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ORIGINAL RESEARCH

Cerebral Amyloid Angiopathy-Related Inflammation: Clinical Characteristics and Treatment Experience

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Objective: The present study aims to analyze the clinical manifestations, laboratory results, neuroimaging features, treatment interventions, and outcomes in a cohort of patients with cerebral amyloid angiopathy-related inflammation (CAA-ri), providing a deeper understanding of this rare subtype of CAA and enhancing diagnostic precision in clinical practice.

Methods: We conducted a systematic retrospective review of clinical records from 13 consecutive patients who met the diagnostic criteria for probable CAA-ri and were evaluated at the First Affiliated Hospital of Zhengzhou University between January 2021 and August 2024.

Results: The study cohort comprised 13 patients (7 males, 6 females; mean age 65.2 years, range 42–81), predominantly presenting with subacute onset (53.8%, n=7). Cognitive impairment (61.5%, n=8) emerged as the most frequent clinical manifestation, followed by headache (46.2%, n=6), epileptic seizures (30.8%, n=4), and focal neurological deficits (23.1%, n=3). Neuroimaging findings across all patients demonstrated asymmetric white matter hyperintensities in conjunction with cortical-subcortical cerebral microbleeds. A subset of patients exhibited cortical superficial siderosis lobar hemorrhage, and/or punctate acute infarction. Among the nine patients who underwent lumbar puncture, five showed elevated cerebrospinal fluid (CSF) pressure and protein levels. All four patients assessed for CSF Alzheimer's disease biomarkers showed reduced Aβ42 and Aβ40 levels, alongside elevated total tau and phosphorylated tau levels. Furthermore, over 70% of the patients who treated with immunosuppressive therapy achieved favorable clinical outcomes.

Conclusion: Clinical manifestations and neuroimaging abnormalities serve as pivotal non-invasive criteria for guiding clinicians in the diagnosis of CAA-ri. Timely initiation of immunosuppressive therapy in CAA-ri patients can lead to favorable outcomes.

Keywords: CAA-ri, clinical manifestations, neuroimaging, immunosuppressive treatment

Introduction

Cerebral amyloid angiopathy-related inflammation (CAA-ri) is a rare and treatable subtype of cerebral amyloid angiopathy (CAA). It is triggered by an autoimmune response to the Aβ deposition in the media and adventitia of small and medium-sized cortical and leptomeningeal brain arteries.[1] Two pathological subtypes of CAA-ri have been identified: non-destructive perivascular inflammation, also termed inflammatory CAA (ICAA), and transmural granulomatous inflammation, designated as Aβ-related angiitis (ABRA).[2,3]

CAA-ri is an aggressive disorder, characterized by distinct clinical manifestations and neuroradiological features. The diagnostic criteria for probable CAA-ri, first proposed by Chung et al in 2011 and updated by Auriel et al in 2016, are

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considered the most accurate way to diagnose this disorder without a brain biopsy for confirmation.[4,5] However, clinicians still encounter challenges in achieving timely and accurate identification of CAA-ri, which consequently influences therapeutic decision-making process. This is attributed to the rarity of morbidity and the requirement to exclude differential diagnoses, including intracranial infection, autoimmune encephalitis, posterior reversible encephalopathy syndrome (PRES), primary angiitis of the central nervous system (PACNS), and primary central nervous system lymphoma (PCNSL). Given the favorable response of CAA-ri to immunosuppressive therapy, delayed diagnosis may lead to postponed treatment initiation and adversely affect the prognosis of CAA-ri patients.

Herein, we have compiled a summary of the distinctive characteristics observed in 13 cases of probable CAA-ri diagnosed at our center, aiming to enhance neurologists' awareness of this uncommon disease.

Methods

A retrospective review was conducted on all CAA-ri patients at the First Affiliated Hospital of Zhengzhou University between January 2021 and August 2024. All cases underwent independent evaluation by two seasoned neurologists, with 13 cases ultimately meeting the diagnostic criteria for probable CAA-ri as proposed by Auriel et al in 2016.[4] The study was approved by the Ethics committee of the First Affiliated Hospital of Zhengzhou University (2021-KY-1059-002) and complies with the Declaration of Helsinki. Written informed consent was obtained from all patients included.

