Sporadic Creutzfeldt–Jakob disease presenting as stroke mimic

Introduction

This article reports a case presenting as acute stroke but subsequently manifesting rapid cognitive and functional decline leading to a revised diagnosis of sporadic Creutzfeldt–Jakob disease. This case highlights the importance of serial investigations when diagnosing this rare condition.

Sporadic Creutzfeldt–Jakob disease is a rare, progressive neurodegenerative illness. Early non-specific symptoms predate higher cortical function deficits which progress relentlessly and often rapidly. The pathological process can present abruptly mimicking acute stroke. Such mimics can usually be ruled out by neuroradiological and electroencephalographic evaluation although diagnostic clarity may require serial studies.

Discussion

Sporadic Creutzfeldt–Jakob disease presents clinically in middle-aged and elderly individuals. Pocchiari et al (2004) have shown the illness duration to be brief leading to death within a few months (median survival 4 months). Rare focal presentations are recognized including the Heidenhain (affecting central vision centres) and Brownell–Oppenheimer (affecting the cerebellum) forms. Abrupt presentation can cause diagnostic confusion with acute stroke. Brown et al (1986) described

Figure 1. Computed tomograph of the brain on initial presentation suggesting infarction of the posterior limb of the left internal capsule.

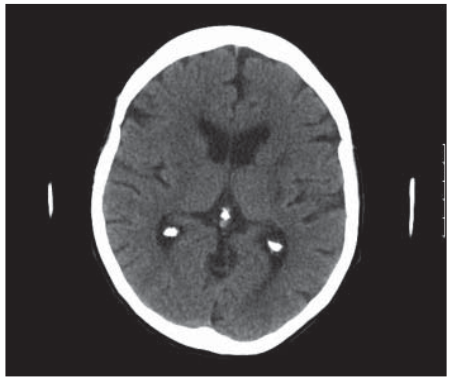
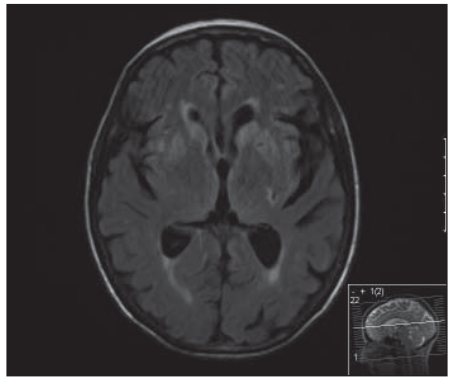


Figure 2. Magnetic resonance imaging of the brain T2 FLAIR indicating increased bilateral signal intensity in the basal ganglia (putamen and caudate nuclei).



Case Report

A hypertensive 75-year-old woman with hypertrophic cardiomyopathy, hypercholesterolaemia and paroxysmal atrial fibrillation was referred to the author's neurovascular clinic with sudden onset word-finding difficulty and right-sided weakness. Examination confirmed mild right pyramidal drift and expressive dysphasia in an otherwise relatively independent patient. Computed tomography (CT) confirmed a lacunar infarct involving the posterior limb of the internal capsule (*Figure 1*).

Neurorehabilitation afforded initial improvement, but her family reported deterioration in concentration, coordination and continence control with emotional lability over the next 4 weeks, prompting hospital admission. On examination, she was bedbound, with severe expressive dysphasia (abbreviated mental test score 0/10), dysphagia, global weakness, hypertonia, hyperreflexia and bilateral upper limb myoclonus (more on the right hand side). Repeat CT confirmed previous findings. Lumbar puncture was acellular with a protein count of 0.42 g/dl. Sequential electroencephalograms (EEGs) demonstrated bilateral periodic lateralized epileptiform discharges which were abolished by intravenous lorazepam suggesting non-convulsive status epilepticus. However, deterioration continued despite antiepileptic therapy.

Magnetic resonance imaging demonstrated bilateral putamen and caudate nucleus enhancement on T2 FLAIR with left temporal lobe cortical ribboning on diffusion weighted imaging (*Figures 2* and *3*). A subsequent EEG showed an evolving picture, with bilateral triphasic waves (1Hz) (*Figure 4*) compatible with a diagnosis of probable sporadic Creutzfeldt–Jakob disease. This was supported with a positive cerebrospinal fluid 14-3-3 protein assay. The patient died 59 days after the initial presentation. No tissue diagnosis was made.

Figure 3. Magnetic resonance imaging (diffusion weighted imaging) of the brain showing left cortical ribboning of the temporal lobe.

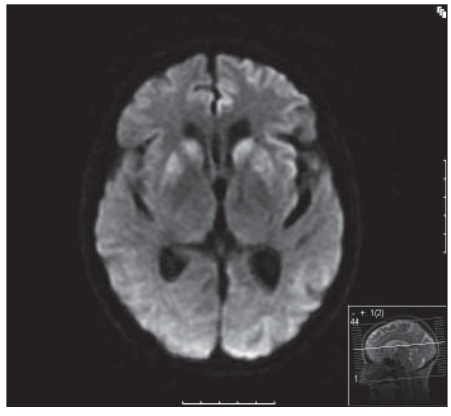
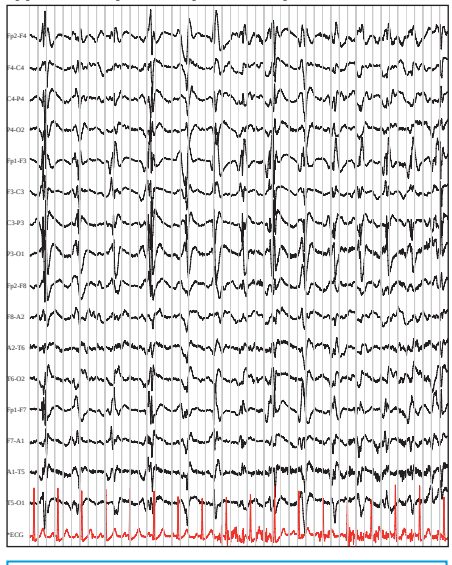


Figure 4. Electroencephalogram showing approximately 1 Hz triphasic complexes.



