Cerebral Amyloid Angiopathy-related Inflammation Presenting with Steroid-responsive Higher Brain Dysfunction: Case Report and Review of the Literature

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Abstract
A 56-year-old man noticed discomfort in his left lower limb, followed by convulsion and numbness in the same area. Magnetic resonance imaging (MRI) showed white matter lesions in the right parietal lobe accompanied by leptomeningeal or leptomeningeal and cortical post-contrast enhancement along the parietal sulci. The patient also exhibited higher brain dysfunction corresponding with the lesions on MRI. Histological pathology disclosed β-amyloid in the blood vessels and perivascular inflammation, which highlights the diagnosis of cerebral amyloid angiopathy (CAA)-related inflammation. Pulse steroid therapy was so effective that clinical and radiological findings immediately improved.

CAA-related inflammation is a rare disease, defined by the deposition of amyloid proteins within the leptomeningeal and cortical arteries associated with vasculitis or perivasculitis. Here we report a patient with CAA-related inflammation who showed higher brain dysfunction that improved with steroid therapy. In cases with atypical radiological lesions like our case, cerebral biopsy with histological confirmation remains necessary for an accurate diagnosis.

Keywords: cerebral amyloid angiopathy, CAA-related inflammation, higher brain dysfunction

Background
Cerebral amyloid angiopathy (CAA) is a common pathology in the elderly characterized by the deposition of amyloid proteins within the leptomeningeal and cortical arteries [1]. Recently, coexisting inflammations in CAA patients, such as vasculitis or perivasculitis, which clinically resemble central nervous system vasculitis, have been recognized as CAA-related inflammation [2,3]. The inflammation typically responds well to steroid therapy [4], and recent studies have pointed out its similarities with meningoencephalitis induced by immunization to Aβ in Alzheimer disease patients [4-6]. Herein we report a patient with CAA-related inflammation who showed convulsion in the left lower extremity and higher brain dysfunction; both were dramatically improved by steroid therapy.

Case presentation
A 56-year-old man first noticed discomfort in his left lower limb in January 2010. After 7 days, convulsion in the left lower limb suddenly occurred, and he was transported to the emergency hospital. Magnetic resonance imaging (MRI) showed increased white matter intensities in the right parietal lobe on T2-weighted and fluid attenuated inversion-recovery (FLAIR) images. T1-weighted gadolinium (Gd)-enhanced images revealed enhanced leptomeningeal lesions along the parietal sulci (Figure 1A-B). No microhemorrhages were observed with Gradient-recalled echo (GRE)-T2* imaging (1.5T). He was referred to our institution.

On admission, neurological exam showed mild hyperesthesia in the left lower limb and mild hypalgesia in the...
left crus. No other abnormal findings were present. Bio-
chemical screening tests were generally normal except
for serum C-reactive protein (0.77 mg/dL), soluble inter-
leukin-2 receptor antibody (462 U/mL), erythrocyte sedi-
mentation rate (26/1 h, 72/2 h), and carcinoembryonic
antigen (4.5 ng/mL). In the cerebrospinal fluid, protein
levels were elevated (72 mg/dl) and the cell count was
mildly elevated (12/μL).

Because a follow-up MRI revealed progression of the
white matter lesions and parenchymal enhanced lesions
without microhemorrhages (GRE-T2* imaging; 3T) (Fig-
ure 1C-G), a brain biopsy was performed in March
2010. Histological pathology showed nonspecific menin-
goencephalitis involving perivasculitis of the leptome-
ninges and cortical gray matter (Figure 2A-D).

Starting in April 2010, the patient complained of diffi-
culty with his handwriting. Neuropsychological tests of
higher brain functions revealed mild constructional apraxia,
line imbalance for words and numbers, difficulty drawing a
figure following oral instructions, and problems with visual
reproduction. No apathy or dementia was observed.

After the episode, further histological analysis with
Congo-red staining disclosed amyloid laden blood vessels.
Immunohistochemical staining for β-amyloid led to the
diagnosis of CAA-related inflammation (Figure 2F-G).

Steroid pulse therapy (methylprednisolone 1 g/day for 3
days) was performed. The abnormal Gd-enhanced findings
immediately improved with gradually decreasing FLAIR
findings, and the higher brain dysfunctions also gradually
resolved (Figure 3).
Figure 2  Histological and immune-histological examination of brain biopsy. Microscopic examination showed nonspecific meningoencephalitis involving perivasculitis of leptomeninges (arrows) and cortical gray matter (A). The cellular infiltrate was mainly composed of CD-3-positive T-lymphocytes (B) and CD-68-positive macrophages (C) with minimal CD-20-positive B-lymphocytes (D). PAS staining showed no deposits (E). Congo-red staining revealed amyloid positive blood vessels (F); the amyloid was disclosed to be amyloid-β by immunohistochemical staining (G).
Figure 3 Clinical course of treatment with steroid. Abnormal T1 Gd-enhanced findings immediately improved in the fifth course of steroid pulse therapy, accompanied by a gradual decrease of FLAIR findings and a gradual improvement in higher brain function. As the MRI lesions improved (05/28), the descriptions of the 3D-house and sunflower were made more vivid (05/25). Because T1 Gd-enhanced lesions almost disappeared after the fifth course of the steroid (05/28), we stopped the steroid therapy, and the lesion relapsed (06/04). However, after the initiation of oral steroid therapy, no relapse was observed either clinically or radiologically (08/17).
After the fifth course of steroid pulse treatment, the T1-enhanced lesions had almost disappeared, and we stopped the treatment. However, 2 weeks later, the lesions had relapsed on a follow-up MRI, although no clinical signs were observed. We performed pulse steroid therapy again, followed by oral methylprednisolone therapy (70 mg/day). After the oral steroid therapy was initiated, no relapses were observed either clinically or radiologically. Two months later, the oral steroid was tapered at a rate of 5 mg/week, and he was discharged on a regimen of methylprednisolone 30 mg/day.

**Discussion**

CAA is defined by the deposition of amyloid proteins within leptomeningeal and cortical arteries, arterioles, and capillaries [1]. Recently, a subset of patients who presented with seizures, subacute cognitive decline, or headaches with hyperintensities on T2-weighted or FLAIR MRI images with microhemorrhages were described as having CAA-related inflammation [2,3]. Neuropathologic examination has generally revealed angiitis of CAA-affected vessels and peripheral inflammation, presenting as vasculitis or perivasculitis [7]. Both pathologic forms can co-exist, and it has been suggested that the prognosis is better for the perivascular type [8]. This inflammation appears to represent an autoimmune response to vascular β-amyloid deposits. The mechanism by which this immune response occurs is not well understood, although one possible factor is the increased frequency of apolipoprotein E ε4/ε4 genotype [9].

The clinical spectrum of CAA-related inflammation is mainly composed of rapidly progressive dementia and seizure. Although the initial presentation of our case was seizure and numbness, the subsequent higher brain dysfunction is uncommon. To clarify how often higher brain dysfunction has been observed, we reviewed previous cases including our case (Table 1) [1,3,4,7-37]. In 64 cases, 10 presented with higher brain dysfunction without encephalopathy or dementia (15.3%). The most frequent symptom was aphasia (6 cases: 9.3%), followed by hemineglect (2 cases: 3.1%). One other case was reported of various higher brain dysfunction without mental change or dementia, like our case [23]. In these ten cases with higher brain dysfunction, MRI lesions and the presence of leptomeningeal enhancement were inconsistent, and thus the presentation of higher brain dysfunction was considered to be derived from the observed lesion rather than specific to CAA-related inflammation.

The MRI presentation for CAA-related inflammation was previously described as characterized by large confluent areas of predominantly white matter hyperintense signal on T2-weighted or FLAIR images [34]. These lesions are typically asymmetric and involve one or more cortical lesions without evident preferential laterality. T2-weighted gradient-echo sequence images usually showed multiple scattered cortical or subcortical microhemorrhages [34]. However, these microhemorrhages were not observed in our case, resulting in a delayed diagnosis. In our review, 13 cases were examined by MRI with an echo gradient sequence, and microhemorrhages were not seen in 2 cases including our case (13.3%). A possible explanation is that the inflammation caused by the immunoreactivity to amyloid might precede the vascular change of cerebellar amyloid angiopathy in some cases, such that microhemorrhages were not observed in radiological exams. This suggests that the gradient-echo sequence image might not be adequate for diagnosis of CAA-related inflammation in all cases. Brain biopsy should be considered if CAA-related inflammation is highly suspected from clinical presentation, even if microhemorrhages were not radiologically observed.

Approximately three quarters of all patients described had a good clinical response to corticosteroid therapy. Additionally, patients presenting with CAA and meningeal enhancement seem to have less progressive disease [29]. In our review, the leptomeningeal enhancement status of 42 patients was mentioned, and the clinical courses of 39 patients were described. Among 19 patients with leptomeningeal enhancement, only one patient died (5.3%) and the remaining 18 patients survived. However, among the other 20 patients without enhancement, 7 patients died (35%), suggesting that leptomeningeal enhancement might be a good prognostic factor.

The distinctive pattern of asymmetric MRI lesions in CAA-related inflammation appears to be distinguishable from both non-inflammatory CAA and other causes. This observation raises the possibility that typical MRI findings should prove sufficient to diagnose CAA-related inflammation without necessitating brain biopsy [4]. However, in our case, preoperative imaging did not show the typical microhemorrhages associated with CAA, and the diagnosis could not have been established before biopsy. Therefore, we suggest that cerebral biopsy with histological confirmation remains necessary for an accurate diagnosis.

**Conclusion**

We described a patient with CAA-related inflammation whose higher brain functions were dramatically improved by steroid therapy. Because the improvement of cognitive function paralleled resolution of the lesions seen on MRI, this report demonstrates clinically and radiologically progressive improvement of CAA-related inflammation. Our case also suggests the importance of brain biopsy for diagnosis in a case with atypical radiological findings, because correct diagnosis and treatment are crucial for successful recovery and good prognosis.
| Reference          | n  | Age | Sex | Clinical presentation | MRI lesion | Micro bleeds | MRI enhanced lesion | Pathology | treatment | Outcome |
|--------------------|----|-----|-----|------------------------|------------|--------------|---------------------|-----------|-----------|---------|
| Greenberg et al. 1993 [10] | 1  | 72  | F   | dementia headache      | left frontal | NA           | (-)                 | vasculitis | NA        | NA      |
| Ortiz et al. 1996 [11]   | 1  | 68  | F   | headache               | right temporal/parietal | NA           | (-)                 | vasculitis | steroid   | NA      |
| Fountain et al. 1996 [12] | 2  | 66  | M   | fluent aphasia right hemianopia | bilateral temporal/parietal | NA           | (-)                 | vasculitis | steroid cyclophosphamide | alive relapse (+) |
|                       | 69  | F   |     | headache confusion focal neurology seizure | bilateral confluent multifocal | NA           | NA                 | vasculitis | steroid cyclophosphamide | died relapse (+) |
| Anders et al. 1997 [13] | 2  | 70  | M   | mental status change   | right frontal | NA           | NA                 | vasculitis | NA        | NA      |
|                      | 69  | M   |     | headache lethargy behavior change | bilateral white matter | NA           | (+)                | vasculitis | NA        | NA      |
| Fountain et al. 1999 [14] | 1  | 71  | M   | headache confusion gait difficulty left hand apraxia | right temporal/parietal | NA           | NA                 | vasculitis | cyclophosphamide | alive relapse (+) |
| Scully et al. 2000 [15] | 1  | 63  | M   | behavior change ataxia | bilateral white matter | NA           | (+)                | perivasculitis | cyclophosphamide | alive |
| Oide et al. 2002 [16]   | 1  | 69  | M   | dizziness dementia seizure | bilateral symmetrical periventricular | NA           | NA                 | vasculitis | (-)       | NA      |
| Schwab et al. 2003 [8]  | 2  | 74  | M   | seizure dementia headache | bilateral multifocal | NA           | (+)                | perivasculitis | steroid        | alive relapse (+) |
|                       | 70  | F   |     | dementia headache      | right temporal | NA           | (+)                | perivasculitis | steroid        | alive relapse (+) |
| Tamargo et al. 2003 [17] | 1  | 80  | F   | dementia left-side hemineglect word finding difficulty | bilateral left frontal right parietal | NA           | (+)                | vasculitis | steroid        | alive |
| Oh et al. 2004 [1]       | 2  | 80  | F   | Headache aphasia       | bilateral right parietal/occipital left frontal | NA           | (-)                | perivasculitis | steroid        | alive |
|                        | 77  | M   |     | aphasia                 | left temporal | NA           | (-)                | vasculitis | steroid        | alive |
| Safriel et al. 2004 [18] | 1  | 49  | M   | seizure                 | right occipital/temporal | NA           | (-)                | vasculitis | steroid        | alive |
| Hashizume et al. 2004 [19] | 1  | 65  | M   | headache left hemianopia left-side hemineglect | right temporal/occipital | NA           | (+)                | vasculitis | steroid cyclophosphamide | died |
| Harkness et al. 2004 [20] | 1  | 72  | F   | dementia                 | bilateral frontal | NA           | (-)                | vasculitis | no specific therapy | alive |
| Jacobs et al. 2004 [21]  | 1  | 81  | F   | confusion Balint's syndrome agraphia right-left confusion finger anomia left-side neglect | bilateral parietal/occipital | NA           | (+)                | vasculitis | steroid        | alive |
Table 1: Review of reported cases of CAA-related inflammation (Continued)