The key demographic and clinical data were collected from the hospital registry. Subsequently, we conducted a comprehensive analysis of all patients, focusing on the clinical manifestations of their initial episode, encompassing factors like age, sex, onset form, personal history, and clinical symptoms. The onset patterns were classified into three distinct categories: acute (within 48 hours), subacute (48 hours to 4 weeks), and chronic (longer than 4 weeks). For the initial evaluation, 3-Tesla MRI scans were mandatory, including sequences such as T1-weighted, T2-weighted, fluid-attenuated inversion-recovery (FLAIR), diffusion-weighted imaging (DWI), apparent diffusion coefficient (ADC), and susceptibility-weighted imaging (SWI). Additionally, we collected supplementary data, including cerebrospinal fluid (CSF) test results, ApoE genotype, and details of the respective treatment regimens and outcomes.

All patients had at least one outpatient visit or re-admission after their initial discharge. The final follow-up was conducted via telephone interview on October 30, 2024. Functional outcomes were assessed using the modified Rankin Scale (mRS) at baseline and during follow-up. Patients who received immunosuppressive therapy underwent at least one follow-up MRI examination.

Results

Demographic Data and Clinical Characteristics

A total of 13 patients (female to male ratio ≈ 1:1) were diagnosed with probable CAA-ri.[4] The cohort predominantly comprised elderly individuals, with a mean age of 65.2 years (range 42–81 years) and a median age of 68 years at diagnosis. Comprehensive demographic, clinical, neuroimaging characteristics, and genetic markers were summarized in Table 1.

Table 1 Demographic Characteristics and Clinical Manifestations of All Patients

Characteristic	No./Total No. (%) with Available Information
Mean age	65.2
Female sex	6/13 (46.2)
Personal history	
Hypertension	5/13 (38.4)
Diabetes mellitus	2/13 (15.4)
Stroke history	2/13 (15.4)
Smoking	3/13 (23.1)

(*Continued*)

Table 1 (Continued).

Characteristic	No./Total No. (%) with Available Information
Forms of onset	
Acute onset	3/13 (23.1)
Subacute onset	7/13 (53.8)
Chronic onset	3/13 (23.1)
Clinical symptoms	
Cognitive decline	8/13 (61.5)
Headache	6/13 (46.2)
Focal neurological signs	3/13 (23.1)
Behavioural alterations	1/13 (7.7)
Epileptic seizures	4/13 (30.8)
Hallucinations	1/13 (7.7)
Others	
Vomiting	1/13 (7.7)
Dizziness	2/13 (15.4)
Hypersomnia	1/13 (7.7)
Cerebrospinal fluid (CSF)	209
Mean pressure (mmH_2O)	
Pleocytosis	2/9 (22.2)
Elevated protein (>0.45 g/L)	7/9 (77.8)
OCB	0/9 (0)
Alzheimer's disease biomarkers	
Decreased $A\beta_{42}$	4/4 (100.0)
Decreased $A\beta_{40}$	4/4 (100.0)
Increased t-tau	4/4 (100.0)
Increased p-tau	4/4 (100.0)
APOE genotype	
ε2/-	0
ε3/ε3	0
ε3/ε4	2/4 (50.0)
ε4/ε4	2/4 (50.0)
Magnetic Resonance Imaging (MRI)	
Asymmetric WMH	13/13 (100.0)
Cerebral microbleeds	
Labor microbleeds	13/13 (100.0)
Deep microbleeds	0/13 (0)
Acute or subacute lobe hemorrhage	4/13 (30.8)
Cortical superficial siderosis	2/13 (15.4)
Acute subarachnoid hemorrhage	0/13 (0)
Acute or subacute infarcts	5/13 (38.5)

Hypertension was the most common comorbid condition, affecting 38.4% of the patients. Most patients presented with either acute (3/13, 23.1%) or subacute onset (7/13, 53.8%), while the remaining exhibited chronic onset (3/13, 23.1%). At presentation, the primary clinical symptoms included cognitive decline (8/13, 61.5%), headache (6/13, 46.2%), epileptic seizures (4/13, 30.8%) and focal neurological signs (3/13, 23.1%), such as limb weakness, visual impairment, facial paralysis, dysphagia, and aphasia. Other typical symptoms included behavioral alterations (1/13, 7.7%) and visual hallucinations (1/13, 7.7%). Atypical symptoms, such as dizziness, vomiting, and hypersomnia, were also observed in some patients. Notably, approximately 62% of patients had two or more symptoms mentioned above.

