Cauda equina syndrome of long-standing ankylosing spondylitis

Case report and review of the literature

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Cauda equina syndrome as a neurological complication of long-standing ankylosing spondylitis was first reported in 1961. The syndrome is relatively uncommon and its pathophysiology is still poorly understood. Based on their experience with such a case, the authors review the clinical, electrographic, histological, and radiographic features of the syndrome, including the findings of magnetic resonance (MR) imaging. The addition of MR imaging to the evaluation of patients with ankylosing spondylitis and the cauda equina syndrome not only aids in the diagnosis of the syndrome but may also provide valuable insight into the pathophysiology of this condition.

KEY WORDS • ankylosing spondylitis • cauda equina syndrome • diverticulum • laminar erosion • arachnoidal adhesions

ANKYLOSING spondylitis, also known as von Bechterew's disease, Marie-Strümpell disease, or rheumatoid spondylitis, is a spondyloarthropathy that affects approximately 1% of the population. Diagnostic clinicoradiological criteria were first formulated in 1961 and were revised in 1966. The most important of these criteria are radiographic demonstration of sacroiliitis in the presence of a history of pain and/or limitation of motion. There is a strong male preponderance and a specific association with the human lymphocyte antigen (HLA)-B27 histocompatibility antigen. The primary pathological site of ankylosing spondylitis is at the insertion of ligaments and capsules on bone, whereas in rheumatoid disease the essential pathology resides within the synovium. In addition to spinal symptomatology, some patients may exhibit a peripheral arthropathy and regions of insertional tendinitis. Other nonmusculoskeletal manifestations of the disease include uveitis, chronic prostatitis, pulmonary fibrosis, cardiac valvular disease, and amyloidosis. Neurological complications include solitary nerve root lesions, spontaneous atlantoaxial subluxation with associated myelopathy, cauda equina compression secondary to lumbar spinal stenosis, and traumatic fractures of the multilevel fused vertebral column with resulting nerve-root or spinal cord compression.

A poorly understood complication of long-standing ankylosing spondylitis is a cauda equina syndrome, first reported by Bowie and Glasgow. The first case of this syndrome reported in the North American literature appeared in 1973. Prior descriptions have related the characteristic radiographic findings on conventional radiographs, contrast myelography, and computerized tomography (CT).

Case Report

This 61-year-old white man was admitted for evaluation of a 5-year history of progressive perineal numbness. He described the onset of circumferential numbness of the penis 6 months following a transurethral resection of the prostate. The symptoms prior to prostatic resection were hesitancy and dribbling, soon succeeded by urinary incontinence. Following resection, his
FIG. 1. *Left:* Metrizamide myelogram, anteroposterior view, demonstrating slight dilatation of lumbar and sacral nerve-root sleeves (arrow). *Center:* Myelogram, oblique view, showing adherence (arrows) of nerve roots of the cauda equina to the dorsal lumbar theca adjacent to the thecal diverticula at the L-3 and L-4 vertebral levels. *Right:* Myelogram, lateral view, revealing the accumulation of contrast material (arrows) within the dorsal thecal diverticula of the L-3 and L-4 vertebrae.

Urinary incontinence persisted, and he began to have episodes of fecal incontinence and also noted the onset of penile numbness. Over the subsequent years, numbness progressed to include bilateral involvement of his scrotum and buttocks. During the year prior to admission he noted numbness on the lateral aspect of his left foot. He had no history of lower-extremity pain or weakness. At his most recent urological evaluation, a cystometrogram had revealed a neurogenic bladder.

Medical history revealed that at 20 years of age, this patient had experienced slow progressive onset of neck, back, and hip pain which had been diagnosed as ankylosing spondylitis. Steroid therapy in 1946 provided no relief from his discomfort. In 1959 he underwent spinal radiation which appeared to relieve his pain, and in 1966 he had a subtotal colectomy for ulcerative colitis.

