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Case Report

UNUSUAL DISCOVERY OF DIATEMATOMYELIA IN AN ADULT: A CASE REPORT

***I Assoumane, N Lagha, A Khalifa, M Al Zekri, T Benbouzid, A Sidi Said and A Morsli**
Department of neurosurgery, CHU Bab El Oued Algiers, Algeria
**Author for Correspondence: as_ibrah2006@yahoo.fr*

ABSTRACT
Introduction: Diastematomyelia (DM) or split cord malformation (SCM) is a rare form of spinal dysraphism, in which the spinal cord or conus is split in the sagittal plane, over a number of segments, into two separate parts or hemi cords. It can be associated with many other malformations. The manifestation in adulthood is very rarely reported in the literature. We report a case of a 37 years old lady. *Case presentation*: Madame A M is a 37 years old lady who complained of walking difficulties since 10 years which worsened in the last 3 months. Clinical examination at admission revealed pyramidal syndrome in the lower limbs and urinary disturbance. A lumbar spine CT Scan and MRI revealed a bony septum which split the spinal cord at the level of L2 associated with a low lying cord attached to a lipoma and a neuroenteric cyst. We operated the patient, after resection of the bony septum we released the spinal cord. The follow up included objective clinical assessment. We compared our case with other cases reported in the world literature. *Conclusion*: The Split Cord Malformations cause traction on the spinal cord, which causes ischemia as well the local trauma of the cord. The diagnosis is confirmed by CT scan and MRI. The aim of surgery is the decompression of neural elements, the removal of bony septum and the spinal cord release.

Keywords: *Diastematomyelia, Bony Septum, Dysraphism*

INTRODUCTION

Diastematomyelia (DM) or split cord malformation (SCM) is a rare form of spinal dysraphism, in which the spinal cord or conus is split in the sagittal plane, over a number of segments, into two separate parts or hemi cords (Neville *et al.*, 1994). It can be associated to many other malformations. The manifestation in adult hood is very rarely reported in the literature.

CASE

We reported here a case of a 37 years old lady without past medical or surgical history complaining with walking impairment since 10 years worsening in the last 3 months. Clinical examination at admission revealed a pyramidal syndrome in lower limbs with paraparesis 4/5, bilaterally positive Babinski's sign, urinary disturbance. There were no local stigmas on the lumbo sacral region and no history of recent traumatism. The lumbar CT scan showed a bony septum which split the spinal cord at L2 level (figure1).

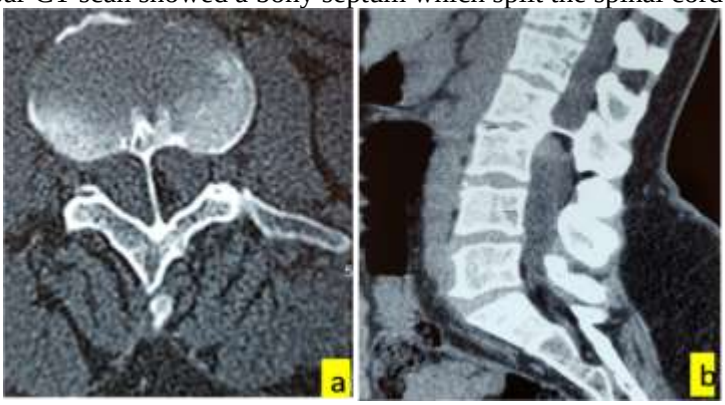

Figure 1: Bone septum at L2 level: a. axial b. sagittal

The lumbar spine MRI confirmed the diagnosis of diastematomyelia showing two hemi cords housed in a single dural tube separated by a median septum at L2 level (figure 2).

