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syndromes with known serum antibodies, such as relapsing polychondritis and Sjögrens, have been subsequently diagnosed in patients presenting with subacute encephalopathy.

The aetiology of a humourally mediated central nervous system condition may require initial compromise of the blood-brain barrier. This could expose the brain to serum factors, such as antibodies, resulting in central nervous system damage after the development of serum antibodies. Such serum antibodies could have been initially raised against skin tissue in our patient, and subsequently cross-react with neurons to mediate cognitive deterioration.

The autoantigens BP180 and BP230 are structural components of dermal hemidesmosomes and are known to be targeted by antibodies in BP. Recent work has shown that BP230 also exists in human CNS neurons, with prominent expression within hippocampal neuronal somae and axons, providing a theoretical common antigen binding site.⁵ Furthermore, IgG from our patient bound to hippocampal somae and axons during the peak of the illness, at a time when the skin biopsy demonstrated features consistent with basement zone IgG deposition. This suggests the concomitant presence of skin and neural antibodies during disease onset. However, serum analysis for BP180 and BP230 was negative in our patient. Hence, our patient may have antibodies to an antigenic target which is common to both skin and brain but is not BP180 or BP230.

This is the first study to directly link the clinical entities of encephalitis and BP through an antibody-based mechanism. It also emphasises the importance of recognising autoimmune encephalopathies and the benefits of prompt immunotherapy.

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Characteristics of abnormal eating behaviours in frontotemporal lobar degeneration: a cross-cultural survey

Frontotemporal lobar degeneration (FTLD) is characterised by behavioural changes, including loss of insight, disinhibition, apathy, mood changes, stereotypic behaviour and abnormal eating behaviour.^{1,2} Although many studies have highlighted the high prevalence of alterations in food preference and eating habits in FTLD and described loss of appetite in dementia represented by Alzheimer disease (AD),³ there have been few systematic studies comparing FTLD subgroups, or contrasting AD and FTLD.^{1,2,4} Eating behaviours are modulated by many factors including personal habits, ethnic culture and climate, such that alteration in eating behaviour in dementias may be confounded by ethnic or cultural factors. Food culture, meal styles and customs differ substantially between Western countries and Japan. People in the UK consume considerably more sweets, and total daily calorific intakes are higher than they are for the Japanese. (Data derived from the Food and Agriculture Organization of the United Nations; <http://faostat.fao.org/>.) Therefore, it is unclear whether altered eating behaviours of FTLD in Western cohorts are an entirely disease-specific effect or whether they are modulated by ethnic-cultural factors. The aims of this study were to investigate changes in eating behaviours in Japanese FTLD and AD patients and to compare the profile of abnormal eating behaviours in Japanese and Western patients using the same instruments.

A total of 163 patients were involved: 72 from Ehime, Japan (18 frontotemporal dementia (FTD), 11 semantic dementia (SD) and 43 AD), and 91 from Cambridge, UK (23 FTD, 25 SD, 43 AD) (fig 1). A detailed description of British patients has been reported previously;⁵ all were of white European ethnicity. All patients were living at home. Patients in the FTD and SD groups fulfilled consensus criteria for FTLD. FTD

with motor neuron disease patients were excluded. The diagnosis of probable AD was made according to the NINCDS-ADRDA criteria. All underwent comprehensive investigation including MRI and/or HMPAO-SPECT, a battery of screening blood tests, neuropsychological and psychiatric evaluations, mini-mental state examination (MMSE) and clinical dementia rating (CDR).

The care-giver-based questionnaire consisted of 36 items investigating five domains: swallowing, appetite, food preference (including sweet food preference and food fads), eating habits (including stereotypic eating behaviours and decline in table manners) and other oral behaviours (including food cramming and indiscriminate eating). It was emphasised that a “symptom” should reflect a substantive change from a patient's premorbid state. If care givers endorsed a particular item, they were asked to rate the frequency and severity, to derive a frequency×severity score.⁵ Patients' present weights were measured, and patients' premorbid weights were ascertained from their previous check-up to estimate the amount of weight change.

As demographic variables, there were no significant differences between patients in Japan and in the UK in age, education and CDR grade. There were significantly more females in the Japanese cohort, and the mean MMSE score of Japanese patients was significantly lower. Figure 1 shows the total scores (frequency×severity) for each domain in the three patient groups in Japan and UK. For all five domains, two-way ANOVAs showed a significant main effect of diagnosis only, no significant main effect of patients' nationality and no interaction between diagnosis and nationalities. In all instances, FTD patients scored significantly higher than AD patients. For appetite change, food preference and eating habits, SD patients also scored significantly higher than AD patients. A weight gain of more than 7.5 kg was found in 30% of the FTD cases and 36% of SD cases in UK, compared with 5.6% of FTD cases and 9.1% of SD cases in Japan.

