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CASE REPORT

Combination of transverse myelitis and arachnoiditis in cauda equina syndrome of long-standing ankylosing spondylitis: MRI features and its role in clinical management

Howard Haw-Chang Lan · Der-Yuan Chen · Clayton Chi-Chang Chen · Joung-Liang Lan · Chia-Wei Hsieh

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Abstract The cauda equina syndrome (CES) is a rare neurological complication of ankylosing spondylitis (AS). Imaging diagnosis of CES in long-standing AS patients (CES-AS) using myelography, computed tomography (CT), and magnetic resonance imaging (MRI) were reported in the literature. They, however, demonstrate only the chronic abnormalities of CES-AS, i.e., dural ectasia, dorsal dural diverticula, and selective bone erosion at the posterior elements of the vertebrae. To our knowledge, imaging features of acute intradural inflammation in CES-AS were not described. We report a patient of CES-AS in whom MRI disclosed acute transverse myelitis and arachnoiditis along the lower spinal cord, and discuss the pathogenesis of CES-AS and the role of MRI in clinical management.

Keywords Ankylosing spondylitis · Arachnoiditis · Cauda equina syndrome · Dural diverticula · Magnetic resonance imaging · Transverse myelitis

H. H.-C. Lan · C. C.-C. Chen
Department of Radiology, Taichung Veterans General Hospital, Taichung, Taiwan, Republic of China

D.-Y. Chen · J.-L. Lan · C.-W. Hsieh
Division of Allergy, Immunology and Rheumatology, Taichung Veterans General Hospital, Taichung, Taiwan, Republic of China

H. H.-C. Lan · C. C.-C. Chen
Central Taiwan University of Science and Technology, Taichung, Taiwan, Republic of China

H. H.-C. Lan · C. C.-C. Chen
Hungkuang University, Taichung, Taiwan, Republic of China

D.-Y. Chen · J.-L. Lan
National Yang Ming University, Taipei, Taiwan, Republic of China

D.-Y. Chen · J.-L. Lan
National Chung Hsing University, Taichung, Taiwan, Republic of China

H. H.-C. Lan (✉)
160 Taichung-Kang Road, Sec. 3, Taichung 407 Taiwan, Republic of China
e-mail: lanhc@mail.vghtc.gov.tw

Introduction

The cauda equina syndrome (CES) is a rare neurological complication of ankylosing spondylitis (AS) [1]. It was first described in 1961 independently by Bowie and Glasgow [2]. Imaging diagnosis of CES in long-standing AS patients (CES-AS) using myelography, computed tomography (CT), and magnetic resonance imaging (MRI) were reported in the literature [3–8]. They, however, demonstrate only the chronic abnormalities of CES-AS, i.e., dural ectasia, dorsal dural diverticula, and selective bone erosion at the posterior elements of the vertebrae. Oh et al. diagnosed transverse myelitis in a patient with long-standing AS based on biochemical abnormalities in cerebrospinal fluid (CSF) when MRI was normal [9]. To our knowledge, imaging features of acute intradural inflammation in CES-AS were not described.

We report a patient of CES-AS in whom MRI disclosed acute transverse myelitis and arachnoiditis along the lower

spinal cord, and discuss the pathogenesis of CES-AS and the role of MRI in clinical management.

Case report

A 50-year-old man visited our outpatient department with the chief complaint of pain and numbness at his right inguinal region for 1 month. He was diagnosed as having AS since he was 20 years old. He had no other history of infection or spinal disease. His rheumatological symptoms were intermittent before the onset of CES. Bilateral numbness along the upper limbs, diagnosed as transverse myelitis, was noted once 15 years ago. Symptoms were relieved completely with methylpredinisolone pulse therapy (1 gm for 3 consecutive days). Bilateral numbness along the lower limbs had developed progressively over the past 1 1/2 years. Symptoms then progressed upward to the pelvis and perineum, and finally to the umbilical region. Progressive urinary incontinence was noted as well for 4 months.

Routine rheumatological and biochemical screening investigations were normal. Electromyography revealed partial motor unit loss and radiculopathy at the S_1 level. Neurological examinations showed decrease in muscle power of the bilateral lower limbs (3/5 by the Medical Research Council scale) and increase in deep tendon reflex of the right lower limb.