| Study Authors       | Age | Sex | Gender | Presenting Symptoms | Neuroimaging | Treatment | Outcome |
|---------------------|-----|-----|--------|---------------------|--------------|-----------|---------|
| Scolding et al. 2005 | 69.3| M   | 3      | Encephalopathy 6 focal neurology 2 seizure 1 headache 2 | NA           | (+) 5 perivasculitis | Died 2   |
| Mikolaenko et al. 2006 | 50  | M   | 3      | Seizure right frontal | NA           | (+) vasculitis | Alive   |
| Wong et al. 2006    | 79  | F   | 9      | Higher brain dysfunction fatigue right frontal/temporal/parietal | NA           | (+) vasculitis | Alive relapse (+) |
| Kimnecom et al. 2007 | 62.3| M   | 9      | Encephalopathy headache 5 seizure 7 aphasia 1 presyncope 1 | NA           | NA (the presence of microbleeds are mentioned but the proportion is not mentioned) | Alive relapse (+) 3 |
| Greenberg et al. 2007 | 63  | M   | 3      | Headache behavioral change cognitive change bilateral multiple | NA           | (+) vasculitis | Alive relapse (+) |
| Marotti et al. 2007 | 57  | F   | 9      | Headache seizure bilateral frontal/temporal/insular right thalamus | (+)          | (+) vasculitis | Died    |
| McHugh et al. 2007  | 80  | F   | 9      | Confusion incontinent urine global aphasia seizure right hemianopia right hemiparesis bilateral frontal | (+)          | (-) perivasculitis | Alive relapse (+) |
| Takada et al. 2007  | 69  | F   | 9      | Headache cognitive decline bilateral right frontal/parietal bilateral parietal/occipital | (+)          | (-) vasculitis | Died    |
| Machida et al. 2008 | 69  | F   | 9      | Cognitive decline bilateral multifocal | (-)          | (+) perivasculitis | Alive relapse (+) |
| Salvarani et al. 2008 | 63*| M   | 6      | Encephalopathy 6 focal neurology 2 headache 3 only aphasia with alexia 1 bilateral 8 multifocal | NA           | (+) 5 (-) 3 vasculitis | Improved 6 died 1 worsened 1 |
| Amick et al. 2008   | 79  | F   | 9      | Transient right sided weakness left occipital/parietal | NA           | (-) vasculitis | Died    |
| Alcalay et al. 2009 | 92  | F   | 9      | Mental status change bilateral multifocal | (+)          | (+) (+) | Steroid | Alive |
| Daniëls et al. 2009 | 80  | F   | 9      | Mental status change right sided hemiparesis dysphasia seizure bilateral left hemisphere right parietal/occipital | (+)          | (-) (+) | Steroid | Alive relapse (+) |
| Greenberg et al. 2010 | 87 | F   | 9      | Seizure cognitive impairment bilateral multifocal | (+)          | NA perivasculitis | Died    |
| Kloppenborg et al. 2010 | 74 | M   | 9      | Increased sleepiness loss of initiative seizure bilateral frontal | (+)          | (+) perivasculitis | Alive   |
| Morishige et al. 2010 | 78 | F   | 9      | Motor aphasia dementia left frontal | NA           | (+) vasculitis | Died    |
| Savoardo et al. 2010 | 76  | M   | 9      | Fatigue confusion bilateral temporal/occipital/parietal | (+)          | (-) (+) | Steroid | Alive |

