pressure.⁵ It should be recognised that in the setting of a cerebellar or brainstem infarction, unilateral proptosis may herald increasing intracranial pressure mandating immediate treatment.

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Normal frontal cortex histology and immunohistochemistry in patients with motor neuron disease

In the light of the association between frontotemporal dementia and motor neuron disease,^{1,2} two recent studies have examined neuropsychological function, and distribution of cerebral blood flow³ or metabolism,⁴ in patients with motor neuron disease without overt clinical evidence of dementia. These authors showed subclinical evidence of the characteristic changes of frontotemporal dementia, with failure on tasks sensitive to frontal lobe function, and hypometabolism, or reduced cerebral blood flow, particularly in the orbitofrontal area.

Brains from patients with motor neuron disease with clinically evident frontotemporal dementia show both microvacuolation of the outer cortical laminae⁵ and ubiquitinated inclusions⁶ in neurons of the superficial layers of the frontal and temporal cortices; inclusions are also seen in hippocampal granule cells.⁶ We have therefore examined the brains of 17 unselected patients referred for routine necropsy to a general pathology department with the clinical diagnosis of motor neuron disease, without dementia. Formal neuropsychological testing had not been carried out. Brains were stained with haematoxylin and eosin for routine histological evaluation, and immunostained for ubiquitin, using standard techniques, for detection of inclusions. Ubiquitinated inclusions typical of motor neuron disease were present in the anterior horn cells of the spinal cord in all cases; frontal cortex was normal, with no evidence of either microvacuolation or inclusions.

Patients with motor neuron disease and frontotemporal dementia usually present with dementia, subsequently developing signs of amyotrophy. It is therefore possible that the neuropathological hallmarks of

microvacuolation and inclusions are a relatively late feature. It will be important to examine pathologically brains from patients with motor neuron disease who have been prospectively tested in life; nevertheless others have failed to demonstrate frontal cortical atrophy in motor neuron disease without dementia,⁷ and it is interesting that patients with motor neuron disease can seemingly have significant neuropsychological and functional deficits without overt evidence of structural or pathological changes.

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Very severe amnesia with acute onset after isolated hippocampal damage due to systemic lupus erythematosus

Neuropsychiatric lupus designates the whole range of behavioural and cognitive impairments occurring in systemic lupus erythematosus. Up to 75% of patients with systemic lupus erythematosus may be affected. Typical manifestations are strokes, seizures, dementia, psychosis, and confusional states.¹ We describe a patient with acute onset of isolated, extremely severe, and persistent global amnesia due to systemic lupus erythematosus.

This 55 year old previously healthy farmer was confused after a nap in the afternoon. He had no known vascular risk factor. At admission, he was alert but confused for time and place and repeatedly asked the same questions about where he was and what had happened to him. Physical examination was normal. He could not repeat three words after a two minute delay. Routine blood tests were normal except for thrombocytopenia ($26 \times 10^9/l$). His CSF had a normal cell count, protein, and glucose content.

His behaviour was remarkable only for a most pervasive amnesia: the patient never

recalled visitors, specifics from test sessions, or daily events. He always recognised his family and long time friends but never the examiners or other people on the ward. He easily oriented himself in his own home but did not find his way around on the ward, where he spent three months. Whereas very remote memory seemed preserved, he was unaware of events of the past 10 to 15 years. He easily recognised the photographs of cattle he had owned more than 15 years ago, but not those he had owned more recently. He never confabulated but always indicated that he did not know the answers to questions. He was initially placid and unconcerned but became depressed after a month; he often cried and complained about his bad memory.

Neuropsychological evaluations established normal oral and written language, arithmetic skills, praxis, finger gnosis, and right-left discrimination as well as constructive abilities, space exploration, and visual recognition of complex material. Intelligence was low average (Wechsler adult intelligence scale, IQ = 91; VIQ = 89; PIQ = 96). Attention and cognitive speed were normal. Executive functions were initially deficient (in particular high rate of perseverations in fluency tasks) but reached the normal range in the course. The main finding was a profound amnesia (table). Although the immediate memory span was normal, he was unable to recall any previously acquired explicit information, independent of the modality of presentation. Both in recall and recognition tasks, he denied any familiarity with the items and had a high rate of false positives in forced choice recognition tasks. An autobiographical interview and a test probing knowledge about Swiss politicians disclosed a temporally graded retrograde amnesia extending 10 to 15 years backwards. Semantic memory was normal, as evidenced by normal naming and verbal fluency. Motor learning in a mirror drawing task was normal.

Brain MRI was performed three times. In the initial MRI 10 days after onset, both hippocampi seemed discretely swollen with blurring of the cortical structure. In the MRIs after three and 10 months, both hippocampi appeared considerably smaller than in the initial scan (figure). No additional lesions were found; in particular, there was no indication of small hemispheric or thalamic vascular lesions.

Transcranial and carotid Doppler ultrasound examination and echocardiography were normal. Thrombocytopenia persisted (15 to $31 \times 10^9/l$). Haemoglobin and leucocyte count, renal function, and liver enzymes were normal. A bone marrow biopsy showed normal haematopoiesis. Antinuclear (1:400), anti-dsDNA, and anti-ssDNA antibodies were raised. No antibodies were detected against ENA-Sm, phospholipid, thrombocytes (anti-GPIIb/IIIa and anti-GPIb/IX), or neurotrophic viruses. No anti-neuronal antibodies were found (sought after five months when antinuclear antibodies were no longer detectable). Complement factor C4 was slightly decreased (135 mg/l), C3 was normal. A skin biopsy contained deposits of IgG, IgM, and C3 along the basal membrane. These findings were consistent with a diagnosis of systemic lupus erythematosus according to the 1982 American Rheumatism Association criteria.

The patient was treated with prednisone (75 mg daily for 12 weeks), cyclophosphamide (150 mg daily for 10 weeks), and