Mr Imaging in Spinal Dysrphism: A Pictorial Review.

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ABSTRACT:

Background: Spinal dysraphism refers to the entire range of spinal column and neural axis anomalies. The objective was to evaluate the role of magnetic resonance imaging (MRI) in characterizing the congenital and developmental disorders of spine.

Methods: MRI images depicting the spinal dysraphism, stored in InstaRAD system in Department of Radiodiagnosis, SSIMS &RC, Davangere during the period of 2018 January to 2018 July were retrospectively reviewed and data were retrieved.

Conclusion: Imaging of spinal dysraphism may appear complicated as it is a group of diverse conditions which can have variable imaging appearance. A systematic approach and correlation with clinical, neuro-radiological and developmental data imaging correlation provides an organized approach in their accurate diagnosis.

Keywords: Spinal dysraphism, Neural tube defects (NTD), Congenital spinal disorders, MRI spinal dysraphism

I. Introduction

Spinal dysraphism is an umbrella term used to describe a collection of congenital abnormalities of the dorsum of embryo that include defects in the vertebrae and underlying spine or nerve roots (1). MR imaging is the modality of choice in children with clinically and/or radiographically suspected spinal dysraphism, which offers a noninvasive technique for evaluating the spine (2). The advantages of MRI over CT includes providing a definitive diagnosis without the hazards of ionizing radiation or intrathecal injection of contrast media. MDCT is helpful in detecting vertebral anomalies and spinal bifida better than MRI, however the underlying complex spinal cord abnormalities will be easily missed(3). This pictorial review aims to familiarize the radiologists and medial colleagues with the spectrum of spinal dysraphism.

II. Aims And Objectives

To evaluate the role of MRI in evaluation of spinal dysraphism.
To familiarize the radiologist and clinical colleagues with spectrum of spinal dysraphism.

III. Materials And Methods

MRI images depicting the spinal dysraphism, stored in InstaRAD system in Department of Radiodiagnosis, SSIMS &RC, Davangere during the period of 2018 January to 2018 July were retrospectively reviewed and data were retrieved. All MR imaging examinations was performed on a 1.5-T magnet MR system(GE SIGNA). The study included spinal dysraphism detected on fetal MRI to occult spinal dysraphism detected incidentally in adult life. The study focuses on spinal cord

IV. Discussion

Spinal Cord Development

A knowledge of normal development of the spine and spinal cord is an essential to understand the complex entity.

Spinal development can be summarized into three basic embryologic stages [1, 2].

1. Gastrulation: occurs during the second or third week of embryonic life. It is the conversion of the embryonic disk from a bilaminar to a trilaminar disk composed of ectoderm, mesoderm, and endoderm.
2. Primary neurulation: Occur during third and fourth week of embryonic life. Notochord and overlying ectoderm interact to form the neural plate. The neural plate bends and folds to form the neural tube, which then closes bidirectionally (Fig. 1).

3. Secondary neurulation: Occurs during the fifth or sixth weeks of embryonic life. Secondary neural tube is formed by the caudal cell mass which is formed by the epithelialization of mesenchymal cells. The secondary neural tube is initially solid and subsequently undergoes cavitation, eventually forming the tip of the conus medullaris and filum terminale by a process called retrogressive differentiation.
Spinal dysraphism:
Spinal dysraphism is a broad term given to a group of anomalies where there are malformations in the dorsum of the embryo. Neural tube defects come under this group as well.

Pathology:
There is often abnormal fusion of the midline embryonic neural tube leading to abnormal development of the vertebral and mesenchymal structures.

Subtypes:
Spinal dysraphism can be broadly divided into two different pathological entities:\n
1) **Open spinal dysraphisms**
   - Myelocle
   - Myelomeningocele
   - Hemimyelomeningocele
   - Hemimyelocele

2) **Closed spinal dysraphisms**
   - With subcutaneous mass
     - Lipomyelomeningocele
     - Lipomyelocele
     - Terminal myelocystocele
     - Meningocele
     - Myelocystocele
   - Without subcutaneous mass
     - Intradural lipoma
     - Filarlipoma
     - Tight filum terminale
     - Persistent terminal ventricle
Myeloceles are rare.

