**1**	**3.5**
Butterflyvertebra	**1**	**3.5**
Sacralagenesis	**0**	**0**

Table 8: Overall distribution of spinal abnormalities in patients with congenital spinal dysraphism

SPINALABNORMALITY	NO. OF. CASES	PERCENTAGE (%)
MMC	**16**	**34**
DDS	**3**	**6.25**
DMM	**3**	**6.25**
ID Lipoma	**4**	**8**
VT	**1**	**2**
TC	**2**	**4**
LMM	**1**	**2**
Scoliosis	**7**	**14.50**
Syringohydromyelia	**11**	**23**
TOTAL	**48**	**100**

Table 9: Distribution of spinal dysraphism according to clinico-radiological classification

TYPEOFSPINAL DYSRAPHISM	NO.OF. CASES	PERCENTAGE (%)
OSD	**16**	**54**
CSD	**14**	**46**

Table 10: Distribution of abnormalities in open spinal dysraphism

TYPEOFOSD	NO.OF. CASES	PERCENTAGE (%)
MMC	15	93
MYELOCELE	0	0
HMMC	0	0
LMMC	1	7

Table 11: Distribution of abnormalities in closed spinal dysraphism

SPINAL ABNORMALITIES	NO.OF. CASES	PERCENTAGE(%)
DMM	3	21.25
LMM	1	7
DDS	3	21.25
VT	1	7
ID lipoma	4	29
TC	2	14.50

Table 12: Level of myelomeningocelein our study

LEVEL	NO. OF. CASES	PERCENTAGE (%)
Cervical	**2**	**12.50**
Thoracolumbar	**3**	**18.75**
Lumbar	**11**	**68.75**

Table 13: Anomalies associated with myelomeningocele in our study

ANOMALIES ASSOCIATED	NO.OF. CASES	PERCENTAGE (%)
SHM	**8**	**27**
TC	**7**	**23**
ACM	**7**	**23**
HC	**8**	**27**

Table 14: Distribution of spinal abnormalities associated with tethered cord

ASSOCIATED ANOMALIES	NO.OF.CASES(n=13)	PERCENTAGE(%)
DDS	2	15.38

MMC	8	61.54
DMM	1	7.7
ID Lipoma	2	15.38

Table 15: Distribution of spinal abnormalities associated with syringohydromyelia

SPINALBNORMALITIES	NO.OF. PATIENTS	PERCENTAGE (%)
MMC	9	60
DMM	1	7
TC	5	33

Table 16: Distribution of spinal abnormalities associated with hydrocephalus

SPINAL ABNORMALITIES	NO.OF. PATIENTS(n=9)	PERCENTAGE (%)
MMC	**8**	**89**
DDS	**1**	**11**

DISCUSSION

The spine and spinal cord can develop congenital malformations called spinal neural tube defects. Occurs between the third and fourth weeks of pregnancy as a result of improper neural tube closure. The term spinal dysraphism (SD) refers to the collective set of abnormalities resulting from ectodermal, mesodermal, and neuroectodermal tissue maldevelopment. Some of these illnesses may be found in older children or even adults, even though the most of the mare identified at birth or in the first few months of life. The numerous imaging methods that help with detection and diagnosis include magnetic resonance imaging, spinal ultrasonography, myelography, and CT myelography.

The use of spine ultrasound (SUS) as a first-line screening in new-borns thought to have spinal dysraphism. Spine ultrasound has the same diagnostic sensitivity as MRI, but it also has the benefit of being portable and not requiring sedation or general anaesthesia, unlike MRI. The resolution of an MRI is greatly influenced by the patient's movement, physiological motion from the pulsing of the cerebral spinal fluid (CSF), and blood flow.[14] USG has been demonstrated to be a useful screening technique before ossification of posterior spinal elements; however, infants with abnormal ultrasonography or who have a neurological problem with normal ultrasound still require MR imaging.[15]

