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Case Report

Spinal Dysraphism Complex: Diastematomyelia with Neurenteric Cysts

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Abstract

Spinal neurenteric cysts are rare congenital lesions often associated with spinal dysraphism. Diastematomyelia is a malformation involving a split spinal cord divided by a bony or fibrous septum into two hemicords. The coexistence of these anomalies, particularly with a dorsal neurenteric cyst, is extremely uncommon. Here, we present the case of a 7-year-old girl who presented with lower limb weakness and gait disturbance. Magnetic resonance imaging (MRI) revealed type II diastematomyelia extending from 12th thoracic vertebra to 3rd lumbar vertebra, a low-lying tethered cord, and a dorsal bilobed neurenteric cyst at the level of the 11th and 12th thoracic vertebrae. Surgical resection of the cyst and bony spur resulted in clinical improvement. This case supports the unified embryological theory of split cord malformations and highlights the importance of MRIs for diagnosis and surgical planning.

Keywords: *Diastematomyelia; Neurenteric Cyst; Spinal Dysraphism; Pediatric Spine; Magnetic Resonance Imaging (MRI)*

Abbreviations

MRI: Magnetic Resonance Imaging; SCM: Split Cord Malformation; CT: Computed Tomography; D12: 12th Thoracic Vertebra; L3: 3rd Lumbar Vertebra; D11: 11th Thoracic Vertebra; L5: 5th Lumbar Vertebrae

Introduction

Spinal neurenteric cysts and diastematomyelia are rare congenital anomalies arising from errors in early embryogenesis. Derived from endodermal tissue, neurenteric cysts are most frequently located along the ventral spinal canal and are often associated with other spinal malformations. Diastematomyelia is a type of split cord malformation (SCM) characterized by a longitudinal division of the spinal cord into two hemicords, which are usually separated by a bony or fibrous spur. The simultaneous occurrence of a neuroenteric cyst and diastematomyelia is rare. MRI remains the imaging modality of choice for providing a detailed evaluation of neural and associated bony abnormalities. CT is complementary for defining osseous anomalies. Here, we present a rare pediatric case of type II diastematomyelia with a dorsally located neurenteric cyst, emphasizing the MRI findings. The case highlights the importance of accurate imaging in diagnosing complex spinal anomalies. Additionally, it emphasizes that one must use a multidisciplinary approach in managing such rare presentations to ensure optimal patient outcomes.

Case Report

A seven-year-old girl was referred to our department for evaluation of a progressive disturbance in her gait, which was the main symptom that prompted medical consultation. She had also experienced lower limb weakness and intermittent back pain for the previous eight months. Her past medical history was unremarkable, with normal growth and developmental milestones. There was no history of trauma, infection, or surgery. There was no family history of neurological disorders, spinal anomalies, or hereditary neuromuscular diseases. Physical examination revealed mild scoliosis and bilateral lower limb hypotrophy without urinary incontinence or bowel dysfunction. A neurological examination revealed decreased tone and strength (4/5) in the lower limbs, hyperreflexia, and a bilateral Babinski sign. There were no cranial nerve deficits or upper limb abnormalities. She was referred to a neurologist, who recommended an MRI of the spine for further evaluation.

The MRI study revealed type II diastematomyelia, which extends from D12 to L3. This condition is characterized by two symmetrical hemicords that are separated by a bony spur and do not exhibit abnormal cord signal. Additionally, the conus medullaris was located at L4, and the anterior filum terminale was thickened. A bilobed intradural-extramedullary cyst was identified at D11–D12, which is consistent with a neurenteric cyst (Figure 1). Additional anomalies included an L5 right hemivertebra, partial sacrococcygeal agenesis, and mild compression of the D12 vertebral body. Finally, a sacrococcygeal mass with cystic and solid components extending to the skin, but not the pelvis, was observed. This mass is most likely another neuroenteric cyst (Figure 2).

