ferred to an organic psychiatric unit for further investigation. The dysphasia resolved, but he became increasingly perseverative 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 + + + +. Further investigation of CSF showed VDRL positive (1:4), TPHA positive (1:1024), and FTA positive + + + +. The isoelectric 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. A 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 source 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 disorder 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.² 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 pretreponemal literature that the onset of general paralysis of the insane could be precipitated by "mental shocks" such as bereavement and illness in the family,^{3,4} 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 usually because the sudden departure of the carer discloses the impairment, but sometimes the stress of the event causes a significant decompensation, 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 that clinicians need to keep in mind particularly when investigating elderly patients. There is evidence that neurosyphilis is becoming clinically less typical,⁵⁻⁷ so serological investigation is all the more important if the 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.^{8,9} 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.⁹

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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 longstanding 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 sometimes 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. Flunarizine likewise produced no benefit.

Blood tests including lactate, ammonia, thyroid function, cholesterol, triglycerides, caeruloplasmin, copper, α -foetoprotein, renal function tests, calcium, magnesium, arylsulphatase A, hexosaminidase, plasma very long chain fatty acids; renal function tests and urinary oligosaccharides were normal. Examination of CSF, including assay for lactate and amino acids, was normal, and there were no oligoclonal bands. Measles and rubella antibodies were not detected. An ECG was normal, EEG was mildly dysrhythmic, and visual 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, caudate nucleus, 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 development 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 Reback¹ and from paroxysmal kinesigenic choreoathetosis,² which may be familial or sporadic. In each of these conditions alternating and bilateral attacks have been described. In paroxysmal kinesigenic choreoathetosis the attacks also tend to be much shorter than those of our patient, unlike familial paroxysmal choreoathetosis, in which they may last several hours. The occurrence of hemiparesis in association

MRI: T2 weighted images (TR2000, TE40) in axial (top) and coronal (bottom) views showing a notable hypointense signal in the thalami and basal ganglia. An abnormal white matter hyperintense signal is seen involving the subcortical white matter (U fibres).

with these attacks, if it occurred, was not stressed. The occurrence of dystonia as a complication of various neurological diseases has been well documented.³ With the possible exception of a report of bilateral subcortical epilepsy by Spiller in 1927⁴ the alternating form has not to our knowledge been reported, although clearly it might be anticipated in bilateral disease.

Although in some respects, particularly from the radiological aspect, the underlying condition bears some resemblance to Hallervorden-Spatz disease (and may represent an atypical form), the features are not entirely characteristic, and MRIs similar to those seen in Hallervorden-Spatz disease have been reported in patients with quite different clinical symptoms,⁵ suggesting that they are not specific. In the absence of a

family history, confirmation of the diagnosis of Hallervorden-Spatz disease cannot be made with certainty in life. Other diagnoses that were considered, including Leigh's disease and pyruvate dehydrogenase deficiency, seem unlikely on the basis of the clinical features and results of investigations.

The presentation in this child also bears a considerable resemblance to the picture seen in alternating hemiplegia of childhood, in which the alternating episodes of hemiparesis are often accompanied by dystonia, and may be associated with a migraine-like headache. The bilateral attacks, in which drooling, severe dysarthria and some respiratory distress occurred, also have some similarities with the less frequent bilateral attacks of alternating hemiplegia of childhood. The onset of alternating hemiplegia of childhood is invariably in infancy, however. This is unlike the situation in our patient, and although other neurological symptoms occur in association with it, they usually develop after the onset of the hemiparetic attacks, and consist predominantly of mental retardation and choreoathetosis. Even when such deficits occur, the MRI remains normal. None the less, the similarities raise the possibility of a common underlying mechanism.

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MATTERS ARISING

Functional brain imaging

Sawle's review (Imaging the head: functional imaging. *J Neurol Neurosurg*

Psychiatry 1995;58:132-44) completely ignores the stable xenon (Xe) CT cerebral blood flow (CBF) method for functional imaging despite a large literature dealing with this subject. We are not aware that radioactive ¹³³Xe gas can be used with the CT scanner for measuring cerebral circulation, as isotopic Xe requires external detectors for monitoring clearance of the commonly used radioactive isotope ¹³³Xe from the brain.^{1,2}

The stable Xe CT CBF method is not an isotopic method, but uses cold Xe which stops x rays as a radio-opaque indicator much like iodine, which has a similar atomic number. Unlike iodine, however, or HMPAO indicator used for SPECT, Xe is freely diffusible and is widely used for functional imaging of the brain³ in three dimensions with unparalleled resolution. Xe CT CBF methodology also has the advantage of being accurately quantifiable for measuring local CBF, in ml blood/100 g brain/min; whereas SPECT imaging with HMPAO, which is not freely diffusible, is only a relative method that cannot provide absolute units of local CBF.

Lassen *et al* have shown that it is possible to measure and quantify regional CBF in three dimensions using ¹³³Xe inhalation, employing a specially designed single emission tomograph, but local resolution is poor.

The stable Xe CT CBF method has unique advantages of correlating changes in perfusion as a brain map, superimposed directly on the actual CT slices from which they were derived. This provides an indicator of local function with local CT morphological estimates of cerebral tissue changes such as atrophy, hypodensities, or infarction.^{5,6}

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