**Examination.** Physical examination revealed rigid ankylosis of the lower cervical, thoracic, and lumbar spine and limitation of chest expansion. No muscular atrophy or fasciculation was present; mental status and cranial nerve examination were normal. The general motor examination was normal, and sensory examination revealed hypesthesia and hynalgia of the genitalia, the perineum, the buttocks bilaterally, and the lateral aspect of the left foot. Deep-tendon reflexes were normal except for absent ankle jerks bilaterally. Abdominal and cremasteric reflexes were intact. Rectal sphincter tone was absent, as were anal and bulbocavernous reflexes. The gait was normal.

Cerebrospinal fluid (CSF) obtained just prior to myelography revealed no cellular, chemical, or serological abnormalities. Values for serum alkaline phosphatase, complete blood count, erythrocyte sedimentation rate, and serological test for syphilis were normal. The patient tested positive for the presence of HLA-B27 histocompatibility antigen.

Nerve conduction velocities of the right and left sural and peroneal nerve were normal. Electromyography of the anal sphincter revealed bilateral positive waves and fibrillations with markedly decreased recruitment and high-frequency discharges. Motor unit potentials were highly polyphasic. Paraspinous muscles at the S-1 vertebral level and below revealed 2+ positive waves and 1+ fibrillations. Normal findings were obtained in the L-5 paraspinous, gastrocnemius, and gluteus maximus muscles bilaterally. The H reflex was absent on both sides. Somatosensory evoked potentials, elicited by stimulation of the sural and dorsal penile nerve, suggested a probable cauda equina or conus medullaris lesion affecting the roots below the S-1 level.

Plain radiographs of the cervical, thoracic, and lumbar spine demonstrated significant bone ankylosis at all levels. Sacral and pelvic radiographs also showed advanced spondylitic changes with complete fusion of the sacroiliac joints. Myelography performed via the lumbar route demonstrated a large lumbar thecal sac and multiple dilated sacral nerve-root sleeves. There was also adhesion and tethering of several nerve roots of
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FIG. 3. Axial T₁-weighted magnetic resonance image demonstrating clumping and adherence (arrows) of nerve roots of the cauda equina to the dorsal lumbar theca lateral to the L-3 diverticula.

FIG. 2. Computerized tomography scan showing bone erosion of the lamina and spinous process of the L-3 vertebra (arrow).

The cauda equina to the dorsal lumbar theca, adjacent to dorsal thecal diverticula at the L-3 and L-4 vertebral levels (Fig. 1). A CT scan of the lumbar spine illustrated bone erosion of the ventral surface of the lamina and spinous processes of the L-3 and L-4 vertebrae (Fig. 2).

Magnetic resonance imaging of the thoracic, lumbar, and sacral regions was performed. No compressive or mass lesions were identified, and special attention was directed to the conus medullaris and cauda equina regions. A large lumbar thecal sac, dilatation of the lower lumbar and sacral nerve root sleeves at the neural foramina, and dorsal lumbar thecal diverticula underlying laminar erosion at the L-3-4 vertebral level were evident. There was clumping of nerve roots and some nerve roots of the cauda equina were adherent to the dorsal theca adjacent to the rostral and lateral aspect of the lumbar thecal diverticula. No evidence of abnormal MR enhancement of bone or soft tissue elements could be identified following intravenous administration of gadolinium diethylenetriamine penta-acetic acid (Gd-DTPA) (Figs. 3 and 4).

Review of the Literature

Over the past 25 years, the development of the cauda equina syndrome has been recognized as a neurological complication of long-standing ankylosing spondylitis. In addition to the first description of the syndrome in three patients by Bowie and Glasgow, four 46 cases have been reported in the literature. The largest series (14 patients) was reported from the Mayo Clinic. Bartleson et al. reviewed a 25-year experience with patients whose diagnoses included both ankylosing spondylitis and cauda equina syndrome.
Several case reports have appeared in the literature with many common clinical and radiographic findings, similar to the findings in our patient. The overwhelming majority (45 of 49 patients) are male. The average age of these individuals at onset of symptoms referable to ankylosing spondylitis is 24 years. A long interval separates the onset of ankylosing spondylitis and the development of a cauda equina syndrome. In 46 previous cases, this interval ranged from 10 to 54 years (average 29 years). With many patients, the ankylosing spondylitis was asymptomatic for years prior to the late onset of neurological complaints. The process tends to involve selectively the lowest lumbar and sacral nerve roots.\textsuperscript{3,5,6,17,34}

