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Clinical and imaging features of a Chinese-speaking man with semantic dementia

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Sirs: A 56 year-old right-handed businessman from Northeastern China was previously well, with no family history of neurologic or dementing illnesses. He first presented with a three-year history of progressive difficulty in naming everyday objects, accompanied by increasing irritability and inability to run his business. He had also developed difficulty understanding certain spoken words two years into his illness. Orientation, memory of day-to-day events, visuospatial orientation and facial recognition remain intact.

On examination, he scored 27/30 in the Mini Mental State Examination. His speech was effortless and fluent, but vacuous in content. Articulation was normal. He was able to repeat words and phrases. He was not able to name a pencil and when shown the picture of a goat, he called it a cow. This man also made frequent errors when reading aloud compound Chinese words, pronouncing their components rather than the overall compound word. For example, the compound word 痰 (meaning 'phlegm') – correctly pronounced as 'tan2' (The number following 'tan' represents intonation) – was read aloud as 炎 (yan2, meaning 'blazing'). He scored within normal range on the Wechsler Adult Intelligence Scale (WAIS III) and had intact calculation, frontal executive and visuospatial function. He scored at a moderate to high level of episodic memory function as evaluated by the Chinese Clinical Scale for Memory [1]. On the Chinese version of the Western Aphasia Battery test, he was only able to name 22/60 objects. Neurological examination of the cranial nerves, upper and lower limbs was normal with no fasciculations.

Magnetic resonance imaging revealed moderate to severe atrophy of the left temporal pole, superior temporal lobe and hippocampus (Fig. 1a–c). Diffusion tensor imaging (DTI) was performed on a 3T Siemens Trio Scanner using a region of interest (ROI) approach. DTI image was acquired with 12 directions at b = 1000 mm^2/s. ROIs were placed at white matter locations beneath the primary speech centers (Brodmann's 44 and 45 for Broca's area and Brodmann's 22 and 39 for Wernicke's area). DTI showed decreased fractional anisotropy values in the left arcuate fasciculus and tracts linked to Wernicke's area, compared to the corresponding areas on the right (Fig. 1d).

In summary, this man presented with a three-year history of a fluent aphasia, accompanied by mild behavioral features but preservation of other cognitive domains, accompanied by left anterior temporal lobe atrophy, satisfying the consensus clinical criteria for semantic dementia [2, 3]. This man's characteristic errors when reading aloud is of interest. Surface dyslexia refers to errors in reading aloud orthographically irregular words, where there is no clear correspondence between the written word and its pronunciation. For example, the word 'pint' is read aloud erroneously 'by sound' to rhyme with 'hint'. In non-phonetic languages such as the Chinese language, the concept of surface dyslexia may manifest itself differently. For example, the compound character 痰 is composed of a radical 疒 and the component 炎. Our patient pronounced the compound word according to its component rather than the word itself. This error in reading a compound component is similar to that described in a Chinese-speaking patient post left perisylvian/temporal lobe infarct, and is most consistent with a Chinese version of surface dyslexia [4].

There has been one other report in the Chinese medical literature describing a progressive syndrome of isolated fluent aphasia with left temporal lobe and hippocampal atrophy [5]. However that patient read normally and more clinical detail is needed before a criterion-based diagnosis of semantic dementia could be made. DTI imaging in our patient found decreased fractional anisotropy values in the left arcuate fasciculus and tracts linked to Wernicke's area. This supports the idea that temporal lobe

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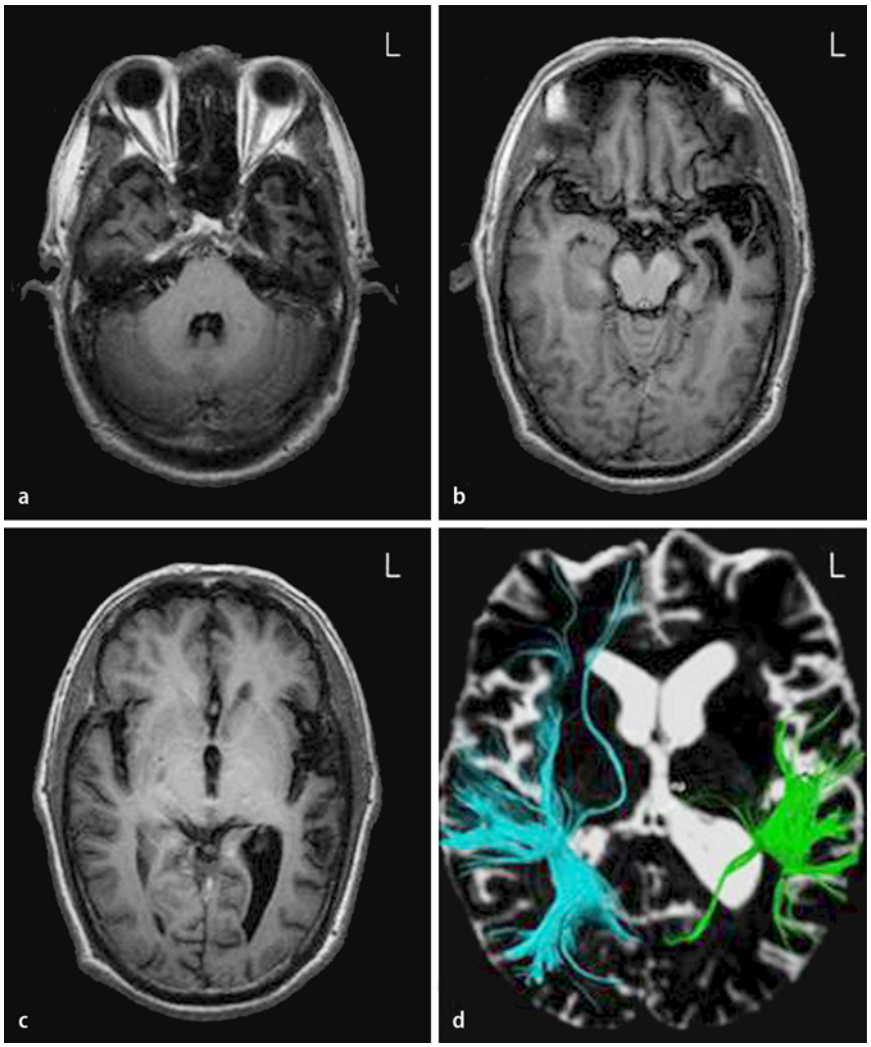

Fig. 1 T1-weighted MRI showing left greater than right temporal lobe atrophy (**a, b, c**). DTI with two dimensional tractography showing reduced connectivity in the left Wernicke's area (**d**)

connections are disrupted in people with semantic dementia, as demonstrated in a previous MRI/PET study [6]. DTI was found to be more sensitive in detecting white matter abnormalities than conventional MRI, with good correspondence with histopathologic findings [7]. To translate DTI into clinical use, further standardization of quantitative measures and norms will need to be established.

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