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Radiological Evaluation Of Spinal Dysraphism Using CTAND MRI

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Abstract

Introduction: Spinal dysraphism is a broad term encompassing a heterogeneous group of congenital spinal anomalies, which results from defective closure of the neural tube during early fetal life. Spinal dysraphism can be classified as Spina Bifida Aperta and Spina Bifida Occulta.

Aim & Objectives: To assess the role of Helical CT and MRI,The identification of various forms of Spinal dysraphism, Characterization of the lesions and associated anomalies.

Methods: The study was conducted in the year 2021,in Department of Radiodiagnosis, Apollo Hospitals, Hyderabad, Telangana, India. The study comprises of 70 patients including 33 males and 37 females, age ranging from birth to 30 years. Clinically the most common cause for referral was swelling in the back predominantly Lumbosacral region. The other symptoms were sensory/motor deficit, bladder/ bowel disturbances, spinal curvature deformities, cutaneous features like dermal dimple, hypertrichosis, silky hair, dermal sinus and capillary hemangioma.

Results: The most common type of spinal dysraphism is open spinal dysraphism accounting for 80% of the total 70 cases. The most common open spinal dysraphism is Myelomeningocele accounting for 94.6% of the total 56 cases followed by Myelocele and Meningocele.

The most common cutaneous manifestation is palpable mass in the back. Neurological manifestations occurred in all cases of open SD while in occult SD 11 of the 14 cases. Lipomyelomeningocele is the most common type of spinal lipoma accounting for 50% of the total cases. Bony and fibrous septum occurred equally in both open and occult SD. In open SD the most common site of occurrence of Diastematomyelia are DL and L regions. In occult SD Diastematomyelia occurred equally in both DL and L regions. Spinar biffila is the most common vertebral anomaly occurring in 97.14% of total cases. Lumbosacral spine is most common site of occurrence in both open and occult spinal dysraphism. Hydromyelia occurred in 31.43%. It was found to be more common in open SD. Hydrocephalus is present in 42.86% of the total cases. Chiari II malformation is associated with open spinal dysraphism in 91.07% of cases. Chiari I malformation is associated with occult spinal dysraphism in 14.29% of cases.

Conclusion: MRI is the imaging modality of choice for evaluation of the soft tissue anomalies of Spinal dysraphism especially spinal cord anomalies. Multiplanar reformatted CT is an excellent imaging modality for characterization of vertebral segmentation defects, spinal curvature anomalies associated with spinal dysraphism. Thus CT and MRI together play an important role in the complete radiological evaluation of spinal dysraphism.

Keywords: MRI sequence comparison, Neural tube defect, Placode, Spina bifida cystica, Spina bifida occulta

Introduction

Spinal dysraphism is the most common Neural tube defect in developing countries like India. The incidence varies from 0.5 to 11 per 1000 live births in different parts of our country, largely affecting the lower socioeconomic strata of the population.[1] The etiology of Neural tube defects is Multifactorial. The interaction of diverse factors related to Genetics, Nutrition and Environment play an important role in the etiopathogenesis. Some of the environmental factors that may contribute to Open Neural Tube Defects are uncontrolled maternal diabetes, and certain prescription medications.[2] It has been proved that deficiency of essential vitamins especially Folic acid during pregnancy results in higher incidence of Neural tube defects which led to prophylactic supplementation of folic acid in the antenatal period. [3]Spinal dysraphism is a broad term encompassing a heterogeneous group of congenital spinal anomalies, which results from defective closure of the neural tube during early fetal life. Spinal dysraphism can be classified as Spina Bifida Aperta and Spina Bifida Occulta.Spina bifida aperta is most common type of spinal dysraphism representing a serious congenital anomaly with severe Neurologic, Musculoskeletal, Genitourinary, and Bowel anomalies.[4] It encompasses three forms namely Myelomeningocele, Myelocele and rarely Meningocele. Females show a higher incidence than Males and most of them present at birth and are immediately taken for surgical repair and hence are rarely imaged in unoperated cases.[5]Spina bifida occulta is characterized by minor Neurological manifestations and presents at a later age. Most distinct clinical findings are cutaneous stigmata like Dermal dimple, Hemangioma, Cutis aplasia, Dermal sinus, or Hairy patch, Rudimentary tail (caudal appendage). Segmentation anomalies of spine are a common feature of spinal dysraphism and along with muscle imbalances due to motor deficits result in Spinal curvature anomalies like Scoliosis, Kyphosis and Lordosis.[6] Scoliosis is the most common type of spinal curvature anomaly. Associated anomalies include Chiari malformations, Hydromyelia and Hydrocephalus. Imaging plays a pivotal role in the diagnosis and management of spinal dysraphism.[7]

