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Role of rehabilitation a case of diastematomyelia

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Abstract

Diastematomyelia (**split cord malformation**) is a rare dysraphic lesion in which a part of the spinal cord is split in the sagittal plane into two hemicords, a bony, cartilagenous or fibrous spur projecting through the dura mater is visible in 33% of cases. Vertebral anomalies (spina bifida) are common. It occurs usually between D9 and S1. Classificatin includes two types: type 1 with a duplicated dural sac, with common midline spur, usually symptomatic, and type 2 with a single dural sac and usually less symptomatic. The majority of patients are presenting with tethered cord syndrome (neurologic deficits in the lower limbs and perineum). MRI is the modality of choice for diagnosis. In symptomatic cases, surgical release of the cord and resection of spur with repair of dura are performed, with good results. We present a case of a pauci-symptomatic type 1 dyastematomyelia, manifested by intermittent and resistant lumbar pain, in which physiotherapy during rehabilitation program have shown to improve pain intensity.
Key words: *spinal cord malformation, dyastematomyelia, lumbar spine,*

INTRODUCTION: Diastematomyelia (also known as a **split cord malformation**) is a rare dysraphic lesion of the spinal cord in which a part of the spinal cord is split in the sagittal plane into two hemicords. Each of them includes the central canal, one dorsal and one ventral root. The splitting of the cord is caused by the presence of an osseous, cartilaginous, or fibrous septum [1].
Diplomyelia, or true duplication of the spinal cord, occurs when the split does not reunite distally to the spur.

EMBRIOLOGY: The defect is a result of an incorrect development of the notochord between the 15th and the 18th day of pregnancy [1] or from an abnormal adhesion between ectoderm and endoderm [2]. It may be isolated or associated with other spinal dysraphisms, such as myelomeningocele, meningocele, spinal lipoma, neuroenteric cysts or dermal sinuses and vertebral abnormalities, such as hemivertebrae, butterfly vertebrae or scoliosis. In the latter case, laminar fusion of the vertebrae associated with a neural arch defect are good predictors of diastematomyelia and occurred at the level of the defect, or at an adjacent level.

Intramedullary tumours associated with diastematomyelia have been rarely described and associated conditions like a tethered cord, inclusion dermoid, lipoma, syringo-hydromyelia and Chiari malformation have also been described in literature. This malformation may also be accompanied by abnormalities of internal organs – kidneys, anus or reproductive organs [1]. Most authors believe that cases of an isolated diastematomyelia are connected with better prognosis.

PATHOLOGY. Diastematomyelia usually occurs between D9 and S1 levels, being more common in the lower cord; 50 % occur between L1 and L3 and 25 % occur between T7 and T12. Sometimes it occurs at multiple levels, but cervical diastematomyelia is a very rare.
In about 50% of patients, the hemi-cords are contained in a single dural sac, while in other half of the patients, they are two separate dural sacs, and bony or fibrous spurs are usually found between the two sacs. An associated bony, cartilagenous or fibrous spur projecting through the dura mater forwards from the neural arch is visible in 33% of cases .

Vertebral anomalies (spina bifida, butterfly or hemivertebrae) are common.

CLASSIFICATION. Split cord malformations are divided into two types according to presence of a dividing septum and single vs dual dural sac.

- type I - duplicated dural sac, with common midline spur (osseous or fibrous) and usually symptomatic

- type II - single dural sac containing both hemicords; usually less symptomatic

Type I is the classic diastematomyelia, characterised by :

- duplicated dural sac
- hydromyelia
- midline spur (osseous or osteocartilaginous)
- vertebral abnormalities: hemivertebrae / butterfly vertebrae / spina bifida / fusion of laminae of adjacent levels
- skin pigmentation / haemangioma / hypertrichosis (hair patch) are common
- patients are usually symptomatic presenting with scoliosis and tethered cord syndrome

Type II is milder than type I, and lack many of the features of latter:

- single dural sac and no spur / septum
- cord divided, sometimes incompletely
- hydromyelia may be present
- spina bifida may be present, rare other vertebral anomalies
- patients are less- or asymptomatic

EPIDEMIOLOGY. Split cord malformations are a congenital abnormality and account for approximately 4% of all congenital spinal defects. Most patients with a diagnosed diastematomyelia are children under the age of 7. In adult patients, such a spinal abnormality is diagnosed less frequently [1]. It is more common in females (3:1) [2].

ANTENATAL DIAGNOSIS: Presence of an extra echogenic focus at ultrasound in the midline between the foetal spinal posterior elements has been described as a reliable sign [3]

CLINICAL PRESENTATION. Patients may be asymptomatic or may present progressive spinal cord dysfunction. The majority of patients with diastematomyelia are symptomatic, presenting with signs and symptoms of tethered cord, manifested by neurologic deficits in the lower limbs and perineum: leg weakness, muscular atrophies, gait troubles, sphincterian disturbances, reflex changes, associated with low back pain and squeletal abnormalities such ascongenital scoliosis and foot deformities.

