Small Vessel Disease

10-Year Follow-Up of a Patient with Cerebral Amyloid Angiopathy

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Keywords

Amyloid angiopathy · Intracranial hemorrhage · Brain magnetic resonance imaging

Abstract

We report the case of long-term follow-up of brain magnetic imaging of cerebral amyloid angiopathy. Cerebral amyloid angiopathy is often considered a major cause of spontaneous intracerebral hemorrhage in the elderly. This case illustrates the markedly progressive clinical and radiological features of the vasculopathic process in 10 years.

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Introduction

Cerebral amyloid angiopathy (CAA) is a common cause of spontaneous lobar intracerebral hemorrhage (ICH) in the elderly [1]. CAA is often asymptomatic but can present cognitive decline or transient focal neurological episodes [2]. The key neuroimaging signatures of the disease on magnetic resonance imaging (MRI) include multiple, strictly lobar-restricted microbleeds, cortical superficial siderosis, white matter hyperintensities, cortical microinfarcts and MRI-visible perivascular spaces in the brain [3]. The process of CAA was a somewhat progressive manner [4, 5]. Nonetheless, long-term interval change of brain imaging of patients with CAA is not known. We report the case of a patient with recurrent ICH showing the progressive course of CAA for 10 years.

Case Report

A 52-year-old woman presented with acute onset of right-side weakness. She had a history of hypertension but no other medical history, alcohol, or drug abuse. Initial brain MRI revealed left thalamic ICH (5 cm³). Fundoscopic examination revealed hypertensive retinopathy. After admission, she was started on amlodipine 5 mg and transferred to rehabilitation. The next year after initial presentation, she was admitted to a local hospital with headache and left-side weakness. Her brain MRI revealed right thalamic ICH (7 cm³) with intraventricular hemorrhage. The patient was stabilized and could walk with assistance after the second ICH event. After 6 months, she was admitted to our hospital with progressive dysphagia, urinary incontinence, memory impairment, and gait disturbance. Her brain MRI revealed hydrocephalus and several microbleeds (Fig. 1). Radionuclide cisternography showed a type IV pattern, and 50 ml of cerebrospinal fluid drainage was done with a dramatic effect. This was consistent with communicating hydrocephalus as a delayed complication after intraventricular hemorrhage, so she had ventriculoperitoneal shunt insertion. After 1 month, she was discharged with minimal dependent ambulation. The next 10 years after the second ICH event, she presented with acute onset of headache and memory loss. She did not recall a recent event and her location at admission. She was reported to have visual hallucinations in the emergency department. Her brain MRI revealed left parietooccipital ICH (6 cm³), multiple microbleeds in both cerebral hemispheres, and cortical siderosis (Fig. 2). Although the biopsy was not performed to confirm β -amyloid deposition, her diagnosis of probable CAA could be made by the Boston criteria [6]. No abnormalities were found in subsequent examinations including apolipoprotein E typing and electrophoresis. After stabilization, the patient was able to return home. At the follow-up visit, she was noted to have improved cognitive function and strength similar to after the second ICH event.

Discussion and Conclusion

CAA is a small to medium vasculopathy most commonly associated with symptomatic intracerebral hemorrhage and microbleeds [7]. CAA is characterized by microbleeds preferentially affecting cortical-subcortical lobar regions (especially the occipital and posterior temporal lobes) [8]. In this paper, we report a patient with recurrent ICH. The first and second thalamic hemorrhage of the patient may be attributed to hypertensive arteriosclerosis, considering the location of the lesion, history of hypertension and results of the fundoscopic examination. However, due to posterior dominant microbleeds, the possibility of superimposed CAA pathology should be considered. Preferential involvement of leptomeninges and the cerebral cortex is a finding that supports the possibility of CAA [3]. The progress of microbleeds after 10 years from the first presentation with a posterior to anterior pattern is shown in this patient. The third lobar ICH event, which occurred 10 years after the second presentation, was attributed to CAA rather than hypertensive arteriosclerosis. The superimposed CAA pathology had progressed to the development of lobar ICH and multiple microbleeds in this patient. In the previous study, it is known that about 30% of hypertensive ICH patients have CAA pathology and coexistence of two pathologies may increase the risk of ICH [9, 10]. Multiple ICH events of this patient can be interpreted in the same context.

There are some case reports showing progression of CAA with brain MRI [4]. However, they proved the decline in connectivity strength with a posterior-frontal connection, the

increase of microbleeds or white matter hyperintensities by less than 5-year follow-up of MRI. Although CAA might be diagnosed with gradient echo sequences of MRI, there was no report of gradient echo sequences of brain imaging of 10-year follow-up [5]. This case captures the evolving and dynamic nature of CAA and the markedly progressive clinical and radiological features of this vasculopathic process.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

M.K.K.: Taking responsibility for the data management and construction of the body of the manuscript. B.-W.Y.: Study conceptualization, organization, execution; review and critique of the analysis and revised the manuscript for intellectual content.

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Fig. 1. A total of 6 lobar microbleeds on baseline gradient echo sequences of magnetic resonance imaging.

Fig. 2. Growth of microbleeds, left temporooccipital intracerebral hemorrhage, and superficial siderosis on gradient echo sequences of magnetic resonance imaging on follow-up 10 years later.