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LETTER TO THE EDITORS

Giant serpentine aneurysm of the anterior cerebral artery mimicking frontotemporal dementia

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Dear Sirs,

A 63 year-old man presented with a progressive 2-year history of personality change. He had little insight into his problems. Having previously been meticulous in self-care, he had gradually developed extreme neglect of personal grooming and hygiene. He showed little interest or motivation but would impulsively act in an irrational, disinhibited or aggressive manner. He had been banned from local supermarkets for inappropriate attire, disruptively singing out loud or filling trolleys with food for which he could not pay. He had developed habitual behaviours such as counting patterns on carpets and repetitive right hand movements. He was doubly incontinent, neglecting to eat and refusing to allow carers into his house. On direct enquiry about physical symptoms, he admitted to dizzy spells and headaches. Cranial nerve examination, including fundoscopy, was normal. He had a left grasp reflex, mild pyramidal left lower limb weakness and an extensor right plantar response.

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He underwent neuropsychological assessment using the Manchester Neuropsychological Profile, an assessment instrument developed for characterization of different forms of dementia, consisting of both locally developed and published tests [8], as well as further assessment of executive function and emotion recognition. He displayed a marked dysexecutive syndrome. He was jocular and facetious in manner. Behaviourally he was inattentive, disinhibited and emotionally labile. He showed poor temper control, making some threatening remarks. He was impulsive and also impersistent, quickly abandoning tasks without external prompts. On tasks of executive function, he showed impaired abstraction, sequencing and response inhibition. Performance was profoundly impaired on a standardised task of facial emotion recognition, particularly for negative emotions such as fear and anger (Table 1). There were no problems in language, visuospatial function, memory or praxis. Mini-mental state examination score was 29/30.

CT brain scan showed a multi-lobular, slightly oval hyperdense mass extending bilaterally displacing the frontal horns of the lateral ventricles. There was some curvilinear calcification peripherally and marked contrast enhancement outlined a lumen (Fig. 1a). There was additional ring enhancement after intravenous contrast administration. CT angiography confirmed a partially-thrombosed giant serpentine aneurysm arising from the anterior cerebral artery (Fig. 1b).

Giant serpentine aneurysms were first described by Segal and McLaurin [7]. The characteristic radiographic features are of giant (>25 mm), partially thrombosed aneurysms with serpiginous intra-aneurysmal channels [1]. The most common site of giant serpentine aneurysms is the middle cerebral artery (MCA), accounting for 50 % of cases, in which focal presentations such as hemiparesis are

Table 1 Performance on tasks of executive function and emotion recognition with 5 % normative cutoff values shown for comparison

	Patient score	5 % cut-off value
Hayling sentence completion test		
Sensible completion response time	22	>27
Unconnected completion (inhibition) total response time	25	>88
Unconnected completion (inhibition) error score	78	>18
Brixton Spatial Anticipation Test		
Error score	33	>27
Recognition of Facial Emotion Test		
Happiness	4/10	<9.4
Surprise	6/10	<5.9
Fear	2/10	<6.7
Sadness	1/10	<6.5
Disgust	1/10	<6.9
Anger	2/10	<5.4

common [3]. In contrast, only 3 % of aneurysms arise from the anterior cerebral artery (ACA) [3]. A 'frontal syndrome' was the presenting complaint in one of the two cases of ACA aneurysms reported [9], although the authors did not characterise this clinical syndrome in more detail. To our knowledge this is only the third reported case of a giant serpentine aneurysm arising from the ACA, and the clinical presentation, while consistent with its anatomical location, has not previously been reported in detail.

Our patient presented with gradual onset of self-neglect, impaired conduct and lack of insight, associated with distractibility and stereotyped behaviour. The initial evaluation was suggestive of a neurodegenerative dementia, and fulfilled core clinical diagnostic criteria for behavioural variant frontotemporal dementia (bvFTD) [5], although lateralised physical signs were a clue to an alternative aetiology. Neuropsychological findings of impersistence and impaired executive function together with emotional lability are suggestive of an orbitofrontal syndrome [2]. Functional imaging studies have implicated hypometabolism of the ventromedial frontal cortex, including orbitofrontal areas, in the pathophysiology of bvFTD [6]. The present case report demonstrates the importance of structural imaging in the evaluation of patients with suspected bvFTD in order to exclude potentially treatable causes [4].

Conflicts of interest The authors report no conflicts of interest.

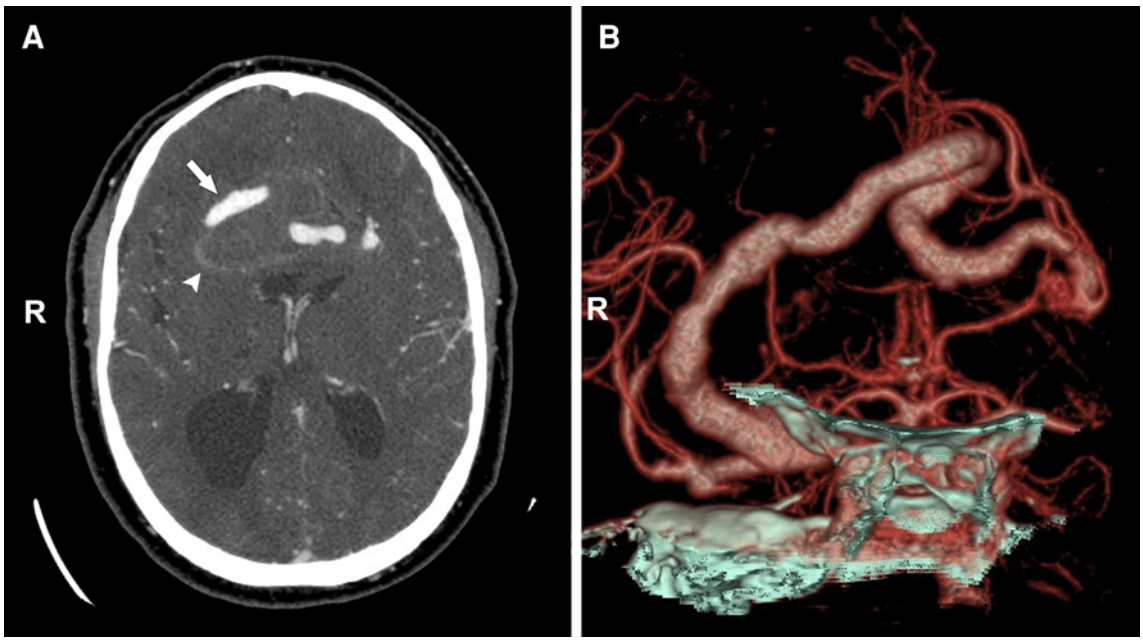

Fig. 1 **a** Axial contrast-enhanced CT brain scan showing a large 5 cm diameter oval mass with a calcified and ring-enhancing margin (*arrowhead*). The lumen of the aneurysm (*arrow*) is surrounded by thrombus. **b** Frontal projection of the volume-rendered reconstruction of the CT angiogram showing the lumen of the giant serpentine aneurysm

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