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Clinical case

Multiple Type II Diastematomyelia in an Adolescent: A Rare Case Report

Diastématomyélie multiple de type II chez un adolescent : à propos d'un cas rare

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Résumé

La diastématomyélie est une malformation congénitale rare du tube neural, caractérisée par une division sagittale de la moelle épinière. Ce rapport décrit le cas d'un adolescent de 13 ans présentant une diastématomyélie multiple de type II, diagnostiquée suite à l'apparition progressive d'une tétraparésie, en l'absence de stigmates cutanés ou vertébraux typiques. L'imagerie par résonance magnétique (IRM) a révélé une duplication de la moelle épinière sur plusieurs niveaux thoraciques (T1–T4/T5 et T6–T8). Le tableau clinique atypique, associant retard psychomoteur, difficultés scolaires et déficit neurologique progressif, souligne la diversité des présentations de cette entité. Une prise en charge chirurgicale précoce a été indiquée pour prévenir l'aggravation des déficits. Ce cas illustre l'importance d'évoquer le diagnostic face à des symptômes neurologiques inexpliqués, même en l'absence de signes cutanés, et confirme le rôle central de l'IRM dans le bilan diagnostique.

Mots-clés : Diastématomyélie, Malformation spinale, IRM, Chirurgie.

Abstract

Diastematomyelia is a rare congenital neural tube defect characterized by sagittal splitting of the spinal cord. This report describes the case of a 13-year-old adolescent with multiple type II diastematomyelia, diagnosed following progressive tetraparesis in the absence of typical cutaneous or vertebral stigmata. Magnetic resonance imaging (MRI) revealed spinal cord duplication across multiple thoracic segments (T1–T4/T5 and T6–T8). The atypical clinical presentation, including psychomotor delay, learning difficulties, and progressive neurological deficits, highlights the diverse manifestations of this condition. Early surgical intervention was recommended to prevent further neurological deterioration. This case underscores the importance of considering the diagnosis in the presence of unexplained neurological symptoms, even without cutaneous signs, and confirms the central role of MRI in the diagnostic workup.

Keywords: Diastematomyelia, Spinal malformation, MRI, Surgery.

Introduction

Diastematomyelia is a rare congenital anomaly of the neural tube, accounting for approximately 3.8% of all spinal dysraphisms [1]. It is defined as a closed form of spinal dysraphism characterized by the presence of a median fibrous or bony spur traversing the spinal canal in the sagittal plane. This septum divides the spinal cord into two distinct hemicords, usually asymmetrical, each with its own nerve roots and meningeal coverings. The lesion most commonly occurs at the lumbar or thoracolumbar level [2].

Prenatal diagnosis is sometimes possible during routine morphological ultrasound, but it remains challenging due to the rarity and variable presentation of the malformation. Magnetic resonance imaging (MRI) is the investigation of choice for confirming the diagnosis, as it provides detailed analysis of the spinal anatomy, the relationships of the cleft with adjacent vertebral structures, and facilitates the topographic and morphological classification of the lesion [2].

When the diagnosis is not established before birth, a meticulous clinical examination is essential. The systematic search for lumbosacral cutaneous stigmata (hypertrichosis, naevus, subcutaneous lipoma, sacral dimple, dermal sinus, etc.) is crucial, as these signs may be the only indicators of an underlying dysraphism [3]. Early identification allows appropriate neurosurgical management, thereby reducing the risk of permanent neurological, urological or orthopaedic sequelae.

We describe a case of multiple type II diastematomyelia in a 13-year-old adolescent, diagnosed following progressive tetraparesis, without cutaneous or vertebral stigmata.

Clinical case

- *Background*

The patient was a 13-year-old adolescent, born at term following an uncomplicated delivery. His psychomotor development was delayed, with independent walking achieved at around 18 months of age, and early learning difficulties that eventually led to discontinuation of schooling at the age of 11 years.

Medically, the patient had a history of two episodes of generalized tonic–clonic seizures at the age of six, which had been treated irregularly with carbamazepine syrup. A family history of epilepsy was also reported in the father.

