

Fetal Diastematomyelia: Rare Case

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Abstract: A ten -month-old infant with Diastematomyelia, as a very rare variety of spinal dysraphism is presented in this article Routine USG in a 36 weeks pregnant woman revealed a spinal abnormality of the fetus in thoracolumbar [D12-L1] regions.

At birth, examination of the newborn showed a small midline swelling and bluish discoloration of skin and scoliosis in the lumbar region. There was associated umbilical hernia.no neurological deficit was noted.

Postnatally 8 month plane radiograph showed multiple deformed dorsal vertebrae and mild scoliosis.MR imaging revealed diastematomyelia was seen at thoracolumbar [D12-L1] regions. It was type I with a bony spur measuring 1.0 X0.5 cm the two hemicords were asymmetrical. There was no cord tethering. The conus ended at the L4 level. . There was associated thoracolumbar spina bifida measuring 1.6 cm with a small meningocele measuring 2.5 X 0.6 cm. multilevel syringohydromelia and multiple deformed dorsal vertebrae and mild scoliosis of dorsal spine was seen. Thus what makes this case different is inspite of having so many microanatomical defects, there is no neurologic deficit. So its advisable to do corrective surgery to avoid further complicatiions- tethered cord syndrome before the baby starts to take steps.

Keywords - Diastematomyelia, myelomeningocele, meningocele , syringohydromelia,

I. Introduction

Diastematomyelia, also known as split cord malformation (SCM) is a congenital spinal anomaly in which there is longitudinal splitting of the spinal cord. The two hemicords may be separated by a fibrous, bony, or cartilaginous septum.[1] Antenatal detection of cord abnormalities by USG can be limited by acoustic shadowing from the spine, fetal position and the amount of liquor, though it is usually quite accurate in the diagnosis of spina bifida, in experienced hands, when correlated with maternal alpha-fetoprotein levels.[2] MRI is a safe and important tool for confirming the presence of spinal cord abnormalities in a fetus when a suspected spinal deformity has been detected on USG.[1,3-6]

II. Case Report

Routine USG in a 36 weeks pregnant woman revealed a spinal abnormality of the fetus in thoracolumbar [D12-L1] regions

At birth, examination of the newborn showed a small midline swelling and bluish discoloration of skin and scoliosis in the lumbar region. There was associated umbilical hernia. No neurological deficit was noted.

Postnatally 8 month plane radiograph showed multiple deformed dorsal vertebrae and mild scoliosis. [Fig 1& 2]

MR imaging revealed Diastematomyelia was seen at thoracolumbar [D12-L1] regions. It was type I with a bony spur measuring 1.0 X0.5 cm. The two hemicords were asymmetrical. There was no cord tethering. The conus ended at the L4 level. . There was associated thoracolumbar spina bifida measuring 1.6 cm with a small meningocele measuring 2.5 X 0.6 cm. multilevel syringohydromelia and multiple deformed dorsal vertebrae and mild scoliosis of dorsal spine was seen. [Fig 3 & 4]

Presently the baby is 10 month old, has same gross examination features of swelling and bluish discoloration of skin and scoliosis in the lumbar region [Fig 5]and umbilical hernia.[Fig 6] Developmental milestones and neurological examination being normal – jerks and power of limbs unaffected.

Thus what makes this case interesting is inspite of having so many micro anatomical defects, there is no neurologic deficit. So it's advisable to do corrective surgery to avoid further complications- tethered cord syndrome before the baby starts to take steps.

III. Discussion

Diastematomyelia (or SCM) is a form of OSD (occult spinal dysraphisms) that appears with duplication of the spinal canal. [7, 8, 9.] In SCM, there is vertical splitting of the spinal cord. Pang *et al.* have proposed a classification for SCM [9]. According to them, type I SCM consists of two hemicords, each contained in a separate Dural tube and separated by an osseocartilaginous septum. Type II SCM consists of a single dural sac containing both hemicords; the two hemicords being separated by a nonrigid fibrous septum. Imaging studies rarely show this fibrous septum. This differentiation has surgical importance as type I split cords are technically more difficult to correct and are associated with more surgical morbidity than type II, especially if there is an oblique septum dividing the cord asymmetrically.[9]

Diastematomyelia may be an isolated finding or may be associated with other spinal dysraphisms such as myelomeningocele, meningocele, lipoma, neurenteric cyst, and dermal sinus. The vertebral anomalies associated with diastematomyelia include hemivertebra, with kyphosis or scoliosis. There may be associated renal, rectal, and uterine malformations. hydromyelia, Klippel–Feil syndrome, hydro-cephalus, and Arnold–Chiari malformation and as well as intradural teratoma in rare cases.[7-11] The most common location of diastematomyelia is in the thoracolumbar region. Rarely, it can affect the cervicodorsal region. There is even a case report in the literature of a basicranial Diastematomyelia.[12]

In our patient, the Diastematomyelia was seen at thoracolumbar [D121-L1] regions. It was type I with a bony spur measuring 1.0 X 0.5 cm. There was associated thoraco lumbar spina bifida measuring 1.6 cm with a small meningocele measuring 2.5 X 0.6 cm. multilevel syringohydromelia and multiple deformed dorsal vertebrae and mild scoliosis of dorsal spine was seen.

