

## Neurological disturbances in ankylosing spondylitis

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### Case history

A 56 year old man presented with a seven week history of tingling in both buttocks associated with numbness of the right buttock which radiated down the back of the leg to the level of the knee. He also gave a four year history of numbness along the heel and sole of the right foot. There was no motor symptoms. No specific urinary or bowel symptoms were evident. In the past he had experienced intermittent low back pain, but he had been asymptomatic in that regard for many years.

On examination there was impaired sensation along the right S2 dermatome and also perianally in the S3 to S5 distribution on the right side. Movement of the spine was limited in all directions. No focal tenderness was elicited. Examination was otherwise normal. Plain radiographs of the lumbar spine were obtained.

### PLAIN RADIOGRAPHIC FINDINGS

Radiographic examination of the lumbar spine

(fig 1) demonstrated bilateral sacroileitis with fusion of the sacroiliac joints. Syndesmophyte formation was present throughout, giving a classic "bamboo" appearance. Calcification of intervertebral discs and pronounced thickening/calcification of the anterior longitudinal ligament were also apparent. The lateral view also demonstrated a marked widening of the spinal canal.

### Differential diagnosis

The patient demonstrates the radiographic features of advanced ankylosing spondylitis (AS) with a fused immobile spine. Widening of the spinal canal is not a feature of uncomplicated AS but the finding may indicate an underlying neurological disorder. Widening of the spinal canal as a result of dural ectasia may occur in other conditions such as Marfan's syndrome, Ehlers-Danlos syndrome and neurofibromatosis. The changes in AS, however, result from involvement of the posterior elements rather than the posterior aspect of the

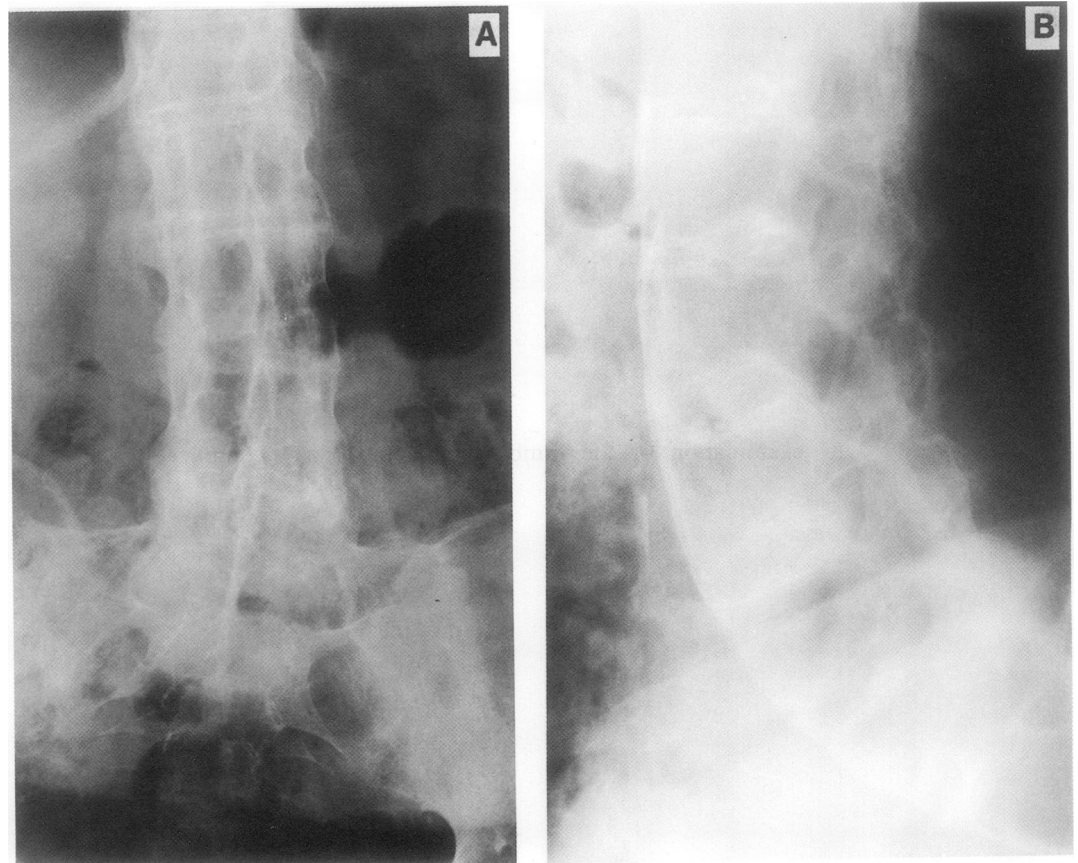


Figure 1 Anteroposterior (A) and lateral (B) radiographs of the lumbosacral spine.