Methods: the study was conducted in the year 2021,in Department of Radiodiagnosis, Apollo Hospitals, Hyderabad, Telangana, India. The study comprises of 70 patients including 33 males and 37 females, age ranging from birth to 30 years. Clinically the most common cause for referral was swelling in the back predominantly Lumbosacral region. The other symptoms were sensory/motor deficit, bladder/ bowel disturbances, spinal curvature deformities, cutaneous features like dermal dimple, hypertrichosis, silky hair, dermal sinus and capillary hemangioma. Inclusion Criteria:All cases of open spinal dysraphism.Cases presenting with lumbosacral swelling.Cases presenting with cutaneous stigmata like Dermal dimple, tuft of hair, Nevi, dermal sinus ect.Cases presenting with congenital scoliosis/ kyphoscoliosis/ lordosis ect. Cases presenting with bladder/bowel incontinence since childhood. Exclusion Criteria:Treated cases.Spinal tumors.

Results

Table 1 Open Spinal Dysraphism

S. NO	Type	Number of cases	Percentage	
			(Out of 70)	(Out of 56)
1	Myelomeningocele	53	75.71%	94.64%
2	Myelocele	2	2.86%	3.57%
3	Meningocele	1	1.43%	1.79%
	Total	56	80%	100%

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Table :1 shows The most common type of spinal dysraphism is open spinal dysraphism accounting for 80% of the total 70 cases. The most common open spinal dysraphism isMyelomeningocele accounting for 94.6% of the total 56 cases followed by Myelocele and Meningocele.

Table 2 Occult Spinal Dysraphism

S.No	*Type*	*Number of cases*	*Percentage*	
			(Out of 70)	**(Out of 14)**
1	Spinal lipomas	6	8.57%	42.86%
2	Diastematomyelia	4	5.71%	28.57%
3	Dorsal dermal sinus	1	1.43%	7.14%
4	Tight filum terminale	1	1.43%	7.14%
5	Anterior sacral meningocele	1	1.43%	7.14%
6	Sacral agenesis	1	1.43%	7.14%
	Total	14	20%	100%

Table :2 Occult spinal dysraphism accounted for 20% of the total 70 cases. The most common type of occult spinal dysraphism is Spinal lipoma accounting for 42.86% of the total 14 cases.

Table . 3:Cutaneous Manifestations Of Occult Spinal Dysraphism

Cutaneous Signs	*Dermal dimple*	*Hyper-trichosis*	*Silky hair*	*Palpable mass*	*Dermal sinus*	*Capillary hemangioma*	*Total*
No.of cases	2	2	1	7	1	1	14
%	14.29	14.29	7.14	50	7.14	7.14	100

Table :3 The most common cutaneous manifestation is palpable mass in the back

Table 4:Neurological Manifestations Of Spinal Dysraphism

Type	*Motor and Sensory deficit*	*Bowel and Bladder incontinence*
Open spinal dysraphism	56	56
Occult spinal dysraphism	6	5

Table :4 Neurological manifestations occurred in all cases of open SD while in occult SD 11 of the 14 cases showed neurological manifestations.

Table 5:Spinal Lipomas

Type	*Number of cases*	*Percentage*
Lipomyelocele	1	16.67%
Lipomyelomeningocele	3	50.00%
Dural lipomas	1	16.67%

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Filar lipomas	1	16.67%
Total	6	100%

Table :5 shows Lipomyelomeningocele is the most common type of spinal lipoma accounting for 50% of the total cases.

Table :6 Diastematomyelia

Type	*Fibrous septum*	*Bony septum*	*Total*	*%*
Diastematomyelia in occult SD	2	2	4	28.57
Diastematomyelia in open SD	5	5	10	71.43
Total	7	7	14	100

Table :6 shows Bony and fibrous septum occurred equally in both open and occult SD.

Table 7: Diastematomyelia: Sites Of Involvement In The Spine

Type	*Cervical*	*Dorsal*	*Dorsolumbar*	*Lumbar*	*Lumbosacral*	*Total*	*%*
Open SD	0	1	4	3	2	10	71.43
Occult SD	0	0	2	2	0	4	28.57
Total	0	1	6	5	2	14	100

In open SD the most common site of occurrence of Diastematomyelia are DL and L regions. In occult SD Diastematomyelia occurred equally in both DL and L regions.

Table 8 Tethering Of Cord

Type	*Tethering*	*No tethering*	*Total*
Spinal lipomas	4	2	6
Diastematomyelia	1	3	4
Open SD	4	52	56
Dorsal dermal sinus	0	1	1
Tight filum terminale	1	0	1
Anterior sacral meningocele	0	1	1
Sacral agenesis	0	1	1
TOTAL	10	60	70
%	14.29	85.71	100

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Table :8Tethering of cord occurred in 14.29% of the total cases.

Table 9:Vertebral Anomalies

	Hemivertebra	***Butterfly vertebra***	***Block vertebra***	***Spina bifida***	***Others***
Open SD	21	23	10	56	2
Spinal Lipomas	2	3	1	6	0
Dorsal dermal sinus	0	1	0	1	0
Diastematomyelia	2	0	1	3	0
Tight Filum Terminale	0	1	0	0	0
Anterior Sacral Meningocele	1	0	0	1	0
Sacral Agenesis	0	1	0	1	0
TOTAL	26	29	12	68	2
%	37.14	41.43	17.14	97.14	2.86

Table :9 Spinar biffila is the most common vertebral anomaly occurring in 97.14% of total cases.

Table 10:Spina Bifida Distribution In Spine

Types	***Spina bifida Cases***	***Distribution in spine***								***Total***	
		Cervical		***Dorsal***		***Lumbar***		***Lumbosacral***			
		No	***%***	**No**	***%***	**No**	***%***	**No**	***%***	**No**	***%***
Open SD	56	4	7.14	12	21.43	18	32.14	22	39.29	56	100
Occult SD	12	1	8.33	2	16.67	4	33.33	5	41.67	12	100
Total	68	5	7.35	14	20.59	22	32.35	27	39.71	68	100

TAble :10 Spina bifida occurs in 97.14% of the total cases. Lumbosacral spine is most common site of occurrence in both open and occult spinal dysraphism.

Table 11: Distribution Of Spinal Dysraphism In Spine

Types	***No***	***Distribution in spine***								***Total***	
		Cervical		***Dorsal***		***Lumbar***		***Lumbosacral***		**No.**	**%**
Open SD	56	4	7.14%	12	21.43%	18	32.14%	22	39.29%	56	100
Occult SD	14	1	7.14%	2	14.29%	4	28.57%	7	50%	14	100

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Total	70	5	7.14%	14	20%	22	31.43%	29	41.43%	70	100

Table :11 The most common site of occurrence of spinal dysraphism is LS spine.

Table 12 Spinal Curvature

Spinal curvature	***Scoliosis***			***Kyphosis***		***Lordosis***	***Total***
Region	***Cervical***	***Dorsal***	***Lumbar***	***Dorsal***	***Lumbar***	***Lumbar***	
OPEN SD	1	6	5	4	2	4	22
OCCULTSD	1	5	4	5	3	3	21
TOTAL	2	11	9	9	5	7	43
%	2.86	15.71	12.86	12.86	7.14	10	61.43

Table :12 The most common spinal curvature anomaly is scoliosis.

Table 13 Hydromyelia Association

Type	***Hydromyelia***		***Total***
	Present	***Absent***	
Open SD	15	41	56
Occult SD	7	7	14
Total	22	48	70
%	31.43	68.57	100

Table :13 Hydromyelia occurred in 31.43%. It was found to be more common in open SD.

Table 14 Hydrocephalus In Spinal Dysraphism

Type	***Hydrocephalus***		
	Present	***Absent***	***Total***
OPEN SD	25	31	56
OCCULT SD	5	9	14
TOTAL	30	40	70
%	42.86	57.14	100

Table :14 Hydrocephalus is present in 42.86% of the total cases. Hydrocephalus is more common in Open spinal dysraphism.