Imaging Features

The neuroimaging characteristics of the cohort presented in Table 2. All patients exhibited cortico-subcortical microbleeds and asymmetric, confluent, patchy subcortical white matter hyperintensity (WMH) lesions confined to the lobes. Cerebral microbleeds (CMBs) were predominantly located in the bilateral hemispheres, with lower frequencies in the cerebellum and brainstem. Most patients had at least 10 CMB lesions. Additionally, subacute lobar hemorrhage was detected in 4 patients (30.8%). While SWI sequences revealed cortical superficial siderosis (cSS) in two patients (15.4%), none of convexity subarachnoid hemorrhage (cSAH) was observed. DWI sequences showed punctate acute or subacute infarcts in 5 patients (38.5%), primarily localized in the cerebral lobes. Figure 1 displays representative neuroimaging findings of CAA-ri.

Cerebrospinal Fluid Results and ApoE Genotype

Among 13 patients, 9 patients underwent lumbar puncture. The mean cerebrospinal fluid (CSF) pressure was 209 mmH_2O (range: 110–370 mmH_2O), with approximately half (55.6%, n=5) demonstrating elevated pressure (normal range: 80 to

Table 2 Imaging Features of the Patients

Patient/ NO.	Subcortical WMH	CMBs	Lobar Hemorrhages	CSS Lesions	Acute Infarcts
1	Bilateral temporal, parietal, occipital lobes	Bilateral frontal, temporal, parietal, occipital lobes	_	_	_
2	Bilateral parietal lobes	Bilateral frontal, temporal, parietal, occipital lobes	_	_	_
3	Bilateral temporal, parietal lobes	Bilateral frontal, temporal, parietal, occipital lobes, cerebellum and brain stem	_	_	_
4	Left frontal lobes and bilateral temporal, parietal, occipital lobes	Bilateral frontal, temporal, parietal, occipital lobes	Right temporal and parietal lobes	_	Left temporal lobe and right periventricular area
5	Bilateral frontal lobes, left parietal lobe and right temporal lobe	Bilateral frontal, temporal, parietal, occipital lobes	_	Right parietal lobe and fissure interhemispheric	_
6	Bilateral frontal, temporal, parietal, occipital lobes	Bilateral frontal, temporal, parietal, occipital lobes	_	_	_
7	Left frontal, parietal lobe and bilateral temporal, occipital lobes	Bilateral frontal, temporal, parietal, occipital lobes	Left temporal lobe, and right occipital lobe	_	_
8	Bilateral centrum semiovales, corona radiates and posterior horn of lateral ventricles	Bilateral frontal, temporal, parietal, occipital lobes and cerebellum	Right frontal lobe	_	Bilateral frontal, temporal, parietal, occipital lobes
9	Bilateral frontal and parietal lobes	Bilateral frontal, temporal, parietal, occipital lobes	_	_	Right frontal, parietal, occipital lobes
10	Bilateral centrum semiovale, corona radiate and posterior horn of lateral ventricles	Bilateral frontal, temporal, parietal, occipital lobes and cerebellum	Right parietal lobe	Bilateral frontal lobes and right parietal lobe	Right temporal lobe
11	Left frontal, temporal lobes and bilateral centrum semiovale	Bilateral frontal, temporal, parietal, occipital lobes and cerebellum	_	_	Bilateral frontal, temporal, parietal, occipital lobes
12	Right frontal, parietal lobes and bilateral temporal, occipital lobes	Bilateral frontal, temporal, parietal, occipital lobes and cerebellum	_	_	_
13	Bilateral frontal, temporal, parietal and occipital lobes	Bilateral frontal, temporal, parietal, occipital lobes	_	_	_

Abbreviations: WMH, white matter hyperintensity; CMBs, cerebral microbleeds; CSS, cortical superficial siderosis.