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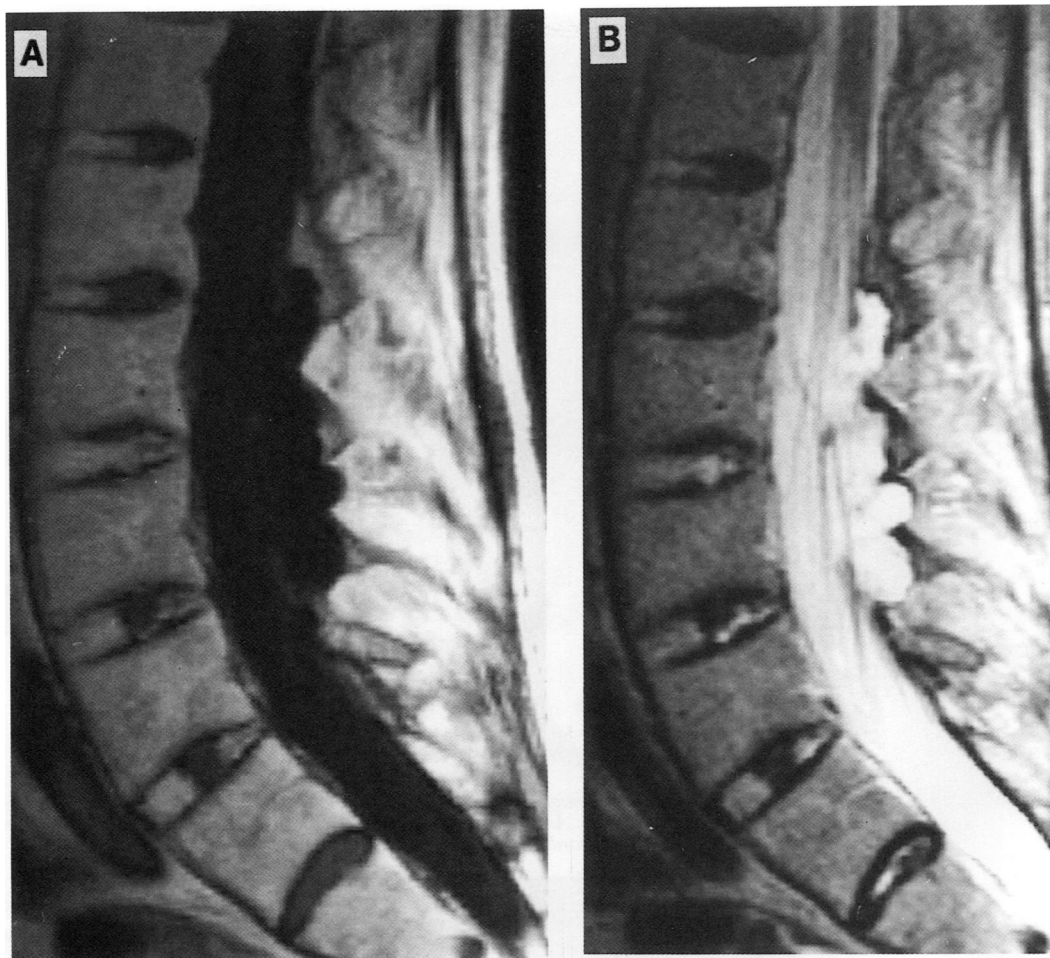


Figure 2 Sagittal T1W (A) and T2W (B) images of the lumbosacral spine. B shows, in addition to details apparent in A, the intermediate signal nerve roots descending on the thecal sac.

vertebral bodies as is usually seen in these other conditions. The clinical assessment of this patient is that of a cauda equina syndrome, which is a well recognised, albeit rare, complication of chronic AS. A similar symptom complex may occur as a result of a compressive lesion of the cauda equina such as a spinal tumour or other extradural mass lesion, including an epidural haematoma which may occur in relation to a spinal fracture, both of which occur with increased frequency in the patient with advanced AS compared with the normal population. Imaging is thus always required for further evaluation.

A magnetic resonance imaging (MRI) examination of the lumbosacral spine (figs 2–4) confirmed the diagnosis. The sagittal images (fig 2) demonstrate a widened thecal sac with dorsal sacculations filled with cerebrospinal fluid (CSF) and are particularly prominent on the right side at the L3 and L4 levels. The posterior aspect of the vertebral bodies is not affected. On the T2 weighted (T2W) image (fig 2B), intermediate signal nerve roots can be seen descending in the high signal CSF. On the axial T1 weighted (T1W) image (fig 3A) there is marked erosion of the right lamina into which projects the dorsal diverticulum. The axial T2W image (fig 3B) shows the intermediate signal nerve roots to better advantage deviating towards the diverticulum. The nerve roots appear free.