- *History of the Present Illness*

The first neurological symptoms appeared insidiously at the age of 11 years, characterized by progressive weakness of the cervical muscles and intermittent headaches. Over the following year, the patient developed motor weakness in the lower limbs, resulting in repeated falls without loss of consciousness. The condition subsequently progressed with the onset of weakness in the upper limbs, accompanied by resting tremors and increasing difficulty in grasping and holding objects.

During the six months preceding admission, the motor deficit worsened significantly, leading to complete loss of ambulation and near-total dependence for daily activities. This progression occurred in a context of gradual weight loss, without fever or other associated infectious symptoms.

- *Clinical Examination*

Upon admission on 13 January 2025, at the Mother and Child Hospital of Bingerville, the patient presented with marked general deterioration and signs of severe malnutrition (body mass index estimated at 12 kg/m^2). Neurological examination revealed aphasia and tetraparesis, with muscle strength graded at 3/5 in the lower limbs and 4/5 in the upper limbs. Deep tendon reflexes were brisk and symmetrical, with bilateral Babinski signs. Diffuse muscle wasting was noted, along with resting tremors affecting the upper limbs.

Physical examination revealed a congenital malformation of the right foot, corresponding to syndactyly of the second toe. Cardiopulmonary, abdominal, and musculoskeletal examinations revealed no other abnormalities.

- *Paraclinical Investigations*

Spinal magnetic resonance imaging (MRI) revealed a duplication of the spinal cord at multiple levels, extending from T1 to T4/T5 and from T6 to T8, consistent with multiple type II diastematomyelia (figures 1, 2, and 3).

The electroencephalogram (EEG) showed an asymmetric sleep pattern, suggesting a possible left temporal encephalopathy.

Biological investigations demonstrated a moderate inflammatory syndrome (C-reactive protein: 11.46 mg/L), microcytic anaemia (haemoglobin: 10.8 g/dL), severe vitamin D deficiency (12.2 µg/L), and hypophosphataemia (1.18 mmol/L). Serum electrolyte levels, renal function tests, and endocrine profile were within normal limits.

- *Management*

Upon admission, multidisciplinary management began, including electrolyte stabilization and progressive nutritional support under close supervision. Given the progressive neurological deficits, early surgical intervention was deemed necessary to prevent further deterioration, in accordance with published recommendations for symptomatic type II diastematomyelia. After stabilization of the general condition, the patient was transferred to a specialized neurosurgical unit for appropriate surgical management.

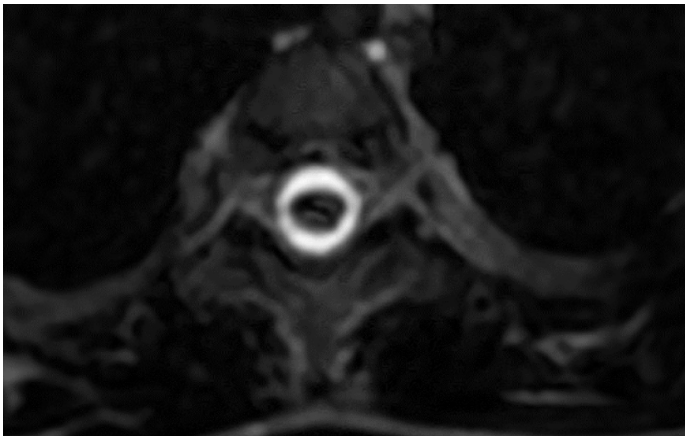

Figure 1: Axial MRI view at the level of the T2 vertebra showing duplication of the spinal cord consistent with type II diastematomyelia.

Figure 2: Axial MRI view at the level of the T4 vertebra showing a second plane of spinal cord duplication.

Figure 3: Sagittal MRI view of the cervical, thoracic, and lumbar spine showing multiple segments of spinal cord duplication.