- Dermal sinus

**Complex dysraphic states**

- Dorsal enteric fistula
- Neuroenteric cyst
- Diastematomyelia
- Caudal agenesis
- Segmental spinal dysgenesis

**Open Spinal Dysraphism:**

Myelomeningocele and myelocele:

These are due to defective primary neurulation with resultant exposure of the neural placode through a midline skin defect on the back. Myelomeningocele account for more than 98% of open spinal dysraphism. In myelocele, the spinal cord is exposed so that nerve tissue lies exposed on the surface of the back without even a covering of skin or of the meninges, the membranous tissue surrounding the brain and spinal cord. Open spinal dysraphisms are often diagnosed clinically, so imaging is not always performed.

When imaging is performed, the main differentiating feature between a myelomeningocele and myelocele is the position of the neural placode relative to the skin surface. Myelocceles are rare. The neural placode protrudes above the skin surface with a myelomeningocele (Figure 1 and figure 2) and is flush with the skin surface with a myelocele (Figure 4 and 6).

Hemimyelomeningocele and hemimyelocele—Hemimyelomeningoceles and hemimyeloceles can also occur but are extremely rare. These conditions occur when a myelomeningocele or myelocele is associated with diastematomyelia (cord splitting) and one hemicord fails to neurulate. The child shows marked asymmetry of innervation of the lower limbs and where one limb is fully innervated there is a normally functioning lower urinary tract.

