Cerebral amyloid angiopathy (CAA) results from amyloid-β protein deposition in the walls of intracerebral small arteries. It is a disease of old age, apparent on autopsy in 34% of patients over age 85 years. Although frequently asymptomatic, symptomatic CAA most commonly presents as spontaneous lobar hemorrhage. A significantly less common presentation is as a transient neurologic event (TNE). We present a case of TNE heralding a diagnosis of CAA in a nonagenarian.

Case Report

A 99-year-old woman with history of hypertension and mild cognitive impairment presented January 2017 to the emergency department (ED) after being found shaking with subsequent confusion at her assisted living facility. The shaking persisted for minutes according to witnesses.

In November 2015, she suffered a fall with concomitant right frontal intraparenchymal hematoma. Conservative treatment was pursued. Ten months later, she presented with a seizure-like episode similar to the current presentation. She was initiated on oxcarbazepine and had remained event free.

In the ED, she appeared confused and hyperactive. Vital signs included blood pressure 200/80 mm/Hg, which spontaneously reduced to 131/53 mm/Hg on repeat. Other vital signs were within normal limits. Besides chronic hearing loss, head and neck examinations were normal. Cardiovascular, pulmonary, and gastrointestinal examinations were benign. Neurologic examination revealed no strength or sensory deficits, but confusion complicated examination. Language testing suggested transient expressive aphasia, which resolved within minutes.

Complete blood count, comprehensive metabolic panel, and urinalysis were noncontributory as were chest x-ray and electrocardiography. Head computed tomography (CT) identified right frontal encephalomalacia consistent with prior hematoma. Electroencephalography indicated diffuse slowing, more focal in the right frontal region. She was admitted to the hospital pending brain magnetic resonance imaging (MRI) and returned to baseline after 18 hours. MRI findings confirmed her diagnosis as indicated in the Figure.

Discussion

TNE describes neurologic symptoms lasting less than 24 hours. The differential diagnosis includes transient ischemic attacks (TIA), seizure, migraine, and syncope, with TIA being most common. While the patient's age and history of hypertension would support an ischemic cause, seizure-like episodes after a history of fall and intraparenchymal hematoma could suggest post-traumatic epilepsy.

CAA is not commonly considered in the differential diagnosis for TNE. In this case, the history of intraparenchymal hematoma could suggest prior spontaneous lobar hemorrhage. Previous brain imaging was limited to CT, which failed to provide etiology. The presence of frontal intraparenchymal hemosiderin staining along with multifocal cortical-subcortical hemosiderosis on brain MRI is highly suggestive of CAA.

CAA was traditionally diagnosed postmortem. Contemporary diagnostic criteria, the modified Boston criteria, combine imaging and clinical characteristics with pathology when available. Application of the criteria for probable CAA to patients with symptomatic intracerebral hemorrhage has 95% sensitivity and 81% specificity. Sophisticated MRI techniques substantially increase the diagnostic certainty.

No disease-modifying therapies exist for CAA. Management hinges upon the presenting clinical syndrome. Intracerebral hemorrhage may require surgery. Rebleeding is common, suggesting a need to work up bleeding diatheses. Importantly, as in other causes of intracerebral hemorrhage, blood pressure control is critical to minimize risk of rebleeding. TNE requires alternative consideration. Since symptoms mimic TIA, patients often receive antithrombotic and statin therapy. Use of antithrombotics may prove catastrophic, however, as nearly 40% of patients progress to lobar hemorrhage within 2 months. Additionally, a link has been demonstrated between low total and low-density lipoprotein cholesterol and increased risk of intracerebral hemorrhage. Although this mechanism is unclear, statin therapy may be approached with caution. These decisions could be significantly altered by distinguishing TIA from...
Microhemorrhages a

Susceptibility Signal Loss at the Cortico-Subcortical Junction in Keeping With CAA is an important cause of cerebral bleeding and cortical dysfunction in older adults. Recognition of its varied presentations, including as TNE, is critical for making informed diagnostic and treatment decisions.

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REFERENCES