Discussion

Spinal cord malformations are among the most common congenital anomalies. They are classified into two main groups: open dysraphisms, represented primarily by myelomeningocele, and occult dysraphisms, a heterogeneous entity encompassing several forms, including diastematomyelia, which is considered one of the most complex types [3].

- *Aetiopathogenesis*

The aetiology of diastematomyelia remains uncertain but is thought to be multifactorial. Several hypotheses have been proposed to explain its development, involving a possible genetic predisposition, certain maternal conditions during pregnancy, and foetal exposure to teratogenic agents or specific drugs that may interfere with neural tube closure [4].

- *Classification and Anatomical Characteristics*

Diastematomyelia is classically divided into two types of approximately equal frequency [5].

Type I refers to a duplication of the spinal cord with two hemicords separated by a fibrous septum, both enclosed within a single dural and arachnoid sac. This form is generally mildly symptomatic or occasionally asymptomatic.

Type II is characterised by a duplication of the spinal cord with two distinct dural sacs, separated by a bony or cartilaginous spur anchored to a vertebral body. This form is more frequently symptomatic [5].

The most common site of involvement is the thoracolumbar region. Scoliosis is present in approximately 60–70% of cases. Other neural tube malformations may coexist, such as syringomyelia, myelomeningocele, or neurenteric cysts. A female predominance has also been reported [2].

- *Clinical Manifestations*

The clinical presentation of diastematomyelia is polymorphic and depends on the type, the level of the lesion, and the extent of the spinal cord division. It may include a pyramidal syndrome (paraparesis or tetraparesis), orthopaedic deformities such as scoliosis or foot abnormalities (notably clubfoot), sphincter disturbances in the form of urinary retention or incontinence, and lumbosacral cutaneous stigmata (dimple, tuft of hair, naevus, or subcutaneous lipoma) [2,8].

According to a Russian study published in 2018, approximately 15% of children with diastematomyelia were asymptomatic, whereas 64.7% presented with paraparesis [7]. A Chinese study also reported a higher frequency of cutaneous stigmata and spinal malformations in type I forms compared with type II [9].

In our observation, the diagnosis was delayed due to the absence of classic cutaneous stigmata—an occurrence noted in up to 15% of cases according to Vissarionov et al. [7] and similarly described by Cheng et al. [9]. The atypical presentation, with cognitive difficulties preceding progressive muscle weakness and tetraparesis, underscores the clinical diversity of diastematomyelia.

- *Radiological Diagnosis*

The diagnosis of diastematomyelia may be suspected antenatally through morphological ultrasound and confirmed by the measurement of biomarkers such as alpha-fetoprotein and acetylcholinesterase [3,10]. A case reported in Spain in 2023 described a diagnosis established at 24 weeks of gestation, with a positive amniotic acetylcholinesterase test [11].

After birth, magnetic resonance imaging (MRI) and computed tomography (CT) are the reference investigations. MRI allows precise determination of the level of spinal cord splitting, its single or multiple nature, and its anatomical relationship with the conus medullaris and surrounding structures [9]. CT, particularly with three-dimensional reconstructions, provides better visualisation of the bony or cartilaginous spur and associated vertebral anomalies, facilitating assessment of the risk of complications such as severe scoliosis or syringomyelia [9].

In our case, spinal MRI revealed multiple type II diastematomyelia extending across several thoracic segments.

- *Therapeutic Management*

The treatment strategy for diastematomyelia, particularly for asymptomatic patients, continues

to be debated. However, for symptomatic type II cases with progressive neurological impairment, most authors recommend early surgical intervention to prevent further deterioration [12]. Some authors advocate systematic prophylactic surgery, even in the absence of clinical symptoms, to prevent secondary neurological deterioration [12,13]. Others adopt a more selective approach, reserving surgical intervention for symptomatic patients or for those with associated malformations, notably orthopaedic or urological anomalies [1].

Postoperative outcomes are generally more favourable in type I cases, where spinal cord separation is less complex. In contrast, type II forms often show an unfavourable evolution in the absence of surgical intervention, supporting the need for early and appropriately targeted management [9].

