

# Anterior dural ectasia associated with cauda equine syndrome in ankylosing spondylitis: a case report

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## Research article

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## Abstract

**Background:** Cauda equina syndrome (CES) is a rare neurologic complication in patients with long-standing ankylosing spondylitis (AS). MRI characteristically shows an enlarged caudal thecal sac, bone erosion and dural ectasia. Dural ectasia is more likely to occur in the posterior elements rather than the vertebral bodies.

**Methods:** We report an uncommon case of CES in AS with formation of anterior dural ectasia. The origin of this rare pathology is analysed and relevant literatures are reviewed.

**Results:** The patient had a 50-year history of AS and presented with weakness of his left leg but without bladder or rectal dysfunction. Radiographs of the full spine showed the classic features of AS with fusion of all spinal ligaments and bilateral sacroiliac joints. CT scanning demonstrated erosion-like lesions with marginal sclerosis in the posterior aspect of the T12 and L1 vertebral bodies. Lumbar MRI revealed anterior dural ectasia into the vertebral body, and ventral herniation of the spinal cord and adhesion to the dural sac.

**Conclusions:** The clinical manifestations of anterior dural ectasia in AS with CES include lower limb numbness and weakness, bladder and rectal dysfunction, impotence, etc. MRI characteristically shows an enlarged caudal thecal sac, bone erosion and anterior dural ectasia at the junction of thoracolumbar and upper lumbar.

## Background

Ankylosing spondylitis (AS) is a chronic inflammatory autoimmune disease of the axial skeleton. It mainly affects the spine and sacroiliac joints, and results in low back pain and progressive stiffness. Neurological complications are uncommon and usually caused by spinal fractures and compression of the spinal cord<sup>[1, 2]</sup>. Cauda equina syndrome (CES) is a rare neurologic complication of long-standing AS. This complication may cause numbness and/or tingling in the buttocks and lower extremities, weakness in the legs, and incontinence of bladder and/or bowels.

Magnetic resonance imaging (MRI) of CES in AS characteristically shows an enlarged caudal thecal sac, bone erosion and dural ectasia<sup>[3]</sup>. Dural ectasia is more likely to occur in the posterior elements rather than the vertebral bodies<sup>[4, 5]</sup>. Here, we report an uncommon case of CES in AS with formation of anterior dural ectasia.

## Case Presentation

A 78 year-old man with a 50-year history of AS had started to experienced gradual onset of weakness and muscle atrophy in the left lower limb for the past 1 years, and finally had to use a cane. After being diagnosed with AS at the age of 28, our patient had seldom visited his physician but he used painkillers as needed. The patient denied any history of trauma, tuberculosis or surgery.

On physical examination, the motion of the thoracic and lumbar spine was severely decreased. There was no leg length discrepancy, but the entire left lower extremity was atrophied: the left thigh circumference was 8 cm smaller than the right thigh, and the left calf was 2 cm smaller than the right side. His neurological examination revealed significant wasting of the left leg and decreased muscle tone. He presented a weakness

of the iliopsoas (3/5), quadriceps muscles (2/5), tibialis anterior (1/5), extensor hallucis longus (1/5) and gastrocnemius (3/5) in left lower extremity. There was normal lumbosacral dermatomal sensation and anal sensation. Patellar and Achilles tendon reflexes were lost on the left side, and the Babinski response was negative.

Except HLA-B27 positivity, routine rheumatologic and biochemical screening investigations were normal. Electromyography (EMG) was abnormal for only the left L5 nerve. Radiographs of the full spine showed the classic features of AS with fusion of all spinal ligaments and bilateral sacroiliac joints (Fig. 1). CT scan demonstrated erosion-like lesions with marginal sclerosis in the posterior aspect of the T12 and L1 vertebral bodies (Fig. 2). Lumbar MRI revealed anterior dural ectasia into the vertebral body, and ventral herniation of the spinal cord and adhesion to the dural sac (Fig. 3).

## Discussion

CES is a well-known but rare and poorly understood complication of AS. This complication may cause numbness and/or tingling in the buttocks and lower extremities, weakness in the legs, and incontinence of bladder and/or bowels. CES manifests late in the course of AS, often at a time when the AS is no longer active<sup>[6]</sup>. The time interval between the onset of AS and the clinical manifestation of CES averages 35 years (range 17–53 years)<sup>[7]</sup>. MRI characteristically shows an enlarged caudal thecal sac, bone erosion and dural ectasia<sup>[3]</sup>. The lesions primarily affect the lumbar region but can also appear in the thoracic region<sup>[8]</sup>.