**Closed spinal dysraphisms**

- Lipomyelomeningocele: These lesions are formed mainly due to a defect in primary neurulation in which mesenchymal tissue enters into neural placode and forms lipomatous tissue. (Figure 17)
- Lipomyelocele: is one of the most common closed spinal dysraphism. It is seen in thoraco-lumbar region and usually presents as a fatty subcutaneous mass. It is twice as common as lipomyelomeningocele. As the spinal canal grows, there is a distortion of nerve roots with growth thereby leading to neurologic deficits, highlighting the importance of early diagnosis. This is a premature separation of surface ectoderm before formation of proper neural tube with the ingress of mesoderm (which forms fatty elements). The mesoderm prevents proper neurulation. (Figure 21)
- Terminal Myelocystoceles: Terminal swelling of the neural axis probably secondary to hydromyelia in-utero. The expanding mass contains dilated cords as well as meninges. CSF and fatty/fibrous tissue that herniates midline through a spina bifida defect. This results in a tethered cord syndrome. (Figure 18)
- Caudal agenesis: Caudal regression syndrome/ Caudal agenesis is a disorder that impairs the development of the lower (caudal) half of the body. It may affect the lower back (including the spinal cord), limbs, genitourinary tract, and the gastrointestinal tract. Maternal diabetes is a major risk factor for the disorder. Most cases are sporadic or are associated with maternal diabetes. The prognosis is poor; some newborns with severe cases do not survive the neonatal period. Those that survive infancy usually have normal cognitive function. (Figure 25)
- Segmental spinal dysgenesis: Segmental spinal dysgenesis (SSD) is a rare congenital spinal anomaly characterized by localized agenesis or dysgenesis of the lumbar or thoraco-lumbar spine, severe congenital kyphosis or kypho-scoliosis, and focal abnormalities of the underlying spinal cord and nerve roots. The malformation is typically segmental; in fact, vertebrae are present both above and below the anomaly and there usually is a lower cord segment within the spinal canal caudally, as if an intermediate segment of the spine and spinal cord were forgotten during embryonic development. (Figure 11)
- Dorsal enteric fistula: The dorsal enteric fistula is due to persistent connection between the endoderm and ectoderm, resulting in splitting of the notochord. The fistula traverses the prevertebral soft tissue, the vertebral bodies, and the spinal canal with its contents. Any portion of this tract may involute or become fibrous, leading to a fistula or a cyst. The dorsal enteric sinus, a remnant of the posterior portion of the tract, has an opening on the skin surface. Dorsal enteric cysts are trapped remnants of the middle portion of the tract, found in the intraspinal or paraspinal compartments. The dorsal enteric diverticulum is a tubular diverticulum arising from bowel and represents a remnant of the anterior portion of the tract, there exists a high incidence of urogenital malformations and anorectal malformations.
- Neurenteric cysts: Spinal neurenteric cysts are a rare type of foregut duplication cyst, accounting for ~1% of all spinal cord tumours. They are usually classified as spinal or intracranial and are associated with vertebral or CNS abnormalities respectively. Neurenteric cysts result from incomplete resorption of the neurenteric canal. The intraspinal cysts are usually intradural extramedullary (80-90%) and ventral in location. They most commonly occur in the thoracic region (~40%).( Figure 19).
- Diastematomyelia: Diastematomyelia, also known as a split cord malformation, refers to a type of spinal dysraphism (spina bifida occulta) when there is a longitudinal split in the spinal cord. Although traditionally it has been distinguished from diplomyelia (in which the cord is duplicated rather than split) the term split cord malformation is advocated to encompass both conditions. Split cord malformations are divided into two types according to the presence of a dividing septum and single vs. dual dural sac:
  1. Type I: duplicated dural sac, with common midline spur (osseous or fibrous) and usually asymptomatic
  2. Type II: single dural sac containing both hemi cords; impairment less marked(figure 14 and figure 15)
- Intradural/ intramedullary lipoma: rare and account for less than 1% of all spinal cord lesions. Usually extramedullary lesions, and are typically found in the lumbo sacral spine with spinal dysraphism. Because of their intramedullary location, following surgical intervention/ debulking/ excision, patients may only exhibit partial recovery. These usually originate from the migration of mesenchymal elements into the neural tube before its complete closure during embryogenesis. Symptoms typically consist of a progressive myelopathy associated with increasing degrees of paralysis (e.g., quadriaparesis/ plegia, paraparesis/ plegia). (Figure 8 )
- Filarlipoma: Fatty filum terminale, also known as lipoma of the filum terminale or filarial lipoma, is a relatively common finding on imaging of the lumbar spine, and in most cases is an incidental finding of no clinical concern. However, in some patients it may be associated with signs and symptoms of tethered cord syndrome. In such cases it is usually associated with a thickened filum and a low-lying conus. Fat is seen within the filum terminale in ~5% of relevant examinations. Lipoma of the filum terminale is formed as a result of a developmental error in meso-dermal cell migration. In most cases, a fatty filum is an incidental and asymptomatic finding. However, in some individuals, it is associated with spinal dysraphism and tethered cord syndrome. (Figure 13)
- Tight filum terminale syndrome: Tight filum terminale syndrome is caused by incomplete involution of the distal spinal cord during embryogenesis. This leads to development of an abnormally thickened filum terminale, which may be associated with lipomas or cysts within the filum. Tight filum terminale syndrome is always associated with spinal cord tethering and an abnormally positioned conus medullaris below L2-3 (normal range, L1-L2). Clinical symptoms are due to stretching of the spinal cord with resulting vascular insufficiency at the level of the conus medullaris. Associated vertebral body deformities and spina bifida are common findings.
- Dorsal dermal sinus: is an epithelium-lined tract from the skin to the spinal cord, caudaequina, or arachnoid. Dorsal dermal sinus is caused by incomplete separation of the superficial ectoderm from the neural ectoderm, resulting in a focal segmental adhesion. Dorsal dermal sinus manifests as a small dimple or pinpoint ostium, which is often associated with an area of hyper pigmented, angiomaticus skin or hypertrichosis and occurs in a midline location or rarely in a paramedian location. Predominantly located in the lumbo sacral region and less often in the occipital region. Dorsal dermal sinus occurring in a paramedian location is often associated with an intraspinal dermoid or epidermoid, which causes compression of neural structures with neurologic symptoms. (Figure 20)
- Persistent terminal ventricle: terminal ventricle of Krause, also known as the 5th ventricle, is an epedymal-lined fusiform dilatation of the terminal central canal of the spinal cord, positioned at the transition from the tip of the conus medullaris to the origin of the filum terminale. It represents the canalization and retrogressive differentiation of the caudal end of the developing spinal cord and regresses in size during the first weeks after birth. Abnormal persistence or cystic dilatation of the ventriculosterninalis (cyst of the medullary conus) can occur and may present symptomatically in adulthood with bladder or bowel sphincter disturbance. (Figure 16)
MYELOMENINGOCELE

Figure 1 (A-C): open spinal dysraphism-melomeningocele – Sagittal T2 (A) and Axial STIR (B) weighted images of lumbosacral spine showing open spinal dysraphism-melomeningocele with tethering of spinal cord (arrow in A) and (arrow in B) and Sagittal T2 (C) image demonstrates hydrocephalus secondary to aqueductal stenosis and cerebellar tonsillar Arnold chiari malformation (arrow)

Figure 2: Myelomeningocele

A. Axial T2 weighted MR image in 1 day old boy shows neural placode (Black arrow) extending above skin surface due to expansion of underlying subarachnoid space(white arrow), which is characteristic of myelomeningocele.