Patients with FTD and SD presented similar abnormal eating behaviours both in Japan and in the UK. Changes in eating behaviours in Japanese patients with both of the FTLD subtypes were significantly more common than AD patients, as was the case in the UK. Therefore, it is clear that patients with FTD and SD exhibit similar abnormal eating behaviours, as is the case with other behavioural and psychiatric symptoms.^{1,2} Changes in eating behaviours in FTLD groups appear to be universal, and although ethnic-cultural factors might modulate these changes to some extent, they are likely to be a direct consequence of the pathology of FTLD. Changes in food preference and eating habits were the main alteration in SD. The FTD group also showed changes in appetite and oral behaviours. These findings are in keeping with prior reports.² The

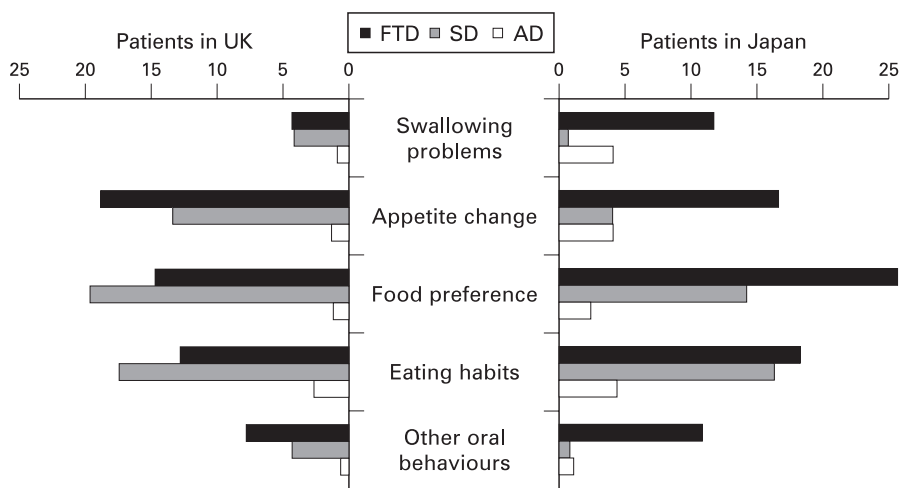


Figure 1 Score (frequency × severity) for each symptom domain in three patient groups in UK and Japan. Frequency: 1, occasionally, less than once per week; 2, often, about once per week; 3, frequently, several times per week but less than every day; 4, very frequently, once or more per day or continuously. Severity: 1, mild, easily controlled; 2, moderate, not easily controlled; 3, marked, embarrassing or otherwise disturb family. FTD, frontotemporal dementia (Japan: n = 18, age 65.5 (SD 7.0) years, F/M 9/9, mini-mental state examination (MMSE) 16.1 (9.4); UK: n = 23, age 61.1 (6.6), MMSE 22.9 (7.4)). SD, semantic dementia (Japan: n = 11, age 66.8 (7.6) years, F/M 7/4, MMSE 15.7 (10.8); UK: n = 25, age 65.1 (7.0), MMSE 17.2 (8.3)). AD, Alzheimer disease (Japan: n = 43, age 70.1 (9.8) years, F/M 30/13, MMSE 13.7 (7.9); UK: n = 43, age 68.3 (7.7), MMSE 20.6 (5.7)).

higher rate of appetite change in British SD may reflect the more advanced disease of the British: four out of 11 Japanese patients were moderate or severe demented cases (CDR ≥ 2), whereas 16 out of 25 were moderate or severe in the UK cohort.

It appears that some abnormal eating behaviours such as appetite increase are modulated by cultural factors. A weight gain of more than 7.5 kg was found in 30% of FTD and 36% of SD cases in the UK, while it was found in less than 10% of FTD and SD cases in Japan. As described above, sugar intake and total calorie consumption differ significantly across Japan and the UK. We suggest that Japanese FTLD patients did not manifest such severe weight gain, because their eating behaviours are not amplified by cultural factors.

The current results highlight the stability of abnormal eating behaviours in FTLD across cultures with significantly different dietary habits and reinforce the view that changes in eating behaviour are diagnostically useful in detecting FTLD.^{1,5}

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Posterior circulation strokes without systemic involvement as the presenting feature of Fabry disease

Fabry disease is a multisystem lysosomal storage disorder with serious effects including cardiomyopathy and renal failure. Although neurological involvement at presentation is unusual, it is increasingly recognised that Fabry disease may present with ischaemic strokes and may be responsible for up to 5% of cryptogenic strokes in young men.¹ Early recognition is vital to permit early therapeutic intervention and family screening, and could prevent clinical progression and recurrent stroke. We report a patient who presented with recurrent brainstem ischaemic strokes due to Fabry disease, with no evidence of systemic manifestations at presentation. Fabry disease should be considered in cases of cryptogenic stroke (especially young men with vertebrobasilar territory symptoms) even without multisystem involvement.

CASE REPORT

In January 2007, a 24-year-old man was admitted with sudden rotatory vertigo and nausea. He reported three previous similar episodes. In 2004, he had diplopia for 5 days; later that year, he experienced transient vertigo and gait ataxia. The third episode occurred in February 2005, when he suffered the abrupt onset of unsteadiness, nausea, slurred speech and right-sided weakness. An MRI scan of the brain at this time showed a lesion of high signal on T2-weighted images in the left pons; MR angiography, carotid duplex scan and transoesophageal echocardiogram (TOE) were normal. Demyelination was considered, but when a cerebrospinal fluid (CSF) examination failed to show oligoclonal bands, a diagnosis of probable cryptogenic stroke was made. He made a good recovery over several weeks and remained symptom-free on aspirin and simvastatin until the present admission.

His general and neurological examinations were unremarkable. His only known vascular risk factor was smoking; there was no history of drug abuse, family history of stroke or premature vascular disease. He did not report painful acroparaesthesias during childhood. Pulse and blood pressure were normal. Routine haematology, biochemistry and cholesterol levels were normal. Detailed thrombophilia and vasculitic screens were negative. T2-weighted MRI of the brain revealed the old lesion in the left pons and a new lesion in the right midbrain compatible with ischaemia (fig 1), but an extensive battery of investigations (including repeat MR angiography and TOE) failed to reveal any cause. There was no signal abnormality in the thalamic pulvinar on T1-weighted images. However, plasma and leucocyte alpha-galactosidase A activity were very