Previous studies have revealed that the formation of dural ectasia and arachnoid diverticulum in AS is closely related to the chronic inflammatory process<sup>[9]</sup>. Liu et al.<sup>[10]</sup> described fibrous tissue with scant lymphoplasma cell infiltration in the dura mater of an AS patient with dural ectasia. Local inflammation leads to a slowly progressive process of bony erosion<sup>[11, 12]</sup>. Furthermore, dural fibrosis causes diminished resorption of cerebrospinal fluid (CSF). Reduced elasticity and compliance of the caudal sac exposed to inflammatory changes is also a factor inducing dural sac enlargement and diverticula formation<sup>[13]</sup>.

The direction of arachnoid diverticula formation is thought to be dependent on the direction of the force of increased CSF pulse pressure and the “give” of the structures surrounding the spinal canal<sup>[6]</sup>. Dural ectasia is more likely to occur in the posterior elements rather than the vertebral bodies<sup>[4, 5]</sup>. Posterior dural ectasia in AS was first documented using myelography by Hauge<sup>[14]</sup>, and has been well described subsequently. However, the cases with anterior dural ectasia are more rare (Table 1)<sup>[6, 11, 15–18]</sup>. These changes seem to occur commonly at the level of the thoracolumbar junction and upper lumbar vertebrae<sup>[17]</sup>.

Table 1

Summary of the previous case reports of anterior dural ectasia associated with CES in AS

Author	Age (y)	Sex	Time since onset of CES (years)	Presenting complaints	Investigation	Lesion site	Description of the lesion	Neural tissue herniation
Byrne et al. (1985) <sup>[15]</sup>	71	Male	2	Difficulty walking, impotence, bladder disturbance	Myelography	L2-L3	Intradural cyst (believed to be arachnoid cyst)	Yes
Ginsburg et al. (1997) <sup>[6]</sup>	65	Male	9	Numbness and weakness in both legs, right buttock and posterior thigh pain, bladder and bowel disturbance, impotence	MRI	T12-L1	Arachnoid diverticula	Yes
Baur et al. (1997) [16]	50	Male	2	Numbness in left buttock and feet, bladder disturbance, faecal incontinence	CT, CTM, MRI	T12-L1	Ventral dural defect	Yes
Hayashi et al. (2009) <sup>[17]</sup>	63	Male	2	Numbness and tingling left leg posteriorly, difficulty walking, constipation	MRI	L1-L2	Arachnoid diverticula	No
Bele et al. (2011) <sup>[11]</sup>	50	Male	0.5	Weakness of right leg	CT, MRI, MRM	L1-L2	Arachnoid diverticula	Yes
Huang et al. (2019) <sup>[18]</sup>	44	Male	0.1	Numbness and weakness of left leg	CT, MRI, contrast-enhanced MRI	L1-L2	Arachnoid diverticula	Yes
Present case	78	Male	1	Weakness of left leg	CT&MRI	T12-L1	Arachnoid diverticula	Yes
CTM = CT myelography; MRM = magnetic resonance myelogram								

Dural ectasia can also be caused by spinal tumor, Marfan syndrome, Ehlers-Danlos syndrome, or neurofibromatosis, but the patient's clinical manifestations and imaging findings did not support the diagnosis of these disease. In our case, anterior dural ectasia and bony erosion of the posterior aspect of T12 and L1 vertebral bodies were seen. MRI also demonstrated ventral shift and adhesion of the conus medullaris and the cauda equine. It is a remarkable fact that our patient presented with weakness of the left leg, but without bladder and rectal dysfunction. The mechanisms of nerve root damage are poorly understood, but some hypotheses have been proposed. These include inflammatory injury from arachnoiditis, gravitational traction injury resulting from the compression of arachnoid diverticula, and traumatic damage from excessive CSF pressure fluctuations [7, 19, 20].

