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Tethered spinal cord syndrome with lumbar segmental stenosis treated with XLIF

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1. Introduction

Tethered cord syndrome (TCS) is a congenital malformation due to errors during embryonic development of neural axis, consisting in attachment of spinal cord to the base of vertebral canal, associated with various forms of dysraphism [1–3]. Most cases are diagnosed in childhood; rarely patients progress to adulthood in absence of neurological deficits. Symptomatic TCS in adults causes insidious and progressive lower limbs weakness and urinary disturbances. Onset or worsening of symptoms in adulthood may be related to vertebral trauma or, rarely, degenerative spine disease (stenosis and/or disc herniation) [4]. In presence of low-lying cord, lumbar disc prolapse or canal stenosis may produce spinal cord compression and myelopathy [5].

Reporting a case of association between TCS and lumbar spine degenerative stenosis causing lumbar myelopathy, a literature review has been performed. Clinical aspects and possible surgical approaches are considered.

Differently to previous surgical solutions consisting in posterior decompression of low-lying spinal cord [4,6,7], combined or not with fixation, we performed lumbar indirect decompression and arthrodesis by XLIF approach (extreme lateral interbody fusion).

2. Case report

A 69 year old man, still working as veterinary in private practice, suffering from spina bifida with sacral meningocele received surgical repair as a newborn. All life long, as referred, he walked with slight spastic paraparesis and presented occasionally sphincter disturbances. He had autonomous social and working life and never did a lumbar CT or MRI.

He was admitted in our Department with a 6-month history of severe low back pain and progressive worsening of preexisting symptoms: 1. paraparesis 2. bladder and bowel dysfunction (urinating independently with retention and constipation). Neurological examination at admission showed neurogenic claudication, spastic paraparesis with weakness in lower limbs, increased deep tendon reflexes (except patellars), sphincter retention, and sensory disturbances with upper level at S1. Independent ambulation was impossible to sustain.

Lumbar MRI (Fig. 1) shows the spinal cord tethered to the previous surgical site of his sacral myelomeningocele associated with severe L3-L4 vertebral stenosis, resulting in spinal cord compression. Stenosis was caused both by ligamentum flavum hypertrophy and L3-L4 disc herniation and osteophytes.

Bone decompression and fixation by posterior approach was proposed but the patient was concerned of healing problems after posterior approach because of his heavy scar from the repair of the myelomeningocele as a baby. Our recommendation to explore the myelomeningocele and micro surgically detether at the dural level was refused by the patient. As possible alternative, he accepted an anterior-lateral approach to L3-L4, with indirect decompression and arthrodesis, minimizing instability by the XLIF technique as an alternative rationale.

In flexed lateral position, under continuous two-projections-X-rays control, an extreme lateral interbody approach for discectomy and arthrodesis (XLIF) at L3-L4 was performed. A stand-alone PEEK trapezoidal cage (Nuvasive, San Diego, CA) filled with gel of hydroxylapatite was inserted in the space, obtaining indirect spinal cord decompression and anterior stability by arthrodesis.

The postoperative period was uneventful and the patient referred improvement of preoperative neurological symptoms. Segmental L3-

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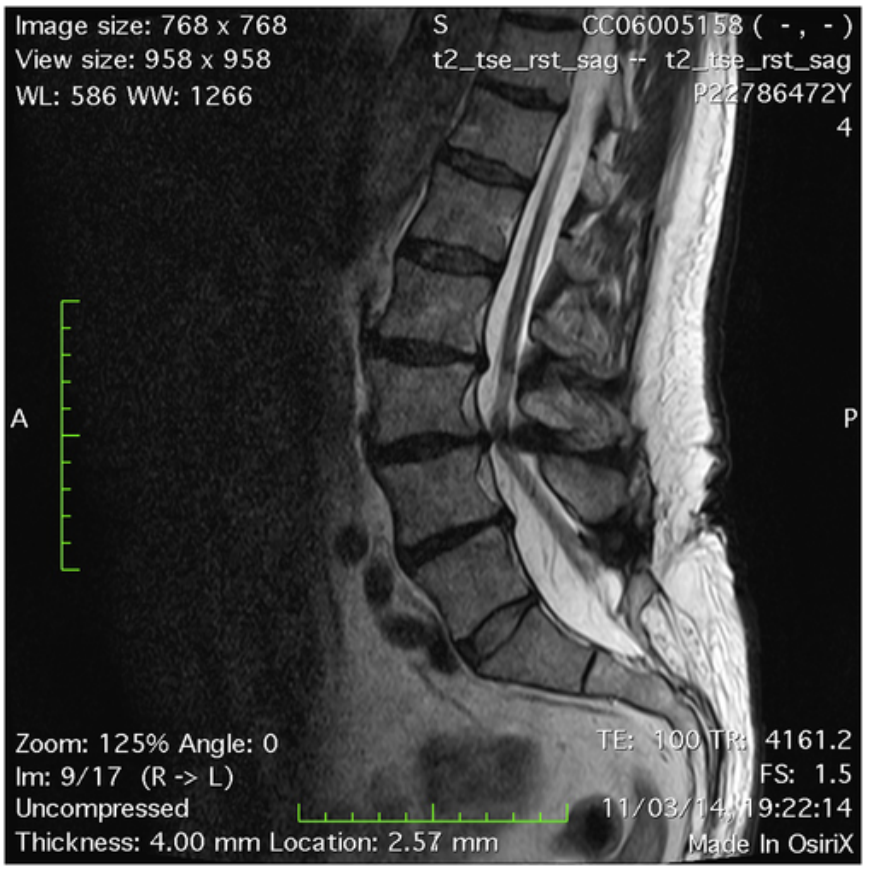

Fig. 1. Preoperative lumbar MRI T2W-images: sagittal view. Signs of previous surgical intervention at sacral level and low lying cord associated with severe canal stenosis at L3-L4, caused by ligamentum flavum hypertrophy, L3-L4 disc prolapse and osteophytes.

L4 CT scan (Fig. 2) and lumbar MRI showed correct positioning of the cage, with a residual slight vertebral canal stenosis. Six months after surgery, the preoperative paraparesis and bladder dysfunction were markedly improved (confirmed by urodynamic studies and the subjective impression of the patient) and patient returned to his previous activity of daily living function.

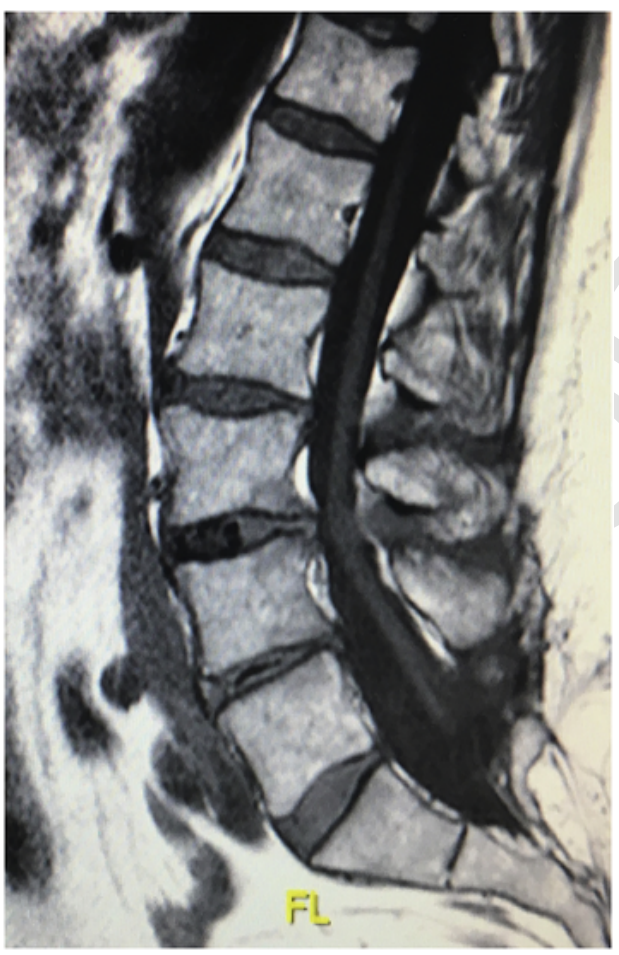

Fig. 2. Postoperative lumbar CT: axial view at L3-L4 level. Interbody PEEK cage containing osteoinductive material.

3. Discussion

TCS is a developmental abnormality resulting in fixation of spinal cord and nerves, associated to spinal musculoskeletal deformities and cutaneous signs [1]. The most common lesions that restrict physiological ascent of spinal cord are: thickened filum terminale, diastematomyelia, myelomeningoceles, intra/extra-dural lipomas, intradural neurenteric, dermoid, epidermoid, or arachnoid cyst, hamartoma, intradural fibrous adhesions and adhesive arachnoiditis [2,3,5,8].

Filum terminalis is an elastic structure that protects the cord against the stretch [3]. The mechanism of TCS in any of the above pathologies is supposed to be stretching and mechanical forces on the spinal cord and nerves, especially in flexion/extension neck movements, reducing regional blood flow with disturbance of oxidative metabolism [3,7].

Moderate entity of traction of spinal cord may remain asymptomatic and result in delayed neurological deficits in adult life, mainly because of repeated conus medullaris micro-traumas during spine bending on the stretched spinal cord [1,3,7].

In their literature review, Aufschnaiter et al. [8] showed that the most common symptoms of TCS in adults are: pain (81.6%), weakness (72.3%) and sensory disturbances (78.7%) of lower limbs, and urinary dysfunctions (68.5%). Only 1.5% of subjects don't experience any neurological deficit.

Two types of adult onset of TCS are described [2]: 1. progressive or sudden neurological deficits in asymptomatic patient, without previous diagnosis of TCS and 2. deterioration of neurological deficit in patient with TCS diagnosis in childhood (as our case).

Adult onset of TCS is still an underrecognized and probably overlooked phenomenon; the association with degenerative lumbar spine diseases is increasingly observed and in focus, however, still not established surgical and diagnostic criteria exist to select appropriate surgical candidates and intervention. Reviewing the international literature, we have found only six published cases (Table 1). All patients had a well known spinal dysraphism diagnosed in infancy, treated or not, and presented progressive worsening of their neurological deficits [4–6] or a new onset of symptoms. Two patients showed lumbar disc herniation [4,5] and other two spinal canal stenosis [6]. Our patient had combined discal-stenotic compression.

Except our case, every patient received surgical treatment with posterior spinal cord decompression. Surgical strategies were laminectomy, facet joints removal, discectomy, posterolateral interbody fusion (PLIF), laminectomy or pedicle subtraction osteotomy (PSO) without arthrodesis [4–6,9].

In our case we obtained lumbar canal “indirect” decompression and interbody arthrodesis by XLIF technique [10]. XLIF consists of lateral retroperitoneal trans-psoas approach to lateral side of vertebral body, firstly described by Ozgur et al. [10]. It is a minimally invasive procedure, safe, fast and reproducible [5] consisting of “indirect” decompression of lumbar canal by discectomy and osteophytectomy and subsequent arthrodesis for interbody fusion with PEEK cage containing osteoinductive materials. Main limitation of XLIF is feasibility at L5-S1 for interposition of iliac crest. XLIF restores disc height and maintains stability by inserting a large interbody implant, preserving ligamentous structures. In this way, the interbody implant contributes to reduction of possible radiculopathy. In opposition, PLIF (posterolateral interbody fusion) and laminectomy lead destruction of capsulo-ligamentous and osseous structures.

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Table 1
Spinal degenerative disease in adult tethered cord syndrome. Literature review.

Authors, years	Age, sex, spinal dysraphism	Neurological symptoms	Degenerative lumbar disease	Surgical treatment
Martinez-Lage et al. [6], 2001 - Case 1	40 y.o. M, SB occulta not treated	LBP + CN	L3-L4 lumbar canal stenosis	Bilateral L3-L4 laminectomy
Case 2	20 y.o. M, sacral MyMe treated in infancy	LBP radiated to LL + CN	L5-S1 lumbar canal stenosis	Bilateral L5 laminectomy
Kawamura et al. [9] (2010) – Case 1	56 y.o. M, SB	LBP + progressive paraparesis and BD	L3-L4 lumbar canal stenosis	Bilateral L3-L4 PSO
Case 2	60 y.o. F, SB	LBP + progressive paraparesis and BD	L3-L4 lumbar canal stenosis	Bilateral L3-L4 PSO
Srinivas et al. [7] (2012)	77 y.o. F, SB	LBP + progressive paraparesis	L2-L3 disc herniation	Bilateral L2-L3 laminectomy
Endo et al. [4] (2014)	43 y.o. M, lumbar SB previously treated	6-month gait dist + numbness in LL, hyperreflexia	L2-L3 disc herniation	Laminotomy, medial facetectomy discectomy and PLF with ABG
Present case	69 y.o. M, SB + sacral meningocele	6-month LBP, BD and worsening paraparesis	L3-L4 lumbar canal stenosis and lumbar disc herniation	L3-L4 XLIF (indirect decompression-fusion with PEEK cage)

Legend: y.o. = years old; M = male; F = female; SB = spina bifida; MyMe = myelomeningocele; LBP = low back pain; CN = claudicatio neurogena; LL = lower limbs; dist. = disturbances; PLF = posterior lateral fusion; ABG = autologous bone graft; BD = bladder dysfunction – PSO = pedicle subtraction osteotomy.

Lateral approach allows disc removal avoiding manipulation of neural structures, in opposition to PLIF, in which this maneuver is unavoidable.

As showed by Rodgers et al. [5], perioperative complications (intraoperative and out to 6 weeks after surgery) related to XLIF technique are uncommon: 6.2%: in a series of 600 patients. In–hospital surgery-related events were 1.5% (wound infections, vascular injuries, visceral injuries); 2.8% in–hospital medical events; 1% out-of-hospital surgery-related events; 0.8% out-of-hospital medical events; 0.7% transient neurological deficits [5].

4. Conclusions

Sudden or progressive onset or worsening of paraparesis with spastic gait, bladder dysfunction, and acute low back pain in patient with history of spinal dysraphism must be considered as possible spinal cord compression for low lying cord related to TCS. Lumbar MRI is mandatory for correct diagnosis. Surgical decompression should be performed as early as possible to ensure neurological recovery. As these scenarios are still unique, surgical approaches have to also consider patient's personal perspectives and concerns as shown in our case. XLIF approach is a surgical option to obtain spinal cord indirect decompression and lumbar interbody fusion.

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