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Cerebral amyloid angiopathy-related inflammation with epilepsy mimicking a presentation of brain tumor: A case report and review of the literature

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ABSTRACT

INTRODUCTION: Cerebral amyloid angiopathy-related inflammation (CAA-ri), a rare and treatable variant of cerebral amyloid angiopathy, lacks specific imaging and clinical features, and requires invasive brain biopsy to confirm the diagnosis. We report the case of a patient with nonconvulsive status epilepticus (NCSE) caused by CAA-ri in the right occipital lobe.
PRESENTATION OF CASE: A 78-year-old man with a history of hypertension and rheumatoid arthritis was admitted to our hospital following an episode of seizures. CT scan showed a low-attenuating subcortical lesion in the right occipital lobe. MRI revealed the lesion as hypointense on T1-weighted imaging (WI) and hyperintense on T2-WI, showing no enhancement on T1-WI contrast-enhanced with gadolinium. In addition, T2*-weighted gradient-recalled echo (T2*-GRE) and susceptibility-weighted imaging (SWI) revealed extensive cortical microbleeds. Biopsy to determine the exact diagnosis revealed histological findings of reactive changes and perivascular inflammatory infiltration associated with amyloid deposition in vessel walls. These findings were consistent with CAA-ri. Corticosteroid therapy with dexamethasone was initiated for a short period as a diagnostic and therapeutic maneuver, resulting in marked reductions in the lesion.
DISCUSSION: CAA is generally associated with intracerebral hemorrhage, dementia, and small cerebral infarctions in the elderly population, but in a small proportion of cases is related to inflammatory responses to vascular deposits of Aβ, as so-called CAA-ri.
CONCLUSION: CAA-ri should be considered among the differential diagnoses for causes of unprovoked seizure onset in elderly individuals, when associated with petechial hemorrhages on T2*-GRE and SWI sequences on MRI.

1. Introduction

Cerebral amyloid angiopathy (CAA), a common small vessel disease of the brain, is characterized by progressive deposition of β-amyloid (Aβ) protein in the walls of small to medium-sized arteries and capillaries in the cerebral cortex and overlying leptomeninges [1–4]. The typical presentation of CAA is spontaneous lobar intracerebral hemorrhage in an elderly patient [2,4]. However, CAA can also manifest as subacute cognitive impairment, dementia, transient neurological symptoms, or epilepsy [1]. This could be a manifestation of an uncommon subtype of CAA, distinguished as CAA-related inflammation (CAA-ri, also known as Aβ-related angiitis) [3,5–7]. CAA-ri represents the coexistence of CAA and vascular inflammation, and is thought to result from an inflammatory response to Aβ protein in the blood vessel walls. Cognitive and behavioral changes are the most common symptoms of CAA-ri, followed by focal neurological signs, headache, and seizures [3,5]. In addition to this, CAA-ri can also mimic brain tumor from clinical and radiological point of view [3,4,8,9]. In this paper, we report an elder case of nonconvulsive status epilepticus (NCSE) due to CAA-ri in the right occipital lobe. It appears like an intraparenchymal tumor at first, and can be finally diagnosed only through biopsy. Therefore, we also demonstrat the pathological features of this unique and cryptic entity.

Abbreviations: CAA, cerebral amyloid angiopathy; Aβ, β-amyloid; CAA-ri, cerebral amyloid angiopathy-related inflammation; NCSE, nonconvulsive status epilepticus; EEG, electroencephalogram; CT, computed tomography; MRI, magnetic resonance imaging; WI, weighted image; FLAIR, fluid-attenuated inversion recovery; Gd, gadolinium; T2*-GRE, T2*-weighted gradient-recalled echo; SWI, susceptibility-weighted imaging; PET, positron emission tomography; H&E, hematoxylin and eosin; DFS, direct fast scarlet; GFAP, glial fibrillary acidic protein.

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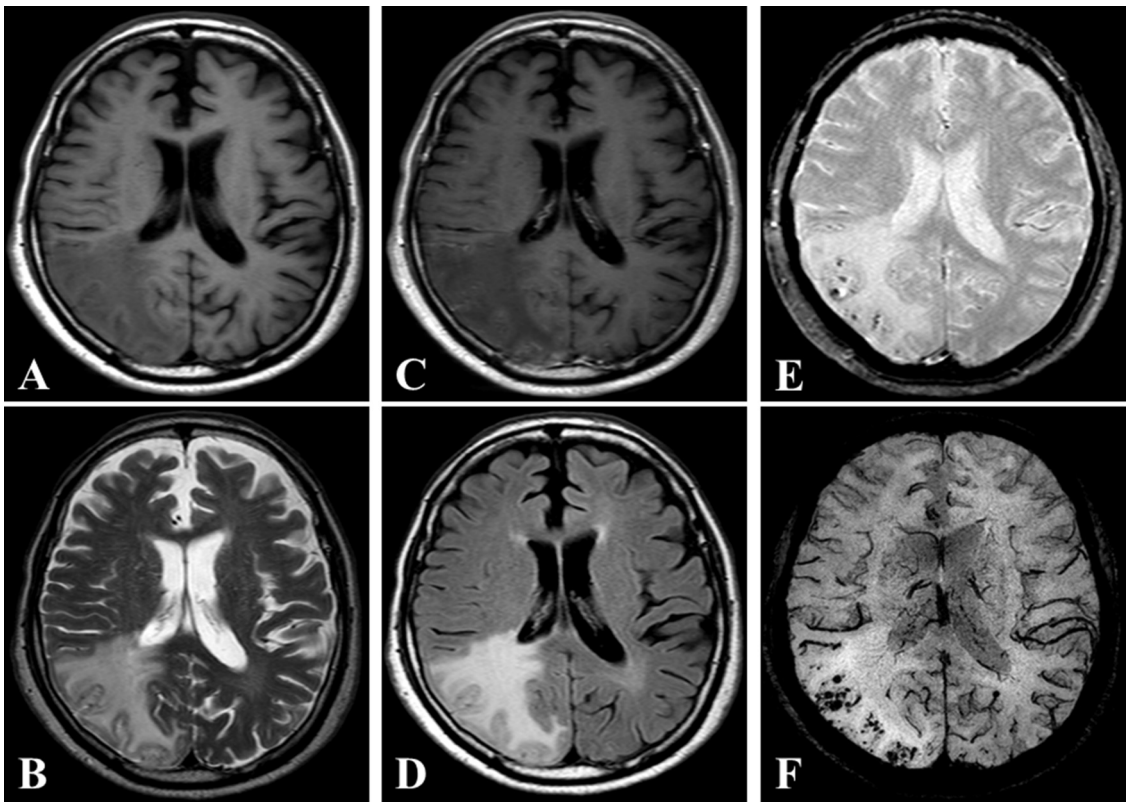