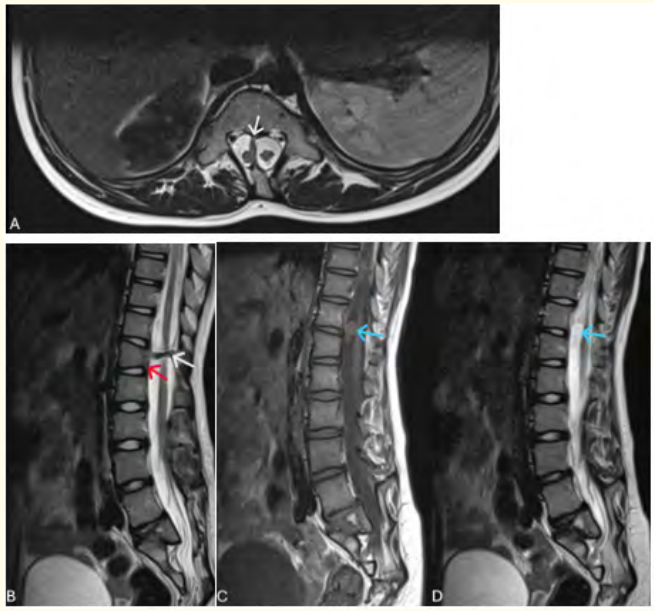

Figure 1: *MRI findings showing type II diastematomyelia and a dorsal neurenteric cyst. (A) Axial T2-weighted image demonstrating two symmetrical hemicords separated by a bony spur extending from the D12 to L3 vertebra, consistent with type II diastematomyelia (White arrow). (B) Sagittal T2-weighted image showing diastematomyelia (white arrow) with mild compression of the D12 vertebral body (Red arrow). (C) Sagittal T1-weighted image showing a bilobed intradural-extramedullary cyst at the D11-D12 level with isointense signal relative to the spinal cord, consistent with a neurenteric cyst. (D) Sagittal T2-weighted image showing the same bilobed cyst with hyperintense signal, clearly delineated from the surrounding cord (Blue arrow).*

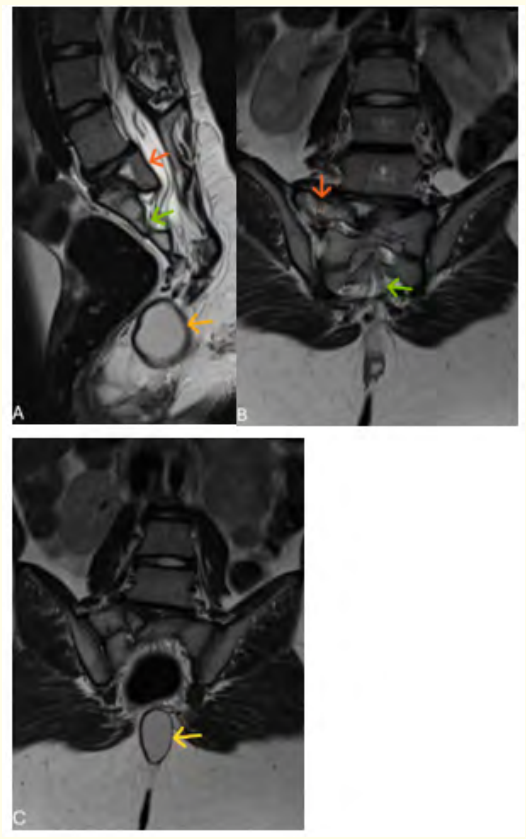

Figure 2: *(A) Sagittal T2-weighted image showing a right hemivertebra at L5 (orange arrow), partial sacrococcygeal agenesis (green arrow), and neuroenteric cystic (yellow arrow). (B) Coronal T2-weighted image demonstrating partial sacrococcygeal agenesis (green arrow) and right hemivertebra at L5 (orange arrow). (C) Coronal T2-weighted image showing a sacrococcygeal neurenteric cyst, exhibiting hyperintense signal on T2, extending to the skin without pelvic involvement (Yellow arrow).*

Due to the variety of spinal anomalies identified on the MRI, a multidisciplinary discussion was held, and the patient underwent staged surgical intervention. The first stage consisted of laminectomy and microsurgical resection of the D11–D12 cyst, as well as removal of the bony spur, to decompress the hemicords and detether the spinal cord. Histopathological analysis confirmed that the cyst wall was lined by columnar epithelium with goblet cells, which is consistent with a neurenteric cyst diagnosis.

Postoperatively, the patient showed progressive improvement with increased motor strength and reduced gait disturbances. She has remained clinically stable during follow-up, and a control MRI is planned to monitor the postoperative outcome and check for recurrence.

Discussion

Spinal neurenteric cysts are rare congenital lesions believed to develop from abnormal communication between the primitive endoderm and ectoderm during the third week of embryonic development. Although they are most often found in the spinal canal, these cysts have also been reported in the brain, chest cavity, abdomen, pelvis, and subcutaneously [1]. Although these cysts are uncommon in children, they can present at any age, and their clinical presentation can vary accordingly.