Table 15 Chiari Association

Type	***Chiari II***	***Chiari I***	***%***

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OPEN SD	51	0	91.07
OCCULT SD	0	2	14.29

Table :15 Chiari II malformation is associated with open spinal dysraphism in 91.07% of cases. Chiari I malformation is associated with occult spinal dysraphism in 14.29% of cases.

Table 16 Comparison Of Ct And Mri In Spinal Dysraphism

S. No	***Characteristics***	***CT***	***MRI***
1	OPEN SPINAL DYSRAPHISM		
	Meningomyelocele	++	++++
	Myelocele	++	++++
	Meningocele	++	++++
2	OCCULT SPINAL DYSRAPHISM		
	Spinal lipomas	++	++++
	Diastematomyelia	++	++++
	Dorsal dermal sinus	++	++++
	Tight filum terminale	+	++++
	Anterior sacral meningocele	++	++++
	Sacral agenesis	++	++++
3	VERTEBRAL ANOMALIES	++++	++
4	DISTRIBUTION IN SPINE	+++	+++
5	SPINAL CURVATURE	+++	+++
6	TETHERING	+	++++
7	CHIARI ASSOCIATION	++	++++
8	HYDROMYELIA	++	++++
9	HYDROCEPHALUS	++	++++

Discussion

Clinical features of spinal anomaly are hair tuft, hypertrichosis, Port-wine stain, visible swelling, haemangioma, dimpling, sinus tract in back, visibly short back, abnormal spinal curvature, deviation of gluteal furrow, human or faun tail, asymmetry of legs/feet, subcutaneous lipoma, sensory loss in limb/limbs, decreased movements in limb/ limbs, dribbling of urine, recurrent urinary tract infection .[8]Combination of two or more cutaneous midline lesions in back is the strongest marker of occult or closed spinal dysraphism .Main objective of study was to evaluate clinically suspected spinal dysraphism by MRI scan; which were accurately diagnosed with their type and extent. MRI findings are well correlated on surgery (20 patients), radiographs/CT (15 patients with 5 having CT) and detailed clinical examination (15 patient). Meningomyelocele is seen in Arnold-Chiari malformation in all 17 cases; which constitutes 100% open spinal dysraphism and 34 % of all patients. Meningomyelocele was reported to constitute 98.8 % of open spinal dysraphism in west.[9] Another objective of study was to explore possibility of finding best sequence pair by comparing different sequence pairs to detect neural tissue and fatty tissue in different spinal anomalies. Similar study of MRI

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sequence comparisons for spinal dysraphism was not found in different internet searches. The only sequence comparison study in MRI of spine was done .MR Three-dimensional gradient steady state sequence- Constructive Interference with Steady State (CISS) with its reformations in multiple planes and at various angles is mainly used to assess intracranial ventricles and cisternal spaces. Predisposing factors of spinal dysraphism are nutritional deficiency of folic acid and mutations in genes involving folic acid metabolism .[10] Recurrence risk after birth of an infant with isolated spina bifida is 3%-5% with recurrence in sibling may occur as spina bifida or other spinal anomaly. Chromosome abnormalities, single gene mutation, maternal diabetes mellitus, prenatal exposure to certain anti-convulsants given to mother are other causative factors; which have different recurrence risks .[11] Few Northern provinces in China have highest prevalence rates of 1 in 200 pregnancies for spina dysraphism; while european countries have lower prevalence rate of < 1 in 2,000 pregnancies.[12] Overall estimated incidence of spinal dysraphism is 1-3/1000 live births In India, as in world over; steady decrease is observed in incidence of spinal dysraphism in the decade of 2001-2010 due to folic acid fortification in pregnancy, antenatal detection by ultrasound and elevated maternal Alpha Feto Protein (AFP) (found in upto 80% cases of spina bifida) as well as medical termination of pregnancy. Limitations are single institution study and not having any previous baseline study of similar type of sequence pair comparisons to determine sample size and robust study design.[13] This study is done to explore the possibility of better choices of sequences in protocol, if study need to be done in short time. This kind of study should be done at multiple institutions to determine best sequence or pair of sequences for accurate tissue characterisation; especially for neural tissue identification, which affect the prognosis of spinal dysraphism.[14,15]

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