Fig. 1. Images from preoperative axial T1-weighted imaging **(A)**, T2-weighted imaging **(B)**, gadolinium (Gd)-enhanced T1-weighted imaging **(C)**, fluid-attenuated inversion recovery (FLAIR) **(D)**, T2*-weighted gradient-recalled echo (T2*-GRE) **(E),** and susceptibility-weighted imaging (SWI) **(F)** show an area of abnormal intensity in the right occipital lobe. The lesion demonstrates no enhancement with Gd. T2*-GRE and SWI. images reveal multiple spotty cortical and subcortical hypointensities in the same region.

Thiswork has been reported in line with the SCARE criteria [10].

2. Case description

A 78-year-old man with a history of hypertension and rheumatoid arthritis was admitted to our hospital with suspected unprovoked seizure. His family had first noted new onset of irritability and confusion as uncharacteristic behaviors. Neurological examination on admission showed mild left-sided hemiparesis and a visual field defect. Laboratory examinations, including cerebrospinal fluid studies, revealed no abnormal findings, and the concentrations of tumor markers remained with normal limits. Routine scalp electroencephalogram (EEG) showed frequent epileptogenic discharges in the right occipital region. Computed tomography (CT) scan of the head showed a low-attenuating subcortical lesion in the right occipital lobe. Magnetic resonance imaging (MRI) demonstrated the lesion as hypointense on T1-weighted imaging (WI) and hyperintense on T2-WI and fluid-attenuated inversion recovery (FLAIR) imaging, without enhancement on T1-WI using gadolinium (Gd) contrast. In addition, T2*-weighted gradient-recalled echo (T2*-GRE) and susceptibility-weighted imaging (SWI) demonstrated extensive cortical microbleeds (Fig. 1). Cerebral angiography did not show any vascular abnormalities. Positron emission tomography (PET) scan of the brain revealed slightly increased uptake of methionine in the right occipital region, consistent with the lesion shown on MRI (maximum standardized uptake value for methionine, 2.4). On initial consideration, the history and results of laboratory examinations and radiological studies seemed most consistent with multifocal glioma, rather than any infectious, an inflammatory disease, or acute stroke. Therefore, the patient was first loaded with levetiracetam for epilepsy control. The dose of levetiracetam was increased after confirming a recurrent episode of intermittent confusion and aphasia as a manifestation of NCSE. In order to obtain the exact histological diagnosis and plan effective treatment for the primary disease, we performed surgical biopsy of the right occipital lesion with the assistance of image-guided navigation. Postoperative histopathology obtained from hematoxylin and eosin (H&E) staining demonstrated reactive changes including vacuolization, which suggests edema and gliosis, and thickened blood vessels (Fig. 2A–C). Most subcortical glial cells were small and pyknotic, indicating ischemic changes. Some subcortical vessels showed evidence of thrombosis and hypertrophied vessel walls. Fresh perivascular hemorrhage and several areas of hemosiderin deposition, and infiltration of small inflammatory cells were also present (Fig. 2D). Lymphocytes and epithelioid macrophages had infiltrated not only around the blood vessels, but also into the vessel walls (Fig. 3A). Immunohistochemical studies were performed using antibodies for CD3, CD20, CD68, Congo red and direct fast scarlet (DFS). Perivascular inflammatory cells mostly comprised monocyte/microglial cells (positive staining for CD68) (Fig. 3B**)**. T lymphocytes (positive staining for CD3) were also present **(**Fig. 3C), whereas a neoplastic increase in B lymphocytes was absent (negative staining for CD20) (Fig. 3D). In addition, Congo red and DFS staining after permanganic acid treatment showed Aβ deposition in the vascular walls (Fig. 4). The H&E staining and immunohistochemical studies revealed morphological characteristics consistent with CAA-ri. Corticosteroid therapy with dexamethasone was started as a diagnostic and therapeutic maneuver. Two weeks after starting this treatment, MRI showed marked size reduction of hyperintense lesions on T2-WI (Fig. 5) and we stopped the administration of dexamethasone. The postoperative course was uneventful, with no relapse of epileptic symptoms. With entirely resolving of speech difficulties and confusion, he