There is currently no standard and uniform treatment for dural ectasia associated with CES in AS. Kotil et al. [21] recommend that asymptomatic or symptomatic patients with dural ectasia should be closely observed without need for immediate surgical intervention. Ahn et al. [22] suggested that steroids are ineffective for patients with CES in AS, whereas non-steroidal anti-inflammatory drugs may improve back pain but do not improve neurological deficits, probably because no active inflammation occurs in the chronic stage. Huang et al. [18] reported a case of anterior dural ectasia with AS, and treated with an oral nonsteroidal anti-inflammatory drug. Neurological deficits remained stable during 2 years of follow-up, consistent with the disappearance of the dural enhancement in the lytic lesion of the vertebral bodies. Their case supports the essential role of inflammation in anterior dural ectasia formation and confirms the effectiveness of anti-inflammatory therapy. Infliximab is a humanized antibody against tumor necrosis factor  $\alpha$  (TNF- $\alpha$ ) that is used in the treatment of active AS. Cornec et al. [23] reported on an AS patient with posterior dural ectasia that was unexpectedly cured by treatment with Infliximab, and subsequent studies have confirmed this dramatic finding [9, 10].

A meta-analysis by Ahn et al. [22] concluded that surgical treatment of dural ectasia with CES might improve the neurologic symptoms or halt the progression of neurologic deficit, and has a better outcome than conservative or no treatment. However, the literature regarding management of anterior dural ectasia is scarce. Surgical options for dural ectasia with CES include detethering of the cord, decompressive laminectomy, and lumboperitoneal shunting (LPS). Liu et al. [10] found that little improvement was achieved after performing de-adhesion of the tethered conus medullaris. Bele et al. [11] suggested releasing the arachnoid adhesions and fibrosis around the cauda equina roots, which may lead to deterioration of neurological function. Decompressive laminectomy provides more space for the neural elements at the cauda equine, but LPS for CES in AS is more efficient than laminectomy [24]. LPS helps in relieving the intradural pressure and has been offered for the more common posterior dural ectasia with CES [24]. Ea et al. [13] reported that five AS patients with dural sac dilation achieved substantial improvement that persisted at follow-up by treatment of LPS. Although it is not clear whether LPS is effective for anterior dural ectasia, Bele et al. [11] suggested performing LPS, as the pathogenesis for an anterior and posterior dural ectasia remains the same.

## Conclusions

CES in AS is a rare clinic entity, while the cases with anterior dural ectasia are more rare. Its clinical manifestations include lower limb numbness and weakness, bladder and rectal dysfunction, impotence, etc. MRI characteristically shows an enlarged caudal thecal sac, bone erosion and anterior dural ectasia at the

junction of thoracolumbar and upper lumbar. Non-steroidal anti-inflammatory drugs may improve back pain but do not improve neurological deficit, while Infliximab or surgical intervention were relatively effective.

## Abbreviations

CES: cauda equina syndrome; AS: ankylosing spondylitis; MRI: magnetic resonance imaging; CSF: cerebrospinal fluid; LPS: lumboperitoneal shunting; TNF- $\alpha$ : tumor necrosis factor  $\alpha$ .

## Declarations

### Ethics approval and consent to participate

This study was approved by the Ethics Committee of Fuzhou Second Hospital affiliated to Xiamen University. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

### Consent for publication

All authors have approved the manuscript for submission in *Journal of Orthopaedic Surgery and Research* and consent for the publication.

### Availability of data and materials

The datasets and materials generated or analyzed during the current study are available from the corresponding author on reasonable request.

### Competing interests

The authors declare that they have no competing interests

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No funds were received in support of this work.

### Authors' contributions

LSQ: designed manuscript, collected patient information and performed literature search. CW: advised on design and corrected manuscript. LGQ: supervised project, provided patient information and corrected manuscript. All authors read and approved the final manuscript.