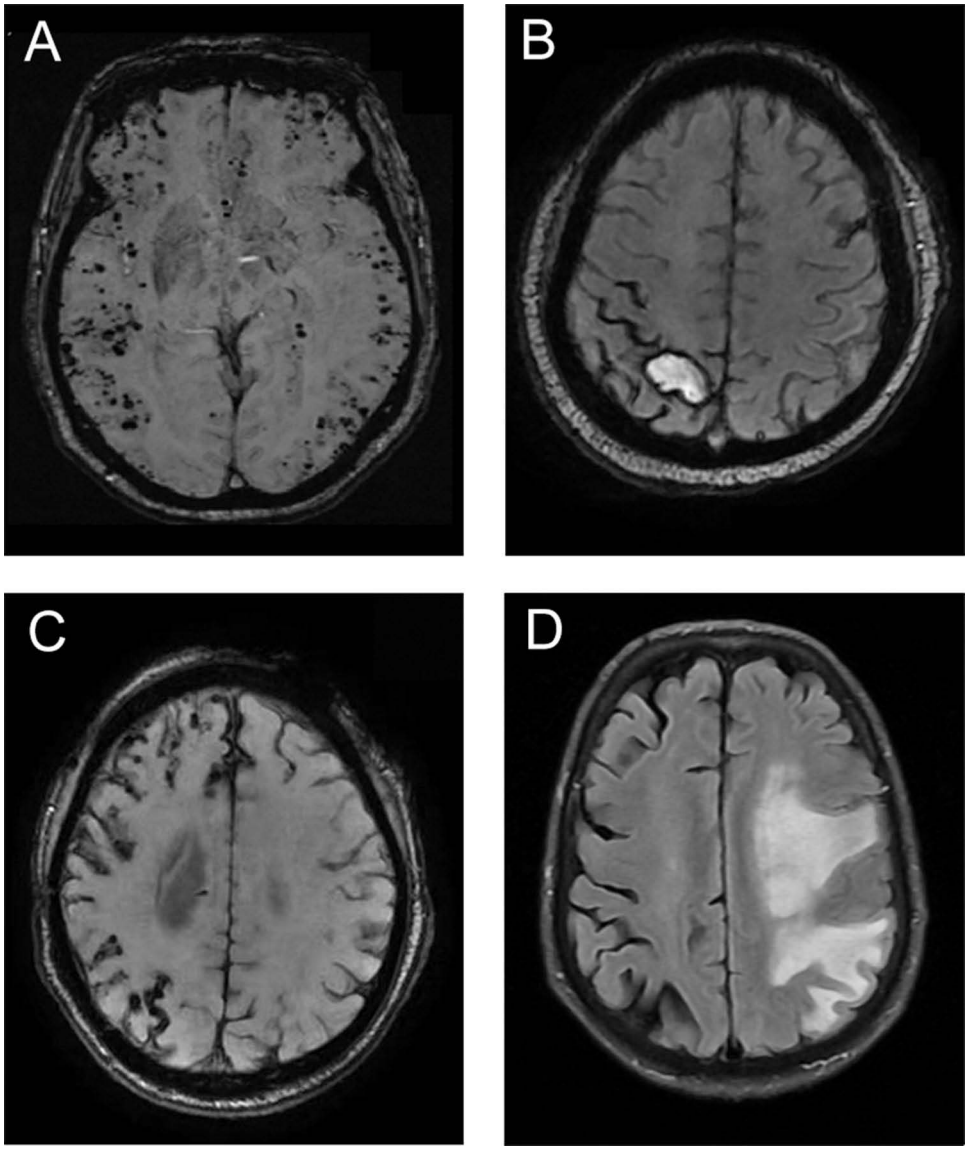

Figure 1 Representative neuroimaging findings of CAA-ri. (**A**) Microbleeds with cortical-subcortical involvement. (**B**) Cerebral hemorrhage in the right parietal lobe. (**C**) cSS in the right parietal lobe and Interhemispheric fissure. (**D**) Asymmetric WMHs in left frontal and parietal lobes.

180 mmH2O). CSF analysis revealed slight pleocytosis in only 2 patients (22.2%). Elevated CSF protein levels (>0.45 g/L) were observed in 7 of 9 patients (77.8%). CSF glucose and chloride levels remained within normal ranges for all 9 patients, and no oligoclonal band detected. In 4 patients, CSF biomarkers for Alzheimer's disease (AD) were evaluated. All CSF samples consistently showed reduced Aβ40 and Aβ42 levels, along with increased total tau (t-tau) and phosphorylated tau (p-tau) protein levels. ApoE genetic testing was performed in 4 patients, revealing two cases of ApoE ε3/ε4 and ApoE ε4/ε4 genotypes, respectively.

Treatment and Prognosis

Table 3 outlines the therapeutic regimens for all patients along with their corresponding clinical and radiological outcomes. In this study, 7 out of the 13 patients received corticosteroids treatment. Conversely, the remaining 6 patients refrained from this therapy due to infection, advanced age, or concerned potential adverse drug effects. Corticosteroids doses and tapering regimens varied among the 7 treated patients, and the prevalent treatment involved administering high-dose intravenous methylprednisolone (IVMP) for 3 to 5 days, subsequently transitioning to oral prednisone (OP)

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Table 3 Treatment and Outcomes of All Patients