B. Sagittal T2 weighted MR images from same patient as in B with myelomenigocele shows neural placode (white arrow) protruding above skin surface due to expansion space(black arrow)
CONGENITAL INFECTED DERMOID CYST WITH FOCAL SPINA BIFIDA

Figure 3. 10 months old male baby with h/o Swelling in nape of neck, noticed 20 days after birth and has recently increased in size, Sag T2 and Sag STIR reveals, Congenital Infected dermoid cyst possibly ruptured with Spinabifidia at C4- Cystic lesion seen in the midline in the nape of neck. The lesion has thick enhancing wall. There is tract( pseudopod like projection) traversing from the lesion into the right side of lamina of C4 vertebral body which is thin with absent spinous process.( focal spina bifida).

OPEN TYPE OF SPINA BIFIDA WITH MENINGOCELE:

Figure 4 (A-C): Open type of spina bifida with meningocele -Lower lumbar neural tube defect in the form of splaying of posterior element and lipomeningocele at L4 vertebral level. Also there is defect in the skin and subcutaneous tissues at that level. Diastematomyelia at L2 - L4 vertebral levels. Tethering of spinal cord roots noted at L3-L4 vertebral level. Patulous spinal canal in lower dorsal and lumbar vertebral levels. (Image courtesy Dr. Kumar Ashok Charan (Joint director, Radiology department, Bokaro General Hospital, Bokaro, Jharkhand, India).
OPEN TYPE OF SPINAL DYSRAPHISM WITH MENINGOCELE.

Figure: 5(A-E) Swelling noted in the sacral region since birth.

Splaying of posterior elements noted from S1- S2 and herniation of posterior dura in the form of a thin tract extending to the subcutaneous plane with small meningocele. Low lying conus medullaris noted extending till the L4/5 level. Capacious spinal canal noted at lumbar and sacral levels suggestive of Open type of spinal dysraphism with meningocele.
MENINGO-MYELOCELE WITH LOW LYING CONUS MEDULLARIS AND TETHERING OF SPINAL

Figure 6: (A-F) 30 year old with h/o Back pain with bilateral lower limb numbness. Operated for meningocele 20 years back-Defect noted in the posterior elements of L4 and L5 vertebrae causing herniation of the dural sac.
- Neural tube defect. There is low lying conus medullaris with tethering of spinal cord at L4 and L5 vertebral level. Capacious spinal canal noted from L3/4 to L5/S1. Long segment syrinx noted in the spinal cord from C3 to L3 levels suggestive of recurrent meningo-myelocele with low lying conus medullaris and tethering of spinal cord at L4 and L5 vertebral level.
(Image courtesy Dr. Kumar Ashok Charan (Joint director, Radiology department, Bokaro General Hospital, Bokaro, Jharkhand, India).

CLOSED SPINAL DYSRAPHISM.

Figure 7: Dermal sinus, 6 year old baby shows Closed spinal Dysraphism. Dermal sinus is seen in the subcutaneous plane predominantly epicentered on right side extending from S3 to S4 vertebral level. Subcutaneous fibrous tract is seen extending up to the posterior dura at that level. No herniation of dura or tethering of cord noted. Bony defect of posterior arch seen extending from S2 to S5 level- Spina bifida occulta.
Figure 8—Intradural lipoma. Sagittal T1-weighted (A) and sagittal T2-weighted fat-saturated (B) MR images in 6-year-old girl show large intradural lipoma (arrows), which is hyperintense on T1-weighted image and hypointense on T2-weighted fat-saturated image. Lipoma is attached to conus medullaris, which is low lying.

Figure 9: MRI of thoracolumbar junction with intradural/ extramedullary neurenteric cyst at T11. (a) Midsagittal MRI demonstrates isointense lesion (arrow) on T1-weighted and (b) hyperintense signal (arrow) on T2-weighted.
Figure 10: Diastematomyelia at L1-L2 to L3-L4 level

Figure 11: Segmental spinal dysraphism MRI L.S spine, T2W seq, sagittal view revealing spinal canal narrowing and complete CSF column cutoff at LV-2 level with gibbus deformity

Figure 12: Axial T2-weighted image shows the right hemi cord exits the spinal canal through a posterior spinal defect and forming a placode (white arrow), thus resulting in hemi myelomeningocele. Note the postoperative changes of closing this open dysraphism.
Figure 13: Filarlipoma A and B. Sagittal (A) and axial (B) T1-weighted MR images in 2 year.

Figure 14: Type 1 diastematomyelia.