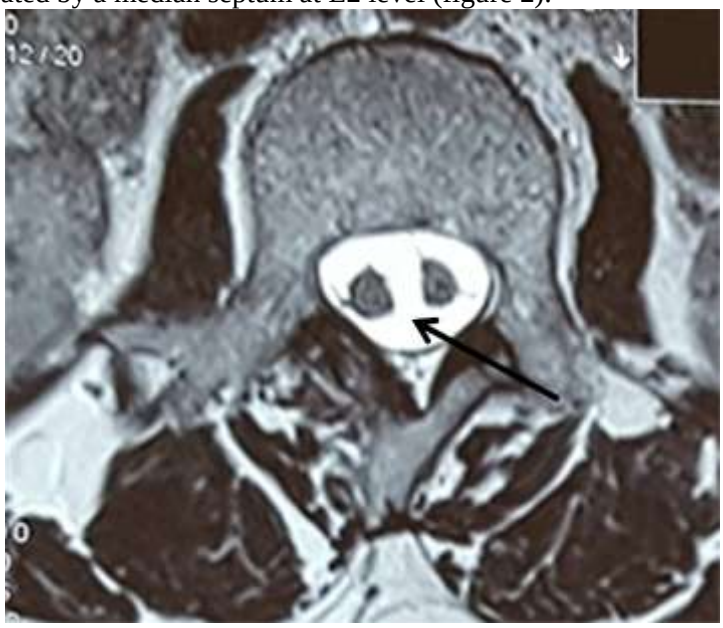

Figure 2: Axial MRI showing two hemi cords housed in a single dural tube separated by a median septum (black arrow)

It is SCM type II of Pang's classification. The diastematomyelia was associated to a lipoma and a neuroenteric cyst (figure 3) with a low lying cord attached by a fatty filum terminale.

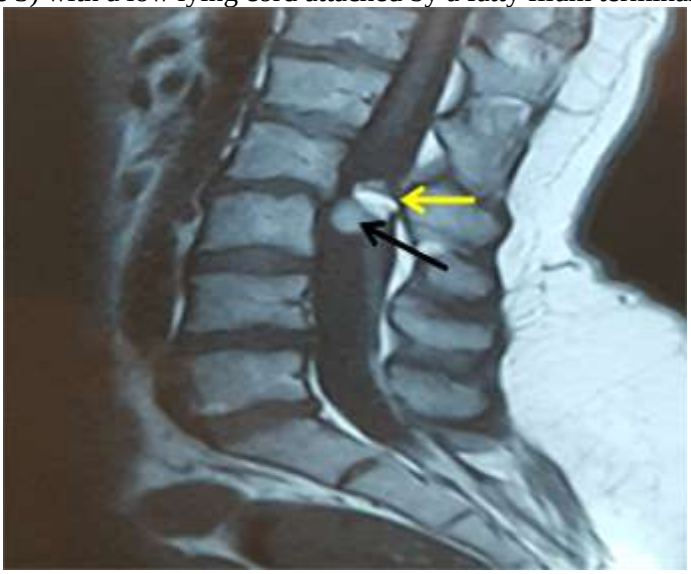

Figure 3: Sagital MRI showing a lipoma (yellow arrow) and a neuroenteric cyst (black arrow) at the conus level and a low lying cord attached by a fatty filum terminale.

We operated the patient under general anesthesia in prone position. We performed a midline incision followed by laminectomy (L1-L2-L3) one level under one level above the spur. Under operative microscope we used a diamond drill to remove the median septum which is fibrous and bony**.** We opened

the dura in a linear fashion with a curve encircling the spur which was drilled until the body of L1. The cyst was evacuated but we did not remove the lipoma because it was attached on the cauda equina (figure 4).

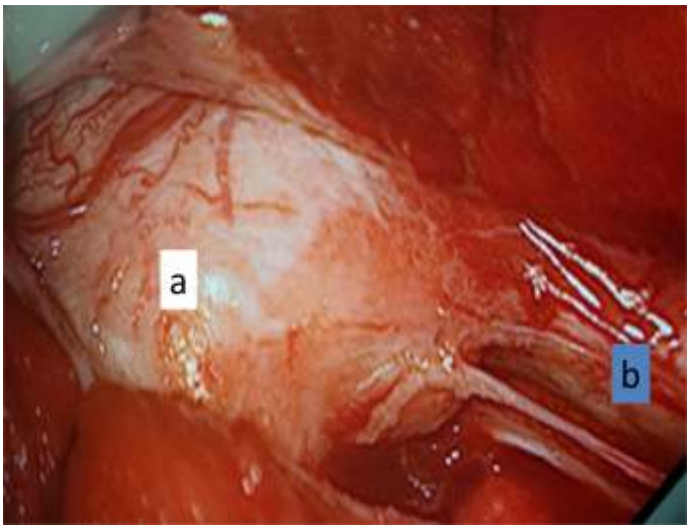

Figure 4: Per operative image showing the lipoma (a) and the fatty filum terminale (b)

A watertight duroplasty was performed using 4-0 vicryl suture. On the second step we released the spinal cord by section of the filum terminale. The post operative outcome was good, with the improvement of bladder disturbance.

