The pathogenesis of the cauda equina syndrome of ankylosing spondylitis is unknown. The long duration between the onset of ankylosing spondylitis and neurological symptoms (average 35 years in the Mayo Clinic series) suggests an initial injury and subsequent chronic inflammatory or degenerative processes. Such a slow process might produce not only the bony erosion and arachnoid diverticula, but also contribute to neural damage. Atrophy of peridural tissues and adherence of dura to adjacent structures, as documented at operation1 and pathologically,4 might reduce elasticity and compliance of the caudal sac so that it can only accommodate to a limited extent without significant changes in intraspinal pressure. In the presence of excess pressure, CSF may fluctuate and, over time, cause nerve root alterations, including adhesion and ependymal proliferation, which might contribute to the pathogenesis of the cauda equina syndrome.

A review of previous cases of cauda equina syndrome associated with ankylosing spondylitis has indicated that neither steroid treatment nor surgical exploration is of proved utility.1 Moreover, instances of clinical deterioration after surgical intervention on the spine have been documented. Neurological improvement after L-3-L-5 laminectomy and marsupialisation of arachnoid cysts has been reported, but in this single case there was evidence of compression of the spinal cord.5,6 Cytolysis and cellularity, often unrecorded in the idiopathic cauda equina syndrome, were found only in ankylosing spondylitis.7

The use of lumboperitoneal shunting is established for the treatment of idiopathic intracranial hypertension and cranial cerebrospinal fluid fistulae, but previous reports of its use in the cauda equina syndrome of ankylosing spondylitis have not been found. In view of our clinical findings, and the desirability of avoiding radical surgical intervention on the spine in ankylosing spondylitis, we suggest that lumboperitoneal shunting merits consideration in patients with ankylosing spondylitis presenting with an idiopathic cauda equina syndrome. If excessive CSF pressure fluctuations are important in pathogenesis, a case may be made for early surgical intervention by lumbo-peritoneal shunting in ambulant patients before the development of nerve damage.

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Diffuse neurofibriillary tangles with calcification in a non-demented woman

In 1994 we proposed the term “diffuse neurofibriillary tangles with calcification” (DNCT) for a new form of presenile dementia.8 This disease is clinically characterised by progressive cortical dementia. Neuropathological features consist of temporal lobe calcification, neuronal loss and astrocytosis, numerous neurofibrillary tangles spread throughout the cerebral cortex but lacking senile plaques, and Fahr’s type calcification. Recently we have reported a case of DNCT without evidence of dementia, and pointed out that DNCT is not necessarily associated with dementia.9 We have experienced a similar case.

A 64-year-old woman was admitted to a mental hospital with anxiety attacks and hypochondrial complaints. Despite mild memory disturbance, dementia was not detected. She had hypochondria and delusions of persecution. She was dependent, and often displayed a negativistic attitude. Personality changes were considerable. At the age of 70 years, she fell down and soon died. At necropsy, there was severe cerebral oedema (the cause of death) was found. The brain weight was 1265 g. Bilateral temporal atrophy was not so severe as in our previous patients with DNCT. Numerous neurofibrillary tangles were present in the hippocampus, entorhinal and transentorhinal cortex, and amygdala, but sparsely distributed in the neocortex. No senile plaques were found. Fahr’s type calcification was present. Because we had no evidence of dementia, this case was not clinically diagnosed as having DNCT.

In this case, neuronal loss and neurofibrillary tangles, which are thought to contribute to dementia, were much less obvious than those in our previous patients with profound dementia. Therefore, we diagnose this patient as having early stage DNCT.

Although Langlois et al did not describe the detailed distribution and degree of neurofibrillary tangles in their patient, it is possible that their case also exhibited early stage DNCT.

As we pointed out, all reported cases except one were Japanese. Recently, DNCT has received considerable attention, and more clinically diagnosed cases of DNCT have been reported in Japan.10,11

The CT and MRI findings, consisting of localised temporal or temporofrontal atrophy and pronounced pallidal and cerebellar calcification, are so characteristic of DNCT that clinical diagnosis is not difficult. More cases of DNCT are expected to be reported, probably from other countries as well as Japan.

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NEUROLOGY

Neurology of adult α-mannosidosis

Neurological findings in adult α-mannosidosis are cerebellar dysfunction, absent tendon reflexes, spasticity, and mental retardation.1,2 We present the most extensive long-term follow-up of patients with α-mannosidosis. We report on three adult patients who had been born to affected parents.

In 1995, a 34 year old white man was seen for evaluation of progressive gait ataxia. Clinical data for this patient at the age of 4 years were published.3 He had presentable milestones from 4 months on; the patient did not sit without support until 2 years, and he first walked and spoke single words at 3 years of age. During early childhood, he was clumsy.

In 1967, mental retardation (IQ 60), hepatosplenomegaly, dysostosis multiplex, coarse facial features, severe deafness, but no ophthalmological abnormalities were described.4 In 1994, MRI displayed periventricular hyperintensities and a left occipital subdural haematoma.5 Based on these findings “lipomucopolysaccharidosis”, subsequently called mucolipidosis I, was diagnosed.6 After a follow up investigation at 12 years, this diagnosis was abandoned. The patient was then classified as having α-mannosidosis.7

Since 1967, the patient has lived with his family and is now employed in a sheltered workshop. The parents were of Ukrainian origin and were first cousins. His three sisters, aged 35, 39, and 40 years are clinically healthy. The patient was mentally retarded but with an alert and pleasant personality, with brachycephaly and coarse facial features (prominent forehead, hypertelorism, wide spaced teeth, and a flattened nasal bridge). His height was 170.5 cm and his weight was 62 kg. He needed assistance to sit up and crutches for walking. He had pronounced kyphoscoliosis. Blood pressure and pulse were normal. Neurologically, there was no deficit on oculoc and facial motor testing, pupils reacted normally on both light and convergent gaze. There was bilateral deafness. The patient had slurred speech, clumsy tongue movements, and spoke sentences of only one to two words. Muscle power and tone were normal but the thigh muscles were wasted.
Diffuse neurofibrillary tangles with calcification in a non-demented woman.

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