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Incidentally Discovered Diastematomyelia in Adults: About Two Case Reports

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Abstract | **Case Report**

Diastematomyelia is a rare congenital abnormality of the spinal cord, generally diagnosed in children,its discovery in adults is very rare. In this paper we report two additional cases to the literature, give a brief overview of the clinical features, and especially shed light on the radiological aspects and role of imaging in this diagnosis.

Keywords: Diastematomyelia – incidental imaging findings - split cord malformation.

INTRODUCTION

Diastematomyelia is a rare form of spinal dysraphism that consists of a closed division of the spinal cord. The hemicords may be situated in their own dural sac (type I diastematomyelia) or in a single dural sac (type II diastematomyelia) [1, 3]. It can be either isolated or accompanied by other spinal cord malformations such as myelomeningocele, syringomyelia, low inserted spinal cord, and in some cases to a neuroenteric cyst, it is often associated with multiple vertebral abnormalities responsible for scoliotic or cypho-scoliotic deformities [1-4].

Usually revealed in children, its discovery in adults is far more uncommon [2, 5]. We report two cases of adult diastematomyelia that were discovered respectively in the course of a malformative scoliosis assessment and following a chronic lower back and radicular pain.

CASE REPORT

Case 1

A 31-year-old patient who was initially admitted for the management of an evolving malformative scoliosis and who benefited from a spinal CT scan that allowed measurement of the angles of curvature objectifying a significant dorso-lumbar scoliosis with double convexity associated with multiples defects of the posterior arches of the lumbosacral vertebrae (Figure 1). The patient further benefited from a medullary MRI that showed a low inserted spinal cord with a terminal cone and lipoma at the level of the 4th and 5th lumbar vertebrae, and revealed a type I diastematomyelia with longitudinal splitting into two hemi-cords at the level of 12th thoracic vertebrae with a common dural sac and separated by a fibrous septum (Figure 2).

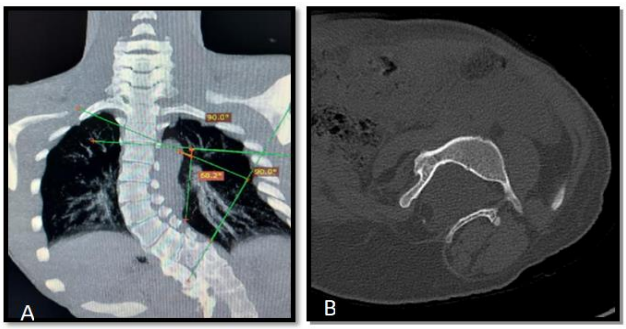

Figure 1: CT scan, A): coronal MIP reconstruction objectifying a significant dorso-lumbar scoliosis with double convexity. B): Axial view showing a defect of the posterior arches of lumbosacral vertebrae

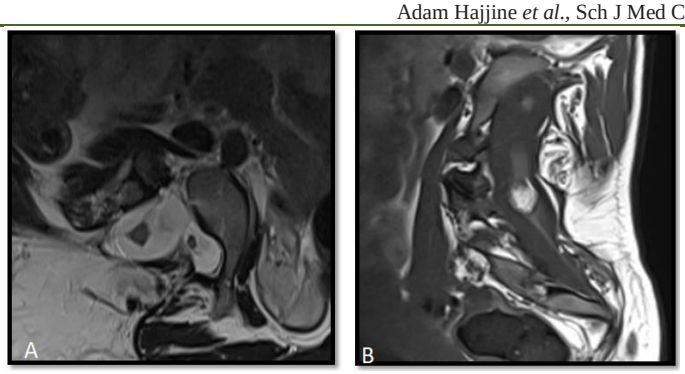

Figure 2: Medullary MRI, A): Axial T2 WI at the level of the 12th thoracic vertebrae revealing a type I diastematomyelia with longitudinal splitting into two hemi-cords with a common dural sac and a fibrous septum. B): Sagittal T1 WI showing a low inserted spinal cord with a terminal cone lipoma

Case 2

A 46-year-old patient, known for a previous lumbar laminoplasty, who presented with persistent back pain and bilateral radicular pain, an MRI of the lumbosacral spine showed a low spinal cord with a terminal cone located at the level of L5 and individualization of two hemi-cords in two dural sacs separated by a bony septum (visualized on a CT scan) suggesting a type II diastematomyelia (Figure 3). A lack of fusion of the posterior arches of the lumbosacral vertebrae was also found. In addition, we find a Protrusion of the L1-L2 intervertebral disc with median hernia reducing the antero-posterior diameter of the lumbar canal with compression of the two hemi-cords. We also note an asymmetrical distribution of the roots of the caudaequina which are adherent to each other and to the theca, suggesting an adhesive arachnoiditis, (Figure 4).