The dorsal diverticula do not encroach on the intervertebral foramina, which are shown to be widely patent with in addition no evidence of any bony encroachment (fig 4).

The diagnosis is that of a cauda equina syndrome complicating longstanding ankylosing spondylitis.

### Discussion

Ankylosing spondylitis may be completely asymptomatic and diagnosed incidentally when a radiograph is performed for some unrelated reason. It may also re-present after a period of quiescence, or present late with complications (table). The cauda equina syndrome (CES) is one such rare complication. First described by Bowie and Glasgow,<sup>1</sup> this occurs in longstanding disease, usually of more than 15 years duration when disease is inactive and the patient typically has been symptom-free for many years. Symptoms are

#### Neurological complications of ankylosing spondylitis

Cause	Complication
Instability	Atlanto-axial subluxation Fracture-dislocation Pseudoarthritis
Inflammation	Cauda equina arachnoiditis Single root lesions
Compression	Ossified intraspinal ligaments Granulation tissue Foraminal stenosis

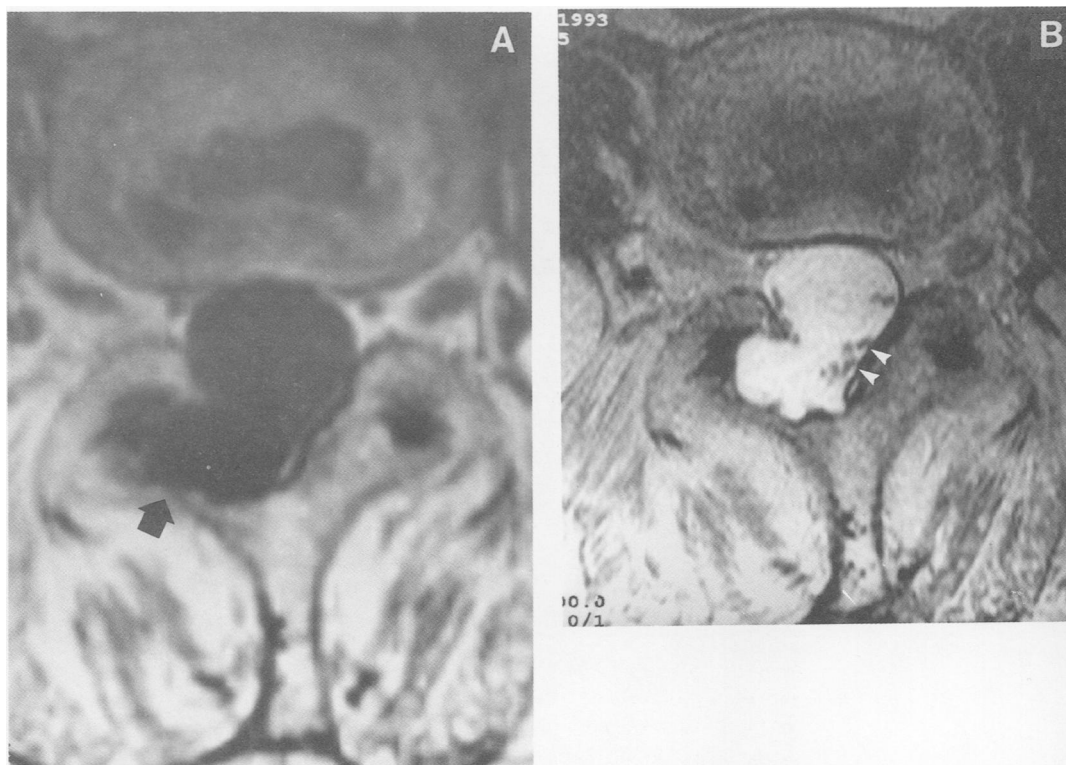


Figure 3 Axial T1W image showing marked thinning of the lamina of L3 (arrow), with a dorsal diverticulum extending into the eroded area. B: Axial T2W image showing high signal CSF filling the diverticulum, and the intermediate signal nerve roots deviating towards the diverticulum (arrowheads).



