

Creutzfeldt-Jakob disease with initial right hemiparesis masquerading as a stroke

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The early stage of sporadic Creutzfeldt-Jakob disease is generally characterised by progressive changes in behaviour and intellectual function. While only a few patients have stroke-like onset, Creutzfeldt-Jakob disease with initial monoparesis has been described. In this report, a patient with an unusual sporadic Creutzfeldt-Jakob disease with typical magnetic resonance imaging findings, positive cerebrospinal fluid 14-3-3 brain protein, sharp-wave complexes in electroencephalogram, and initial right hemiparesis is reported.

Introduction

Creutzfeldt-Jakob disease (CJD) is the most common human prion disease.¹ While between 10% and 15% of cases are familial and a small number are iatrogenic, most (85%) are sporadic. The early stage of sporadic CJD is characterised by changes in behaviour and intellectual function, often followed by ataxia, abnormality of vision, and myoclonic jerks involving either a limb or the entire body.² The minority of cases may have atypical presentations with abrupt onset and prolonged progression.^{3,4} However, CJD presenting with initial neurological symptoms and signs masquerading as a stroke is exceptional.⁵ Previously, CJD with initial monoparesis has been reported.⁶ In this report, a patient with CJD presenting with right hemiparesis is described.

Case report

A 60-year-old woman presented in January 2009 with a 2-week history of right-sided weakness and associated difficulty in writing (she was right-handed). She was also found to be increasingly irritable, with poor concentration and memory. She had a history of hypertension requiring antihypertensive drugs. She did not abuse alcohol, and no relevant family medical history.

At examination, the patient was partially orientated and had a right hemiparesis. Power was 4+ (Medical Research Council scale) in the right arm and 4 at best in the right leg, with a pyramidal pattern. All reflexes were pathologically brisk with positive grasp reflex and equivocal right plantar reflex. There was no myoclonus; and no left-right disorientation, astereognosia, or hemi-neglect were detected.

Routine haematology and biochemistry tests were normal. Blood tests, including lupus anticoagulant, antinuclear factor, antineutrophil cytoplasmic antibodies, anti-extractable nuclear antigen, immunoglobulin pattern, antithyroglobulin, antithyroid microsomal antibody, copper, manganese, and mercury were either normal or negative. Magnetic resonance imaging (MRI) of the brain demonstrated mild cortical hyperintensity at the bilateral frontal and parietal lobes on fluid attenuation inversion recovery (FLAIR) images and T2-weighted hyperintensity, and possible FLAIR hyperintensity at the bilateral caudate nuclei and putamen (Fig). These findings were suggestive of CJD.^{7,8} Computed tomographic cerebral angiogram did not detect any focal vascular lesions. Cerebrospinal fluid analysis was positive for 14-3-3 protein.⁹ Electroencephalography (EEG) investigation showed moderately severe abnormalities of background rhythm with 1 to 1.5 Hz triphasic sharp waves.

Later, the patient became mute together with progressive weakness of all limbs. A late EEG examination 5 days before the death of the patient showed the tracings were near to isoelectric with disappearance of the bursts of sharp or slow waves. She died 14 months after the onset of the illness.

Discussion

Creutzfeldt-Jakob disease occurs worldwide, with a uniform incidence of approximately 1 patient per million population per year.¹⁰ A surveillance study of CJD across 12 provinces in China has found that Chinese patients with CJD have similar epidemiological and clinical

Key words

Brain diseases; Creutzfeldt-Jakob syndrome; Electroencephalography; Paresis; Stroke

Hong Kong Med J 2010;16:487-8

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擬似中風的克雅二氏症患者發病初期出現右側輕偏癱

偶發性克雅二氏症患者發病初期一般會出現智力衰退和行為異常的現象。只有少數患者有類似中風的病症，文獻亦曾記載有克雅二氏症患者發病初期出現單肢輕癱。本文報告一名病人患有不尋常的克雅二氏症，病人的磁力共振影像不正常，腦脊液14-3-3蛋白呈陽性，腦電圖呈尖慢複合波，發病初期亦曾出現右側輕偏癱。

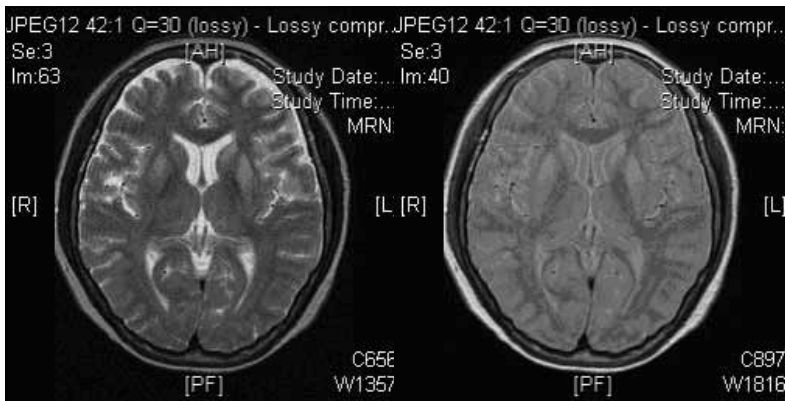


FIG. Magnetic resonance images of the brain showing cortical hyperintensity at the bilateral frontal and parietal lobes and hyperintensity at the bilateral caudate nuclei and putamen

characteristics to those in the rest of the world.¹¹ Of the patients with sporadic CJD, generally 80% are aged between 50 and 70 years, with the mean age of onset at approximately 62 years. The patient in this report was aged 60 years at onset.