A review of the previously reported cases reveals that the most common initial neurological symptoms were a sensory disturbance (in 16 patients), urinary and/or rectal sphincter disturbance (in 14), lower-extremity or perineal pain (in 10), and weakness of the lower extremities (in three). The simultaneous presentation of a combination of symptoms occurred in five patients. During the course of cauda equina syndrome, 47 patients developed sensory disturbances. 44 developed urinary or rectal sphincter disturbances, 27 had motor deficits, and 23 complained of pain.\textsuperscript{3,5,6,17,34}

Prostatic resection for symptoms presumed to result from obstruction due to prostatic hypertrophy is reported in many of these patients. Following surgery, however, they often note little improvement or even observe progression of urinary incontinence. This problem, in turn, may be followed by complaints of sensory disturbance and/or fecal incontinence,\textsuperscript{3,17,19,30,33,34,38} as in our patient.

In the 49 patients reviewed, abnormalities were found in 21 of the 22 individuals undergoing electromyography, and neurogenic lesions were suggested in those cases. Although the electromyogram, nerve conduction velocities, and somatosensory evoked potentials identified the level of the lesion in our patient, such localization is not always achieved.\textsuperscript{3,15,17,20,25,30,32,34,38}

The CSF examination in our patient revealed no chemical or cellular abnormality. Of 27 previous cases with CSF examinations, only seven showed mild increases in protein levels. No elevation in white blood cell count has been noted. Some authors have reported the presence of erythrocytes in the CSF, but this is usually attributed to traumatic puncture.\textsuperscript{3,4,15,17,19-21,23,24,26,28,30,34}

**Management of Cases**

**Differential Diagnosis**

The differential diagnosis of the cauda equina syndrome includes all mass lesions of the conus and cauda equina region, such as intramedullary, intradural, and extradural neoplasia.\textsuperscript{10,11} Other entities to be distinguished include vascular malformations and benign compressive lesions such as thoracolumbar disc herniation and developmental/acquired spinal stenosis.

**Radiological Findings**

In addition to excluding the above-mentioned lesions, myelography demonstrates the characteristic radiographic findings of patients with this syndrome. Of 49 patients described in the literature, 38 underwent myelography and four had CT without myelography. Among the patients undergoing myelography, 18 were reported to have an enlarged caudal thecal sac, 22 had dorsal arachnoid diverticula, two had dilatation of lumbosacral nerve root sleeves, and one was reported to have an "intraspinal expansive process." Seven patients were reported to have normal myelograms.\textsuperscript{3,5,6,17,34} All patients who underwent only CT showed laminar erosion, signifying the presence of dorsal thecal diverticula. In the first case presented by Bowie and Glasgow,\textsuperscript{4} myelography demonstrated "prominent diverticula along the lumbar nerve root sheaths but was otherwise normal." The description given of their second case alluded to what may have been dorsal arachnoid diverticula. However, the findings were regarded as an exaggeration of the normal appearance seen in the supine position and were thought not to be significant.\textsuperscript{4}

The first clear radiographic description of the characteristic dorsal diverticula was provided by Matthews\textsuperscript{24} in 1968. The autopsy findings in one of his patients led to reappraisal of the myelogram in that case. Initially, the myelogram, which was performed in the prone position, was interpreted to show only an unduly capacious sacral canal. Upon review of the films, two collections of contrast medium were visualized lying posteriorly in arachnoid diverticula. In his next patient, myelography was deliberately performed in the supine position, revealing similar persistent accumulations of contrast material lying posterior and lateral opposite the L-4 vertebra. These observations led to the subsequent recommendation that myelography in these patients be performed in the supine position.\textsuperscript{6,17,18,20,28,30,33}

