

## Cerebral Amyloid Angiopathy or Frontotemporal Dementia? A Case Study

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### Abstract

A 67 year old patient developed progressive behavioral abnormalities and executive dysfunction corresponding to the diagnosis of behavioral variant of frontotemporal dementia (bvFTD). Unexpectedly, MRI showed multiple vascular ischemic and hemorrhagic lesions as typically found in sporadic cerebral amyloid angiopathy (SCAA). Based on clinical, laboratory and imaging findings the diagnosis of possible SCAA was made. SCAA may represent another important differential diagnosis among the various underlying disease pathologies of the bvFTD syndrome.

**Keywords:** behavioral variant frontotemporal dementia; cerebral amyloid angiopathy; behavioral abnormalities; dysexecutive syndrome

### Introduction

The behavioral variant of frontotemporal dementia (bvFTD) is characterized by neuropsychiatric abnormalities (disinhibition, apathy, loss of empathy, stereotyped behavior, hyperorality) and neuropsychological deficits (predominantly dysexecutive impairments), combined with frontal and/or anterior temporal atrophy [1]. Several conditions may mimic bvFTD due to their overlapping clinical presentation, such as, e.g. Alzheimer's disease (AD) [2], vascular cognitive impairment [3], particularly in patients with subcortical vascular lesions [4,5], mixed dementia [6] and also psychiatric conditions [7,8]. Sporadic cerebral amyloid angiopathy (SCAA) is an important clinical entity which is present in up to 40% of elderly brains and 80% or more in patients with concomitant Alzheimer's disease [9,10]. SCAA is associated with meningeal or parynchymal amyloid- $\beta$  deposition within cerebral blood vessels causing non-traumatic lobar hemorrhages, microbleeds, focal superficial or sulcal siderosis and diffuse white matter lesions. Spontaneous microbleeds are found in neocortical, posterior regions, and later progress to other cortical areas [9,11]. Due to variable involvement of neural structures, SCAA presents heterogeneously. Clinical correlates of vascular dysfunction in SCAA include recurrent focal neurological symptoms and signs, and progressive cognitive and functional decline [12-14]. The cognitive impairment of SCAA through hemorrhagic and ischemic pathomechanisms comprises mainly memory, executive functions and perceptual speed [15]; it may progress to MCI [16] and eventually to dementia [10]. Prominent psychopathology may also be present in SCAA [17,18]. SCAA appears as a distinct cerebrovascular condition, or comorbid with AD [19,20]. Here we report a case of SCAA whose striking behavioral abnormalities and neuropsychological test results closely resembled bvFTD.

### Case Report and Methods

The patient is a 67 year old right handed male (11y formal education), founder and manager of a large business, with a 2 year history of progressive behavioral abnormalities and cognitive dysfunction. He has no family history of dementia or strokes and reported no vascular risk factors such as hypertension, cardiac, metabolic or vessel disease. He never complained of headache. Over the past 24 months he had two short episodes of a sudden, slight left sided hemiparesis with full recovery within hours which were interpreted as TIAs. Neuropsychiatric abnormalities comprised apathy and social withdrawal, loss of spontaneous ideas, motivation and long-standing interests, excessive periods of daytime sleep and neglect of hygiene and

clothing. He developed a new preference for sweets, alcohol and sexual encounter, a peculiar attachment to schedules and stereotypes like, e.g. starting his car repeatedly over the day to prevent fading of the battery. These traits were in strong contrast to his premorbid personality which was described as active, interested, highly motivated, conscientious, flexible, well organized and behaviorally controlled. Topographical disorientation and problems with more complex, e.g. financial or household tasks were noted as cognitive impairments. Progressive memory decline was reported over the past 12 months, but there was no history of amnesic episodes before that time. The patient remained completely anosognosic despite the progression of impairments and his functional decline. There was no gait disorder, bladder dysfunction or history of seizures. Neurological examination was normal and the Hachinski score was 2/10. Because of the striking neuropsychiatric and cognitive abnormalities, the patient was diagnosed with possible bvFTD and underwent a detailed assessment of psychopathology, neuropsychology, as well as MRI, FDG-PET, EEG and laboratory investigation.

### Results

Assessment of behavior and cognition (Tables 1 and 2). Behavior was rated on standard scales by a knowledgeable family member and partly also by self-rating. In sum, the patient was characterized as apathetic, poorly motivated, hyperoral, with problems to plan, self-organize, execute and monitor more complex routines and to handle personal care. Also notified were problems to make proper decisions or to interpret the behavior of other people correctly. Self and proxy ratings differed considerably in the judgement of executive abilities, behavioral control and apathy. Neuropsychological testing revealed severe impairments of global cognition (MMSE), verbal and figural memory (CERAD [21]) and executive functions (FAB [22]) verbal and figural fluency, trail making tests, clock drawing, Stroop color-word task

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|  |                               |     |                |
|--|-------------------------------|-----|----------------|
| Hospital Anxiety and Depression Scale [48] | Anxiety                       | 4   | Cut-off: 11    |
|  | Depression                    | 0   | Cut-off: 11    |
| Apathy Evaluation Scale [49]               | Patient                       | 35  | Possible       |
|  | Caregiver                     | 49  | Present        |
| The Dysexecutive Questionnaire [50]        | Self-rating                   | 13  | PR 18          |
|  | Caregiver's rating            | 41  | PR 69          |
| Frontotemporal Dementia Scale [51]         | Behavior                      | 1   |                |
|  | Outing and shopping           | 2   |                |
|  | Household chores and phone    | 0   |                |
|  | Finances                      | 1   |                |
|  | Medication                    | 1   |                |
|  | Meal preparation and eating   | 3   |                |
|  | Self-care and mobility        | 3   |                |
|  | Total score/28                | 11  | Moderate stage |
| Frontotemporal Behavioral Scale [52]       | Self-monitoring dyscontrol    | 1   |                |
|  | Self-neglect                  | 1   |                |
|  | Self-centered behavior        | 1   |                |
|  | Affective disorders           | 1   |                |
|  | Total score/4                 | 4   | FTD present    |
| Clinical Dementia Rating [27]              | Memory                        | 1   |                |
|  | Orientation                   | 1   |                |
|  | Judgement and problem solving | 1   |                |
|  | Community affairs             | 0,5 |                |
|  | Home and hobbies              | 1   |                |
|  | Personal care                 | 1   |                |
|  | Composite score               | 1   | Mild dementia  |
| FTDL-modified CDR [53]                     | Language                      | 0,5 |                |
|  | Bahviour and personality      | 1   |                |

Table 1: Assessment of behavior.

[23]). On a gambling task (Game of Dice Task [24]) the patient made many risky decisions. Recognition of facial emotions [25] and more complex mental arithmetics [26] were also impaired. Object naming was defective and comprehension (Token test) was mildly impaired, whereas other standard language functions were normal. Dementia severity (CDR [27]) was estimated as mild.

### Structural MRI

MRI (Siemens Magnetom Symphony TIM 1.5 Tesla Scanner) showed no signs of large vessel pathology (TOF) but markedly confluent white matter hyperintensities, particularly in the frontal, left temporal and right temporo-parietal areas (Figure 1A). Infarctions and posthemorrhagic cavities were found in the frontal area, basal ganglia, left and right temporal regions (Figure 1B) and the PICA territory. There were several microbleeds and areas of superficial siderosis as well as old bifrontal laminar hemorrhages (T2\*, Figure 1C). TOF-MRA showed no evidence for sclerosis in the circle of Willis. Hippocampal atrophy was rated as 3 on the Scheltens score, Figure 1D).

