ORIGINAL RESEARCH PAPER
Volume - 9 | Issue - 6 | June - 2020 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

IMAGING SPECTRUM IN CLOSED SPINAL DYSRAPHISM: STUDY FROM A TERTIARY CARE CENTER

Radiology

Anna Premchandar Abhijith Resident, Department of Radiology and Imageology, NIMS, Hyderabad

Abhishek J. Arora* Associate Professor, Department of Radiology and Imageology, NIMS, Hyderabad. *Corresponding Author

Jyotsna Yarlagadda Professor, Department of Radiology and Imageology, NIMS, Hyderabad

ABSTRACT

Closed spinal dysraphism is defective fusion of the midline neural, bony and mesenchymal structures with a skin covering. It is one of the most common congenital disorders associated with morbidity and mortality with estimated incidence of approximately 0.05 to 0.25 per 1000 births. In our study, we retrospectively analyzed 30 patients with closed spinal dysraphism and categorized them according to their characteristic features. Closed spinal dysraphism was found more commonly in females than males, and also in our study diastometamyelia was the most common abnormality and tethered cord syndrome, the least common among them.

KEYWORDS

INTRODUCTION

Spinal dysraphism is defined as incomplete fusion of midline mesenchymal, bony and neural structures. It is one of the most common congenital disorders associated with morbidity and mortality with estimated incidence of approximately 0.05 to 0.25 per 1000 births.(1) It is broadly classified into Closed and Open type. Open Dysraphism is where the neural tissue is exposed to the external environment without skin covering it, where as in closed type the neural elements have a skin covering. Closed types remain asymptomatic at birth and are suspected in the presence of high-risk cutaneous markers or when patients present with neurological deficits later in life.

Brief note on Embryology: Spinal cord development can be summarized in three basic embryologic stages – gastrulation (2–3 weeks), primary neurulation (3-4 weeks) and secondary neurulation (5–6 weeks). (2,3)

Gastrulation is the process of conversion of bilaminar disc into a trilaminar disc initiated by primitive streak. Primitive node, a depression at the cranial end of streak, contains cells that are important for organizing the embryonic axes. Epiblast cells migrate towards and through the streak and node, detach and form endoderm and mesoderm. Nonmigrating cells of epiblast constitute the ectoderm of which some cells migrate cranially in the midline to form the prechordal plate and notochord, which initiate the process of neurulation by inducing the formation of the neural plate (4).

Primary Neurulation: Lateral borders of neural plate elevate into neural folds which later fuse in the midline to form the neural tube. The open regions of the neural tube called the anterior (cranial) and posterior (caudal) neuropores close by the zippering process. This neurulation process is called primary neurulation which is responsible for establishing brain and spinal cord regions down to the sacral levels on neural tube closure, overlying non-neural epidermal cells form the ectodermal layer of the skin. The neural tube separates from the overlying ectoderm, a process called dysjunction. (4)

Secondary Neurulation: The cord caudal to S2 level is formed by this process. Totipotent mesodermal cells called caudal cell mass or tail bud coalesce to form neural tube which then epithelialize, reorganize around a lumen and finally become continuous with the cranial part of the tube initially formed by primary neurulation (4). Part of the caudal cell mass undergoes both regression and differentiation (a process called retrogressive differentiation) to form filum terminale, terminal ventricle, tip of conus medullaris, and most of the sacrum, coccyx and coccygeal medullary vestige. (5)

Imaging modalities

Plain Radiographs (anteroposterior and lateral views) are mandatory for evaluation of the vertebral column. Radiographs are used as preliminary screening examination that can help to guide the further imaging work up. The findings seen on plain radiographs include focal spina bifida, widened spinal canal, scoliosis, segmentation anomalies like block vertebrae, butterfly vertebrae and other congenital abnormalities. (2,6)

Ultrasonography is primarily useful in the antenatal diagnosis of spinal dysraphism, and is also of some use in infant and neonate. Prenatal sonography can detect the open widened neural arch with flared laminae (7)

