Coronal T2 weighted MRI showing a high signal intensity as a cortical area in the inner bank of the left parietal operculum.

Two somatosensory projection areas, S I and S II, have been shown to exist in the brains of mammals. 

Possible cortical overlap between postencephalitic Parkinsonism and progressive super-nuclear palsy.

Encephalitis lethargica is a transmissible disease of the CNS which occurred as a pandemic between 1916 and 1928 and subsequently a few sporadic cases have been reported. Close neuropathological similarities exist between postencephalitic Parkinsonism and progressive super-nuclear palsy though the clinical presentation is quite different. Particularly, postencephalitic encephalitis lethargica in childhood, developed Parkinsonism and later, progressive neurological disability, including frequent falls, neck extension, and super-nuclear down gaze paralysis. Despite the dire nature of progressive super-nuclear palsy on clinical examination, the neuro-otological investigations showed some distinctive features.

In 1917, aged six years, the patient was diagnosed as having “sleepy sickness”, from which he seemed to recover fully. In 1959, 42 years after his encephalitis lethargica, he began to have excessive sleepiness such that he might fall asleep while playing cards or when at work. At about the same time, he was also noted to have indistinct speech and a resting tremor of his right hand and oculo- gyric crises occurred. Three years later he developed a parkinsonian rigidity and he was noted to have pinpoint pupils with full extraocular movements, slow dysarthric speech, extrapyramidal signs in his limbs, and sluggish limb reflexes. His Parkinsonism remained static until 1986, when he started on a small dose of levodopa with benefit. In 1979, aged 68, he began to fall and developed troublesome blepharospasm, with impairment of extraocular movements. In 1981, he had a small craniotomy and after an excessive dose of sinemet. In 1986, he was readmitted to the National Hospital for Neurology and Neurosurgery for further investigation. His postural reflexes were severely impaired. He was extremely slow to initiate gait, but once started he could walk fairly fluently and with only mildly reduced steps. A general poverty of facial expression and bradykinesia, together with a resting tremor of the lower jaw and right hand were noted. There was pronounced axial rigidity with neck extension, but power, coordination, and limb reflexes were normal, with the exception of absent ankle jerks and extensor planter responses. Speech was slow and monotonous. The pupils were pinpoint, but circular in shape, and reacted briskly to both direct and consensual light. There was a bilateral blepharospasm triggered by glabella tapping, together with increased facial jerks, were noted.

Neuro-otological examination disclosed smooth pursuit eye movements in the horizontal plane of about 30° to 40° to both right and left, but only 10° upwards and no downward pursuit from the midposition. Doll’s head manoeuvres significantly impaired both the horizontal and vertical eye movements, indicating super-nuclear involvement. Volitional saccades were absent in the vertical plane, whereas slow, stepped saccades were elicited at about 45° right and left of the midposition. There was no convergence and no spontaneous or positional nystagmus. Optokinetic nystagmus showed a good response to clockwise and ocular beats on counter-clockwise rotation, but there was no response in the vertical plane. Horizontal eye movements were recorded with conventional DC electro-oculography. Hypometric, slow saccades were pronounced down gaze nystagmus lasting four minutes, particularly at the lowest frequency (0.2 Hz; peak velocity 20°/s), but at higher frequencies and velocities, there was a symmetric large reduction of smooth pursuit (gain 49% at 0.3 Hz and 39% at 0.4 Hz). Small, dysrhythmic, but preserved optokinetic responses were found on both clockwise (gain 82%) and counter clockwise (gain 77%) stimulation. Rotary chair induced extra–ocular and super–nuclear and lateral gaze evoked nystagmus, but neither were found in the presence or absence of optic fixation. Vestibulo-ocular function tests showed a deviation of the eyes to the side opposite the visual stimulus, but no nystagmus was seen. Caloric testing showed large amplitude responses in the horizontal plane, with no impairment of the responses to the left side of optic fixation. Bilateral irrigation at 20°C for 40 seconds resulted in three minutes of pronounced upbeat nystagmus, whereas bilateral irrigation at 44°C showed pronounced down gaze nystagmus lasting four minutes. Auditory brainstem evoked potentials showed waves of small amplitude with bilateral delay of wave III and V consistent with eight nerve or intracranial brainstem dysfunction. The percentage of extra-ocular movements was 40% at 0.3 Hz and 39% at 0.4 Hz and mild hydrocephalus consistent with cerebral atrophy. During the patient’s admission sinuset was stopped, which did not lead to any appreciable deterioration in the patient’s condition. He died of extrapyramidal crisis on October 1987, aged 76, he developed a marked alteration in his affect in that whereas throughout his life he had always complained of many symptoms he rather uncomplainingly and extremely anxious. He died the next year, aged 77. No postmortem examination was carried out.