### Volumetric analysis

High-resolution structural scans were obtained with a T1-weighted magnetization-prepared rapid gradient echo sequence and were segmented into 184 subcortical and cortical as well as 70 white matter brain regions using FreeSurfer (version 5.3) [28]. Standard deviation

|   |   | Raw score               | Percentiles/Cut-off |
|---|---|-------------------------|---------------------|
| Global cognition  | MMSE                                      | 16                      | PR 0                |
|   | Verbal memory, sum of learning trials 1-3 | 4                       | PR 0                |
|   | Verbal free recall                        | 2                       | PR 1,4              |
|   | Recognition correct minus false positives | 6                       | PR 1                |
|   | Intrusions                                | 0                       | PR 79               |
| Dementia screening (CERAD, [21])                              | Constructional praxis                     | 10                      | PR 21               |
|   | Constructional praxis recall              | 4                       | PR 0                |
|   | Animals/min                               | 5                       | PR 0                |
|   | S - words/min                             | 2                       | PR 0                |
|   | Trail A                                   | 99 s                    | PR 0                |
|   | Trail B                                   | Prematurely terminated  |                     |
| Language (Aachener Aphasia Test, [54])                        | Token - Test errors                       | 15                      | PR 74               |
|   | Repetition (trial 2-5)                    | 119                     | PR 99               |
|   | Comprehension Total Score                 | 85                      | PR 59               |
| Short-term Storage  | Digits forward/backward                   | 06-Mar                  | PR 28 / 5           |
|   | Conceptualization                         | 1                       |                     |
| Frontal Assessment Battery [22]                               | Lexical Fluency                           | 0                       |                     |
|   | Motor programming                         | 0                       |                     |
|   | Sensitivity to interference               | 1                       |                     |
|   | Inhibitory Control                        | 1                       |                     |
|   | Environmental autonomy                    | 3                       |                     |
|   | Total Score/18                            | 6                       | Cut-off: 13         |
|   | Figural Fluency [55]                      | Total number of designs | 20                  |
| Total number of unique designs                                |   | 10                      | PR 3                |
| Figural conceptualization                                     | Clock Drawing Task                        | 8                       | Cut-off: 11         |
| Inhibition of Interference [23]                               | Stroop Colour Word Test                   | 72 s                    | PR<10               |
|   | Risky Decision                            | 9                       |                     |
| Game of Dice Task [24]  | Unrisky decisions                         | 9                       |                     |
|   | Net Score                                 | 0                       | PR 14               |
|   | Addition                                  | 4                       |                     |
| Number Processing and Calculation Battery, short version [26] | Subtraction                               | 3                       |                     |
|   | Multiplication                            | 3                       |                     |
|   | Division                                  | 1                       |                     |
|   | Total Score/20                            | 11                      | Cut-off: 14         |
|   | Anger                                     | 7                       | Cut-off: 4          |
|   | Disgust                                   | 1                       | Cut-off: 6          |
| Facial Expressions of Emotion [25]                            | Fear                                      | 1                       | Cut-off: 3          |
|   | Happiness                                 | 10                      | Cut-off: 9          |
|   | Sadness                                   | 6                       | Cut-off: 5          |
|   | Surprise                                  | 4                       | Cut-off: 6          |
|   | Grand total/66                            | 29                      | Cut-off: 41         |

Table 2: Neuropsychology.

of patient's volumetric measures were calculated by z-transformation using mean centering and unit-variance scaling of age adjusted healthy male subjects (n=59). The analysis revealed a pattern of cortical, subcortical and white matter atrophy. Significant bilateral atrophy (>2 SDs below control mean) was found in the thalamus and amygdala



Figure 1A: Confluent white matter hyperintensities and ventricular enlargement (FLAIR).