Patient/ NO.	Clinical Symptoms	Initial Treatment (at First Diagnosis)	Initial Clinical Response (at First Discharge)	Initial Radiological Response (2–6 Months After Initial Treatment)	Follow-Up
1	Headache	IVMP 500mg/day for 5 days → OP 60mg for 1 months	Improvement	Improvement	Relapse in the 12th month after withdrawal of corticosteroids; improvement after reintroduction
2	Headache, cognitive decline	IVMP 500mg/day for 3 days → OP 60mg for 7 months → Mycophenolate	Improvement	Improvement	Relapse in the 4th month after decrement of corticosteroids; improvement after reintroduction
3	Cognitive decline, dizziness focal neurological signs	IVMP 120mg/day for 10 days	Improvement	Improvement	Relapse in the 4th month after withdrawal of corticosteroids; improvement after reintroduction
4	Cognitive decline, focal neurological signs and visual hallucinations	_	Deterioration	_	Die of multiple organ failure
5	Headache	_	Improvement	_	Improvement
6	Cognitive decline, behavioral alterations and seizures	IVMP 500mg/day for 5 days → OP 60mg for 2 months	Improvement	Improvement	Improvement
7	Headache, dizziness	_	Improvement	No change	Improvement
8	Headache, cognitive decline and limb weakness	IVMP 500mg/day for 5 days → OP 60mg for 2 months	No improvement	Aggravation	Died of cranial hypertension
9	Cognitive decline and seizures	_	No improvement	_	Lost to follow-up
10	Headache, cognitive decline and seizures	_	No improvement	_	Died of cerebral hemorrhage
11	Focal neurological signs	_	No improvement	_	Died of ischemic stroke
12	Cognitive decline	IVMP 500mg/day for 3 days → OP 60mg for 3 months	Deterioration	No change	Stability
13	Seizures	IVMP 80mg/day for 3 days → OP 60mg/day for 1.5 months	Improvement	Improvement	Recovery

Abbreviations: IVMP, intravenous methylprednisolone; OP, oral prednisone.

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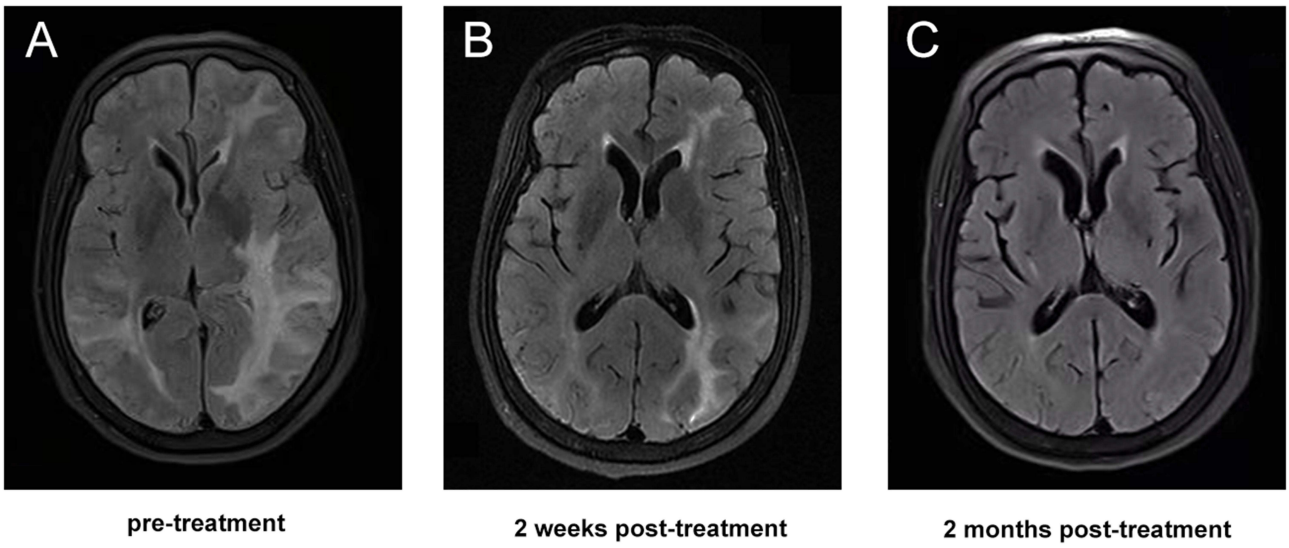

Figure 2 Radiological progression in a representative CAA-ri patient. MRI findings in a 62-year-old woman (patient NO.1) at pre- and post-treatment time points. (**A**) Initial T2/FLAIR imaging revealed marked asymmetric patchy hyperintensities in both cerebral hemispheres. (**B**) At 2 weeks after corticosteroid treatment, FLAIR abnormalities decreased. (**C**) Two months later, follow-up imaging showed further regression of the abnormal lesions.

