A Case of Cerebral Amyloid Angiitis Presenting as Posterior Reversible Encephalopathy Syndrome

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Case Presentation

- 73-year-old male presenting with right homonymous hemianopsia, gait instability, and blood pressure 176/95.
- Non-contrast CT head showed hypodense edema in the posterior lobes. CTA head and neck ruled out acute vascular blockage.
- MRI on admission showed stable posterior T2-weighted signal intensity of bilateral parietal and occipital lobes with sulcal effacement and mass effect (Figure 1).
- MRI with contrast was recommended to evaluate for malignancy. MRI with contrast revealed numerous foci of signal abnormality of bilateral parietal and occipital lobes with sulcal effacement and mass effect (Figure 1).
- MRI with contrast showed enlarged sulci and subcortical white matter—consistent with vasogenic edema (Figure 2).
- MRI with contrast revealed numerous foci of signal abnormality suggestive of chronic hemorrhage, and unchanged T2 hyperintensity in the posterior hemispheres (Figure 2). The differential diagnosis now included chronic microangiopathy in addition to PRES.
- Admission was recommended but the patient opted for outpatient management.
- Unfortunately, four weeks later he experienced acute mental status change and seizure. BP on presentation 141/65.
- MRI with contrast showed stable posterior T2-weighted hyperintensities and countless new foci of susceptibility artifact, concerning for interim hemorraghes (Figure 3).
- Subsequent brain biopsy revealed numerous small hemorrhages and thickened vessel walls with perivascular lymphocytes (Figure 4a). Congo red stain was positive for perivascular amyloid (Figure 4b). Electron micrograph showed tangled, unbranched filaments measuring between 7.5-11.1 nm, characteristic of amyloid filaments. He received a course of Solu-Medrol followed by a prednisone taper and later started mycophenolate.

Discussion

CAA is a small vessel vasculitis that occurs in a subset of patients with cerebral amyloid angiopathy. The typical presentation is rapidly progressive cognitive decline with possible seizures, headaches, focal neurologic signs, and cortical microhemorrhages. PRES is a clinicoradiologic syndrome related to multiple etiologies. Clinical features include visual disturbance, seizures, altered mental status, and hypertension. The pathophysiology of PRES is unclear and likely heterogeneous depending on the etiology.

This case suggests that CAA and PRES can co-occur and may even share an etiologic link. Our patient’s initial presentation was consistent with PRES (visual disturbance, hypertension), but he progressively demonstrated features of CAA (cognitive decline, seizure, microhemorrhages, biopsy findings). The imaging findings that led to his initial diagnosis of PRES and persisted even as his symptoms resolved—confluent areas of increased T2-weighted signal of the bilateral posterior lobes representing edema of the subcortical white matter—are in fact characteristic of either condition. Regarding the mechanism of these findings, failure of vascular autoregulation and endothelial dysfunction in CAA may produce the clinical and radiologic features of PRES.

Here we highlight a potential cause of PRES that requires brain biopsy for definitive diagnosis. The co-occurrence of these two conditions is rare but it has been previously reported, and we identified only one report of biopsy-proven CAA in the setting of PRES. The incidence of this phenomenon is not known but the present report is not an isolated case. Physicians should strongly consider the possible value of a brain biopsy in patients with PRES, especially if refractory to standard treatment.

References