Progressive super-nuclear palsy primarily affects the saccadic system; vestibular and optokinetic stimuli typically produce a tonic drift of the eyes in the direction of the slow phase of the induced response with no saccadic intrusions to produce the skew nystagmic pattern. With progression of the disease, reflex eye movements become deranged as the ocular motor neurons themselves become involved. Various eye signs have been reported, including encephalitis lethargica. Symptoms and signs localised to abnormalities in the pons and midbrain, and characteristic of progressive super-nuclear palsy, have been described in the acute stage of encephalitis lethargica, but, although various oculopalys have been noted in postencephalitic Parkinsonism, the characteristic progressive super-nuclear eye move-
ment abnormalities seen in progressive supranuclear palsy have not been described. Although ocular findings in our patient suggest a progressive supranuclear palsy, the atypical findings of caloric and optokinetic nystagmus, rather than tonic slow eye deviation, suggest preservation of the lateral rectus muscles in this system, enabling the production of reflex saccades, but disconnection of these rectus muscles from the frontal and parietal regions controlling voluntary saccades. These findings are not usually noted in progressive supranuclear palsy.

There are various diagnostic possibilities in this case. Increased neurological disabilities in later life attributable to basal ganglia dysfunction are described in patients with postencephalitic parkinsonism. However, this hypothesis could not explain the loss of responsiveness to levodopa and the emergence of severe gaze palsies in this patient. A further possibility is that he had postencephalitic parkinsonism and then developed unrelated progressive supranuclear palsy. This, however, would seem rather unlikely in view of the different clinical pictures. Our patient could have progressive supranuclear palsy, but in the light of his history of an encephalitis, ocular crises, a resting tremor and, in particular, the very long duration of the disease with, together with the atypical neuro-otological findings, this diagnosis alone would seem unlikely. Finally, there are reports in the medical literature of other conditions such as diffuse Lewy body disease with considerable impairment of volitional saccades, but relative preservation of reflexive saccades, together with sparing of smooth pursuit function. Similarly, supranuclear palsies mimicking progressive supranuclear palsy have been documented in vascular disease.

We are unable to classify this patient within one of the currently accepted categories of parkinsonian syndromes. The case raises some unanswered questions regarding the range of pathological effects of the encephalitis lethargica virus, the natural history of postencephalitic parkinsonism almost 60 years after the original infection, the effect of environmental or other infectious agents on the natural history of this condition. Thorough clinical, neuro-otological, and neuropathological investigation of similar cases may clarify these issues.

Failure of presumed hepatic myelopha-

In March 1992, over a period of about one month he developed stiffness and weakness of both limbs, which have persistently worsened. In August 1992, his liver function tests: bilirubin 26 mmol/l (normal < 17), alkaline phosphatase 202 u/l (normal 30-140), albumin 28 g/l (normal 35-50), alanine aminotransferase 45 u/l (normal < 40), and gamma glutamyl transferase 281 u/l (normal < 52). There was no evidence of viral hepatitis. Serum very long chain fatty acids were slightly raised but the skin fibroblast assay was normal, which excluded adrenoleucodystrophy. A liver biopsy confirmed microcystic cirrhosis, consistent with alcohol related liver disease, and an angiogram showed severe splenomegaly and a major portosystemic shunt from the left gastric vein to the azygous vein.

There was no response to the combina-
tion of a protein restricted diet and lactulose or to attempts to reduce the flow through his shunt with oral nitrates or to symptomatic treatment with baclofen. Despite increasing doses of diazepam. In August 1993 he had a further episode of reversible encephalopathy, and, since there was no evidence of ongoing alcohol intake, he was accepted for liver transplantation on the basis of liver failure alone. A suitable donor organ became available in December 1993, and a successful transplant was performed without major complications. His liver function returned to normal within about 6 months. Despite this, however, there has been no change at all in the function of his lower limbs in the 18 months since his transplant. He continues to be bothered by fatigue despite increasing doses of diazepam and baclofen, and is largely wheelchair dependent.

We think that this is the first patient with hepatic myelopathy who has received a liver transplant. The transplant was performed because of liver failure and from that point of view it has been a success. Disappointingly, there has been no improvement in his neurological symptoms or signs despite normalisation of his liver function. This implies that the spinal cord damage is irreversible, which is compatible with the axonal loss seen in some cases. It is unlikely that there will be any significant neurological improvement at this stage. However, it is unclear whether successful transplantation earlier in the course of the disease when, perhaps, there may already have been demyelination of the cord, would have been beneficial. It is also possible that without transplantation he would have continued to deteriorate neurologically. Finally, it is conceivable that the myelopathy is nothing to do with this patient's liver disease but, within the limits of modern investigation, we can find no other explanation.
Possible clinical overlap between postencephalitic parkinsonism and progressive supranuclear palsy.

P P Pramstaller, A J Lees and L M Luxon

*J Neurol Neurosurg Psychiatry* 1996 60: 589-590
doi: 10.1136/jnnp.60.5.589

Updated information and services can be found at:
http://jnnp.bmj.com/content/60/5/589.citation

**Email alerting service**

Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

**Notes**

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/