Paraneoplastic necrotising myelopathy in a case of AIDS with lymphoma

Paraneoplastic necrotising myelopathy is a rare and still obscure neurological complication of malignancy. Spinal cord affection is common in patients with HIV infection, but necrosis of the spinal cord in AIDS is rare and usually caused by opportunistic infections. Our patient had AIDS and immunoblastic lymphoma with a necrotizing myelopathy, not due to an infectious or vascular cause.

A 29 year old homosexual patient had been known to be HIV positive since 1987. In 1990 he developed AIDS and Pneumocystis carinii pneumonia. Azidothymidine had to be discontinued due to anaemia. In December 1991 he started complaining of a slowly ascending numbness, which began in both feet and reached the T10 level in April 1992. At the same time he developed a weakness in his legs, dull pain on the right, and reduced pain and temperature sensation in the left leg, as well as left masticatory and mandibular herpetic zoster. Lumbar puncture showed 43 cells/μl, mostly lymphocytes, plasma cells, and several cosinophilic granulocytes, and protein (5.9 g/l). The serum/CSF protein ratio indicated a severe disturbance of the blood-brain barrier. T2 weighted spinal MRI showed thickening and enhanced water content of the thoracic cord, maximum at the T12 level. A spinal infiltration of a lymphoma was suspected, but examinations of bone marrow, repeated lumbar punctures, or radiographs of the lungs did not show evidence for a lymphoma at that time. Repeated serological tests for neurotropic viruses, Lyme disease, Toxoplasmosis, syphilis, borreliosis, cryptococcosis, listeriosis, and candidosis were negative in serum and CSF. The neurological deficits were rapidly progressive and treatment with acyclovir and corticosteroids had no effect. Further MRI a month later no longer showed the intramedullary lesion. The patient was discharged six weeks after admission with a flaccid paraplegia, an incomplete loss of sensibility below T8, and complete incontinence. At the end of September 1992 he was readmitted with a lymphoma of the lung and cerebral involvement. The paraplegia had become spastic and the level of sensibility loss was now at T6. He was still completely incontinent and had decubitus ulceration. The lymphoma was not treated and the patient died two months later of respiratory failure due to pulmonary problems.

Postmortem examination showed an immunoblastic lymphoma of both lung and bronchopneumonia of the left lung, and pleural effusions on both sides. There was cerebral involvement of the lymphoma in both hemisphere and opportunistic infections of the brain and no signs of increased intracranial pressure.

The thoracic meninges stuck to the cord, which was very soft in the thoracic region. The cross section of the cord was confined to the thoracic cord. On both ends of the softened part of the cord there was a roughly 1 cm long region of pencil-like softening. There was no macroscopic evidence for angiitis, vascular obstruction, or tumour infiltration. Histologically, the thoracic cord displayed the aspect of complete coagulation necrosis with few macrophages. No vascular disease, tumour, viral, bacterial, fungal, or parasitic infection was detected in the spinal cord. No fibrin thrombi were seen despite a study of numerous sections. Only a few mononuclear inflammatory infiltrates were present. These were generally well preserved, whereas the anterior roots showed a substantial loss of myelinated nerve fibres (figure). In the cervical cord, the fasciculus gracilis exhibited a complete loss of myelinated nerve fibres, whereas the fasciculus cuneatus was not affected. The lumbar and sacral cord was well preserved. Immunohistochemistry for herpes simplex 1 and 2 viruses and cytomegalovirus as well have had affected the dorsal roots. The histological features were identical with paraneoplastic necrotising myelopathy, first described by Nonne1 quoted in Folliis and Netky2. Although the lymphoma was not discovered at the first admission, it is likely to have existed at that time. To our knowledge paraneoplastic necrotising myelopathy has not been described in AIDS before.