and gradually tapering the dosage over several weeks or months to discontinuation (patient NO.2 switched to mycophenolate mofetil owing to steroid myopathy). Adjunctive treatments encompassed intracranial pressure reduction, antiepileptic drugs, cognitive enhancement and anti-infectives. Compared with non-corticosteroid-treated patients, CAA-ri patients treated with IVMP showed a higher rate of initial clinical improvement at discharge (33.3% vs 71.4%). After 2–6 months of IVMP therapy, patients underwent at least one MRI scan to evaluate the early radiological response. As expected, neuroimaging improvements correlated with clinical outcome improvements.

Patients were followed up for a mean duration of 2.6 years (range: 0.75–3.5 years), with one patient lost to follow-up. By the end of the follow-up period, among corticosteroid-treated patients, one achieved complete recovery, one died from high intracranial pressure, and five had mRS scores that remained improved or stable Notably, three patients experienced relapses after corticosteroid withdrawal or dose reduction but subsequently improved with treatment reinitiation. Among non-corticosteroid-treated patients, two improved, three died from cerebral hemorrhage, ischemic stroke, or respiratory failure, and one was lost to follow-up. Pre- and post-treatment MRI examples are shown in Figure 2.

Discussion

CAA-ri is an underrecognized and potentially reversible neurological disorder. Here, we report a series of 13 patients diagnosed with this condition at our center, summarizing their clinical, laboratory, and neuroimaging characteristics, treatment approaches, and disease progression, while comparing our findings with those reported in the literature. The mean age of our cohort was 65.2 years, consistent with CAA-ri's predilection for older adults, though younger than that of non-inflammatory CAA patients.[6] Gender distribution was nearly equal. Historically, CAA-ri has been characterized as an acute or subacute condition; however, this criterion was removed from the 2016 updated diagnostic criteria.[4] Notably, 23.1% of our patients presented with chronic onset, which may contribute to increased misdiagnosis risk and prolonged diagnostic delays. Cognitive decline was the most common symptom, followed by headache, epileptic seizures, focal neurologic deficits, and behavioral alterations. Atypical symptoms, such as involuntary movements, sudden memory loss, cerebral herniation due to severe edema, and Parkinson's disease-like mental manifestations, have been reported.[7]

Neuroimaging features represent critical criteria for CAA-ri, enabling non-invasive diagnosis without brain biopsy. Typical radiological findings supporting CAA-ri include non-hemorrhagic marker (asymmetric cortico-subcortical WMH) and hemorrhagic markers (cortico-subcortical CMBs, cSS and lobar hemorrhage). In our study, asymmetric WMH and cortico-subcortical CMBs were the most important combinations guiding clinical suspicion and subsequent

diagnosis of CAA-ri. CMBs are the most prominent hemorrhagic marker in both CAA and CAA-ri, but they are more abundant in CAA-ri. By contrast, lobar hemorrhage is more common in CAA patients.[8] An interesting finding in our cohort was that all detected microbleeds were superficial, with no deep microbleeds observed. Additionally, the distribution of CMBs in CAA-ri did not follow the regional pattern of occipital dominance.[6] Szalardy et al reported that 66.7% of WMH tended to co-localize with CMBs.[9] In our study, CMBs had a broader distribution than WMH. The reported proportions of cSS vary widely across studies (17–50%),[10,11] with our study documenting a proportion of 15.4%. Lobar intracerebral hemorrhage was evident in approximately one-third of CAA-ri patients in our cohort, consistent with a multicenter cohort study.[11] A case report has suggested that CAA-ri should be considered in cases with recurrent lobar hemorrhages, particularly within a short time frame.[12] Additionally, the presence of cortical micro-infarctions on DWI suggests small blood vessel involvement. However, we found that stroke-like symptoms were incongruent with actual cerebral infarcts demonstrating diffusion restriction. This disparity may potentially arise from the fact that punctate infarcts located in the cerebral lobes do not involve functional areas.

