Case 7725

Diastematomyelia associated with spina bifida occulta, butterfly vertebra, pseudomeningoceles and tethered cord in young adult
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Section: Neuroradiology
Case Type: Clinical Cases
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Patient: 23 years, female

Clinical History:

The patient has been active in sports and dancing since childhood and did not have any complaints until the age of 22. During the last 1.5 years she had leg and low back pain, which was the reason for MRI study.

Imaging Findings:

The patient has been active in sports and dancing since childhood and did not have any complaints until the age of 22. During the last 1.5 years she had leg and low back pain, which was the reason for MRI study.

An MRI study of the lumbosacral spine was performed with 1,5T MR scanner in three orthogonal planes using T1 FLAIR, T1 FSE, FSE T2, FAST STIR and 3D Myelography sequences. The examination revealed congenital cleft within L2 vertebral body – butterfly vertebra and an osseous spur originating from the posterior bony elements of the L2 and traversing the entire spinal canal. The spinal canal is divided by the spur in two unequal parts (dext < sin). At this level the spinal cord is also split by osseous spur in two halves. The hemicords and dural sacs reunite above and below the cleft. There is incomplete closure of the posterior bony elements of the L2 covered by skin consistent with spina bifida occulta, low conus medullaris position at the L4 level whereas the filum terminale extend to the sacral region, and is fixed at the S1-S2 level resulting in tethered cord. At the level S2-S4 dura is interposed between subarachnoid space, and large intrasacral dorsal right and left side pseudomeningoceles not containing neural roots are seen. The possible communication site between the dural sac and the pseudomeningoceles seems to be at the S2 level.

Discussion:

Diastematomyelia (Split Cord Malformation–SCM) is segmental incomplete duplication of the spinal cord in which each hemicord has a single set of anterior and posterior nerve horns and roots. Fibrous, osteocartilaginous or osseous spur splits spinal cord into two hemicords. Hemicords reunite above and below cleft. The commonest location is thoracolumbal cleft (85% between T9 and S1). There are two types of diastematomyelia. The first type has a midsagittal bony or osteocartilaginous spur separating the spinal cord into two parts, each with its own arachnoid and dural sheath. The second type has no spur, but has a split spinal cord that lies within a single, common arachnoid and dural sheath. Females are more often affected than males. SCM arise from adhesion between the embryonic ecto- and endoderm. This leads to formation of an accessory neurenteric canal. The endomesenchymal tract condenses around accessory canal, bisecting the developing notochord and causing two hemineural plates to form. The result is a split spinal cord.

The patients become symptomatic often in adults. The most common symptoms are progressive kyphoscoliosis in
older children, adults; discrete atrophy or weakness in one or both lower extremities, progressive paraparesis; gait disturbances, orthopaedic foot problems; neurogenic bladder, sphincter incontinence, bowel dysfunction. Low back pain is the predominant symptom in adults.

Differential diagnosis of diastematomyelia is duplicated spinal cord (diplomyelia), which consists of two complete spinal cords, each with two anterior and two posterior horns and roots. Diplomyelia occurs exceedingly rare, seen in presence of spinal canal duplication.

Plain films and CT scan show widening of the spinal canal with bony septum. MR imaging is the modality of choice as it provides direct visualization of the split cord and helps in identifying associated abnormalities. There is often a cutaneous stigma, most commonly a patch of hair or dermal sinus; haemangioma, lipoma, nevi and skin dimples. There is a high incidence of spinal segmentation anomalies: hemivertebrae, block or butterfly vertebrae and narrow intervertebral disk spaces. Cutaneous stigmata and segmentation anomalies occur at the same level as the split cord, representing an important clue for the verification of lesion’s level. Other associated abnormalities: intersegmental laminar fusion with spina bifida, which is pathognomonic for diastematomyelia; spinal dysraphism (myelocoele/myelomeningocele, hemimyelocele; tethered spinal cord, thickened filum terminale; syringohydromyelia one or both hemicords above diastematomyelia; congenital scoliosis; Chiari-II malformation. The clinical symptomatology vary from relatively asymptomatic to those similar to tethered cord.

SCM patients need surgical intervention to avoid irreversible damage of nerve tissue. Early therapy affects the clinical outcome by halting the progression of vascular damage to the cord whereas develops stable or progressive disability if untreated. Late onset or previously stable patients may become symptomatic following minor back injury or surgery requiring spinal manipulation. Surgery consists of bony or cartilaginous spur resection (cartilaginous structures can develop into fibromatous tumours) or fibrous septum, correction of tethered cord. Following surgery stabilizes or improves > 90 % of patients. Scoliosis has been rarely affected by surgical untethering.

**Differential Diagnosis List:** Diastematomyelia associated with spina bifida occulta, pseudomeningoceles and tethered cord

**Final Diagnosis:** Diastematomyelia associated with spina bifida occulta, pseudomeningoceles and tethered cord

**References:**

Description: reveals increased anteroposterior diameter of the spinal canal. There is an osseous spur between the posterior border of the L2 vertebral body and its posterior bony elements traversing the entire spinal canal. The conus medullaris ends at the L4 level. Large intrasacral dorsal pseudomeningocele not containing neural roots at the S2 – S4 level. Origin:
**Description:** demonstrates an osseous spur originating from the posterior bony elements of the L2 and traversing the entire spinal canal. The spinal canal is divided by the spur into two unequal parts. The spinal cord is also split in two halves. There is incomplete closure of the posterior bony elements of vertebra covered with skin. **Origin:**
Figure 2

Description: MR sagittal FLAIR T1 WI reveals increased anteroposterior diameter of the spinal canal. An osseous spur between the posterior border of the L2 vertebral body and its posterior bony elements traversing the entire spinal canal, and narrow intervertebral disk space at the level L2 – L3, also large intrasacral dorsal pseudomeningocele at the level S2 – S4 are seen. Origin:
Description: MR axial T1 FSE WI demonstrates an osseous spur originating from the posterior bony elements of the L2 and traversing the entire spinal canal. The spinal canal is divided by the spur into two unequal parts. The spinal cord is also split in two halves. Origin:
Description: shows a wide distal spinal canal and thecal sac. Low conus medullaris position at the level L5 is seen. The filum terminale extend to the sacral region, consistent with tethered cord at the level S1-S2. At the level S2 – S4 there are large intrasacral dorsal pseudomeningoceles not containing neural roots. At the level S2/S3 dura is interposed between subarachnoid space and pseudomeningoceles. The possible communication site between the dural sac and pseudomeningoceles seems to be at the S2 level. Origin:
Description: reveals osseous cleft at the level L2 and two hemicords. Origin:
Description: demonstrates osseous cleft at the level L2, and large intrasacral pseudomeningoceles not containing neural roots at the level S2 – S4. Origin:
Figure 5

Description: shows the butterfly L2 vertebra

Origin: