Phosphocreatine (PCr), relative to PDE deteriorates. Angiography, performed hyperaemic (0-98 (0.08) and the SPECT Perfusion hemiplegia counts an area with tissue same (0.04)).

The changes in this patient must have started by day 3 to account for the acidosis seen spectroscopically. The improvement in perfusion by day 25 (despite additional operative trauma, and postoperative vasospasm), suggests that any change can reverse in three weeks. This is similar to the time course of angiographic vasospasm. Striking radiological changes in the large vessels may have been detected away from the possibility of coexistent changes in the small vessels. Such changes may be as important, or in some patients like this, more important than changes in the smaller arteries. This may explain discrepancies between angiographic vasospasm and cerebral ischaemia.

This concept of small vessel changes does not render angiographic vasospasm unimportant. If small vessel resistance increases, any vasospasm affecting large vessels may compromise the circulation much more than if the small vessels are unaffected. Findings of impaired cerebrovascular reactivity accounting for discrepancies between angiographic vasospasm and cerebral ischaemia may also be explained. These adaptive responses occur primarily at a level of the small vessels, so measuring changes in cerebrovascular reactivity may be a functional assessment of the small vessel changes detected in these imaging studies.

In summary, the vascular changes complicating subarachnoid haemorrhage may be more extensive than is appreciated angiographically, extending to the small vessels. If such changes coexist with angiographic vasospasm, this could explain why angiographic changes correlate poorly with clinical ischaemia.


**Neurosyphilis presenting with dissociative symptoms**

A 62 year old man was admitted as an emergency to a medical ward with a five day history of expressive dysphasia. The onset was acute and the dysphasia variable, becoming more pronounced when he was anxious. He had a 20 year history of insulin dependent diabetes, but there was nothing else of note. There was new onset of hypertension. His wife was also in hospital; she had been diagnosed as having probable Creutzfeldt-Jakob disease three weeks before the onset of his difficulties. His deterioration after initial presentation had been rapid, and she also had pronounced expressive dysphasia in the late stages of her illness. Before admission, the man had an episode of expressive dysphasia after visiting his wife in hospital and another while on the ward visiting her. His wife’s resuscitation status and the possibility of a postmortem had been discussed with the patient within 24 hours of the onset of his speech problems.

According to a mental state examination carried out after admission, he was very anxious and agitated; the agitation and dysphasia prevented cognitive and neurological assessment at this point. Physical and neurological examination were normal. Investigations at this time showed that full blood count, plasma viscosity, urea and electrolytes, liver function tests, thyroxine, and thyroid stimulating hormone were all within normal limits; plasma glucose was raised at 14.2 mmol/litre. A chest radiograph and ECG were normal, as was a carotid scan. In view of the circumstances of the presentation, a provisional diagnosis of a dissociative disorder was made and a psychiatric opinion sought.

Ten days after admission, he became increasingly agitated, emotionally labile, and tearful, and his speech was so slurred that cognitive assessment was still not possible. In view of this deterioration, he was transferred to an acute general psychiatric ward. His wife died eight days later. He seemed to take this news well, becoming less agitated and dysphasic. Cognitive function testing at this point showed significant impairment (mini mental state examination = 18/30). Brain CT showed advanced cerebral atrophy with no focal abnormalities, and of note there was no psychiatric activity in the left temporal and anterior regions on the EEG. Autoantibody screening was normal. In view of these findings, he was trans-
ferred to an organic psychiatric unit for further investigation. The dysphasia resolved, but the perseveration in his speech and actions, confabulating frequently, and his behaviour was socially disinhibited. He began to express grandiose ideas. Neuropsychological assessment showed pronounced frontal lobe impairment.

At this point, syphilis serology showed venereal disease research laboratory test (VDRL) positive (1:32), TPHA positive (1:256), and fluorescent treponemal antibody (FTA) positive 4++. Further investigation of CSF showed VDRL positive (1:4), TPHA positive (1:1024), and FTA positive 4++. The radiologic focusing pattern to detect oligoclonal IgG was negative for serum and positive for CSF. A diagnosis of neurosyphilis was made, and oral doxycycline (100 mg thrice daily) was commenced.

His wife did not undergo a postmortem.

In view of the patient’s diagnosis, reexamination of his wife’s stored serum was carried out to repeat test for syphilis in serum was negative. As part of the investigation of her illness, CSF had been examined, showing normal cells and proteins. Tests for syphilis were not performed on CSF.

The cause of the patient’s primary infection is unknown. He had been happily married for 40 years. Of possible relevance in his employment history was the fact that he had served in the army for two years in Germany as a transport driver.

This case is remarkable for the apparent coincidence of two unusual causes of dementia in a married couple. There is a possibility that the patient’s wife might also have had neurosyphilis; this diagnosis has been reported as presenting as Creutzfeldt-Jakob disease.1 Her negative serum VDRL and normal CSF cells and protein make this unlikely, but in the absence of postmortem confirmation of the diagnosis or antemortem CSF examination it is not possible to be certain. Examination of CSF to exclude neurosyphilis should always be carried out in patients with possible Creutzfeldt-Jakob disease.

This case is also a reminder that hysterical dissociation is a very unsafe diagnosis to make in older adults. Even genuinely dissociative symptoms in elderly patients are usually indicative of an underlying organic cerebral disorder, and this should always be investigated. The initial diagnosis of a dissociative disorder in this patient was based on the circumstances surrounding the onset, the absence of focal neurological signs, and the presentation with symptoms which mimicked his wife’s. The cognitive impairment only became apparent as the dysphasia and agitation resolved.

The dramatic onset of the patient’s symptoms in the context of his wife’s terminal illness is interesting. It was commonly reported in the pregonorrheal literature that the onset of general paralysis of the insane could be precipitated by “mental shocks” such as bereavement and illness in the family,1 but this clinical finding has never been systematically studied. The acute presentation of people with cognitive deficits is not uncommon when a spouse falls ill or dies. This is a cause of sudden departure of the carer discloses the impairment, but sometimes the stress of the event causes a significant decancellation, as seems to have occurred in this case.

Another lesson to be drawn from this patient is that neurosyphilis is not a historical curiosity, but something the clinician must keep in mind particularly when investigating elderly patients. There is evidence that neurosyphilis is becoming clinically less common, but serological investigation is all the more important if its diagnosis is not to be missed. Routine screening of all elderly patients is currently out of favour, although still recommended for those with an organic illness.6 This patient, together with others in whom neurosyphilis has presented as a functional disorder, does raise the question of whether these patients should be screened also, particularly those with an atypical illness that is unresponsive to treatment.

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Alternating paroxysmal dystonia and hemiplegia in childhood as a symptom of basal ganglia disease

We report a 13 year old girl with an unusual clinical presentation of long-standing progressive cerebellar and pyramidal disorder, episodes of alternating paroxysmal dystonia lasting up to an hour, and hemiparesis of the involved side or sides, reminiscent of alternating hemiplegia of childhood. Magnetic resonance imaging showed hypointensity of the basal ganglia, particularly the globus pallidus, similar to that seen in Hallervorden-Spatz disease, raising the possibility that this may represent an atypical form of the condition.

The patient, now aged 13, was born at term after a normal pregnancy and delivery, to non-consanguineous parents: there were no perinatal problems. At two months of age she was noted to be hypotonic. She had delayed motor milestones, sitting at nine months and walking at 22 months, and was said to be “always clumsy”. Tremor and ataxia were noted at two years and have been slowly progressive, particularly since the age of eight: by 12 years she needed to use a frame. Examination showed her to have pyramidal signs in all four limbs as well as considerable titubation and cerebellar signs.

At the age of 10 she developed episodes of painful dystonic posturing of the arm and leg, usually on the right but CSF, including that on the left, associated with hemiparesis. These initially occurred during sleep and were preceded by a cry. They were often associated with a contralateral headache. They lasted 15 to 45 minutes and ended abruptly, and she could have several in a month, sometimes more than one a day. They were at first helped by carbamazepine but later recurred, during waking as well as in sleep. After withdrawal of the carbamazepine she developed distressing bilateral attacks that were also associated with drooling and difficulty in breathing: these settled after reintroduction of the medication. She did not respond to benzodiazepines. Flumazenil likewise produced no benefit.

Blood tests including lactate, ammonia, thyroid function, cholesterol, triglycerides, carotid Doppler in carotid arteries, blood urea, renal function tests, calcium, magnesium, arylsulphatase A, hexosaminidase, plasma very long chain fatty acids; renal function tests and urinary oligosaccharides were normal. Examination for syphilis, including assay for lactate and amino acids, was normal, and there were no oligonuclear bands. Measles and rubella antibodies were not detected. An ECG was normal, EEG was mildly dysrhythmic, and auditory evoked responses were both delayed. Nerve conduction studies and nerve and muscle biopsies were normal. Cytochrome oxidase and pyruvate dehydrogenase activity were normal. Repeated CT was unremarkable, but MRI showed notable symmetric low intensities in the basal ganglia, particularly in the globus pallidus, but also the putamen, red nucleus, substantia nigra, and thalamic pulvinar. There were also changes in the white matter signals in the region of the U fibres (figure).

Our patient presents an unusual clinical picture, in which the diagnosis and treatment of a paroxysmal alternating or sometimes bilateral dystonia associated with weakness supervened on the background of a progressive neurological disorder involving cerebellar, pyramidal, and extrapyramidal systems, the aetiology of which remains unclear despite extensive investigation. At their onset, the possibility that these episodes represented a seizure disorder was considered, but this has not been confirmed either by the clinical course or by EEG video-monitoring. The attacks were not precipitated by movement, nor was there any family history. They thus differed from the familial paroxysmal choreoathetosis described by Mount and Reback2 and from paroxysmal kinesigenic choreoathetosis, which may be familial or sporadic. In these conditions alternating and bilateral attacks have been described. In paroxysmal kinesigenic choreoathetosis the attacks also tend to be much shorter than those in our patient, unlike familial paroxysmal choreoathetosis, in which they may last several hours. The occurrence of hemiparesis in association