A Case of Cerebral Amyloid Angiopathy-Related Inflammation (CAA-RI) Presenting as a Generalized Tonic Clonic Seizure

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Received January 07, 2021; Revised February 08, 2021; Accepted February 22, 2021

Abstract  Cerebral Amyloid Angiopathy (CAA) is characterized by amyloid beta-peptide deposits within the small to medium-sized vessels of the brain and leptomeninges. CAA is an important cause of intracerebral hemorrhage in older adults. Cerebral Amyloid Angiopathy Related Inflammation (CAA-ri) is, however, a rare variant of CAA that results from an autoimmune response to the deposits and is characterized by acute or subacute encephalopathy, headache, or focal neurological deficits. We present a case of a 62-year-old female who presented with a generalized tonic-clonic seizure witnessed by a family member. The event was preceded by a worsening of her dementia in the past few months. The patient had features suggestive of CAA-ri on Magnetic Resonance Imaging of the brain and was treated with high dose IV steroids. With an improvement in her cognitive symptoms and no further seizure episodes, she was discharged on oral steroids.

Although CAA is well studied and well documented, its subset CAA-ri is uncommon with its clinical course and complications mentioned infrequently in medical literature. In conclusion, CAA-ri is underdiagnosed because of its rarity and remains a potentially treatable cause of subacute cognitive decline and seizures demanding further research in this area.

Keywords: format, microsoft word template, style, insert, template


1. Introduction

CAA is represented by the by the deposition of amyloid-beta peptides within the small and medium-sized vasculature of the brain and leptomeninges [1,2]. CAA-ri is characterized by inflammation that is postulated to be due to the body’s, inflammatory response to the aforementioned amyloid deposits [2,3,4,5]. The incidence of the disease is equal in both men and women with onset of symptoms usually in the 7th Decade of life. The most widely reported symptoms are cognitive and behavioral changes, other less common symptoms include focal neurological signs, headaches, and seizures [4]. We present a case of a patient with CAA-ri presenting with a generalized tonic-clonic seizure with a background of subacute cognitive decline and worsening dementia in the preceding months.

2. Case

We present a 62-year-old female with a past medical history significant for anxiety, depression, fibromyalgia, hypothyroidism, lupus, and dementia who presented with an event concerning for a seizure-like episode. The patient's last known normal was the night before admission. On the day of the presentation around 7:30 in the morning the patient had a generalized tonic-clonic seizure. Witnessed by her grandson this episode lasted only a few minutes before spontaneously resolving. There was tongue biting associated with the event but no report of bowel or bladder incontinence. The patient was confused after the event and EMS was contacted and subsequently, the patient was brought to the hospital. The patient did not have any further seizures in her return to her neurological baseline during the rest of her hospital...
3. Discussion

Cerebral Amyloid Angiopathy (CAA) is a fairly common finding associated with the deposition of Amyloid Beta along the walls of small to medium-sized arteries, arterioles, and capillaries in the cerebral cortex, as well as leptomeningeal arteries [2]. This can occur most commonly in patients with Alzheimer’s Dementia as well as a sporadic non-pathological finding in older, otherwise healthy individuals. It is a condition both well studied and documented. There is a subset of CAA that is associated with pathologic vascular inflammation and subsequent manifestations in the form of seizures, headaches, and subacute cognitive decline [6]. While there have been some isolated case reports and studies regarding this particular subset of CAA, this is still an area that requires a considerable amount of research. While it is underdiagnosed, because of its rarity, it remains a potentially treatable entity. This subset is further typed into CAA related inflammation (CAA-RI). This is a condition characterized by perivascular nondestructive inflammatory infiltration [4]. Yet another type is a vasculitic transmural, sometimes granulomatous, inflammatory infiltrate, namely Aβ-related angiitis (ABRA) [7]. Though rare, these two conditions can sometimes coexist within the same patient as evidenced in our case. CAA-RI presents predominantly with Subacute cognitive decline and seizures [8]. ABRA has shown to present more with new-onset headaches, neuropsychiatric manifestations, focal neurological deficits, and epileptic seizures [6]. The median age for diagnosis for both CAA-RI, ABRA is > 50 years. In terms of imaging findings, CAA-RI shows MRI findings consistent with shifting multifocal white matter T2 hyperintensities abnormalities colocalized with petechial hemorrhages on SWI [9]. In ABRA, T2-weighted (T2W) or fluid-attenuation inversion recovery (FLAIR) images show hyperintensities with minimal gadolinium enhancement. Most patients also have the presence of scattered cortical and subcortical microbleeds at GRE images [10,11]. There are considerable differences between the two conditions, however. ABRA presents a younger age than CAA-RI. It also has a lower incidence of strokes or focal neurological deficits, a decreased risk of intracerebral hemorrhage, and a low frequency of altered cognition [6]. Both conditions have shown good response to corticosteroids and immunosuppressive therapy [12]. Brain Biopsy has long been considered the gold standard for diagnosis of these conditions however recent literature suggests that diagnosis can be made based on the clinical picture, MRI findings, and response to corticosteroids [13].

References


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