Plain radiographs revealed the typical abnormalities of advanced AS at the spine and sacroiliac joints. MRI of the spine showed dural ectasia with multiple dorsal dural

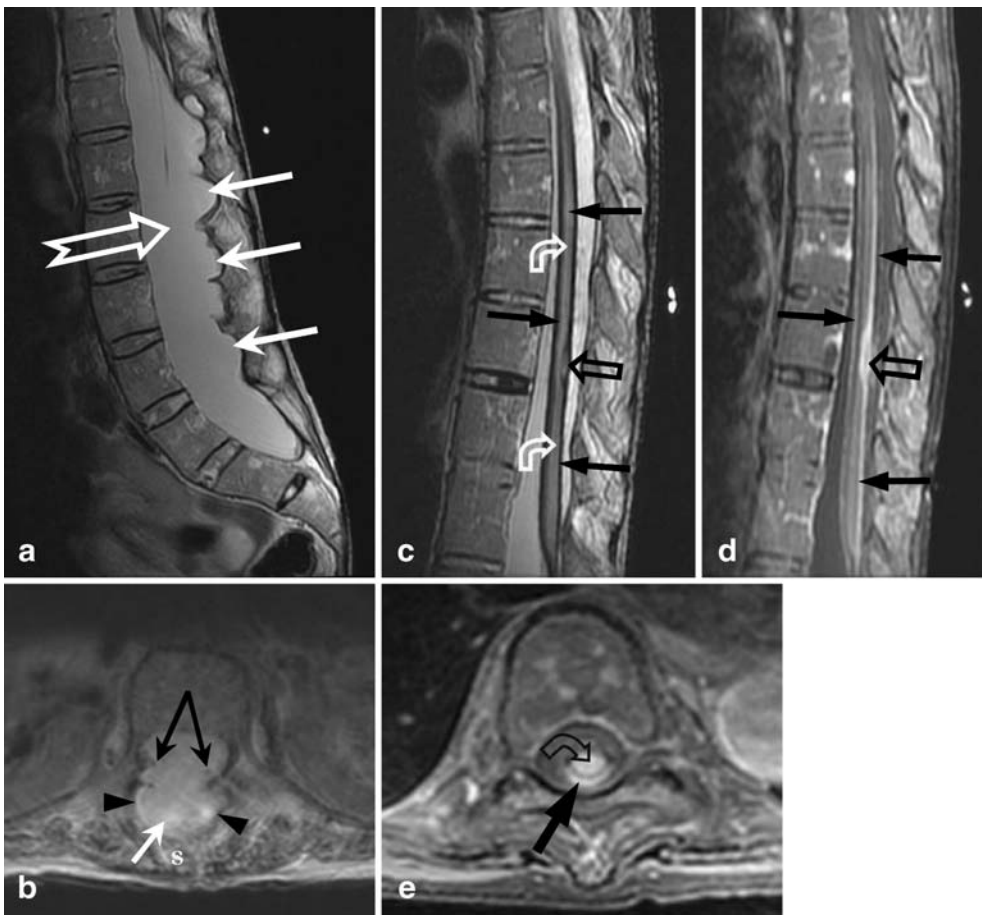

Fig. 1 MRI before treatment. **a**–**b** Lumbar spine. **a** Fast spin echo T2-weighted image (FSE T2WI; TR/TE = 3,000/110 ms), sagittal section. **b** Fast spin echo T2-weighted image (FSE T2WI; TR/TE = 4,267/75 ms), axial section. Note the dural ectasia (*white open arrow*) and the dorsal dural diverticula (*white arrows*). These findings cause pressure erosion on the posterior elements of the vertebra (*arrow heads*). The lumbosacral nerve roots (*black arrows*) were adherent to the dilated dural sac. **c**–**e** Lower thoracic spine. **c** Fast spin echo T2-weighted image (FSE T2WI; TR/TE = 3,000/110 ms), sagittal section. **d** Spin echo T1-weighted image (SE T1WI; TR/TE = 600/15 ms), sagittal section with fat saturation and intravenous administration of the contrast agent gadolinium diethylenetriaminepentaacetic acid (Gd-DTPA). **e** Spin echo T1-weighted image (SE T1WI; TR/TE = 500/18 ms), axial section with fat saturation and intravenous administration of the contrast agent Gd-DTPA. Note that the arachnoid membrane was thickened with contrast enhancement (*black arrows*). An enhancing arachnoid nodule was noted just above the conus medullaris of the spinal cord (*black open arrows*). The findings were diagnosed as acute active arachnoiditis. The spinal cord was swollen with focal ill-defined contrast enhancement (*curve open arrows*) and was diagnosed as acute transverse myelitis. *S*: spinous process