References

- Zerr I, Kallenberg K, Summer DM, et al. Updated clinical diagnostic criteria for sporadic Creutzfeldt-Jakob disease. *Brain* 2009;132:2659-68.
- Collinge J. Human prion diseases: aetiology and clinical features. In: Rossor MN, editor. *The dementias*. Oxford: Butterworth-Heinemann; 1998.
- Hohler AD, Flynn FG. Onset of Creutzfeldt-Jakob disease mimicking an acute cerebrovascular event. *Neurology* 2006;67:538-9.
- Szabo K, Achtnichts L, Grips E, et al. Stroke-like presentation in a case of Creutzfeldt-Jakob disease. *Cerebrovasc Dis* 2004;18:251-3.
- Panagariya A, Jain RS, Sharma AK. Stroke like presentation of Creutzfeldt Jakob disease: an unusual variant. *J Assoc Physicians India* 1999;47:548-50.
- Obi T, Takatsu M, Kitamoto T, Mizoguchi K, Nishimura Y. A case of Creutzfeldt-Jakob disease (CJD) started with monoparesis of the left arm [in Japanese]. *Rinsho Shinkeigaku* 1996;36:1245-8.
- Shiga Y, Miyazawa K, Sato S, et al. Diffusion-weighted MRI abnormalities as an early diagnostic marker for Creutzfeldt-Jakob disease. *Neurology* 2004;63:443-9.
- Meissner B, Kallenberg K, Sanchez-Juan P, et al. MRI lesion profiles in sporadic Creutzfeldt-Jakob disease. *Neurology* 2009;72:1994-2001.
- Hsich G, Kenney K, Gibbs CJ, et al. The 14-3-3 brain protein in cerebrospinal fluid as a marker for transmissible spongiform encephalopathies. *N Engl J Med* 1996;335:924-30.
- Meissner B, Körtner K, Bartl M, et al. Sporadic Creutzfeldt-Jakob disease: magnetic resonance imaging and clinical findings. *Neurology* 2004;63:450-6.
- Shi Q, Gao C, Zhou W, et al. Surveillance for Creutzfeldt-Jakob disease in China from 2006 to 2007. *BMC Public Health* 2008;8:360.
- Ng PW. Case 77: A rapidly fatal degenerative disease. In: Kay R, editor. *Casebook of neurology*. Hong Kong: Lippincott Williams & Wilkins; 2002.
- Weihl CC, Roos RP. Creutzfeldt-Jakob disease, new variant; Creutzfeldt-Jakob disease and bovine spongiform encephalopathy. In: *Central nervous system infections, Neurologic Clinics*. Philadelphia: WB Saunders Co; 1999.
- Machado A, Ribeiro M, Rodrigues M, et al. Sporadic Creutzfeldt-Jakob disease causing a 2-years slowly progressive isolated dementia. *Behav Neurol* 2009;21:175-9.
- Prusiner SB. Shattuck lecture—neurodegenerative diseases and prions. *N Engl J Med* 2001;344:1516-26.

As this patient presented with hemiparesis, a cerebrovascular event was suspected initially. It was only after the appearance of the classical symptoms and signs of CJD, and MRI and EEG examinations, that a diagnosis of probable sporadic CJD was made.¹² Unusual presentation of CJD with initial stroke-like symptoms has been described in the literature, including a patient who presented with monoparesis of the left arm.⁶ The patient in our case was unusual in that the disease started with right hemiparesis. While sporadic CJD is invariably fatal and the usual average survival from diagnosis is about 8 months,¹³ this patient survived for 14 months. At the other extreme, a few people have survived for as long as 2 years after diagnosis,¹⁴ but this should be accepted with caution.

Spongiform encephalopathy is thought to be associated with the conversion of a normal cellular protein, PrP^c, to an abnormal isoform, PrP^{Sc}.¹⁵ Physical conformation of the protein occurs with altering of its helical structure and increasing the proportion of the β pleated sheet. Native prion protein alters shape and becomes misfolded on exposure to the abnormal protein, causing infectivity and propagation of prions. This mass of misfolded proteins disrupts cell function and causes cell death. To date, there is no specific treatment for CJD.

The purpose of reporting this patient is to alert clinicians that sporadic CJD can mimic a stroke in the early stage, and atypical features such as prolonged progression can occur. By recognition of these unusual features, it is hoped that correct diagnosis can be made early, and any iatrogenic or human-to-human transmission can be prevented.