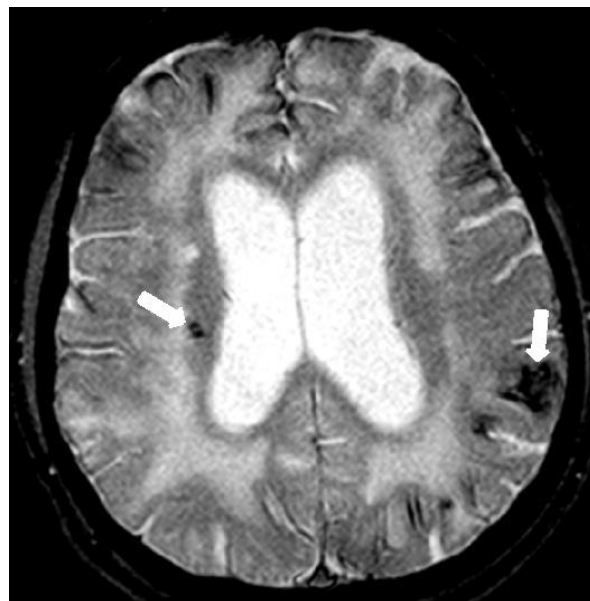


Figure 1C: Microbleeds and siderosis (T2).

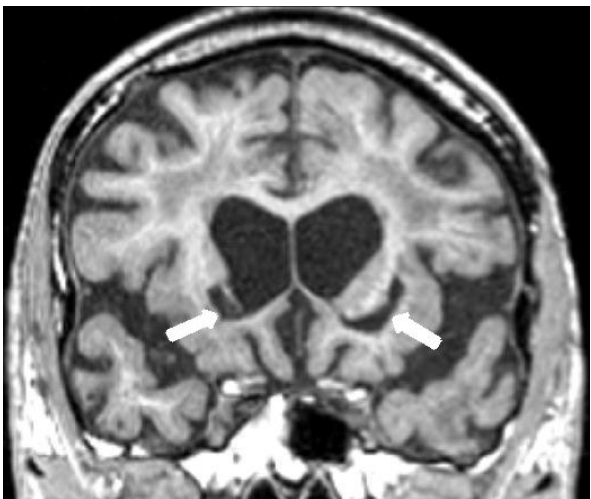


Figure 1B: Bilateral post-hemorrhagic cavities in basal ganglia - internal capsule region (T1).

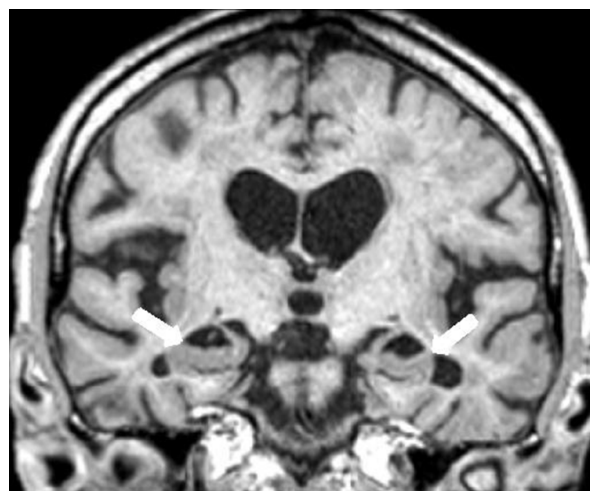


Figure 1D: Hippocampal atrophy (T1).

(range -2 to -2.8 SD), in cortical areas (superior and inferior frontal, inferior and middle temporal, orbital, cingulate, precuneus, superior parietal and angular region, hippocampal formation, cerebellum; range -2 to -6.6 SD) and in the white matter (orbitofrontal, fusiform, inferior and superior temporal, parahippocampal, superior frontal; range -2 to -8.25 SD) of both hemispheres; atrophy of grey and white matter was more advanced in the right hemisphere.