- *Contribution of the Reported Case*

Our observation highlights the diagnostic complexity of atypical forms of diastematomyelia, particularly in the absence of cutaneous stigmata. It underscores the need to maintain a high level of clinical suspicion when faced with any unexplained progressive neurological symptoms, in order to facilitate early diagnosis and ensure appropriate multidisciplinary management.

Conclusion

Diastematomyelia is a rare spinal malformation, generally diagnosed during the antenatal period or in childhood, usually in the presence of suggestive neurological manifestations. The absence of typical cutaneous or orthopaedic stigmata may, however, delay its identification, emphasizing the critical value of magnetic resonance imaging, which remains the gold standard for diagnosis. Management should be individualized according to the clinical presentation and neurological evolution. Increased vigilance and early use of imaging are essential to reduce the risk of disabling functional sequelae. Finally, further studies are needed to harmonies diagnostic and therapeutic protocols and to optimize the prognosis of affected patients.

- *Ethical Approval and Institutional Authorization*

Ethical approval for publication of this case was granted by the Medical and Scientific Directorate of the Mother and Child Hospital of Bingerville, Côte d'Ivoire. Institutional authorization for the use of anonymized clinical data and images was obtained prior to submission.

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Conflict of interest : None

References

[1] Ozturk E, Sonmez G, Mutlu H, Cakir B, Gurel S, Basekim CC, et al. Split-cord malformation and accompanying anomalies. J Neuroradiol. 2008;35(3):150-6.

[2] Achour A, Abdelaali M, Hmida B, Maatouk M, Mnari W, Saad J, et al. Imagerie des diastématomyélies : étude d'une série de 27 cas. Feuillets Radiol. 2019;59(1):6-13. Disponible sur : https://sfip-radiopediatrie.org/wp-content/uploads/2020/02/2019LILLE_CO06-imagerie-des-diastematomyelies.pdf

[3] Zerah M, Kulkarni AV. Spinal cord malformations. Handb Clin Neurol. 2013;112:975-91.

[4] Campbell LR, Dayton DH, Sohal GS. Neural tube

defects: a review of human and animal studies on the etiology of neural tube defects. Teratology. 1986;34(2):171-87.

[5] RADEOS. Diastématomyélie [Internet]. 2020 [cité 25 août 2025]. Disponible sur : https://www.radeos.org/maladie/fiche-diastematomyelie_248.html

[6] Maebe H, Viaene A, De Muynck M. Diastematomyelia and late onset presentation: a case report of a 72-year-old woman. Eur J Phys Rehabil Med. 2018;54(4):618-21.

[7] Vissarionov SV, Krutelev NA, Snischuk VP, Alam M, Kravchenko AP, Zheng YP, et al. Diagnosis and treatment of diastematomyelia in children: a prospective cohort study. Spinal Cord Ser Cases. 2018;4:109.

[8] Eid K, Hochberg J, Saunders DE. Skin abnormalities of the back in diastematomyelia. Plast Reconstr Surg. 1979;63(4):534-9.

[9] Cheng B, Li FT, Lin L. Diastematomyelia: a retrospective review of 138 patients. J Bone Joint Surg Br. 2012;94(3):365-72.

[10] Arbelo-Pérez P, de Ganzo-Suárez T, de Luis-Escudero JF. Prenatal diagnosis of diastematomyelia: a case report and literature review. Reprod Sci. 2023;30(12):3563-7.

[11] Bothara A, Srivastava J, Pillai AA, Gadgil N, Relia R, Jain N, et al. A Case of Type 2 Diastematomyelia With Spina Bifida in a Pediatric Patient. Cureus. 2024;16(2):e54923. doi:10.7759/cureus.54923.

[12] Mahapatra A, Gupta D. Split cord malformations: a clinical study of 254 patients and a proposal for a new clinical-imaging classification. J Neurosurg. 2005;103(6):531-6.

[13] Ersahin Y, Mutluer S, Kocaman S, Demirtas E. Split spinal cord malformations in children. J Neurosurg. 1998;88(1):57-65.

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