The unified theory of embryogenesis proposes that all split cord malformations originate from one basic ontogenetic error occurring around the time when the primitive neurenteric canal closes. This basic error is the formation of an ‘accessory neurenteric canal’ through the midline embryonic disc that maintains communication between the yolk sac and amnion, enabling continued contact between ectoderm and endoderm within the canal. This abnormal fistula causes regional ‘splitting’ of the notochord and the overlying neural plate.[13] The altered state of the emerging split neural tube and the subsequent ontogenetic fates of the constituent components of the endomesenchymal tract ultimately determine the configuration and orientation of the hemicords, the nature of the median septum, the coexistence of various vascular, lipomatous, neural and fibrous oddities within the median cleft, the high association with open myelodysplastic and Cutaneous lesions and the seemingly unlikely relationship with fore- and midgut anomalies .[13]

Prenatal knowledge of spinal cord anomalies is important for prenatal counseling as well as surgical treatment. Since MRI shows these lesions better and with less inter-observer variation than USG, in patients with suspected spinal anomalies, either diagnosed on USG or based on clinical and laboratory criteria, fetal MRI should be used prior to further management.[1]

IV. Figures.



fig 1 plane radiograph showed multiple deformed dorsal vertebrae and mild scoliosis to the right



fig 2. plane radiograph showed multiple deformed dorsal vertebrae and mild scoliosis to the right

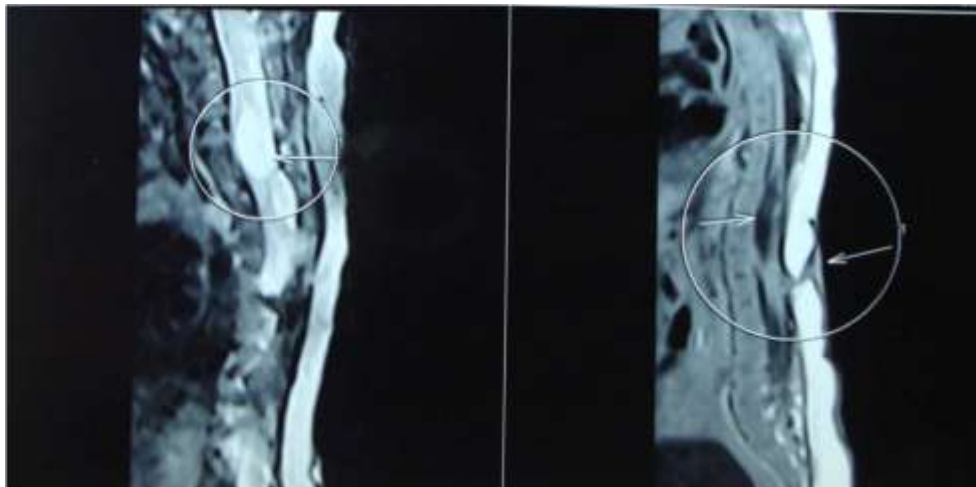


Fig 3 & 4- MR imaging revealed diastematomyelia was seen at thoracolumbar [D12-L1] regions. It was type I with a bony spur measuring 1.0 X 0.5 cm. The two hemicords were asymmetrical. There was no cord tethering. The conus ended at the L4 level. . There was associated thoracolumbar spina bifida measuring 1.6 cm with a small meningocele measuring 2.5 X 0.6 cm. multilevel syringohydromelia and multiple deformed dorsal vertebrae and mild scoliosis of dorsal spine was seen.



Fig 5 MR imaging revealed diastematomyelia was seen at thoracolumbar [D12-L1] regions. It was type I with a bony spur measuring 1.0 X 0.5 cm. The two hemicords were asymmetrical. There was no cord tethering. The conus ended at the L4 level. . There was associated thoracolumbar spina bifida measuring 1.6 cm with a small meningocele measuring 2.5 X 0.6 cm. multilevel syringohydromelia and multiple deformed dorsal vertebrae and mild scoliosis of dorsal spine was seen.



Fig 6 umbilical hernia.



Fig 7 swelling and bluish discoloration of skin and scoliosis in the lumbar region.

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