Magnetic Resonance Imaging (MRI) provides an accurate and noninvasive method of evaluating spinal dysraphism making it the modality of choice. The excellent contrast resolution, wide field of view and multiplanar images allow evaluation of the entire spinal cord, contents of the back mass. T1-weighted (T1W) images (sagittal, axial and coronal plane) demonstrate the malformation in the majority of the cases. T2W images are helpful in the demonstration of syrinx and associated pathologies like dermoid and epidermoid cyst. (6, 8)

MATERIALS AND METHODS

Our study consisted of retrospective observational analysis of all the patients presenting with closed spinal dysraphism in the Department of Radiology of our institute. 30 patients were evaluated from over 3 years period from January 2017 to December 2019. MRI imaging was done in 1.5T MRI and 3T MRI. Coronal, Sagittal and Axial sections of T1 and T2 weighted imaging sequences was done in each case. The features were analysed in each case and characterization was done.

OBSERVATIONS AND ANALYSIS

Retrospective analysis of 30 patients, ranging in age from 10 days to 52 years (mean age of 19 years) with diagnosis of closed spinal dysraphism was done. Out of 30 patients 21(70%) were females and 9(30%) were males. Closed spinal dysraphism without subcutaneous mass was seen in 21 patients (70%) while with subcutaneous mass was seen in 9 (30%) of them. Demographic profile of the different types of closed spinal dysraphism is given below in the table 1 and figures1 and figure 2 below.

Table1: Percentage of the cases in each type of closed spinal dysraphism

TYPE OF DYSRAPHISM	NO. OF CASES	% OF CASES
Lipomyelomeningocele	2	6.66
Lipomyelocele	4	13.3

Meningocele	3	10.0
Filar/intradural lipoma	4	13.3
Diastematomyelia	9	30.0
Caudal regression	4	13.3
Tethered cord syndrome	1	3.33
Dorsal dermal Sinus	3	10.0

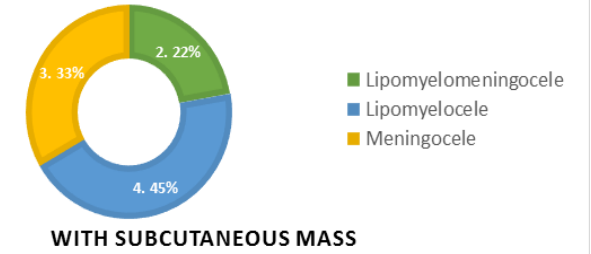


Figure 1: Distribution of the cases with subcutaneous mass

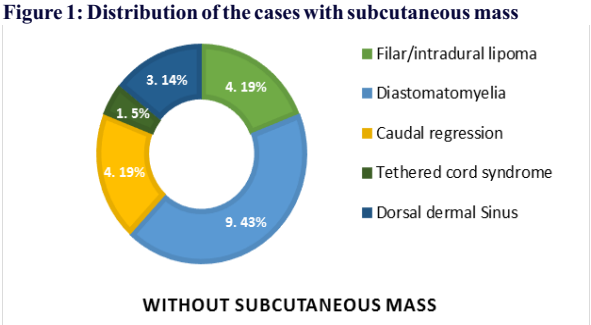


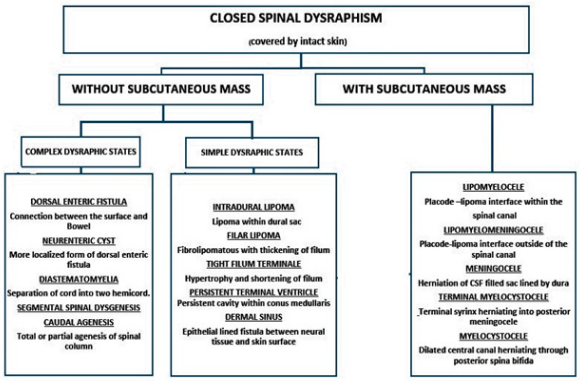
Figure 2: Distribution of the cases without subcutaneous mass

DISCUSSION

Spinal dysraphism is defined as incomplete fusion of midline mesenchymal, bony and neural structures. It is one of the most common congenital disorders associated with morbidity and mortality with estimated incidence of approximately 0.05 to 0.25 per 1000 births.

In our study closed spinal dysraphism was seen commonly in females than males. Most of the patients had no subcutaneous mass. The patients were categorized based on the classification proposed by Tortori-Donati P et al (2), as given in flow chart below.