### Acknowledgements

Not applicable

### Authors' information (optional)

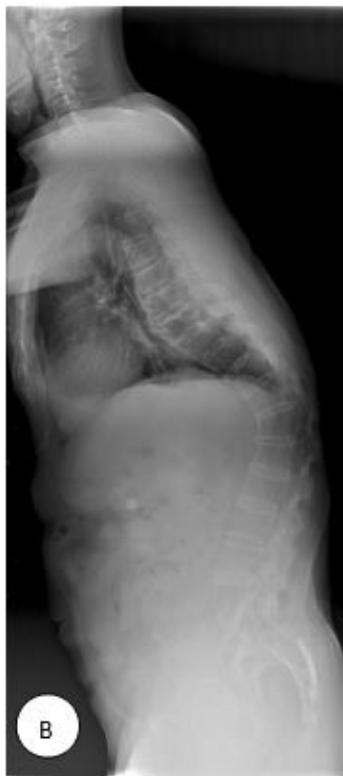
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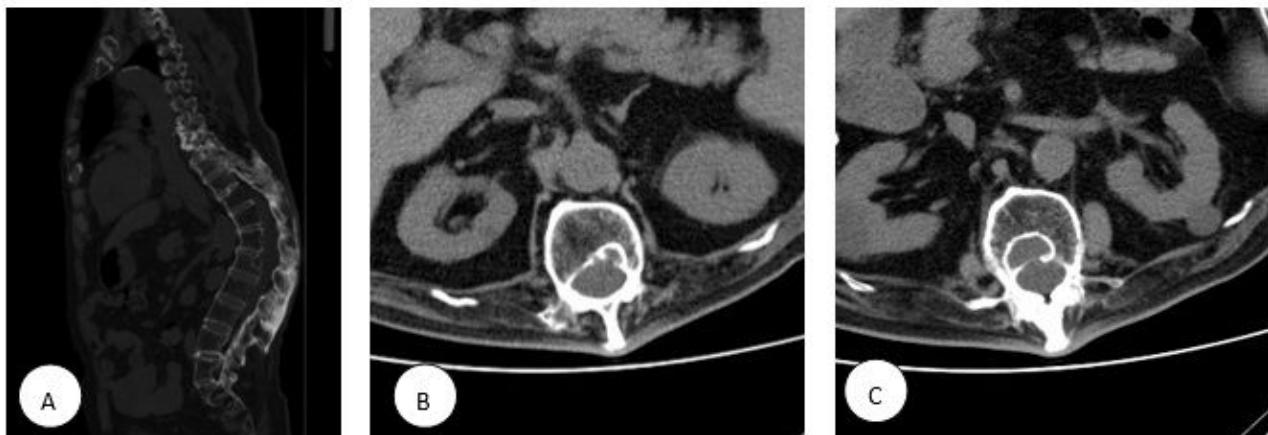
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## Figures



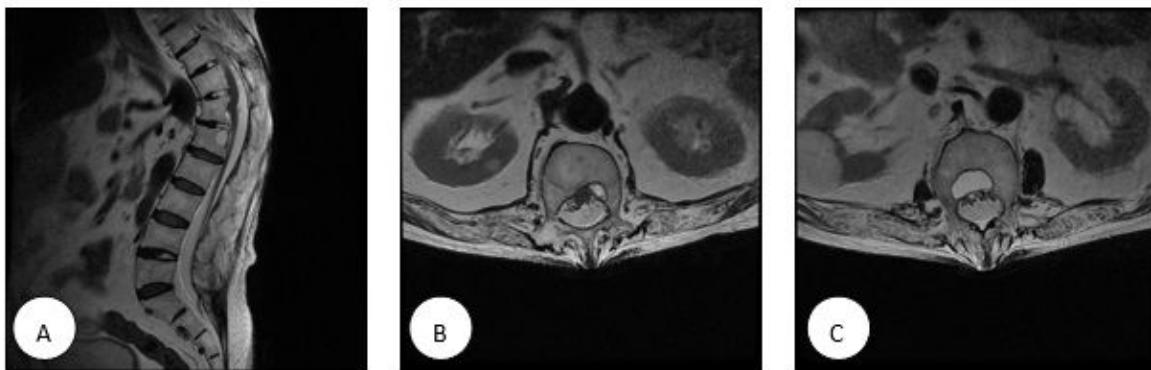
**Figure 1**

Radiographs of the full spine, anteroposterior (A) and lateral (B) view, showing the classic features of AS with fusion of all spinal ligaments and bilateral sacroiliac joints. Kyphosis at the thoracolumbar junction is apparent.



**Figure 2**

Sagittal CT scan image (A) demonstrating erosion-like lesions in the posterior aspect of the T12 and L1 vertebral bodies. Axial CT scan obtained at the T12-level (B) and L1-level (C), showing a smoothly marginated lytic lesion eroding the posterior vertebral border with extension into the body. Posterior spinal elements appear normal.



**Figure 3**

Sagittal T2-weighted MR image of the lumbar spine (A) showing the cystic ectatic lesion occupying the posterior aspect of the T-12 and L-1 vertebral bodies. Axial T2-weighted MR image obtained at the T12-level (B) and L1-level (C), showing the cystic lesion in the dorsal part of the vertebral bodies with ventral herniation of the spinal cord and nerve roots.