Figure 4 Sagittal T1W image through the right intervertebral foramina, demonstrating that there is no diverticular or bony encroachment.

usually sensory in nature, affecting lumbosacral nerve roots and frequently include urinary and bowel symptoms. Motor symptoms, if present, are usually mild. Despite its relative rarity, it is a condition which has been well described, particularly as a result of the newer imaging techniques of computed tomography (CT) and magnetic resonance imaging (MRI).<sup>2-9</sup>

The hallmark of CES in AS is the presence of dorsal dural diverticula associated with a widened thecal sac and scalloped erosions of the laminae and spinous processes of the lumbar spine. Erosions of the laminae may be detected on plain radiographs, but this is often in retrospect as they are difficult to appreciate unless looked for specifically, as in this patient and that of Young *et al.*<sup>8</sup> In the past, imaging assessment relied on myelography which is fraught with problems because of the difficulties in puncturing the theca in the presence of bony ankylosis and soft tissue calcification, and difficulty mobilising the rigid patient to visualise the dorsal diverticula. CT is undoubtedly the imaging modality of choice to show bony fusions and, more specific to the CES, the scalloped and eroded laminae and spinous processes. CT will show the CSF filled dorsal diverticula but MRI, with its inherent soft tissue contrast, is far superior in this regard. While T1W images are satisfactory, the T2W scans will not only show the high signal CSF in the outpouchings, but also exquisitely demonstrate the course of the nerve roots and will show their location within the spinal canal and also their position relative to each other (figs 2B, 3B). Intravenous paramagnetic contrast medium (Gadolinium-DTPA) was not given at this MRI examination. Other cases

in which it was administered did not demonstrate any abnormal enhancement.<sup>5 7</sup>

The actual cause of the CES in AS has not been specifically defined, although various theories have been proposed. The nerve root symptoms are presumably related to the structural changes seen in the spinal canal. An arteritis and demyelination have been suggested as causes of the syndrome, but these seem unlikely to account for the laminar erosion and dural ectasia. Arachnoiditis, occurring early in the disease, does, however, seem to play an important role in problems that may later develop. Tullous *et al*<sup>5</sup> suggested that primary ligamentous inflammation in AS leads to inflammation of contiguous structures such as the meninges and dorsal bone elements of the spine. This produces dorsal meningeal inflammation with adhesion formation. Such arachnoid adhesions may lead to blind pouches into which CSF is forced by the pumping action of arterial pulsation, with consequent enlarging arachnoid cysts together with pressure erosion of the adjacent bone.<sup>10</sup> A resorption defect of the CSF was demonstrated by Confavreux *et al*.<sup>11</sup> CSF resorption normally occurs through arachnoid villi in epidural veins. If this does not occur, then this may contribute to thecal sac enlargement and other structural changes, and rapid CSF pressure fluctuations may thus not be damped efficiently and this could contribute to nerve root injury. The structural changes primarily affect the lumbar region, but may also be present in the thoracic region.<sup>12</sup> Inflammatory changes and arachnoid adhesions are likely to occur also in the cervical spine, but here, pressure effects will be less than in the lumbar region and hence diverticula formation and bony pressure erosion are unlikely to occur. A postmortem study of the lower spinal canal with CES showed that while many nerve roots were normal, others showed fibrosis and loss of myelin.<sup>10</sup> Appearances of the nerve roots at operation include fibrosis and chronic arachnoiditis embedding atrophic nerve roots,<sup>13 14</sup> with small sacral roots splayed and adherent to the dura.<sup>15</sup> Fibrous tissue and scattered lymphocytes surrounding the nerve roots have also been described.<sup>16</sup> It does appear, therefore, that there is a varied picture of nerve root damage in the cauda equina as a result of fibrosis, variable demyelination and atrophy. The dorsal diverticula did not encroach on the intervertebral foramina in this patient, as has been shown by others<sup>13 17</sup> (fig 4), or compress the nerve roots, although such compression has been reported in three cases<sup>7 18 19</sup> and postulated in one.<sup>20</sup>

No specific management for these patients has been defined. Surgery, with decompression of the cysts, either at laminectomy<sup>7 18 21 22</sup> or via a lumbo-peritoneal shunt<sup>11 20</sup> has met with varying success. It seems likely, however, that while the cysts may contribute to the pathogenesis of the condition by interfering with pressure gradients of the CSF, an equally and perhaps more important factor is the result of an inflammatory process that became inactive some long time ago, leaving a residuum of irreversible change.

CT and MRI are complementary imaging techniques in the evaluation of the patient with AS and neurological complications. While MRI has the advantages of superb soft tissue contrast, the multiplanar facility and absence of irradiation, problems may be encountered either where there has been previous surgical instrumentation giving rise to extensive signal void and artefactual change, or with the kyphotic patient who may not fit into the scanner. In the former case, CT will not be useful either, and this may be the rare occasion when myelography may still be required, depending on the clinical question being asked. In the latter situation, imaging the patient with CT in the lateral decubitus position, together with sagittal reconstruction, may provide the required information.

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