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Transverse myelitis in a patient with long-standing ankylosing spondylitis

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ABSTRACT

Ankylosing spondylitis is reported to involve not only the joints but other organs as well. Among these extra-articular involvements, uncommon complications associated with nervous system such as single root lesions, compression of the myelum and cauda equina syndrome have also been documented. Here we present a patient with long-standing ankylosing spondylitis who developed spastic paraparesis. Extensive study to find the cause of a spastic paraparesis failed and therefore led to the conclusion that this patient was suffering from transverse myelitis. Similar reports in the past have been attributed to an association with multiple sclerosis; however, we suggest that the findings support the diagnosis of a rare complication of ankylosing spondylitis with an unknown etiology.

Introduction

The neurologic complications of ankylosing spondylitis (AS) are rare and include solitary lumbosacral or thoracic nerve root lesions, spinal cord compression secondary to atlantoaxial subluxation, and injury to the spinal cord due to the susceptibility of the rigid spine to trauma (1-3).

Rarely, sensory and motor root signs arise due to the cauda equina syndrome (4-6). The pathogenesis of the cauda equina syndrome in AS remains unknown but may be due to demyelination, post-irradiation ischemia, or compression from spinal arachnoiditis. Single root lesions with pain resembling sciatica are described, but there is little evidence of persistent damage to single lumbar roots (3). Some reports have suggested an association between ankylosing spondylitis and multiple sclerosis (3, 7-10).

We present a case of transverse myelitis with a favourable outcome after steroid treatment in a patient with long-standing AS.

Case report

A 26-year-old man with a 14-year history of ankylosing spondylitis presented in 1999 with the rapid onset of motor weakness and a tingling sensation in both legs. He also complained of difficult voiding and fecal incontinence. Symptoms developed rapidly over several days, but then remained static. He had previously had uveitis, but no diplopia or other visual problems. There was no fever and he did not smoke or use alcohol. He was otherwise well with no significant medical history. He was treated with indomethacin, sulfasalazine, and methotrexate for his lower back pain and morning stiffness. He had never received radiotherapy. He had no family history of arthritis or neurological disease.

On examination, his vital signs were normal. He had a fixed spine with an antero-posterior Schober index of 1.5 cm. The distance from ground to finger was 18 cm. The wall to occiput distance was 6 cm. A chest expansion test showed 2.5 cm. He also had increased thoracic kyphosis characteristic of ankylosing spondylitis and slight restriction of flexion and rotation of his cervical spine. His pupils reacted equally to light and a fundoscopic examination was normal. There was no abnormality in his upper limbs and no wasting of thigh and calf muscles. He showed paraparesis, sensory loss below the level of the 10th thoracic cord, bilateral hyperreflexia and bilateral extensor plantar responses in lower extremities.

Radiography of the cervical, lumbar and dorsal spine revealed typical findings of AS, including vertebral body squaring, syndesmophytes, apophyseal joint sclerosis, ankylosis and atlantoaxial subluxation. Cardiovascular examination was normal and blood pressure 120/80. Full blood count was normal with slightly increased erythrocyte sedimentation rate of 31 mm/h (Wintrobe method). Analysis of the cerebrospinal fluid revealed a mild lymphocytic pleocytosis and elevated myelin basic protein (clear color, protein 0.2 g/l, glucose 67 mg/dl, no oligoclonal bands, negative VDRL). HLA B27 was positive. Viral assays for infectious agents such as influenza, measles, varicella, rubeola, mumps, Epstein-Barr virus and cytomegalovirus and mycoplasma were negative. Cranial magnetic resonance imaging (MRI) was normal. MRI of the cervical, thoracic, lumbar spines showed a normal cord with no

extradural compression. There was no evidence of malignancy. The visual evoked potential (VEP), brainstem auditory evoked potential (BAEP) and median nerve sensory evoked potentials (MNSEP) were normal but the posterior tibial nerve sensory evoked potential was abnormal. Although the MRI findings did not show significant changes, neurological symptoms and examinations suggested transverse myelitis. High dose steroid pulse treatment was effective with rapid improvement leading to complete clinical recovery after four weeks.

After 6 months of follow up, he is maintaining good performance in daily life and no new neurological symptoms or signs have emerged.

Discussion

AS is a chronic inflammatory disease that primarily affects the spine. The nervous system may be involved in longstanding AS as a result of atlantoaxial subluxation or spinal fractures and by development of a cauda equina syndrome. In addition, an increased incidence of MS in patients with AS has been reported (3, 7-10). We have described a patient with longstanding AS who developed a sudden spastic paraparesis. Since there were no features of extrinsic compression on MRI and a lymphocytic pleocytosis in CSF study, these appear to be due to transverse myelitis. The patient had not received radiotherapy, which has been reported to cause fibrosis of the spinal cord, nor were there other features of active inflammation. He showed an increase in CSF myelin basic protein, and it could be suggested that the disease process is associated with demyelination. Although demyelination or vascular events are possible etiologies, the nature of the cord lesion is still not clear.

Unlike the spinal cord vasculitis that has been reported in polyarteritis nodosa and systemic lupus erythematosus (11), bony impingement on the spinal arteries feeding the cord is suggested to play a major role in the vascular event in the AS.

Up to 40% of transverse myelitis cases are associated with an antecedent infection or recent vaccination. Many infectious agents have been implicated, including influenza, measles, varicella, rubella, mumps, and Epstein-Barr virus and cytomegalovirus, as well as, mycoplasma (14). However, none of the infectious agents mentioned above were isolated from our patient and there wasn't any specific clinical symptom indicating infection. Moreover the patient did not have any predisposition to vascular disease, such as hypertension, diabetes or cardiac problems.

It is difficult to demonstrate a definite association between MS and AS, as many patients are probably never seen at a hospital or may, in fact, never seek medical care. Neither the oligoclonal band in the CSF nor cerebral plaques on brain MRI were found in our patient and therefore he failed to meet the diagnostic criteria for MS. Furthermore our patient showed a normal VEP, BAEP, and MNSEP. It is unlikely that he represents a case of a subclinical manifestation of a spinal form of MS. The possibility remains that he may have had localized spinal cord demyelination which was not visible on MRI.

The coexistence of AS with MS has been reported to occur at a much higher incidence than might be expected by chance (3, 7-10). Thomas *et al.* reported 2 cases of MS among 45 patients with AS. In addition they identified a case of spastic paraparesis in a patient who had received radiotherapy 20 years before and suggested this interval was too long to be attributed to radiation myelopathy (7). Khan and Kushner studied 196 patients with long-standing AS and reported 2 cases of MS as well, which showed widespread central nervous system involvement and classical transient attacks, but without laboratory or radiological confirmation (8). However, sufficient evidence is not currently available to support a disease association of AS and MS.

Atlantoaxial subluxation, a well recognized complication in about 2% of AS patients, was noted in this case. The prevalence of atlantoaxial subluxation in ankylosing spondylitis was higher than previously reported in other settings (13). Although clinically significant neurological complications are not frequent in patients with AS, atlantoaxial subluxation may cause severe spinal cord compression requiring surgical fusion. However, findings on MRI did not show any sign of specific cord compression due to atlantoaxial subluxation and the patient did not complain of any neurological symptoms.

Appropriate diagnosis for this disorder has not been documented up to date, but various clinical findings support the diagnosis of transverse myelitis in AS. Therefore, we suggest that transverse myelitis may be a separate neurological manifestation associated with AS.

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