MRI has significantly improved the diagnosis of various illnesses and increased the potential of earlier and case-specific treatment due to its multiplanar imaging and tissue characterization capabilities. Understanding the aetiology and neuroradiological picture of these defects requires knowledge of normal embryonic developmental stages. Furthermore, the MRI image of these anomalies is complex and frequently defies memorization of lists of neuroradiological features by rote. The current study involved30 patients in total, representing all age groups, whose final MRI examination diagnosis was spinal dysraphism. On the basis the clinical history, spinal dysraphism was suspected in all 30 instances. A 1.5 Tesla super conducting magnet from a Philips MRI machine was used after a clinical examination and related history-taking.

AGE DISTRIBUTION OF SPINAL DYSRAPHISM

The age of the patients in present study ranged from 4 months to 11 years. Maximum number of patients were in the age group of 1-5 years, accounting for 43.3%. In a study by Kumari MV etal., age of the patients ranged from17daysto13 years.[16]

Most of the children are below 1 year of ageand by Nafees M et al.,age of the patients ranged from 16 days to 37 years and most of them were below 6.4 years.[17] The study done by Mohamed Fathy Dawodh et al., in which 18 are female patients and 14 are male patients.[18]

SEX DISTRIBUTION OF SPINAL DYSRAPHISM

It is thought that Females are more likely than males to experience spinal dysraphism. Most studies seem to support the sex difference. A study done by De Wals P et al. in 1997, found the females account for between 55-70% of neural tube abnormalities.

CUTANEOUS MARKERS

In a study of 14 cases of spinal dysraphism by David Guggisberg et al, the most common lesions associated with spinal dysraphism were lipomas with overlaying port wine stains (n = 5, 35.7%) and deviations of the gluteal fold (n = 6, 42.8%). The two most common spinal malformations were spinal lipoma and tethered spinal cord. In our study, 14 patients with closed spinal dysraphism, or 46.6% of cases, had cutaneous indicators. 4 cases had dorsal dermal sinus, 3 cases had hypertrichosis and one case had a port-wine stain.

Dermal sinus and Hypertrichosis were the most common cutaneous markers noted which accounted for 87.50%. Hypertrichosis was present in 3 out of 3 cases of diastematomyelia [100%]. This is in agreement with the study by Miller et al,[19] in which hypertrichosis was present above the spinal dysraphismin17 patients out of 43 cases of diastematomyelia [39.5%].

LEVEL OF SPINAL DYSRAPHISM

A study conducted by Assaad A et al showed that almost all cases with subcutaneous mass with underlying spinal abnormalities occur in the lumbar

and sacral spine.[20] In a study by Kumar R et al,[100] the lumbosacral region is the commonest site of occurrence of spinal dysraphism followed by the lumbar region.

However, in our present study lumbar spine (n=22,73.33%) is most commonly involved followed by the thoracolumbar spine (n=4,13.33%), cervicothoracic spine [n=2,6.67%], cervical and lumbosacral spine [n=1,3.34%] in decreasing order of frequency. The cervical spine is least commonly involved [n=1, 3.34%].

SCOLIOSIS

SCOLIOSIS

In a study conducted by Prahinski et al,[21] 30% [9 out of 30] of patients with congenital spinal curvature abnormalities had identifiable spinal anomalies at MRI examination. In this study, spinal curvature abnormalities were seen in23%ofcases [n=7] which is in keeping with the study by Prahinski et al.In a study done by Erfani et al,[22] intraspinal abnormalities were found in 41.3% [19 out of 46] of congenital scoliosis at MRI examination which correlates with our study.

VERTEBRAL ABNORMALITIES

In the present study, of all the vertebral anomalies in patients with congenital spinal lesions, spina bifida is the commonest (n=15, 89.50%), followed by hemivertebra (n=1,3.5%) and butterfly vertebra (n=1,3.5%) and block vertebra (n=1,3.5%). None of the cases had sacral agenesis.

