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Unmasking the Inflammatory Face of Cerebral Amyloid Angiopathy: MRI Clues from a Rare Case

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Abstract: ***Background**: Cerebral amyloid angiopathy-related inflammation (CAA-RI) is a rare but treatable subtype of cerebral amyloid angiopathy (CAA), characterized by a perivascular inflammatory response to amyloid-β deposition in cerebral vessels. Accurate diagnosis is crucial, as it responds well to immunosuppressive therapy. **Case Presentation**: We report a case of a 50-year-old female presenting with headache, fever and GTCS type seizure. MRI revealed areas of cortical and subcortical hyperintensities of parieto-temporal -occipital lobes and cortical and subcortical micro bleeds involving bilateral cerebral hemispheres with leptomeningeal enhancement on post contrast study. No evidence of systemic infection or neoplastic process was found. Based on clinical and radiologic criteria, a diagnosis of probable CAA-RI was made. **Conclusion**: CAA-RI should be considered in elderly patients with subacute neurological decline and distinctive MRI features. Early recognition can prevent invasive diagnostics and guide appropriate immunotherapy.*

Keywords: CAA-RI, cerebral amyloid angiopathy, MRI, microbleeds, vasculitis, neuroinflammation

1. Introduction

Cerebral amyloid angiopathy-related inflammation (CAA-RI) represents an immune-mediated response to β-amyloid deposits in small-to-medium-sized cortical and leptomeningeal vessels. Though rare, its identification is vital due to the potential for significant recovery with corticosteroid therapy. MRI is central to the diagnosis, often revealing asymmetric subcortical white matter changes and micro-haemorrhages. We present a case demonstrating classic imaging features of CAA-RI and discuss its close differentials.

2. Case Presentation

We report a case of a 50-year-old female with a GTCS Type of seizures Headache and fever. No visual complaints and no other known comorbidities.

Investigation:

Laboratory results:
Normal inflammatory markers, negative autoimmune and infectious workup, and no neoplastic markers in blood.
Initial non-contrast CT of the head revealed few cortical hyper densities noted in bilateral occipital and parietal regions. To further evaluate the etiology, a brain MRI was performed.

MRI Findings:

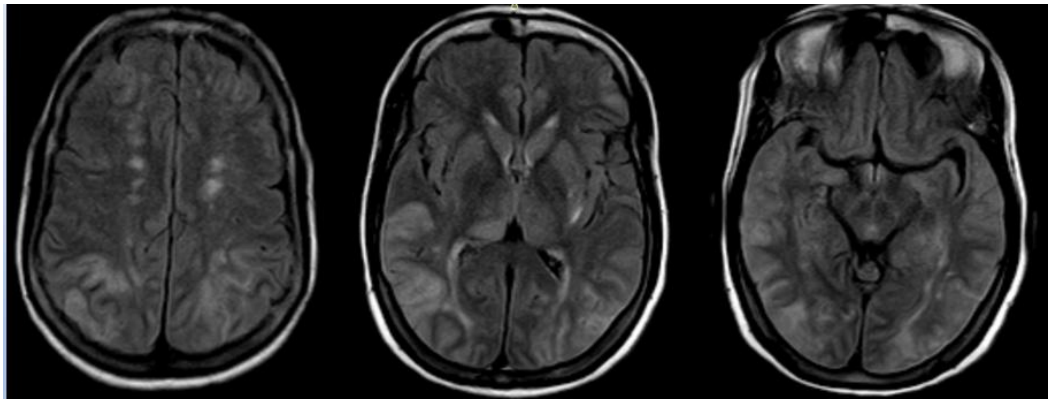

Figure 1: Areas of flair cortical and subcortical hyperintensities noted involving bilateral parieto-temporal –occipital lobe

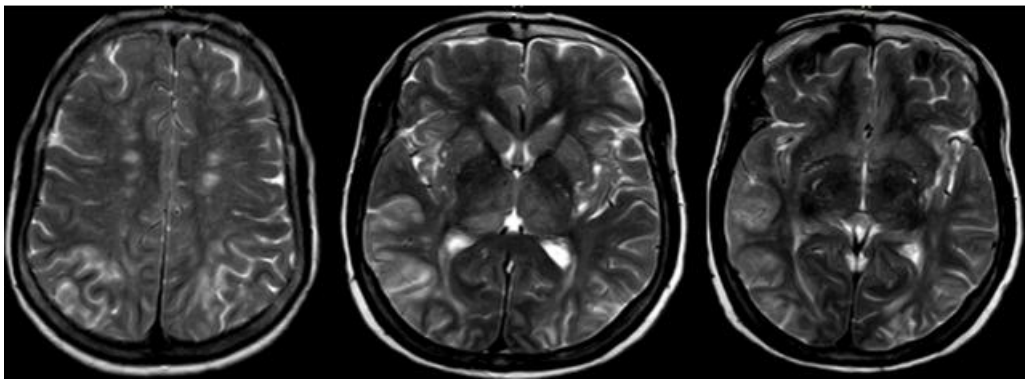

Figure 2: Areas of T2 cortical and subcortical hyperintensities noted involving bilateral parieto-temporal –occipital lobe

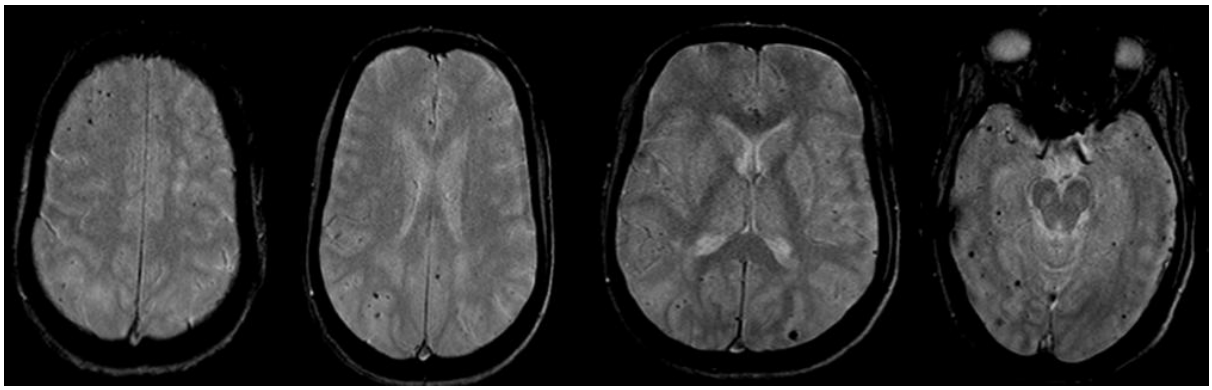

Figure 3: Multiple discrete variable sized foci of blooming on GRE in cortical and sub cortical regions of bilateral cerebral hemispheres. However basal ganglia and brain stem are spared