Neurenteric cysts usually appear during the second or third decade of life and affect more males than females. In pediatric cases, the average age at presentation is 6.4 years. Adults commonly present with localized pain, myelopathy in cervical or thoracic lesions, and radicular symptoms in the cervical or lumbar regions [2]. The intermittent nature of symptoms is thought to result from changes in cyst volume driven by periodic fluid shifts due to hemodynamic or osmotic variations [3].

Most neuroenteric cysts are located along the ventral aspect of the upper cervical and lower thoracic spinal cord, with approximately 90% intradural-extramedullary and the remainder distributed between extradural and intramedullary sites [5]. They belong to the spectrum of occult spinal dysraphism, which also includes lipomas, lipomyelomeningoceles, meningocele manqué, dermal sinus tracts, inclusion cysts (dermoid and epidermoid), terminal syringohydromyelia, myelocystocele, and split cord malformations (SCMs) [1].

Patients suspected of having neuroenteric cysts should undergo imaging evaluation, preferably with MRI, which provides superior delineation of cyst morphology and its relationship to adjacent neural structures compared to CT [2]. MRI also helps eliminate potentially misleading bony artifacts often seen on CT, while CT remains essential for assessing associated osseous malformations. Furthermore, CT myelography can reveal the characteristic "meniscus sign," caused by partial contrast blockage in intradural-extramedullary cysts or complete obstruction in intradural-intramedullary cysts [4].

On MRI, these lesions typically appear isointense to slightly hyperintense compared to cerebrospinal fluid on T1-weighted sequences and iso- to hyperintense on long relaxation time sequences. Such signal characteristics are attributed to the protein-rich fluid content within the cysts and have been frequently described in the literature [2].

Among the spectrum of occult spinal dysraphism, diastematomyelia represents a particularly rare anomaly of the vertebral axis. Diastematomyelia is characterized by a longitudinal split of the spinal cord into two hemicords. Each hemicord possesses its own central canal, as well as dorsal and ventral horns with corresponding nerve roots. The exact underlying cause remains uncertain [6]. Two forms of split cord malformations (SCMs) are recognized. In SCM Type I (diastematomyelia), the spinal cord is divided into two hemicords contained within separate dural sacs, separated by an osseous or cartilaginous septum. In SCM Type II (diplomyelia), both hemicords lie within a single dural sheath [7].

Clinical manifestations of diastematomyelia may include cutaneous abnormalities overlying the spine, neurological deficits, and orthopedic deformities. The symptoms are generally nonspecific and resemble those observed in other types of spinal dysraphism [6].

Traditionally, plain radiography and myelography were employed for diagnosis. However, advances in imaging have established computed tomography (CT) and magnetic resonance imaging (MRI) as the primary modalities for assessing diastematomyelia. CT provides optimal visualization of the bony spur and associated vertebral anomalies, including characterization of the spur's composition-whether osseous, cartilaginous, or fibrous. In contrast, MRI offers superior delineation of the presence and extent of the split spinal cord [6].

Neurological deterioration in patients with diastematomyelia is primarily attributed to traction on the typically low-lying spinal cord, a condition exacerbated by the presence of a bony septum and aggravated during growth. Consequently, neurological signs related to cord splitting-such as lower limb hypotrophy, clubfoot, and neurogenic bladder-may occur [8].

This case lends further support to the unified embryogenesis theory proposed by Pang., *et al*. which attributes all forms of split cord malformations (SCMs) to a shared developmental mechanism [9]. According to this theory, an error during the third week of embryogenesis results in abnormal adhesion between ectoderm and endoderm, creating an accessory neurenteric canal. This canal becomes enveloped by mesenchyme, forming an endo-mesenchymal tract that divides the developing notochord and neural plate. Mesenchymal tissue surrounding this tract differentiates into a fibrous or osseocartilaginous septum, while splitting of the neural plate

produces two hemicords. In some cases, the endodermal lining of the tract may develop into neurenteric cysts under the influence of inductive signals [3].

Conclusion

This case highlights an unusual association of type II diastematomyelia with a dorsal neurenteric cyst and complex spinal dysraphism. MRI provided critical information for diagnosis and surgical planning by delineating both the cystic lesion and the split cord malformation. The coexistence of these anomalies further supports the unified embryogenesis theory, which attributes their development to a single error during the third week of gestation. Early recognition and timely surgical treatment are essential to prevent progressive neurological deterioration and to improve functional outcomes in pediatric patients with complex spinal dysraphism.

Conflict of Interest

The authors declare that they have no conflict of interest.

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