#### FDG-PET

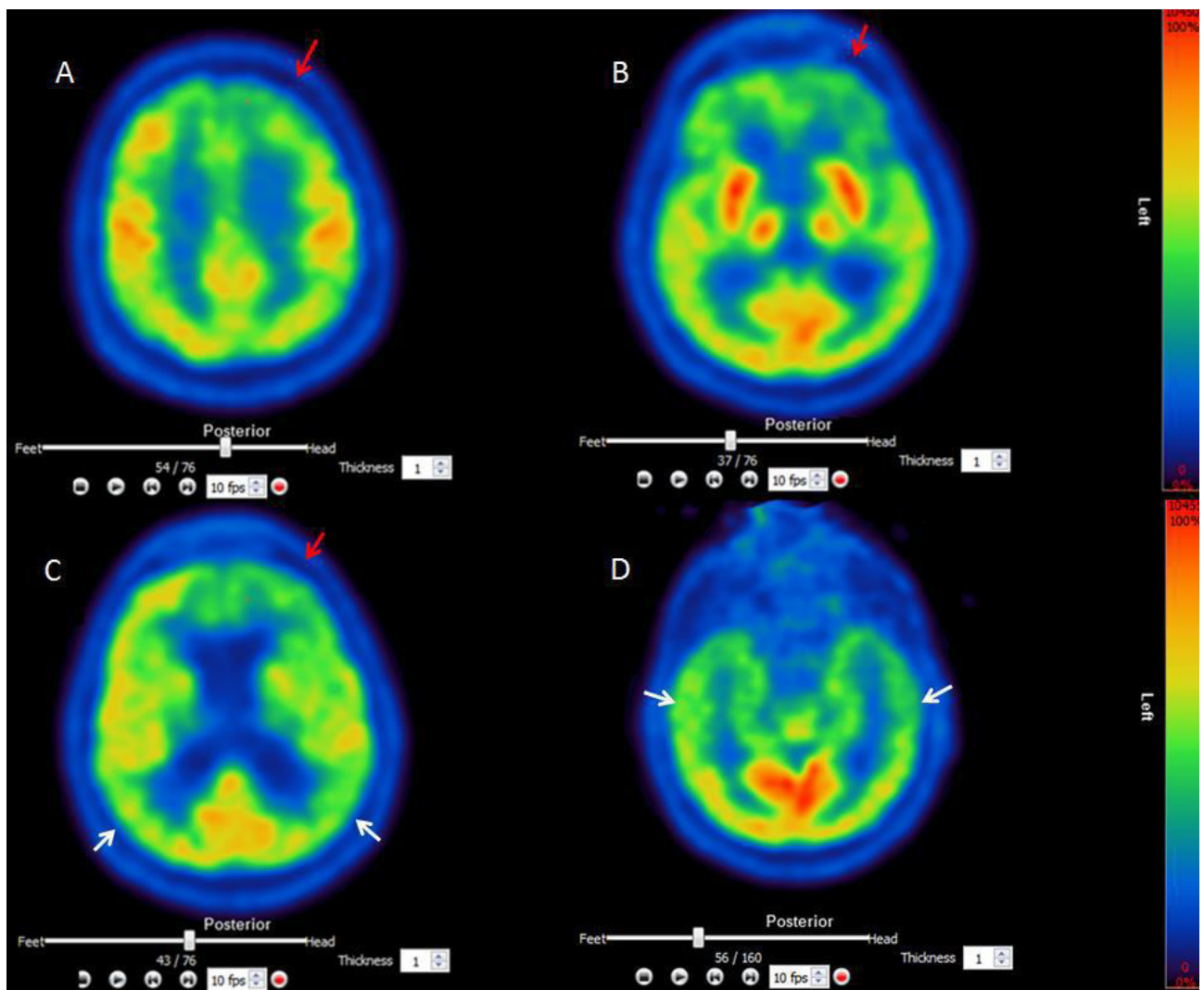
[18F]-FDG Brain PET images showed markedly decreased FDG uptake in frontal, temporal and parietal brain areas (Figures 2A-2D.)