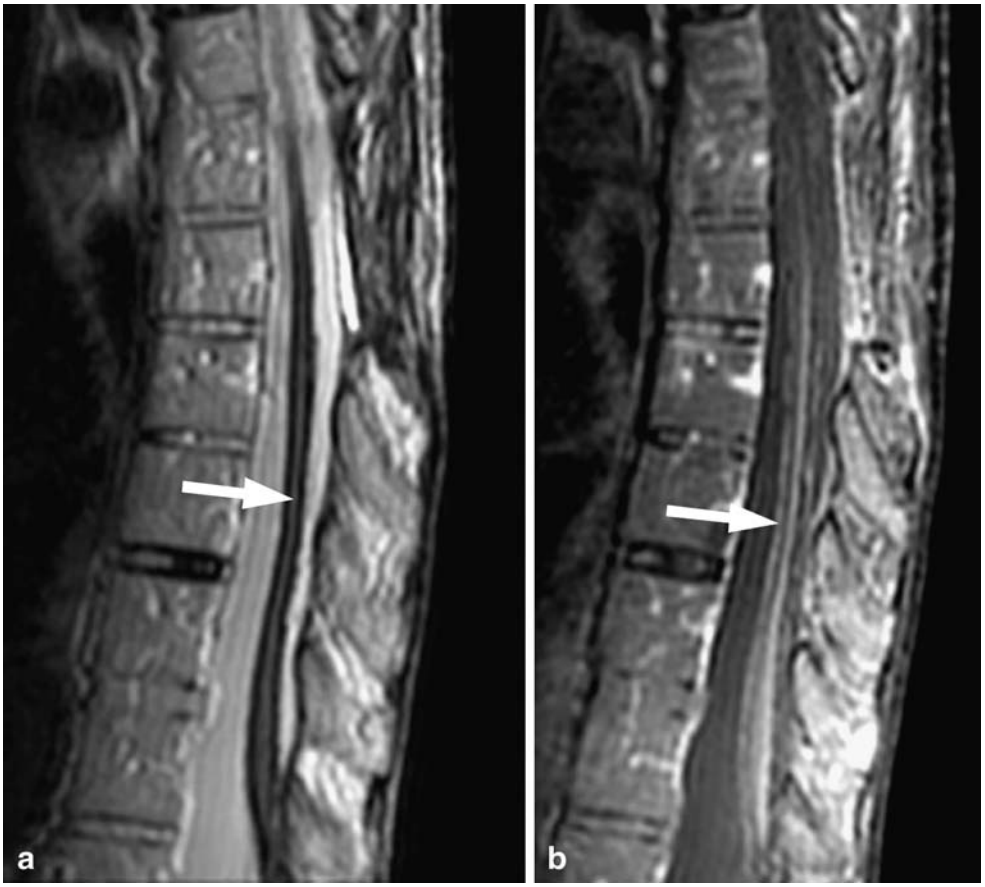

Fig. 2 Follow-up MRI of the lower thoracic spine 6 months after treatment. **a** Fast spin echo T2-weighted image (FSE T2WI; TR/TE = 3,000/110 ms), sagittal section. **b** Spin echo T1-weighted image (SE T1WI; TR/TE = 600/15 ms), sagittal section with fat saturation and intravenous administration of contrast agent Gd-DTPA. Note a segmental thinning of the spinal cord without high signal intensity on T2WI or enhancement on T1WI after intravenous administration of Gd-DTPA proximal to the cornus medullaris, which was diagnosed as spinal cord atrophy. The arachnoid membrane remained thickened with less enhancement (*arrows*). The arachnoid nodule was gone

diverticula, pressure erosion on the laminae, and spinous processes of the vertebrae along the lumbosacral spine (Fig. 1a,b). Nerve roots were adherent to the dilated dural sac (Fig. 1b). The arachnoid membrane of the lower spinal cord was thickened and contrast-enhanced. Contrast enhancement also revealed an arachnoid nodule on the lower spinal cord (Fig. 1c,d). Findings were compatible with a diagnosis of acute arachnoiditis. The lower spinal cord was edematous with focal contrast enhancement (Fig. 1e), which was diagnosed as transverse myelitis.

Analysis of the CSF after MRI disclosed an intrathecal inflammation (mild lymphocytosis, protein 160 mg/dl). The patient received intrathecal injections of methotrexate (10 mg) and decadron (5 mg), and lumboperitoneal (LP) shunting. Symptoms partially improved with sensation returning down to the knee region and no further urinary incontinence. The muscle power of the bilateral lower limbs was also improved (4/5). However, no further improvement was noted after 6 months. MRI of the spine 6 months after treatments revealed atrophy of the spinal cord (Fig. 2). The arachnoid nodule had disappeared. The arachnoid membrane remained thickened. No change was found in dural ectasia, dorsal dural diverticula or nerve root adhesion.

