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Magnetic Resonance and CT Imaging of Diastematomyelia

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Summary. The clinical, MRI and CT features of diastematomyelia with an uncommon clinical course are reported. Possible pathogenetic aspects in the late onset of symptoms are discussed implying vascular factors. MRI provides direct visualization of the split cord and low conus, confirming that frontal images are preferable.

Key words: Diastematomyelia – Late onset – Pathogenesis – Magnetic resonance imaging (MRI) – CT

Introduction

Diastematomyelia is a developmental disorder with longitudinal splitting of the spinal cord over a variable number of segments and sometimes an interposed mid-line septum within the split. The congenital anomaly is probably due to incomplete fusion of the embryonic neural tube and is accompanied by skeletal and cutaneous defects and anomalies [14]. These include abnormal width of the spinal canal, vertical fusion of vertebral bodies, split vertebrae, bony or fibrous spurs or septa, lipomas or dermoid cysts and low conus, tethered by a fibrous band to the dura [1, 4, 5, 8, 12, 15]. The neurological features typically consist of a peripheral paresis of one or both legs, occasionally combined with central paresis, bladder dysfunction, dissociated or global sensory loss and back pain, radiating to the legs [9]. The sensory defect often causes a trophic lesion of the lower extremities. Mostly, diastematomyelia is diagnosed in early childhood, and only rarely do the first symptoms appear later in life [2, 3, 13].

Case Reports

Clinical Course. During the last trimester of her first pregnancy at 23 years of age (1957), a previously healthy woman suffered from an incomplete paraparesis with sudden onset and a slow restitution. Since 1974 she had noticed a steadily increasing weakness in her legs, with occasional falling due to a transient loss of power in her legs. On examination (1984) she exhibited a moderate scoliosis, an incomplete lesion of the lower spinal cord and cauda equina with a pronounced paraparesis, weak ankle jerks despite considerable spasticity, slight sensory disturbances and frequent radicular pain. Following medication with a low molecular dextran over 10 days the disability slightly but significantly decreased.

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Neuroradiological Findings

Conventional radiographs of the spine exhibited moderate scoliosis and fusion of the vertebrae L-3 to L-5.

Myelography including tomographic images and CT myelography demonstrated diastematomyelia with low conus and division of the spinal cord, extending from the level of the L-1 vertebra down to L-5, with a very short septum at the level of L-3/4 (Fig. 1). Overlapping sections allowed image reconstruction in the frontal plane (Fig. 2).

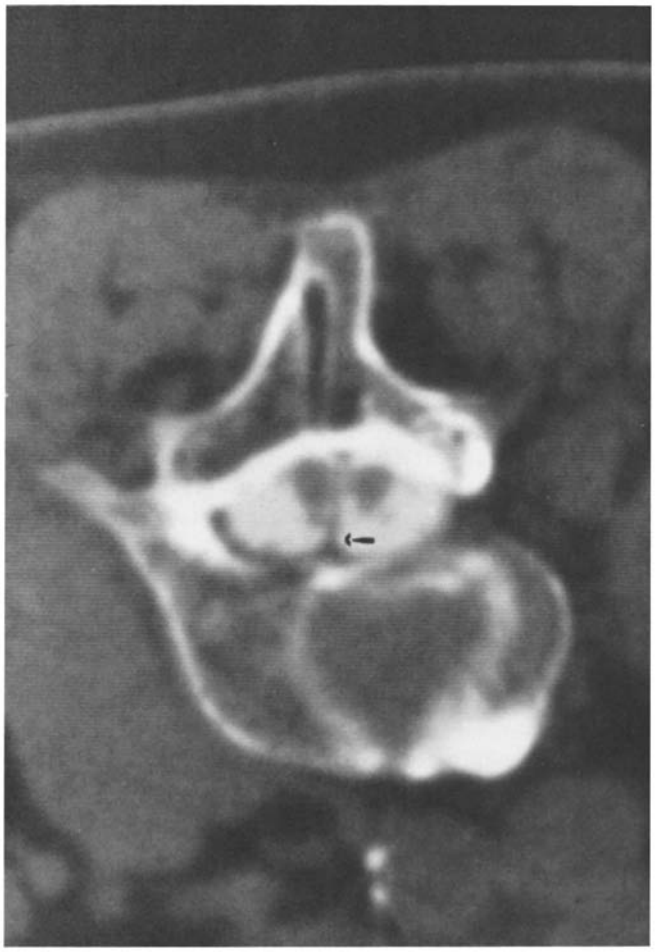

Fig. 1. CT myelography, original axial cut at the level of the L-3 vertebra. In the widened dural sac the separated parts of the split spinal cord and the fibrous septum *(arrow)* are demonstrated