IMAGISTIC FEATURES. With modern imaging techniques, spinal dysraphism is being diagnosed in adults with increasing frequency, often as an incidental finding. Radiographies shows multi level spina bifida , widening of interpedicular distance, which could be remote from the site of the spur; associated scoliosis or anteroposterior narrowing of vertebral bodies. Spinal CT scan shows all the features seen on plain films and may demonstrate the bony septum. Spinal MRI is the modality of choice for assessing children with split cord malformations, showing the cord and the presence of hydromyelia or other associated anomalies.

DIFFERENTIAL DIAGNOSIS. Usually there is little in the way of a differential diagnosis when the MRI aspect is typical. Dyplomyelia is considered part of the same spectrum by many authors. The differential diagnosis includes tethered spinal cord, thickened filum terminale, neurenteric cysts, congenital dermal sinus tract, anterior sacral meningocele, spinal lipomas, and dermoid and epidermoid tumors.

TREATMENT. No treatment is needed in asymptomatic cases. For tethered cord cases, surgical release of the cord and resection of spur with repair of dura are the techniques used. Prognosis after surgical treatment is good: 90 % of patients improve following surgery and 10% are rethering.

CASE PRESENTATION. We present the case of an 27 years old lady, school teacher, which presents for intense lumbar pain. She had a history of asymptomatic spina bifida from the age of 5, and of multiple episodes of lower urinary tractus infections. Her back pain problem started 3 years ago,when she undergone lumbar spine radiographies that confirmed the presence of spina bifida at L5-S1 level.Since than, repeated episodes of low back pain were reported by the patient.

The actual lombalgia started 3 weeks ago, is intense and persistant, is worse in the morning at awakening, and increases after prolonged ortostatic position. The pain was resistant at symptomatic treatment with OTC (paracetamol, metamizole) or at nonsteroidal antiinflamatory drugs (NSAIDs).
Her clinical examination shows no skin changes in the lumbar area, hyperlordosis with reduced spine mobility, tenderness of the lumbar paravertebral muscles. The neurological examination shows a normal gait, absence of motor deficit al lower limb level, the osteo-tendinous reflexes were normal, plantar reflexes were in flexion; no objective sensory troubles were noticed; elongation maneuvers were negative. The patient did not present sphincterian troubles.

Urinalysis did not show traces of urinary infections, abdominal ultrasound was normal. The rest of laboratory investigations were normal. Lumbar spine radiographies showed hyperlordosis, mild scoliosis, spina bifida at L4 level, discopathy at L3-L4 and L4-L5 levels (Figure 1 and Figure 2)

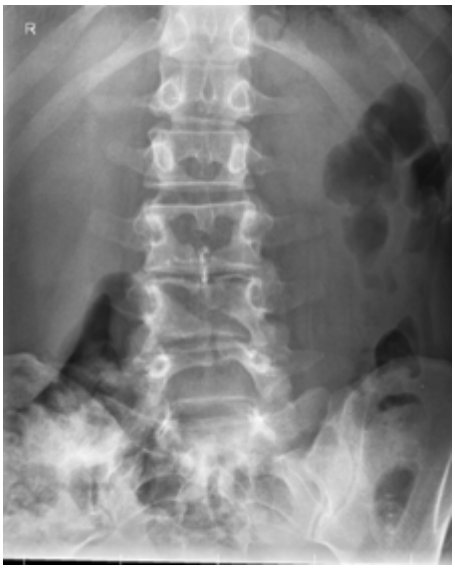


Figure 1: PA radiograph of lumbar spine

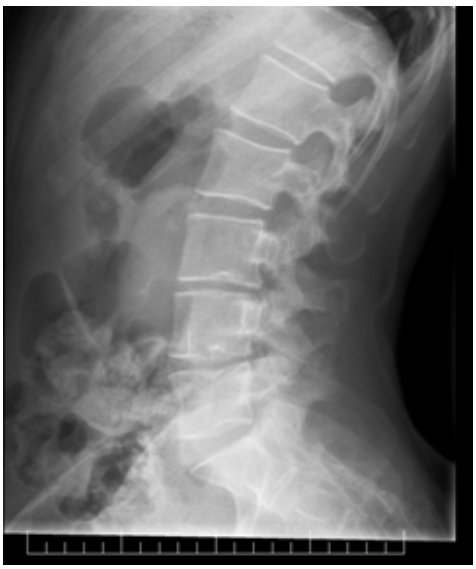
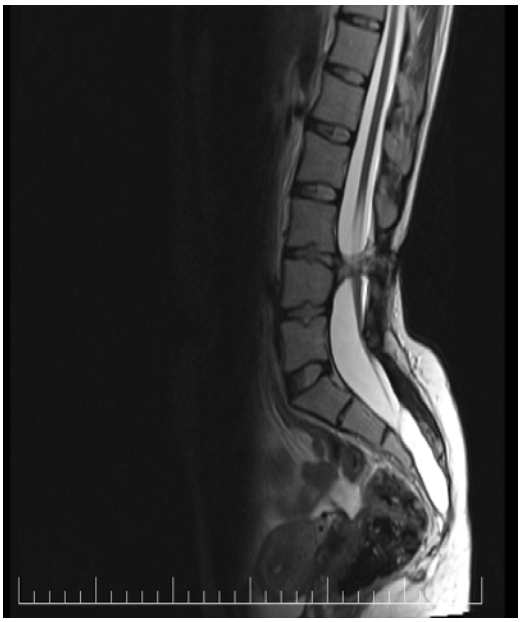