Discussion

CES-AS is a rare, poorly understood complication, which develops 17–53 years (average 35 years) after the onset of AS [7]. Symptoms of CES include sensory impairment of the lower limbs and perineum, and sphincter disturbance, usually when AS is quiescent and laboratory tests are normal [7].

The pathological hallmarks of CES-AS are atrophy, demyelination, and adhesion of the nerve roots, dural ectasia, and dorsal dural diverticula extending into the eroded posterior elements of the lumbosacral canal [10, 11]. The cause of dural ectasia and dorsal dural diverticula are unknown although they were attributed to atrophy of peridural tissues, adherence of dura to adjacent structures, excessive pulse pressure in CSF, and defective resorption of lumbar CSF [10, 12].

The pathogenesis of nerve root damage in CES-AS is unknown. Theories as to its mechanism include damage from arachnoiditis, nerve root compression from dural diverticula, excessive pulse pressure in CSF, demyelination and injury related to radiotherapy or vascular insufficiency, and transverse myelitis [4, 7, 9, 10]. Although an active inflammatory process was not found in postmortem

examination, Matthews posited that inflammation may present at the onset of neurological symptoms and probably resolve over time [10]. In this case, both active transverse myelitis and arachnoiditis coexisted with the chronic dural abnormalities of CES-AS. We assumed that intradural inflammation in the lumbosacral spine might be the primary causative factor of CES-AS, i.e., the acute component, while atrophy and adhesion of the lumbosacral nerve roots, dural ectasia, and dorsal dural diverticula might be the exacerbating or chronic component. Multiple acute episodes or a long-standing inflammation might be needed to develop CES-AS.

Imaging diagnosis of CES-AS was based on the findings of dural ectasia, dorsal dural diverticula, dural calcification, and pressure erosion at the posterior elements of the vertebrae [4, 10]. Radiological investigations of CES-AS included plain radiography, myelography, CT, and MRI [3]. Plain radiography might show widening of the spinal canal, but this feature could be overlooked because of ankylosis of the facet joints [5]. Myelography, the first imaging technique used to diagnose CES-AS, may illustrate the dural ectasia and dorsal dural diverticula well but be difficult to perform in AS and might induce a flare up of neurological symptoms [13]. CT characteristically discloses the chronic abnormalities of CES-AS, i.e., dural diverticula, dural calcification, and bone erosions involving the posterior elements of the vertebrae [6, 14]. Presence of such bone erosions in patients with long-standing AS is virtually pathognomonic of CES [3].

MRI had been suggested as the imaging of choice for diagnosing CES-AS [4, 5]. Both acute and chronic components of CES-AS could be displayed with MRI, including the bone erosion at the posterior elements of the vertebrae [4]. In addition, superior to CT, MRI can show nerve root adhesion as thickened, irregular, distorted linear low signal shadows attaching in the lumbosacral dural sac [4]. To identify unreported acute abnormalities of CES-AS [10], MRI of both lumbar and lower thoracic spine with intravenous administration of a contrast agent is mandatory. Transverse myelitis might show up as focal lesions of high signal intensity on the T2-weighted image (T2WI) and in contrast enhancement in the spinal cord. The involved arachnoid membrane might be thickened but of low signal intensity on the T2WI and in contrast enhancement.

Although no effective treatment is known for CES-AS [5], progression of the neurological symptoms were successfully arrested by LP shunting, based on the theory attributing nerve root damage to excessive pulse pressure in CSF and to nerve root compression from the dural diverticula [15]. Corticosteroid and non-steroid anti-inflammatory agents were thought be ineffective for CES-AS [5]. That might be because there is no active inflammation in the chronic stage of CES-AS. Anti-inflammatory therapy may be necessary in the acute stage of CES-AS to avoid further neurological damage, especially when intradural inflammation is evident on MRI. In our patient, although it might be difficult to know whether the neurological improvements were due to anti-inflammatory therapy or LP shunting, or both, the acute intradural inflammation of CES-AS on MRI was resolved after anti-inflammatory therapy. Neurological restoration might depend upon the extent of initial damage to nerve roots. We believe that early diagnosis of acute intradural inflammation and introduction of anti-inflammatory therapy might be the solution to preventing CES-AS.

In conclusion, MRI of both the lumbar and lower thoracic spine with contrast enhancement is necessary for the diagnosis of CES-AS. The presence of acute abnormalities of CES-AS on MRI might indicate the introduction of anti-inflammatory therapy. LP shunting might be helpful in arresting the progress of neurological symptoms in the chronic stage of CES-AS.

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