Lipomas with dural defect – Lipomyelocele and Lipomyelomeningocele: Lipomyelocele occurs due to defective primary neurulation due to the premature disjunction of the ectodermal and neuroectoderm resulting in allowing the mesenchyme to enter the neural tube. The mesenchyme later forms the lipomatous tissue. Clinically, they present with a subcutaneous fatty mass lesion in the intergluteal region. T1W images show high intensity fat on the dorsal aspect of the neural placode which is continuous with the lipomatous tissue. Lipomyelocele can be identified with the neural placode-lipoma interface in the spinal canal. Lipomyelomeningocele can be identified with neural placode - lipoma interface outside the spinal canal. (Figure 3)(6)



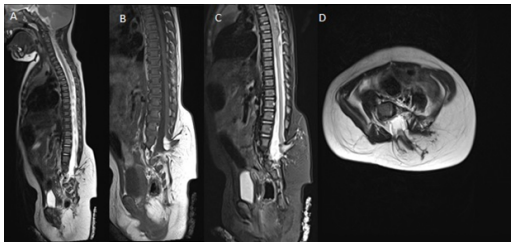


Figure 3 (A to D): Lipomeningomyelocele in a 1-month old female. Sagittal(A&B) T1W and Sagittal(C)T2W fat suppressed and axial(D) T1W. show excessive fat posterior to lower lumbar spine with defect in the postero spinal canal at L5-S1,S1-S2. There is herniation of meninges, CSF Fat and nerve roots through it into subcutaneous plane with overlying large adipose tissue component extending from L3 to S3 vertebral levels. The lipoma and neural placode interface seen outside the spinal canal.

Meningocele: Meningocele is the herniation of CSF filled sac lined by dura and arachnoid mater. The spinal cord is not seen within the Meningocele, however, may be seen tethered to the neck of the meningocele. Posterior meningocele is due to herniation through the posterior spina bifida. It is commonly seen in the lumbosacral region and anterior meningocele is seen in the presacral location (9)

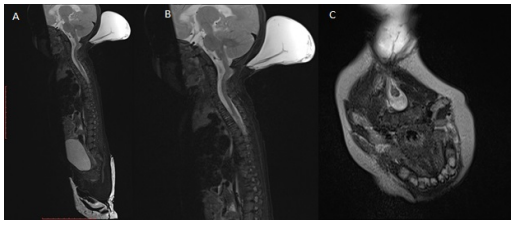


Fig 4.(A to C). Cervical Myelomeningocele in a 6 months old female. Sagittal (A&B) and Axial(C) T2W images showing defect in the posterior elements of C1 and C2 with protrusion of CSF and meninges through the defect with formation of large cyst . It is covered by a thin layer of skin and neural components are seen

Intradural Lipoma: It is the lipoma seen inside the dural sac. The differentiating factor from lipomyelomeningocele is the intact dura. They are commonly seen in the lumbosacral region associated with tethered cord syndrome. On MRI lipomas follow the signal intensity of subcutaneous fat on all sequences.(8)

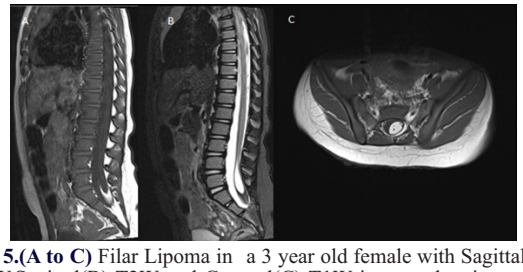


Fig 5.(A to C) Filar Lipoma in a 3 year old female with Sagittal(A) T1W,Sagittal(B) T2W and Coronal(C) T1W images showing a low lying cord with hyperintense lesion on T1W at the filum terminale.

Filar Lipoma: Filar lipoma is the fibrolipomatous thickening of the filum terminale (Figure 5). This occurs due to the defect in the secondary neurulation. On MRI it appears as a hyperintense on T1 and T2 weighted images with a thickened filum terminale. This condition may be associated with the low-lying cord or cord tethered to the duramater. (10)

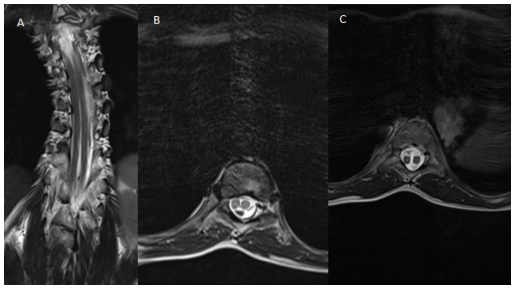


Fig 6. (A to D) . Diastometamyelia in a 11-year-old male with coronal

(A) and axial(B&C) showing scoliosis of the mid thoracic vertebrae with convexity towards left and splitting of spinal cord noted from T7 to L1 level with single dural sac.

Diastematomyelia: It is also known as the split cord malformation. It occurs due to defective midline integration resulting in formation of separate neural plate with intervening primitive streak tissue. There are two types of diastematomyelia. In Type I the intervening primitive streak develops into bone and cartilage, resulting in two hemi cords in different dural sacs separated by an osseocartilaginous septum (Figure 6). In type II diastematomyelia the primitive streak is reabsorbed and the hemi cords lying within same dural sacs. A high lying hairy tuft over a child back is commonly seen in these patients. (11)

Caudal Agenesis: It is characterized by partial or total agenesis od spinal column and it is divided into two types in type I both notochord and caudal cell mass resulting in high and abnormal termination of conus medullaris. In type II type only caudal cell mass is affected with normal primary neurulation and hence only caudal part of the conus medullaris is absent. Vertebral dysgenesis is less severe in these cases and these patients present with tethered cord syndrome. (Figure 7) (12)

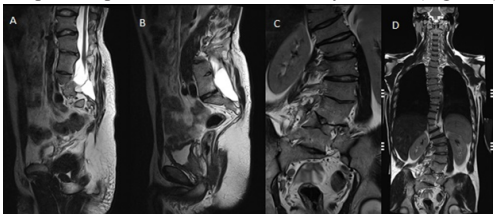


Fig 7. (A to D) Sacral agenesis in a 22-year-old male with Sagittal(A&B) T2W and Coronal(C&D) T2W images showing scoliosis of lumbar vertebra with convexity towards left and thoracic vertebrae with convexity towards right. Block vertebrae at D2-D3 and D10-D11 level. Lower sacral and coccygeal vertebrae are not visualized. Low lying cord tethering noted

Dermal Sinus: Dermal sinus is the fistulous communication between central nervous system or the meningeal covering and skin. It occurs due to the focal incomplete disjunction of ectoderm and cutaneous ectoderm. (Figure 8) Patients presents with a midline dimple with associated cutaneous stigmata like hairy nevus, hemangioma or hyperpigmentation. The tract ascends and opens into the spinal canal. CNS infection is commonly seen in these patients due the fistulous communication and an early surgical repair is required (13).

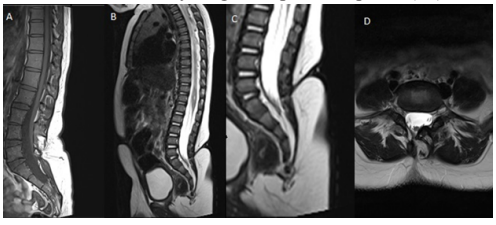


Fig 8. (A to D) Dorsal dermal sinus in a 5 months old female with sagittal(A) T1W, sagittal(B&C) T2W images showing evidence of sinus tract at lumbo sacral region extending anteriorly and inferiorly through the skin, subcutaneous and interspinous plane up to dural space.

Mehta DV et al showed, that out of 50 total patients, various closed spinal Dysraphism abnormalities seen in their study were as follows: diastematomyelia 9(18%), dorsal dermal sinus seen in 4(8%), Intraspinal Lipoma 3(6%), Lipomeningocele 1(2%), Caudal Regression Syndrome 1(2%). Similar to their study, diastematomyelia was the commonest (30% in our study) type of dysraphism in our study too. Caudal regression was the least common abnormality in their study, whereas isolated tethered cord syndrome was the least common type in our study (14). Kumar R et.al studied 155 patients prospectively (143) and retrospectively (12). The male to female ratio was 1.5:1 in their study, however, the ratio was more skewed towards female in our study (2.3:1). Mean age at presentation was 5.7 years. Out of 155 cases of spinal dysraphism, 119 had open spinal bifida and 36 had occult spina bifida [split cord malformation (SCM) without overt MMC sac (pure SCM) in 29 (19%) and midline dermal sinus in 7 (4.5%)]. Lipomeningomyelocele constituted 73 of the 113 cases of MMC (65%) but our study showed only 6.66%. Diastematomyelia constituted most of the cases of spinal dysraphism in our study. Being a tertiary care center and a thriving scoliosis clinic in orthopedic department of our institute could be one of the reasons of getting high numbers of the same. Kumar et al. also had twenty cases of MMC (18%), which had associated SCM (complex spina bifida). The total number of cases with SCM was 49 (32%) which was nearly the same percentage of cases (30%) in our study. (15)

LIMITATIONS

As the cases were taken from a single tertiary care center the number of cases and different types of cases were less. The study has a less sample size compared to the studies that have been done which is one of the limitations of the study. The study is a complete retrospective study and therefore clinical and ultrasound correlation was not possible in any of the cases.

CONCLUSION

Spinal dysraphism is group of developmental abnormalities occurring due to defective closure of midline mesenchymal, bony and neural tissue. Spinal dysraphism includes numerous entities that vary in complexity and imaging appearance. Clinical, embryological and imaging correlation provides an organized approach in their diagnosis. MRI is the single imaging modality in majority of the cases, helps in classifying them and planning the medical and surgical management of the patient.

REFERENCES

1. V Warder DE. Tethered cord syndrome and occult spinal dysraphism. Neurosurg Focus 2001; 15:10:e1.
2. Tortori Donati P, Rossi A, Cama A. Spinal dysraphism: A review of neuroradiological features with embryological correlations and proposal for a new classification. Neuroradiology 2000; 42:471 91
3. Rufener S, Ibrahim M, Parmar HA. Imaging of Congenital Spine and Spinal Cord Malformations. Neuroimaging Clin N Am 2011; 21:659 76.
4. Sadler TW. Embryology of neural tube development. Am J Med Genet Part C Semin Med Genet 2005;135:2 8.
5. Melvin EC, George TM, Worley G, Franklin A, Mackey J, Viles K, et al. Genetic studies in neural tube defects. NTD Collaborative Group. Pediatric Neurosurgery 2000;32:1-9.
6. Rossi A, Biancheri R, Cama A, Piatelli G, Ravegnani M, Tortori-Donati P. Imaging in spine and spinal cord malformations. Eur J Radiol. 2004;50:177–200
7. Silverman FN, et al. Spinal dysraphism. In Silverman FN, Kuhn JP (Eds): Caffey's Pediatric X-ray Diagnosis (9th edn). Lippincott Williams and Wilkins 1993; 1:312-24.
8. Tortori-Donati P, Rossi A, Biancheri R, Cama A. Magnetic resonance imaging of spinal dysraphism. Top Magnetic Resonance Imaging. 2001;12:375–409
9. Lee KS, Gower DJ, McWhorter JM, Albertson DA. The role of MR imaging in the diagnosis and treatment of anterior sacral meningocele. Report of two cases. J Neurosurg. 1988;69:628–631.
10. Uchino A, Mori T, Ohno M. Thickened fatty filum terminale: MR imaging. Neuroradiology. 1991;33:331–333
11. Pang D, Dias MS, Ahab-Barmada M. Split cord malformation: Part I: A unified theory of embryogenesis for double spinal cord malformations. Neurosurgery. 1992;31:451–480.
12. Kanev PM, Park TS. Dermoids and dermal sinus tracts of the spine. Neurosurg Clin N Am. 1995;6:359–366
13. Estin D, Cohen AR. Caudal agenesis and associated caudal spinal cord malformations. Neurosurg Clin N Am. 1995;6:377–391.
14. Mehta DV. Magnetic Resonance Imaging in Paediatric Spinal Dysraphism with Comparative Usefulness of Various Magnetic Resonance Sequences. Journal of clinical and diagnostic research: JCDR. 2017; 11 (8): TC17-TC22.
15. Kumar R, Singh SN. Spinal dysraphism: trends in northern India. Pediatric neurosurgery. 2003;38 (3): 133.