The first evidence of laminar erosion by CT was published by Kramer and Krouth,\textsuperscript{25} who believed that this erosion reflected the presence of dorsal arachnoid diverticula. Because CT provides considerable data in these patients, Young, \textit{et al.},\textsuperscript{38} proposed that myelography is not necessary in the absence of clinical and electromyographic evidence of localized pathology. They also recognized that myelography may even be harmful, basing this on their experience with one patient who deteriorated symptomatically following myelography. Similar opinions concerning myelography in these patients have been expressed by other authors.\textsuperscript{3,6,17,34}

The MR findings in this condition include an expanded lumbosacral thecal sac and multiple dorsal thecal diverticula lying within the bone erosions of the lamina and spinous processes of the lumbar vertebrae. Magnetic resonance imaging also reveals dilatations or diverticula of scattered lumbosacral nerve-root sleeves and adhesions of nerve roots of the cauda equina to the dorsal lumbar theca adjacent to the diverticula (Figs. 3 and 4).
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Pathogenesis

The pathogenesis of the cauda equina syndrome in ankylosing spondylitis is unknown. Theories of the mechanism of nerve-root injury include damage from arachnoiditis and nerve-root compression from expanding thecal diverticula. Other theories propose injury related to previous radiation therapy or from vascular insufficiency. These theories have been based on the results of operative and necropsy cases. Pathological descriptions were first reported by Hauge. In the patient he presented first, surgical exposure revealed firm grayish tissue filling the subdural space and surrounding the cauda equina. Microscopic examination revealed fibrous connective tissue surrounding the nerve roots. In his next patient, Hauge found atrophic lumbosacral muscles, thin dorsal arches of the L-5 and sacral vertebrae, no epidural tissue, and thin dura. The sacral nerve roots were found to be adherent to a thickened arachnoid, which, in turn, was also adherent to the dura. The exposed nerve roots appeared grossly atrophic. Tissue obtained from a nerve-root sleeve histologically showed fibrous connective tissue and scattered lymphocytes. Encouraged by the report of Bowie and Glasgow on three patients with ankylosing spondylitis and cauda equina syndrome, Lee and Waters performed a lumbosacral laminectomy on a similar patient. They found small sacral roots splayed out and loosely adherent to the dura.

Matthews published a detailed report of the postmortem findings in a patient with ankylosing spondylitis and cauda equina syndrome. The findings included general widening of the spinal canal and numerous diverticula extending deeply into eroded lamina, the spinous processes, and the roof of the sacral canal. The dorsal diverticula did not encroach on the neural foramina. The diverticula were lined by a membrane formed of fibrous connective tissue and scattered lymphocytes. Encouraged by the report of Bowie and Glasgow on three patients with ankylosing spondylitis and cauda equina syndrome, Lee and Waters performed a lumbosacral laminectomy on a similar patient. They found small sacral roots splayed out and loosely adherent to the dura. Matthews published a detailed report of the postmortem findings in a patient with ankylosing spondylitis and cauda equina syndrome. The findings included general widening of the spinal canal and numerous diverticula extending deeply into eroded lamina, the spinous processes, and the roof of the sacral canal. The dorsal diverticula did not encroach on the neural foramina. The diverticula were lined by a membrane formed of fibrous connective tissue and scattered lymphocytes. Matthews also speculated that the nerve roots are damaged because of the original arachnoiditis and/or compression by expansive arachnoid diverticula, which may be produced by arterial pulsations of the CSF. Lee and Waters reported chronic arachnoiditis in association with a vascular lesion of the cord in their patient. Radiation therapy in patients with ankylosing spondylitis, although initially implicated as an etiology for the process, is not currently thought to play a role in the pathogenesis.