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2. http://www.jneuroinflammation.com/content/8/1/116
### Table 1 Review of reported cases of CAA-related inflammation (Continued)

| Case    | Age | Sex | Symptom(s)                          | Imaging Features | Treatment | Outcome |
|---------|-----|-----|-------------------------------------|------------------|-----------|---------|
| Cano et al. 2010 [35] | 76  | M   | transient motor aphasia transient headache | bilateral temporal (+) | NA (-) (-) | alive |
| DiFrancesco et al. 2011 [36] | 68  | M   | memory loss mood disorder            | bilateral multifocal (+) | (-) NA steroid | alive |
| Chung et al. 2011 [37] | 83  | F   | seizure                             | bilateral multifocal | NA NA vasculitis steroid | died |
|         |      |     | headache mild hemiparesis sensory loss | right parietal/occipital (+) | NA vasculitis steroid | alive |
|         |      |     | hemianopia                           |                  |            |         |
| our case | 56  | M   | Seizure sensory disturbance higher brain dysfunction | bilateral multifocal | (+) perivasculitis steroid | alive relapse |

From the literature, we extracted the cases of CAA-related inflammation in which an MRI was evaluated. If autopsy or biopsy was examined, the cases without inflammation were excluded. All cases satisfy the diagnostic criteria of definite or probable CAA-related inflammation proposed by Chung et al. [37]. In 64 cases, 10 presented with higher brain dysfunction without encephalopathy or dementia (15.3%). The most frequent symptom was aphasia (6 cases: 9.3%), followed by hemineglect (2 cases: 3.1%). One case besides the current presented with various higher brain dysfunction without mental change or dementia [23]. In these 10 cases with higher brain dysfunction, MRI lesions and the presence of leptomeningeal enhancement were inconsistent. Thirteen cases were examined with MRI with an echo gradient sequence, and microhemorrhages were not seen in 2 cases, including our case (13.3%).

The leptomeningeal enhancement status of 42 patients was mentioned, and the clinical courses of 39 patients were described. Only one patient among 19 patients with leptomeningeal enhancement died (5.3%); however, 7 of 20 patients without enhancement died (35%), suggesting that leptomeningeal enhancement might be a factor in good prognosis. *: calculated mean
Consent
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

List of abbreviations
AJL: amyloid Jr, ADC: apparent diffusion coefficient, CAA: cerebral amyloid angiopathy; FLAIR: fluid attenuated inversion-recovery; Gd: gadolinium; MRI: magnetic resonance imaging; GRE: gradient-recalled echo.

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Authors’ contributions
HS designed this article and direction for investigations and drafted the manuscript. AU, TK, SY, EK, TY, YM, TH, and MU contributed to interpretations of clinical, radiological and pathological details. All authors read and approved the final manuscript.

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All authors are members of the Department of Neurology, Faculty of Life Sciences, Kumamoto University, and TK was also a graduate student of the Brain Research Institute, University of Niigata until March 2011.

Competing interests
The authors declare that they have no competing interests.

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