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rapid neurological onset in as many as 20% of cases, with pyramidal signs at presentation in 2%. McNaughton and Will (1997) described acute focal neurological presentations in 5.6% of definite or probable sporadic Creutzfeldt–Jakob disease cases. A few case reports have also described stroke-like presentations but with incompatible radiological findings. Interestingly, Szabo et al (2004) described a case strongly suggestive of acute stroke clinically in which serial magnetic resonance imaging studies revealed compatible features that later evolved into persistent 'cortical hyperintensity' lesions characteristic of sporadic Creutzfeldt–Jakob disease.

In the present case, the initial presentation was sudden and clinically indistinguishable from acute stroke. The disease progressed rapidly, however, from focal deficits to increasingly global loss of function. Electroencephalographic studies initially caused diagnostic confusion, being suggestive of non-convulsive status epilepticus although no clinical improvement was seen with antiepileptic therapy. Shapiro et al (2004) have described refractory non-convulsive status epilepticus in sporadic Creutzfeldt–Jakob disease. Classical findings of sporadic Creutzfeldt–Jakob disease were only seen on a later electroencephalogram. Such diagnostic confusion, first with acute stroke and then non-convulsive status epilepticus, was only resolved with repeated evaluation. This report highlights the evolving clinical picture typical of sporadic Creutzfeldt–Jakob disease and the need for serial investigations to arrive at a firm diagnosis. **BJHM**

Brown P, Cathala F, Castaigne P, Gajdusek DC (1986) Creutzfeldt-Jakob disease: clinical analysis of a consecutive series of 230 neuropathologically verified cases. *Ann Neurol* **20:** 597–602

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Pocchiari M, Puopolo M, Croes EA et al (2004) Predictors of survival in sporadic Creutzfeldt–Jakob disease and other human transmissible spongiform encephalopathies. *Brain* **127:** 2348–59

Shapiro JM, Shujaat A, Wang J, Chen X (2004) Creutzfeldt-Jakob disease presenting as refractory nonconvulsive status epilepticus. *J Intensive Care Med* **19**(6): 345–8

Szabo K, Achtnichts L, Grips E et al (2004) Stroke-like presentation in a case of Creutzfeldt-Jakob disease. *Cerebrovasc Dis* **18:** 251–3

Digital ischaemia: vasculitis *vs* trauma

A 56-year-old woman with rheumatoid arthritis and Raynaud's phenomenon presented to outpatients with a 7-day history of three painful, dusky-coloured fingers of her right hand. She described relief from pain when placing her hand in warm water. On examination, the radial pulse was diminished in amplitude, the middle three fingers were cyanotic (*Figures 1* and *2*) and cold to palpation, and the colour improved on warming the skin. Cardiovascular and neurological examinations were otherwise unremarkable. The initial diagnosis was of rheumatoid vasculitis and she was admitted for intravenous prostacyclin and heparin.

Her autoimmune profile was negative for antiphospholipid (including enzyme-linked immunosorbent assay) and anticardiolipin antibodies. Investigation with Doppler studies and magnetic resonance angiography revealed 75% stenosis of the distal radial artery, consistent with an arterial thrombus. Four months previously, she had been admitted to the intensive treatment unit where an arterial cannula was administered for invasive blood pressure monitoring.

The radial artery is the commonest site for arterial cannulation. Arterial occlusion occurs in 30% of patients within 1 day of cannula removal (Davis and Stewart, 1980); in 0.1% there follows severe ischaemia (Scheer et al, 2002). Risk factors include cannula size and female sex (Davis and Stewart, 1980); autoimmune conditions may further contribute (Rose, 1993). **BJHM**

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Figure 1. Palmar aspect of right hand demonstrating dusky-colouring of the three middle fingers.

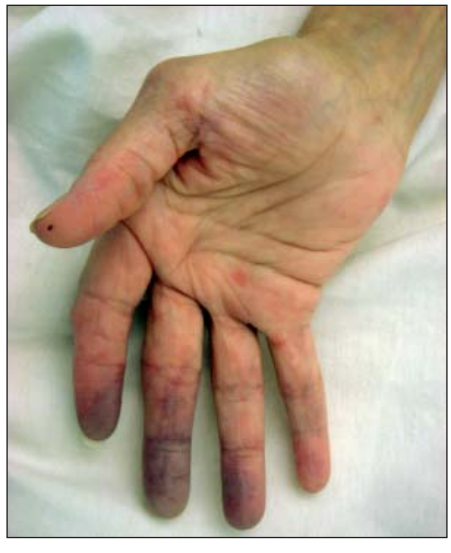
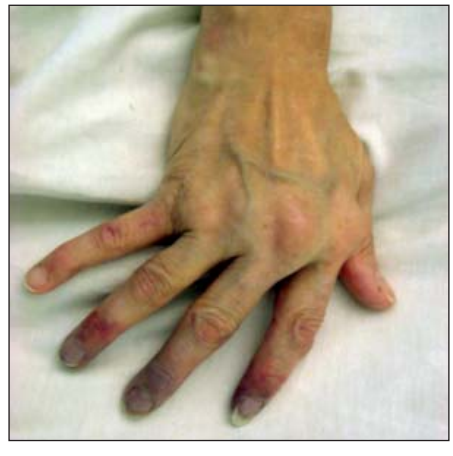


Figure 2. Dorsum of right hand.



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