Treatment

No treatment is known to be effective in the cauda equina syndrome of long-standing ankylosing spondylitis. No clinical improvement has been reported after treatment with corticosteroids or nonsteroidal anti-inflammatory agents. Furthermore, Milde, et al., described a patient who presented with cauda equina syndrome and was treated for 15 years with cortisone and indomethacin for pain associated with ankylosing spondylitis. Surgical exploration has been of no proven utility in this condition, and in some patients, it has led to a worsening of neurological deficits.

Of 49 reported cases, the cauda equina syndrome was progressive in 31 patients following initial evaluation. A stable course was described in nine patients. There was no mention of follow-up findings in the remaining nine cases.
Discussion

Our observations would suggest that MR imaging may obviate the need for both myelography and CT. An MR image rules out compressive and mass lesions of the conus medullaris and cauda equina and clearly demonstrates the characteristic radiographic findings in patients with the cauda equina syndrome of long-standing ankylosing spondylitis. It also avoids some of the difficulties and complications that have been reported in performing myelography in these patients. Based on imaging, previous operative and necropsy findings, and a review of the pathological findings in patients with ankylosing spondylitis, we are able to propose a theory which explains the structural lesions, symptomatology, and etiology of nerve-root injury in this condition.

From pathological studies, it is well recognized that the primary pathological site in ankylosing spondylitis is at the insertion of ligaments and joint capsules into bone. Ligamentous inflammation, followed by fibrosis with or without subsequent ossification, occurs at this site and is associated with inflammation, fibrosis, and destruction of the underlying bone which correlates radiographically to bone erosion. These pathological changes, as described by Cruickshank, occur in the cartilaginous joints of patients with ankylosing spondylitis. Radiographic changes such as these have also been found in the spinous processes, the pelvic bones, the greater trochanter, and the feet. Wilkinson and Bywaters have drawn attention to the fact that the location of these bone lesions is nearly always related to tendon insertions. Engfeldt, et al., found nonspecific inflammation in the areolar tissue adjacent to spinous processes. From biopsies taken of the iliac crest and over the greater trochanter, Ball also found inflammatory lesions at ligamentous attachments and erosive lesions of cortical bone. In addition, he found perivenous collections of lymphocytes in the adipose and loose fibrous tissues adjacent to ligaments, which he suggested were representative of a nonspecific reaction to tissue damage. It has also been found that the ligamentum flavum and interspinous ligaments may be involved, resulting in calcification/ossification of these structures; calcification of the meninges has also been reported.

Based on pathological findings in patients with ankylosing spondylitis, MR imaging, and previous operative and necropsy reports of patients with a cauda equina syndrome secondary to long-standing ankylosing spondylitis, it is most likely that primary ligamentous inflammation leads to inflammation of contiguous structures such as the meninges and dorsal bone elements of the spine. This produces dorsal meningeal inflammation with subsequent arachnoid adhesion formation and erosions of the lamina and spinous processes with secondary diverticula development. Elevated hydrostatic pressure upon the lumbar theca as well as pulse pressure waves, as eluded to by Matthews, probably contribute to this erosive process and to the development of dorsal thecal diverticulae in the lumbosacral region. The paramagnetic contrast agent gadolinium, thought to produce MR enhancement based on blood-brain/meningeal barrier abnormalities, may prove valuable in identifying this early active inflammatory process. This proposed process would also explain the neurological symptoms and findings by involving the most medially and dorsally located nerve roots of the cauda equina.

Nerve-root injury is probably related to arachnoiditis with subsequent neuritis, vasculitis, adhesion formation, and tethering, or a combination of these entities. Early evaluation of patients with long-standing ankylosing spondylitis, who present with urinary and rectal sphincter disturbances, subtle neurological symptoms, and/or signs related to early radicular syndromes, is important. Data obtained early in the process will provide information which may aid in understanding the pathogenesis and pathophysiology of this condition. We believe that MR imaging is the diagnostic modality of choice, and we recommend its use in the initial evaluation of patients with ankylosing spondylitis presenting with a cauda equina syndrome.

References

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