DISCUSSION

The classification of Pang (1992) recommends the term split cord malformation (SCM) for all double spinal cords: Type I SCM consists of two hemi cords; each contained within its own dural tube and separated by a dura-sheathed rigidosseo cartilaginous median septum. Type II SCM consists of two hemi cords housed in a single dural tube separated by a non rigid, fibrous median septum (Pang *et al.,* 1992). The most widely accepted theory about embryogenesis of SCM was originally proposed by Bremer in 1952 taking into account a dorsal intestinal fistula. From the archenteron that gives rise to the gut, a diverticulum develops that, upon expansion, separates the notochord and the neural plate into two parts. If this diverticulum opens at the skin level, it gives rise to the dorsal enteric fistula that is an open form of split notochord syndrome. If the endodermal elements disappear totally, there remains a fibrous or osseous septum between the two hemi cords (Bremer *et al.,* 1952). The key to understand these malformations lies in properly appreciating the embryonic relationship between the developing neural tube and the endoderm during early development. This complex defect occurs during the 4th week of embryological development. Both types are thought to be caused by an abnormal, persistent neurenteric canal between the yolk sac and amnions and thus enables contact between the ectoderm and endoderm within the canal. This abnormal fistula splits later the neural canal and the notochord by forming an endomesenchymal tract. The persistence of parts of the tract, the entrapment of different structures within it, or both, explains the subsequent formation of associated malformations. For instance, "the endodermal remnants predispose to the formation of neurenteric cysts and intestinal duplication; the majority of SCMs has low-lying conus and may have additional tethering lesions
(Carmen *et al.,* 2011; Martin *et al.,* 2007). There are many theories trying to explain the symptom development in diastematomyelia. The neural damage could result from sustained traction by the bone spur, fibrous bands and adhesions, or by other abnormalities, such as tight filum terminale (Neville *et al.,* 1994). Many authors have identified definite events or incidents that precipitated the development of symptoms in adult diastematomyelia, such as traumatism, spinal canal stenosis or spondylosis (Neville *et*

al., 1994; Richard et al 1987; Goina et al 2004; Carmen et al 2011) but it is not the case of our patient. Symptomatic cases often present with progressive signs and symptoms and are nonspecific. The common presentations are lumbar radiculopathy and myelopathy. Other presenting symptoms include sensory and motor deficits, skeletal and foot deformities, and bladder and bowel disturbances (Iraj *et al.,* 2016). Dorsolumbar and lumbar regions were the most common sites (Sumit Sinha et al; Carmen *et al.,* 2011). The diagnosis has been more frequent since the advent of newer imagistic methods. MRI showed soft tissue lesions and the computed tomography is helpful in evaluating the nature of the bony septum. Diastematomyelia can be associated with myelomeningocele, syringomyelia, low lying cord, lipomas or neuro enteric cyst, skin stigmata, Skeletal anomalies (Pang et al ,1992; Sumit Sinha *et al.,* 2005;). Our case is also associated with a lipoma, neuroenteric cyst and low lying cord. Early surgery is the key issue in the management of SCM because once a neurological deficit appears there is a lower chance of complete recovery. Some authors suggest that surgery should be done in asymptomatic patients to prevent future neurological deterioration (Pang *et al.,* 1992; Sumit Sinha *et al.,* 2005; Borkar *et al.,* 2012; Yusuf *et al.,* 2013).The post operative complications consist of cerebrospinal fluid (CSF) leakage, transient unilateral lower limb paresis, paraparesis, wound infection, urinary retention, subcutaneous CSF collection, and neuropathic pain in the lower limbs (Yusuf *et al.,* 2013).

CONCLUSION

The Split Cord Malformations cause traction on the spinal cord, which causes ischemia as well the local trauma of the cord. The diagnostic is confirmed by MRI scans. The aim of surgery is the decompression of neural elements, removal of bony septum.

Conflict of interest*: non*

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