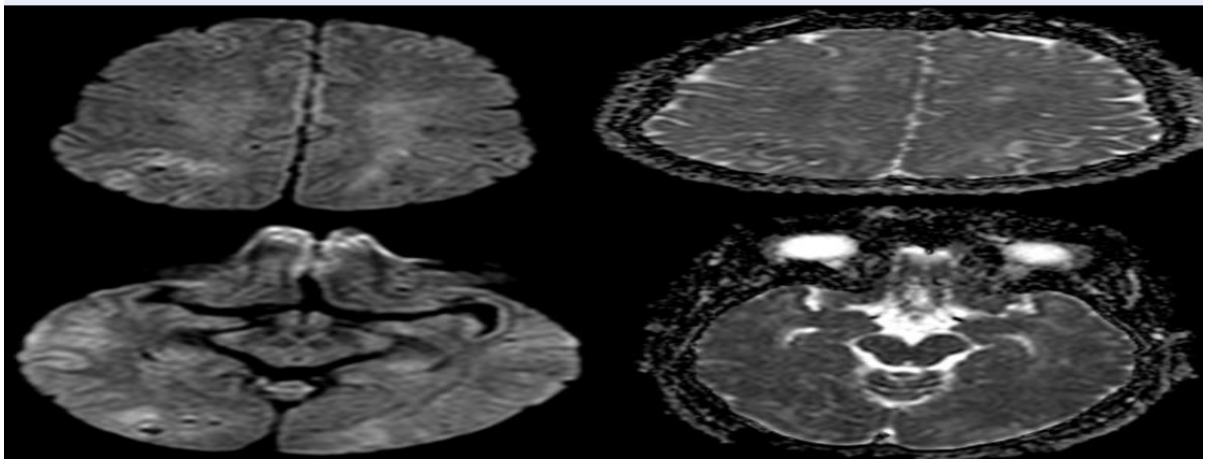

Figure 4: Diffusion-weighted imaging (DWI): No acute ischemic changes

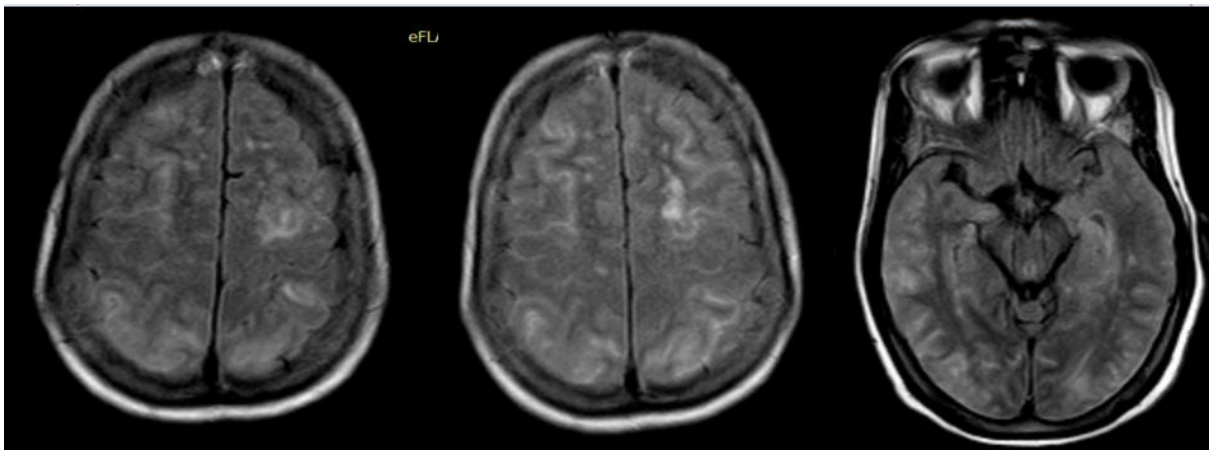

Figure 5: Subtle flair hyperintensity noted in sulcal spaces of bilateral cerebral hemispheres

Differential diagnosis:
Posterior reversible encephalopathy syndrome, primary CNS vasculitis, infectious encephalitis.

Management and Follow-Up:
The patient was started on High-Dose Corticosteroids Methylprednisolone: 1 g IV daily for 3–5 days and significant improvement was seen within 4-5 day and was later put on maintenance therapy for 4–6 weeks Prednisone: 1 mg/kg/day orally

3. Discussion

Cerebral amyloid angiopathy is a cerebrovascular disorder caused by the accumulation of cerebral amyloid-β (Aβ) in the tunica media and adventitia of leptomeningeal and cortical vessels of the brain.

Present with headache, focal neurological symptoms, seizures, and decreased conscious state. While definitive diagnosis requires histological confirmation, neuroimaging—especially magnetic resonance imaging (MRI)—plays a critical role in identifying characteristic features that support a diagnosis of probable CAA using validated criteria, such as the modified Boston criteria.

Cerebral Amyloid Angiopathy-Related Inflammation (CAA-RI) typically presents with **asymmetric**, patchy, or confluent white matter hyperintensities on FLAIR imaging, most prominently in the **parietal and occipital lobes**. A hallmark feature of CAA-ri on MRI is the presence of **multiple cortical microbleeds**, best visualized on **susceptibility-weighted imaging (SWI)** or *T2-GRE** sequences. These microbleeds are classically located in the **cortical and subcortical areas**, particularly posteriorly, and are not seen in other mimics like PRES. There may also be **minimal, no enhancement or leptomemingeal enhancement** with gadolinium, and **restricted diffusion is usually absent**, helping to exclude acute infarction. Mass effect may be mild despite extensive edema.

In contrast, **PRES** is characterized by **symmetrical**, often **bilateral** hyperintensities on FLAIR, predominantly affecting the **parieto-occipital lobes**, but it may also involve the frontal lobes, cerebellum, and brainstem. The edema in PRES is **vasogenic**, showing **high ADC values** with no diffusion restriction, and usually does **not enhance** after contrast. Importantly, **microbleeds are uncommon** in classic PRES, although hemorrhagic variants exist. PRES typically lacks the chronic cortical microbleeds seen in CAA-ri. Additionally, PRES lesions are more likely to **resolve completely** on follow-up imaging once the underlying cause (such as hypertension or renal dysfunction) is corrected.

Primary CNS Vasculitis, on the other hand, can show a **more variable** and **multifocal pattern** of T2/FLAIR hyperintensities that may affect both cortical and **deep white matter regions.** These lesions often have a **scattered distribution** and may involve the **basal ganglia, thalami, and corpus callosum**. Contrast enhancement is more common in CNS vasculitis than in CAA-ri or PRES, and can appear as **nodular, patchy, or leptomeningeal enhancement**. Unlike CAA-ri, **microbleeds are uncommon**, unless the vasculitis has led to secondary haemorrhage. Vessel wall imaging or angiography may show segmental narrowing or beading of vessels, which supports a vasculitis process.

4. Conclusion

CAA-RI should be included in the differential diagnosis for elderly patients with subacute neurological symptoms and asymmetric white matter lesions on MRI. Recognition of characteristic imaging features can guide early treatment with corticosteroids, potentially preventing unnecessary biopsies and improving outcomes.

Declarations

Patient Consent:
Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Conflicts of Interest:
The authors declare no conflicts of interest.

Funding:
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Reference

[1] Auriel E, Charidimou A, Gurol ME, et al. Validation of clinic-radiological criteria for the diagnosis of cerebral amyloid angiopathy-related inflammation. *JAMA Neurol*. 2016;73(2):197-202.
[2] Chung KK, Anderson NE, Hutchinson D, Synek B, Barber PA. Cerebral amyloid angiopathy-related inflammation: three case reports and a review. *J Neurol Neurosurg Psychiatry*. 2011;82(1):20-26.