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) argues against a shared inflammatory process, and does not suggest immune abnormalities. Matthews suggested that arterial pulsations transmitted to the CSF might produce not only the bony erosion and arachnoid diverticula, but also contribute to neural nerve damage. Atrophy of peridental tissues and adherence of dura to adjacent structures, as documented at operation and pathologically, might reduce elasticity and compliance of the caudal sac so that sudden increases in Cauda equina pressure fluctuations. Such excessive pulse pressure in CSF may, over the course of many years, produce the arachnoid diverticula and bony erosions and also have a deleterious effect on nerve roots. The impairment of the cauda equina syndrome more often afflicts those with mild ankylosing spondylitis who remain ambulant may be a reflection of this pathogenetic mechanism. Intrathecal shunting for subarachnoid bleeding may also suggest such pathological pressure oscillations and hence might retard progression of the neuropathy.

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. Moreover, instances of clinical deterioration after surgical intervention on the spine have been documented. Neurological improvement after L3-5 laminectomy and marsupialisation of arachnoid cysts has been reported, but in this single case there was evidence of compression of thecal sac. Therefore, it is distinctly unusual to find in the idiopathic cauda equina syndrome of ankylosing spondylitis.

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.

Thanks are due to Dr Tony Finnegan, Sally Oak Hospital, Birmingham, for electrophysiological studies, and Dr Mark Davies, Woodlands Hospital, Birmingham, for MRI scans.

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Diffuse neurofibribrillary tangles with calcification in a non-demented woman

In 1994 we proposed the term “diffuse neurofibrillary tangles with calcification” (DTNTC) for a new presentation of presenile dementia.1 This disease is clinically characterised by progressive cortical dementia. Neuropathological features consist of temporal lobe, orbital and transentorhinal cortical atrophy, diffuse neurofibrillary tangles spread throughout the cerebral cortex but lacking senile plaques, and Fahr’s type calcification.

Recently we reported a case of DTNTC without evidence of dementia, and pointed out that DTNTC is not necessarily associated with dementia.2 We have experienced a similar case.

A 64 year-old woman was admitted to a mental hospital with anxiety attacks and hypochondriacal 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 generalised cerebral oedema (the cause of death) was found. The brain weighed 1250 g. Bilateral temporal atrophy was not so severe as in our previous patients with DTNTC. 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 of the lack of evidence of dementia, this case was not clinically diagnosed as having DTNTC.

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 DTNTC.

Although Langstrom 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 DTNTC.

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

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

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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 a patient with adult α-mannosidosis. The patient was 30 years old at his first visit, and neurological examination revealed no abnormalities. However, he presented with somnolence and obtundation from the late teenage years on, and then classified as having α-mannosidosis.1 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 brachycephalia and coarse facial features. Based on these findings “lipomucopolysaccharidosis”, subsequently called mucolipidosis I, was diagnosed.1 After a follow up investigation at 12 years, this diagnosis was also confirmed and he was then classified as having α-mannosidosis.1

J Neurol Neurosurg Psychiatry: first published as 10.1136/jnnp.61.1.116-a on 1 July 1996. Downloaded from http://jnnp.bmj.com/ on May 12, 2022 by guest. Protected by copyright.
Electromyography of the serratus anterior and subscapularis muscles: description of a technique

Weakness of the muscles of the shoulder girdle, especially of the serratus anterior muscle, results in winging of the scapula and is most often caused by neuralgic amyotrophy or degenerative myopathies—for example, facioscapulohumeral muscular dystrophy. Several techniques have been proposed for needle EMG of the serratus anterior muscle: The needle can be inserted in the muscle superficially to the fourth or fifth rib in the medial or posterior axillary line. In our experience, this technique usually samples only a few muscle fibres and there is a risk of inducing a pneumothorax by penetrating the intercostal space. Delagi et al suggested inserting the needle just lateral to the inferior angle of the scapula. If the needle is inserted too superficially, the EMG is recorded from the lattissimus dorsi muscle. If the insertion is too cranial the teres major muscle is encountered. As winging of the scapula is the most frequent reason for examination of the subscapularis muscle, which contributes to shoulder stability and internal rotation of the humerus (see figure). Needle EMG of this muscle is usually performed using wire electrodes inserted in the posterior axillary line with the subject in a supine position and the arm held in an abducted and externally rotated position. If the needle is inserted too superficially, it will be in the rhomboid major muscle, which also inserts at the medial border of the scapula. Active adduction of the scapula (rhomboid muscle) and of the humerus (subscapular muscle) allows differentiation of these muscles. In patients with facioscapulohumeral muscular dystrophy, we found more abnormal EMG activity in the subscapular muscle. Thus the investigation of this muscle seems to be of some diagnostic value in patients with winging of the scapula.

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Sagittal MRI (T1 weighted) showing prominent vermian hypoplasia of the cerebellum and enlarged cerebellomedullary cistern.

Tendon reflexes were elicited at a low level and ankle jerks were absent. There were no pyramidal tract signs. A finger to nose test was normal, whereas a heel to shin test was inaccurate. The patient could not rise from the prone position without help. His gait was broad based and ataxic, and tandem gait was not possible. There was no Romberg’s sign. Sensation for touch and pain was preserved. Position and vibration sense could not be tested. Electromyography (M tibialis anterior, M extensor digitorum brevis) and nerve conduction studies (tibial, peroneal and median nerves) gave normal results. Formal testing showed an IQ below 50 on the Columbia mental maturity scale, but an IQ of 90 on the block design test of the revised Wechsler adult intelligence scale. Incipient lenticular opacities were seen on slit lamp examination. Visual evoked potentials were normal. Audiometric examination and acoustic evoked potentials disclosed bilateral combined hearing loss. An EEG disclosed alpha rhythms without seizure activity. Electrocardiography and echocardiography were without abnormalities. Brain MRI showed cerebral atrophy, hypoplasia of the lower vermis, and an enlarged cerebellomedullary cistern (figure). Radiography showed coxa valga deformity with degenerative arthritis in both hips and dysostosis multiplex in his spine and pelvis. Urinary examination showed an abnormal pattern of oligosaccharide excretion; α-mannosidase was very low in serum (0.0268 mU/ml; normal: 0.109–1.173). Other routine laboratory investigations, including measurement of immunoglobulins, gave normal results.

Our patient does not fit the conventional classification, as features of type I (early onset; recurrent infections) and type II α-mannosidosis (prolonged survival) were found. Interestingly, no vacuolated lymphocytes could be found in his or her mother’s blood at follow up on three different occasions. The neuropsychological results are best interpreted as the dissociation between preserved visuospatial abilities and impaired language due to hearing difficulties. Stable scores for general intelligence and visuospatial abilities over time argue against a cognitive decline in α-mannosidosis. Ataxia was progressive also in other cases of adult α-mannosidosis. Cerebellar signs in this condition may be the consequence of precocious age-related attrition in cerebellar neurons due to prior storage of compounds containing mannose.

We are grateful to Professor H-D Weiß, who kindly permitted publication of the MRI.

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*Needle EMG of the subscapular muscle with the upright standing patient pressing his hands against a wall.*