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Total tau protein immunoreactivity in lumbar cerebrospinal fluid of patients with Alzheimer's disease

Alzheimer's disease is the most frequent cause of dementia in elderly people. The definite diagnosis can only be established by (semi) quantitative histopathological examination of neurofibillary tangles. Neurofibrillary tangles are com posed of three, men, mean age 68.3 years (SEM 4.2 years) with major depression (DEPR) according to ICD-10 and DSM-III R; (3) 20 patients (11 men, nine women, mean age 58.7 (SEM 3.4) years) with other neurologi cal disorders without AIDS (4 and 5) 16 young subjects (12 women, four men, mean age 37.8-2.5 years) and 10 elderly subjects (five women, five men, mean age 62.3 (SEM 3.9) years) without CNS disease, in which lumbar puncture was performed for exclusion purposes. There was a highly significant (P < 0.005) increase in concentrations of tau protein in the Alzheimer's disease group compared with the elderly depressive patients, patients with other neurological disorders, and young and elderly controls (figure). The elderly depressive patients had a non-significant increase compared with the young controls, the difference compared with young controls was significant (P < 0.01). Elderly and young controls were not different. Patients with other neurological disorders had significantly higher values (P < 0.005) than both young and elderly controls. There was a significant negative correlation of MMSE score with total tau protein concentrations when all patients with Alzheimer's disease were included (r = -0.6106, P < 0.01). However, the correlation was non-significant (r = -0.2830, P > 0.05) when only patients with MMSE greater than 10/30 points were tested.

There exists, hitherto, no study of total tau protein immunoreactivity in CSF of neuropsychiatric disorders which includes all of the patient groups investigated in the pre-

Bilateral infarction of the rostral pontine tegmentum as a cause of isolated bilateral supranuclear sixth nerve palsy related to hypertension

The difficulty in locating in vivo the paramedian pontine reticular formation (PPRF), which is the immediate premotor centre for saccade generation, has been a major obstacle to a better clinical understanding of many neuro-ophtalmological syndromes.1 2 Lateral rectus weakness from a pontine lesion is usually associated with other neurological signs.1 We report a hypertensive man with an isolated gaze disorder consisting of a severe bilateral abduction palsy and a subclinical bilateral gaze palsy—that is, bilateral conjugate slowness of saccades—with no other identifiable aetiology, who had a bilateral pontine infarction involving the rostral PPRF, seen on MRI.

A 64 year old man with a history of mild hypertension and hyperlipidaemia was referred for evaluation of painless horizontal diplopia of sudden onset. There was no history of head trauma, diabetes mellitus, or any other systemic disorder. Blood pressure was 150/100 mm Hg. Neurological examination was normal except for a complex type of gaze deficit—namely, mild bilateral conjugate slowness of saccades with an additional severe right lateral rectus weakness and pronounced esotropia of the left eye on straight gaze. No nystagmus or internuclear ophthalmoplegia was seen and the rest of the extraocular movements were full. “Doll’s head” manoeuvre (vestibular ocular reflex) in the direction of the palsy was intact. Pupil sizes and light reflexes were normal in both eyes. Sensation in the first division of the trigeminal nerve was intact. Facial and masticator muscles were of normal strength and there were no signs of long tract involvement. Results of routine laboratory tests, including CSF analysis, were unremarkable. Brain MRI (0-5 T) showed only minor bilateral hypointensities at the upper pontine tegmentum, without definite relevance. One week later he suddenly complained of dizziness and a more intense diplopia on horizontal gaze. His neurological examination showed a newly developed complete paraly- sis of abduction of the left eye. A second MRI disclosed bilateral hypointensities at the rostral pontine tegmentum compatible with infarction (figure). The oculomotor function resolved two months later with only a persistent mild bilateral slowness of saccades and no lateral rectus weakness.

Our patient’s brain MRI showed a bilateral infarction with involvement of the rostral pontine tegmentum containing the PPRF, the structure responsible for the gen-

Axial T2 weighted MRI obtained with a 0.5 Tesla unit shows bilateral hypointensities at the rostral pontine tegmentum compatible with infarction.
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*J Neurol Neurosurg Psychiatry* 1996 60: 237-238
doi: 10.1136/jnnp.60.2.237-a