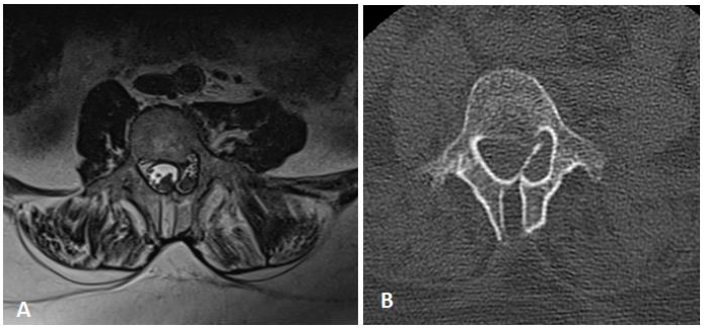

Figure 3: A) Medullary MRI on axial T2WI showing two hemi-cords in two dural sacs separated by a bony septum that is visualized on a CT scan (B) Suggesting a type II diastematomyelia

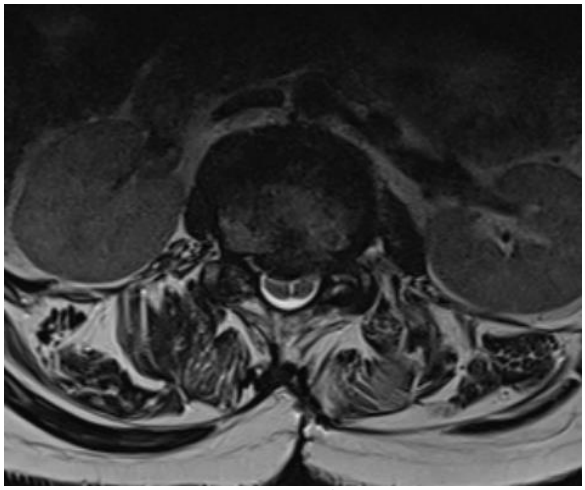

Figure 4: Medullary MRI with axial T2WI view showing a protrusion of the L1-L2 intervertebral disc with median hernia reducing the antero-posterior diameter of the lumbar canal with compression of the two hemi-cords

DISCUSSION

Diastematomyelia is a very rare congenital malformation of the spinal cord that usually occurs between the third and fourth week of pregnancy and is characterized by extensive longitudinal division of the vertebral canal and its contents with a double spine appearance. Various etiopathogenic theories have

attempted to explain its origin; It may result from an abnormal closure of the neural gutter, a duplication of the medulla, a persistence of an abnormal neural canal, or from a hydrodynamic process. Two types of diastematomyelia are described according to the relationship between the hemi-cords and their arachnoid and dural envelopes; type I: Common arachnoid and dural envelopes with the presence of simple fibrous adhesions and absence of bone spur, It is often isolated and rarely symptomatic and Type II: double arachnoid and dural envelopes separated by a central spur of bone or cartilage, these forms are symptomatic [1, 2, 4].

The topography can involve every level of the spine but is particularly predominant in the lumbar region [1]. Diastematomyelia is discovered in the prenatal period thanks to the advancement of ultrasound [1, 4-6]. Their discovery in adulthood is quite rare. Asymptomatic patients are incidentally diagnosed during routine radiological examinations [2], or during direct trauma to the spine; in a fall, during stretching after physical exercise, during prolonged standing, or during a long vehicle ride, in the other hand symptomatic forms can manifest by neurological signs such as spinal and/or radicular pain, walking disorders, amyotrophy, paresis of the lower extremities, sexual and sphincter disorders, and flaccid paraplegia, orthopedic signs such as scoliosis and foot deformities and cutaneous signs such as clumps of hair, lumbosacral mass, café au lait spots, angiomas and lipomas. Other spinal and spinal cord abnormalities are often associated with diastematomyelia including myelomeningocele, syringomyelia, low cord, attached cord, neurenteric cyst, and vertebral malformations [1-4].

Ultrasound provides a prenatal diagnosis by demonstrating an enlargement of the medullary canal, curvature anomalies of the spine and the presence of two hemi-cords. Standard radiographs can show vertebral anomalies including spina bifida, scoliosis, butterfly wing vertebrae, vertebral block, interpedicular widening and disc dysplasia, the presence of a bone spur in the vertebral canal is almost pathognomonic of diastematomyelia but inconstant [2, 4, 6]. CT allows a better study of the bone structures and subarachnoid spaces. It provides information on the fibrous, osteofibrous, bone spur, and the medullary division, its extent, size and shape of the two hemicords, the aspect of the medullary cone, and allows the assessment of associated spinal abnormalities [1, 3]. MRI offers a much better analysis of intracanal structures, by analyzing the two spinal cords, their size, the location of the division, its single or multiple nature, the presence of ectopy of the medullary cone and possible associated intracanal malformations [1, 2, 4, 5].

Surgical management is essential in the event of gradual neurological deterioration, especially when there is a bone septum or an associated malformative abnormalities, requiring surgical intervention, it consist of the removal of the osteo-fibrous septum, a laminectomy next to the malformative defect, surgical removal of adhesions, and the section of the terminal filum in the presence of a low attached cord, in cases where the patient is clinically stable, without any progression of neurological disorders, some authors suggest a prophylactic intervention. The absence of the bony spur on imaging and the general absence of neurological symptoms may lead to therapeutic restraint [1-3].

CONCLUSION

Spinal dysraphism is a broad spectrum of spinal malformative abnormalities, diagnosed prenatally or in childhood, but rarely in adulthood. MRI remains the technique of choice for the diagnosis of spinal malformations.

Disclosure of Interest: The authors declare that they have n conflicts of interest concerning this article.

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