Most patients who underwent lumbar puncture had elevated CSF pressure and protein levels. However, lymphocytic pleocytosis—an expected finding in inflammatory processes—was infrequently observed. Although CSF testing for AD biomarkers was performed in only 4 patients, results consistently showed decreased Aβ42 and Aβ40 levels, along with increased t-tau and p-tau levels. These abnormalities are consistent with the typical biomarker changes seen in CAA.[2] The ApoE ε4 allele, particularly ε4/ε4 homozygosity, plays a key role in Aβ fibrilization and amyloid deposition.[13] A meta-analysis has shown that amyloid-related imaging abnormalities (ARIA) following anti-Aβ immunotherapy are more prevalent in ApoE ε4 carriers than non-carriers, indicating that the ApoE ε4 genotype may exacerbate inflammatory processes.[14] Kinnecom et al found that up to 76.9% of 14 patients with a pathological diagnosis of CAA-ri were homozygous for the ApoE ε4/ε4 genotype.[2] In our study, testing of 4 samples revealed a 50% prevalence of ApoE ε4/ε4 homozygosity and 50% ApoE ε3/ε4 heterozygosity. Although CSF AD biomarkers and ApoE genotype are not included in the diagnostic criteria, they may serve as potential non-invasive diagnostic clues to improve CAA-ri diagnostic accuracy.

Existing evidence indicates that timely immunosuppressive therapy should be initiated in patients with probable or possible CAA-ri prior to pathological confirmation.[15,16] Although there are no explicit recommendations regarding medication selection, dosage, or treatment duration, empirical high-dose intravenous corticosteroids for 3–5 days followed by a prolonged tapering of oral steroids are most used in clinical practice. In addition, other immunosuppressive therapies such as cyclophosphamide, azathioprine, mycophenolate, intravenous immunoglobulin, or plasma exchange, may be warranted in cases unresponsive to corticosteroid therapy.[15,17] In our cohort, patients who received corticosteroids treatment showed a favorable outcome, with most demonstrating significant reduction of WMH during follow-up. Two patients experienced relapse after corticosteroid withdrawal, but their condition improved again following treatment reinitiation. This highlights the sustained efficacy of corticosteroids in managing CAA-ri. During follow-up, increasing cortical microbleeds and siderosis were observed in patients treated with immunosuppressants, suggesting that immunosuppressive therapies resolve the acute inflammatory phase of CAA-ri rather than the chronic course of CAA.[18] A minority of published cases have reported spontaneous remission of neurological symptoms and/or imaging abnormalities without immunosuppressants.[19,20] Collectively, the findings support that immunosuppressive therapies improve clinical outcomes in patients with CAA-ri.

Our study has a several limitations: (1) As a single-center study, it may be subject to selection bias. (2) Being retrospective, some data may be incomplete due to inherent limitations of this study design. (3) The decision to administer glucocorticoid therapy was not randomized but was based on patient age, disease severity, or co-infection, potentially introducing bias into treatment outcome evaluation. (4) The lack of pathological confirmation in our study may lead to missed diagnosis. During case review, we identified a 38-year-old female whose symptoms and imaging features were highly consistent with clinical and radiological diagnostic criteria for CAA-ri. Despite the well-established lower age threshold for CAA-ri, which typically occurs in older adults, this patient was excluded from the study for failing to meet the diagnostic criterion "age ≥40 years" derived from Chung et al.[1] In such instances, brain biopsy is particularly critical.

Conclusions

CAA-ri is a rare neurological disorder in older adults. Accurate identification based on clinical manifestations and imaging characteristics is critical for prompt diagnosis of CAA-ri when brain biopsy is not feasible. For patients with probable or possible CAA-ri, early immunosuppressive treatment can alleviate neurological deficits, resolve imaging abnormalities, and improve patient outcomes.

Data Sharing Statement

The data are available from the corresponding author on reasonable request.

Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, study design, execution, acquisition of data, analysis, and interpretation, or all these areas; took part in drafting, revising, or critically reviewing the article; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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Disclosure

The authors declare that they have no competing interests.