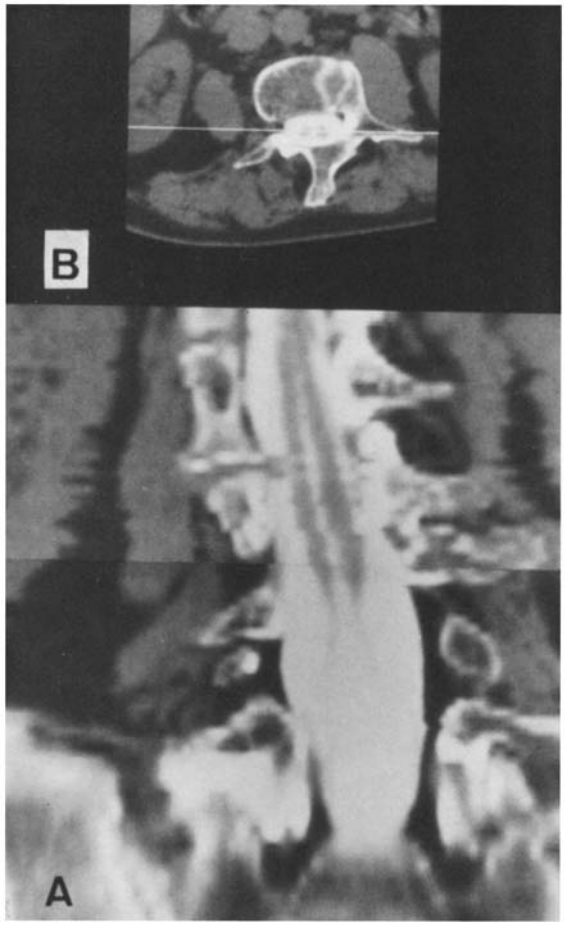

Fig. 2A, B. CT myelography, image reconstruction in the frontal plane (**A**), the longitudinal splitting of the spinal cord is visualized in its whole extension. The orientation of the calculated image is shown in (**B**)

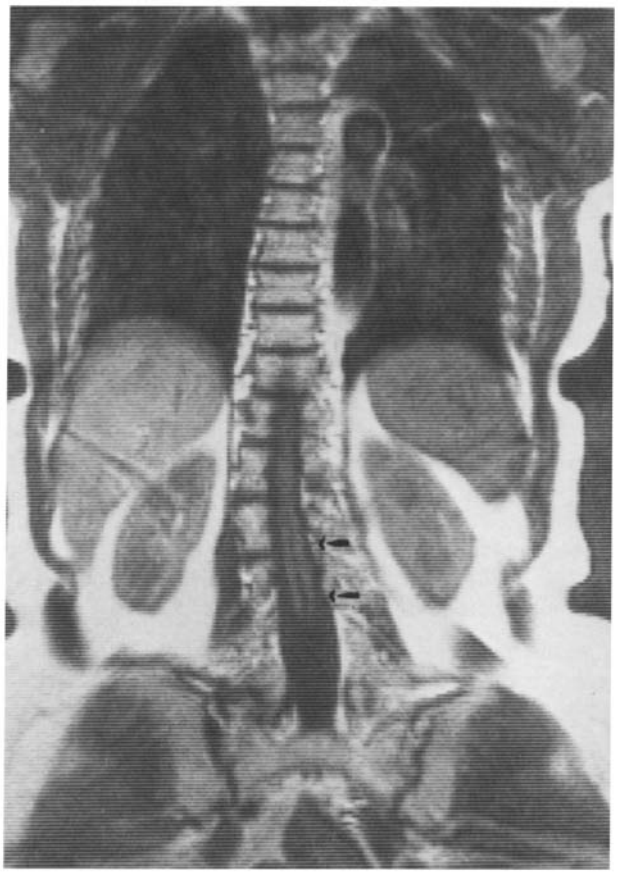

Fig. 3. MRI, frontal section, due to the physiological curvature of the spine only the lower parts of the spinal cord are localized in this cut. Diastematomyelia is clearly visible *(arrows)*

Magnetic resonance images (MRI) were obtained with a Magnetom at 0.5 Tesla. Spin-echo technique (SE) was used with short repetition- and echo-times for optimal discrimination of the CSF (long T1 and T2 values) and the spinal cord (short T1 and T2 values). The splitting of the cord and the position of the separated parts in the lumbar region is well shown on ventral sections (Fig. 3) but also visible on axial cuts similar to CT myelography. Very small details such as the fibrous septum could not be discriminated.

Discussion

Diastematomyelia is a congenital defect that usually causes neurological complications in early childhood. The disability commonly deteriorates progressively probably due to increasing pressure and traction caused by septa or fibrous bands tethering the cord while the body is growing [4, 6, 8, 15]. Only rarely does diastematomyelia remain asymptomatic throughout life [7] or not cause neurological impairment before adulthood. English and Maltby [2] felt that additional degenerative disease of the vertebral axis is pathogenetically relevant in late onset cases. Sheptak [14] presumed that trauma can initiate deterioration. In our patient, the sudden onset of the paraparesis during the last trimester of pregnancy, repeated transient loss of posture and the beneficial effect of low molecular dextran indicate a vascular component in the pathogenetic process. Perhaps the altered circulation during pregnancy, especially the increased venous pressure in the inferior vena cava territory, is a risk for spinal hypoperfusion in diastematomyelia when segmental stenosis and fibrous traction already impair the venous blood flow. A similar mechanism is thought to produce spinal dysfunction during pregnancy in cases of previously silent vertebrospinal hemangioma [10].

Neuroradiological evaluation showed the typical anomalies of diastematomyelia [4, 5, 11, 12]. A bony or cartilaginous spur is not a constant finding [11, 12], in this case a very short fibrous septum at the level of the L-3 vertebra was detected. Modern CT equipment allows image reconstruction perpendicular to the original section. But in extensive anomalies as in our case numerous cuts are necessary causing considerable exposure to ionizing radiation.

Even in its current state of development MRI of the spinal cord has shown remarkable diagnostic potential. Superior to CT, primarily multiplanar imaging provides optimal sections depending on the orientation of the lesion. Syringomyelia, for example, is easily detectable on a mid-saggital cut, for diastematomyelia the frontal plane is optimal as demonstrated in the present case. With this technique the anomalies of the spinal canal and cord were well visualized, but no additional information compared with CT examination was obtained. Further improvement of spatial resolution and choice of adequate imaging parameters may help to detect and discriminate

small defects in detail and finally, perhaps, reveal more understanding about metabolism and circulation and thereby about the fundamental pathogenetic processes.

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