OPEN AND CLOSED SPINAL DYSRAPHISM

In the present study, we followed the clinico-radiological classification of 2000 for the categorization of spinal malformations. Accordingly, in the present study, we found 16 cases of open spinal dysraphism and 14 cases of closed spinal dysraphism.

In a study by Tortori Donati et al2 closed spinal dysraphism is more common than open spinal dysraphism 633 v/s 353 [64.2%] cases. In our study of 30 cases, 46% of cases [n=14] are closed dysraphisms whereas 54%[n=16] are the open type. In our study of open spinal dysraphism, meningomyelocele is the commonest one [n=15, 93%] followed by lipomeningomyelocele [n=1,7%].

MENINGOMYELOCELE

In our study meningomyelocele constituted about 34% of the patients. It is shown that meningomyeloceleis the common estopen spinal dysraphism accounting for 98.8% of the cases.[23] In the present study also meningomyelocele was the commonest open spinal dysraphism accounting for 87.5% of the cases.

In our cases of meningomyelocele, the spinal cord showed syringohydromyelia in 8 patients (27%), and tethered cord in 7 patients (23%), which correlates with the study done by Kumar R, Singh SN et al,[24] states that syringohydromyelia is the common association in MMC.Sagittal MRI sections in the cranio-cervical region show tonsillar herniation i.e., Chiari type II malformation in all 7 patients (23%).

DORSAL DERMALSINUS

A dorsal dermal sinus is an epithelium-lined fistula that extends inwards from the skin surface and connects with the CNS and its meningeal coating.[23] In the presentstudy, adorsal dermal sinus was noted in 3patients accounting for 21.25%of all the CSDs. It was associated with low-lying conus and tethered cord. Mid-sagittal MR image showed a thin hypointensestripe in the subcutaneous fat in agreement with Rossi A et al.[23]

DIASTEMATOMYELIA

The incidence of diastematomyelia in our study was21.25% while a study done by Mc Comb JG et al,[25] shows its incidence to be 20-40% of the cases.

In our study, the peak occurrence of diastematomyelia is seen in 1-20 Years age group constituting about 85.7% of cases. In a retrospective study by Y.C.Ganet al,[26] out of 17 children with diastematomyelia operated during 1989–2004, there were nine girls and eight boys. In the present study, female predominance (n=2,67%) of diastematomyelia is noted with male to female ratio of 1:2.

MRI plays an important role in the characterization of Diastematomyelia into split cord malformation I and II based on the type of septum and nature of dural covering. Size, extent and site of re-joining of the hemicords are best depicted in coronal plane. Fibrous septum is best depicted in MRI and it was surgically removed preferably by laminoplasty rather than laminectomy. Thus, MRI was helpful in presurgical planning.

However, in our study spina bifida [n=8, 18.5%] is the most commonly associated abnormality with syringohydromyelia followed by tetheredcord [n=5,33%], diastematomyelia [n=1, 7%].

CHIARIMALFORMATION

Open defects are associated with abnormalities such as hydrocephalus, Arnold-Chiari, syrinx. In our study Arnold-Chiari type 2 constituted 5 (16.7%). In a study by Kumari MV et al,[16], Arnold- Chiari type 2 constituted 6 (15.7%) and in a study by KumarRetal.,Arnold-Chiaritype 2 constituted 62 (45%) patients.[27]

In our study included a total of 7 patients with Chiari malformation out of which 5 were females and 2 were males, with a female-to-male ratio of 2.5:1. This is in agreement with Hadley DM112 who stated that Chiari malformations affect females twice as often as males.

CONCLUSION

MRI plays an important role in the characterization of Diastematomyelia into split cord malformation I and II based on the type of septum and nature of dural covering. Size, extent and site of re-joining of the hemicords are best depicted in coronal plane. Fibrous septum is best depicted in MRI and it was surgically removed preferably by laminoplasty rather than laminectomy. Thus, MRI was helpful in presurgical planning.

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