#### Laboratory findings

Routine laboratory findings including measures of coagulation were all normal. Analysis of CSF revealed normal total tau (201 pg, n.v. 149+/-149) and phospho-tau (34 pg, n.v. 30+/-5), but low  $\beta$ -amyloid (246 pg, n.v. 744+/-20). The APOE genotype was  $\epsilon$  2/2. Serum lipids (cholesterol, LDL, HDL and triglycerides) were all in the normal range. EEG was moderately abnormal with intermittent slowing of Alpha (6-8 Hz) and a high content of intermittent irregular diffuse delta-theta waves.

#### Discussion

Progressive neuropsychiatric and dysexecutive features as core symptoms in a relatively young patient strongly suggest the diagnosis of bvFTD, which is commonly caused by tau, TDP-43 or Pick pathology.



**Figure 2:** (A-D) FDG-PET. Decreased FDG uptake in the dorsolateral frontal lobe (left>>right) and anterior cingulate (A, B and D, red arrow), in posterior parietal areas (C, white arrow) and in both temporal lobes (left>right) (D, white arrow).

However, other neurological disorders such as atypical AD [29], subcortical ischemic vascular [4] or mixed type dementia [6,30] may have a similar clinical presentation. SCAA is frequently found in elderly subjects; it often appears together with other neuropathologies, mostly AD. It is a known risk factor for cognitive decline [13,31]. Although definite SCAA can only be diagnosed by post-mortem examination of the brain, a probable clinical diagnosis can be made with imaging support [32,33]. In our case, the diagnosis of SCAA was based on clinical (reversible ‘amyloid spells’), behavioral, cognitive and imaging abnormalities. Behavioral abnormalities were apathy, altered food preferences, disinhibition and stereotyped behaviors. Dysexecutive impairment was pronounced and mainly comprised flexibility, inhibitory control, risk estimation, effortful self-initiation and psychomotor speed. Moreover, episodic memory and emotion recognition were also defective. MRI revealed major leukoaraiosis, microbleeds and siderosis. A volumetric analysis showed combined atrophy in subcortical structures (basal ganglia, amygdala), white matter tracts and cortical regions (frontal, temporal, parietal, hippocampal cortex). FDG-PET showed regions of hypoperfusion mainly in the

frontal, but also temporal and parietal lobes. This lesion pattern matches the clinical picture due to disrupted communication of brain regions [34], necrosis, vascular dysfunction and SCAA-related cortical thinning [35]. The patient’s APOE genotype was  $\epsilon 2/2$ ; both APOE  $\epsilon 4$  and  $\epsilon 2$  alleles are associated with more severe SCAA [36]. Decreased CSF levels of  $\beta$ -amyloid implicate vascular amyloid deposition [37,38]. However, approximately 80% of all subjects with moderate to severe SCAA have a pathologic diagnosis of AD [13,36,39] and the question of SCAA as distinct condition vs. SCAA associated with AD remains unclear without further examination [11].

Although clearly resembling bvFTD, the exact diagnosis, type and number of underlying pathologies remain unclear in the present case without neuropathological confirmation. However, in addition to bvFTD and atypical AD [29,40], SCAA appears to be an important differential diagnosis in patients with progressive neuropsychiatric symptoms and executive impairment [17,18,41]. The diagnosis of SCAA is crucial and can have essential therapeutic consequences with regard to thrombolytic or anticoagulation therapy [42,43],

blood pressure control [44] and prognosis due to their risk of further spontaneous brain hemorrhages and consequent dementia [45]. MRI depicting vascular lesions currently represents the best disease marker of SCAA [46]. Our patient never had symptoms of a documented stroke and the disease course was slowly progressive. This suggests that MRI should also be performed in cases without stroke-like presentation. Details regarding the overlap between SCAA with other degenerative syndromes [47] are presently unclear and need further study.

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