References

1. Chwalisz BK. Cerebral amyloid angiopathy and related inflammatory disorders. *J Neurol Sci*. 2021;424:117425. doi:10.1016/j.jns.2021.117425
2. Kinnecom C, Lev MH, Wendell L, et al. Course of cerebral amyloid angiopathy-related inflammation. *Neurology*. 2007;68(17):1411–1416. doi:10.1212/01.wnl.0000260066.98681.2e
3. Scolding NJ, Joseph F, Kirby PA, et al. Abeta-related angiitis: primary angiitis of the central nervous system associated with cerebral amyloid angiopathy. *Brain*. 2005;128(Pt 3):500–515. doi:10.1093/brain/awh379
4. Auriel E, Charidimou A, Gurol ME, et al. Validation of clinicoradiological criteria for the diagnosis of cerebral amyloid angiopathy-related inflammation. *JAMA Neurol*. 2016;73(2):197–202. doi:10.1001/jamaneurol.2015.4078
5. Chung KK, Anderson NE, Hutchinson D, Synek B, Barber PA. Cerebral amyloid angiopathy related inflammation: three case reports and a review. *J Neurol Neurosurg*. 2010;82(1):20–26. doi:10.1136/jnnp.2009.204180
6. Corovic A, Kelly S, Markus HS. Cerebral amyloid angiopathy associated with inflammation: a systematic review of clinical and imaging features and outcome. *Int J Stroke*. 2018;13(3):257–267. doi:10.1177/1747493017741569
7. Wu JJ, Yao M, Ni J. Cerebral amyloid angiopathy-related inflammation: current status and future implications. *Chin Med J*. 2021;134(6):646–654. doi:10.1097/CM9.0000000000001427
8. Renard D, Tatu L, Collombier L, et al. Cerebral amyloid angiopathy and cerebral amyloid angiopathy-related inflammation: comparison of hemorrhagic and DWI MRI features. *J Alzheimers Dis*. 2018;64(4):1113–1121. doi:10.3233/JAD-180269
9. Szalardy L, Fakan B, Maszlag-Torok R, et al. Identifying diagnostic and prognostic factors in cerebral amyloid angiopathy-related inflammation: a systematic analysis of published and seven new cases. *Neuropathol Appl Neurobiol*. 2024;50(1). doi:10.1111/nan.12946
10. Theodorou A, Palaiodimou L, Safouris A, et al. Cerebral amyloid angiopathy-related inflammation: a single-center experience and a literature review. *J Clin Med*. 2022;11(22):6731. doi:10.3390/jcm11226731
11. Antolini L, DiFrancesco JC, Zedde M, et al. Spontaneous ARIA-like Events in Cerebral Amyloid Angiopathy-Related Inflammation: a Multicenter Prospective Longitudinal Cohort Study. *Neurology*. 2021;97(18):e1809–e1822. doi:10.1212/WNL.0000000000012778
12. Su Y, Dong Y, Cheng X. Recurrent intracerebral haemorrhages as main manifestations in cerebral amyloid angiopathy-related inflammation. *Stroke Vasc Neurol*. 2024;9(6):738–740. doi:10.1136/svn-2024-003100
13. van der Lee SJ, Wolters FJ, Ikram MK, et al. The effect of APOE and other common genetic variants on the onset of Alzheimer's disease and dementia: a community-based cohort study. *Lancet Neurol*. 2018;17(5):434–444. doi:10.1016/S1474-4422(18)30053-X
14. Jeong SY, Suh CH, Shim WH, Lim JS, Lee JH, Kim SJ. Incidence of amyloid-related imaging abnormalities in patients with Alzheimer disease treated with anti-beta-amyloid immunotherapy: a meta-analysis. *Neurology*. 2022;99(19):e2092–e2101. doi:10.1212/WNL.0000000000201019
15. Regenhardt RW, Thon JM, Das AS, et al. Association between immunosuppressive treatment and outcomes of cerebral amyloid angiopathy-related inflammation. *JAMA Neurol*. 2020;77(10):1261–1269. doi:10.1001/jamaneurol.2020.1782
16. Bravo GA, Cirera LS, Torrenta LR. Clinical and radiological features of cerebral amyloid angiopathy-related inflammation. *Neurol Sci*. 2021;42(12):5353–5358. doi:10.1007/s10072-021-05490-x
17. Alokley AA, Alshamrani FJ, Alabbas FM, Nazish S. When brain biopsy solves the dilemma of diagnosing atypical cerebral amyoild angiopathy: a case report. *Am J Case Rep*. 2021;22:e933869. doi:10.12659/AJCR.933869

18. Aghetti A, Sene D, Polivka M, et al. Cerebral amyloid angiopathy related inflammation with prominent meningeal involvement. a report of 2 cases. *Front Neurol*. 2019;10:984. doi:10.3389/fneur.2019.00984
19. Cancelloni V, Rufa A, Battisti C, et al. Diagnosis, treatment, and follow-up of patients with cerebral amyloid angiopathy-related inflammation. *Neurol Sci*. 2022;43(11):6381–6387. doi:10.1007/s10072-022-06299-y
20. Banerjee G, Alvares D, Bowen J, Adams ME, Werring DJ. Minimally symptomatic cerebral amyloid angiopathy-related inflammation: three descriptive case reports. *J Neurol Neurosurg Psychiatry*. 2019;90(1):113–115. doi:10.1136/jnnp-2017-317347