Figure 2: LL radiograph of lumbar spine



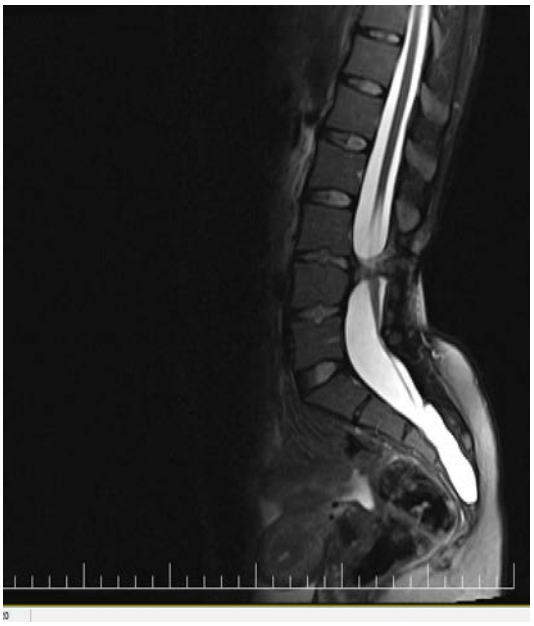


Figure 3: Lumbar spine MRI in sagital plane showing the cartilaginous spur at L3-L4 level and tethered cord (conus medullaris at L4 level)

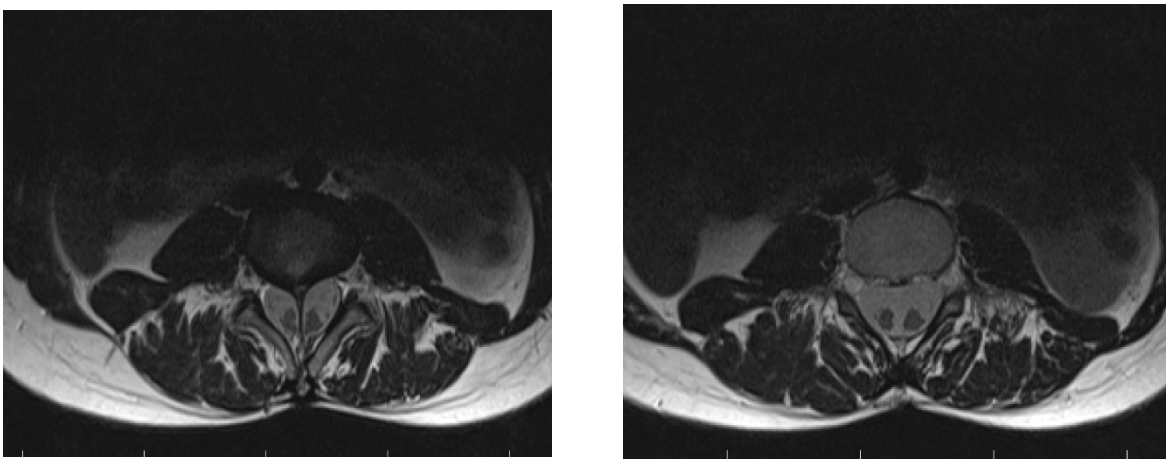


Figure 4: Lumbar spine MRI axial plane showing splitting of the spinal cord

The persistence of the lumbar pain and its characters imposed an thorough investigation with a spinal MRI. This showed a dysraphic lesion of the lumbar spinal cord: dyastematomyelia (Figure 3) and tethered cord, with low conus medularis until L4 level (Figure 3). Splitting of the spinal cord was present above L4 level (Figure 4).

According to the classification, it was a type I dyastematomyelia, with a fibrous midline spur and duplicated dural sac; spina bifida L4 is associated. Tehtered cord was present, with termination of conus medullaris at L4 level.

The patient was pauci-symptomatic, and conservative treatment was proposed.

The patient requires periodical clinical and MRI monitoring for the tethered cord syndrome, with could produce early sphincterian disturbancies, and in evolution peripheral and central motor deficits, sensory deficits and muscular atrophies in lower limbs [4].

The aim of the treatment was minimizing the pain, improvement of spinal mobility and a better life quality. The patient was included in a rehabilitation program, which comprises kinesitherapy – back strengthening exercises, massages, electrotherapy and laser-therapy, in association with the use of short course NSAIDs and analgetics.

After 10 sessions of physiotherapy, the intensity of pain decrease from 6 to 3 on VAS pain scale [5], showing an important improvement. The patient was discharged in a good condition, with mild intensity pain. She was referred to a continuation of her rehabilitation program in an outpatient facility and was encouraged to continue this program in a balnear resort [6,7].

In conclusion, physiotherapy treatment have showed good effects on lumbar pain in a case of a dysraphic malformation of the spinal cord. Case particularity was the lack of neurologic symptoms in a type 1 dyastematomyelia.

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