A-C, Sagittal T2-weighted MR (A), axial T2 weighted MR (B), and axial CT with bone algorithm (C) images in 6 year old boy show two dural tubes separated by osseous bridge (arrows), which is characteristic for type 1 diastematomyelia.
Figure 15: Type 2 diastematomyelia.

A-C, Sagittal T1 weighted (A), coronal T1 weighted (B), and axial T2 weighted (C) MR images in 9 year old girl show splitting of distal cord into two hemicords (white arrows, B and C) within single dural tube, which is characteristic for type 2 diastematomyelia. Incidental filum lipoma (black arrows, A and B) is present as well.

Figure 16: Persistent terminal ventricle.

A and B, sagittal T2-weighted (A) and sagittal T1 weighted contrast-enhanced (B) MR images in 12 months old boy show persistent terminal ventricles as cystic structure (arrows) at inferior aspect of conus medullaris, which does not enhance.
Figure 17: Hemilipomyelomeningocele. T2 weighted sagittal (A), T1 weighted sagittal (B), T2 weighted axial (C) and T1 weighted axial (D) images in a case of hemilipomyelomeningocele. There is diastematomyelia with bony septum (white arrow) suggestive of type I diastematomyelia. There is defect in the posterior spinal canal on the right side through which the spinal canal contents herniate with intact overlying skin and subcutaneous tissue (arrowhead). There is expansion of subarachnoid space (+) anterior to the cord pushing the neural placode-lipoma interface posteriorly to lie outside the boundaries of spinal canal (arrow).

Figure 18: Cystic dilation of the distal central Multiple levels spina bifida with absent posterior neural arch. Canal with trumpet-like flaring syringocele and meningoceles that herniate through the vertebral defect - terminal myelocystocele.
Figure 19: Neurenteric cyst in 3 year old girl.

A and B, Sagittal T2 weighted (A) and axial T1 weighted (B), MR images show bilobed neurenteric cyst (arrows) extending from central canal into posterior mediastinum. C, Three dimensional CT reconstruction images shows osseous opening (arrow) through which neurenteric cyst passes. This opening is called the Kovalevsky canal.

Figure 20: (a) Coronal magnetic resonance imaging (MRI) of the spine showing the L5 hemivertebra. (b) Sagittal MRI showing the conus medullaris ending at the L2/3 level. The is no significant thickening or hyperintensity of the filum terminale. (c) Dermal sinus tract (black arrow) is demonstrated as a dark band within the subcutaneous fat. The tract is seen terminating at the sacral dimple, marked by a fat-containing tablet (White arrow).

Figure 21: Lipomyelocele, Axial T1 images showing an intraspinal lipoma associated with the distal spinal cord that is continuous with the subcutaneous fat through a posterior sacral bony defect at the S3 level.
**Figure 22:** Post-operative dermoid. In a post-operative case of spinal dysraphism, T2 weighted sagittal, T1 weighted sagittal and T1 weighted axial images of lumbosacral spine showing a T1 and T2 hyperintense lesion in the conus medullaris and filum terminale consistent with dermoid formation (black arrow).

**Figure 23:** Sacro-coccygeal teratoma (type III). T2 weighted sagittal (A), T2 weighted axial (B), T1 weighted axial (C) and T1 weighted fat suppressed axial post contrast (D) images show a large heterogeneous presacral lesion. It has a large internal component which is predominantly fat (arrowhead) appearing hyperintense on both T1 and T2 weighted images with signal suppression on post contrast fat saturated image. It also has a small complex solid cystic external component (arrow) appearing hyperintense on T2 weighted and hypointense on T1 weighted with enhancement on post contrast images.
ANTERIOR MYELOCOELE/SACRAL TERATOMA

Figure 24: (A-D) Open spinal dysraphism—melomeningocele – Sagittal T2 (A) and Axial T2(B) weighted images of lumbosacral spine showing anterior myelocoele /sacral teratoma.

Figure 25: Caudal agenesis. T2 weighted sagittal (A), T1 weighted sagittal (B) and T2 weighted coronal (C) MR images showing type II caudal agenesis. There is non-development of distal sacral vertebra (white arrow in A) with abnormal termination of conus medullaris (black arrow). The child also has associated left hydronephrosis (dashed arrow in C).
V. Conclusion

Imaging of spinal dysraphism may appear complicated as it is a group of diverse conditions which can have variable imaging appearance. A systematic approach and correlation with clinical, neuro-radiological and developmental data imaging correlation provides an organized approach in their diagnosis.

References

[1]. Medical Dictionary for the Health Professions and Nursing © Farlex 2012