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Case Report

Cerebral amyloid angiopathy revealed by severe renal failure: A case report ☆

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ABSTRACT

Sporadic cerebral amyloid angiopathy is a common condition in the elderly, characterised by the accumulation of amyloid Aβ peptide in the walls of small cerebral arteries, leading to intracranial haemorrhage and cognitive impairment. We present the case of a 65-year-old woman admitted for sudden intracranial hypertension and severe renal failure requiring dialysis.

This case illustrates an uncommon presentation of cerebral amyloid angiopathy in a patient with concurrent end-stage renal disease, highlighting the complex interplay between systemic and cerebrovascular pathology. The diagnostic challenge posed by overlapping neurological and uraemic symptoms underscores the importance of multidisciplinary evaluation in such cases.

Introduction

Sporadic cerebral amyloid angiopathy (CAA) is a common condition in elderly individuals, characterized by the deposition of amyloid β-peptide in the walls of small cerebral arteries, leading to intracranial hemorrhage and cognitive decline. We report a unique case of a 65-year-old woman who presented with acute intracranial hypertension and severe renal failure requiring dialysis. This case is noteworthy due to the uncommon coexistence of acute renal failure and CAA-related intracranial hypertension, suggesting a potential interplay between systemic and cerebrovascular pathologies. The diagnostic process was further complicated by the overlapping neurological and clinical features of uraemia observed upon admission. This case underscores the importance of a

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multidisciplinary approach in resolving diagnostic complexities and optimizing patient management.

Case description

We describe the case of a 65-year-old woman with a 20-year history of diabetes mellitus managed with gliclazide and a 10-year history of hypertension treated with amlodipine. She had no history of anticoagulant or antiplatelet therapy. The patient presented with acute headache, vomiting, blurred vision, and altered consciousness. On examination, she was hypertensive (180/90 mmHg), tachycardic (110 bpm), and conscious, with stable respiratory status. Diuresis was preserved, but significant proteinuria was noted. Neurological examination revealed left hemiplegia.

Initial noncontrast CT imaging, performed emergently for suspected intracranial hypertension, showed no abnormalities. Laboratory evaluation revealed severe acute kidney injury with elevated urea (2.8 g/L; normal range 0.15–0.45 g/L) and creatinine (107 mg/L; normal range 6–12 mg/L). Urinalysis, C-reactive protein (CRP), procalcitonin, and cerebrospinal fluid analysis were unremarkable. Renal ultrasound revealed bilateral renal atrophy, and anemia was confirmed with a hemoglobin level of 8.5 g/dL (normal range 12–16 g/dL). These findings were consistent with end-stage renal disease, likely secondary to diabetic nephropathy.

Despite partial improvement in renal function after 3 dialysis sessions, the patient's neurological status remained unchanged. A contrast-enhanced CT scan subsequently revealed a right parietal hematoma and multiple cerebral microbleeds, consistent with cerebral amyloid angiopathy (CAA) (Fig. 1).

With a view to etiology of the intraparenchymal hematoma, magnetic resonance imaging (MRI) with cerebral angio-MRI showed a right parietal parenchymal hematoma (Fig. 2), with evidence of microbleeds on SWAN sequences (Fig. 3).

Neurosurgical advice recommended clinical monitoring and blood pressure control without surgery. A biopsy of the accessory salivary glands suggested kappa light-chain amyloidosis (Fig. 4). Serum protein electrophoresis showed hypogammaglobulinemia with increased α-globulins, and bone marrow examination was normal.

The patient was treated with a 3-day course of high-dose Methylprednisolone, followed by oral corticosteroids, nephroprotective therapy, preventive anticoagulation, and adjuvant treatments. She underwent motor rehabilitation with good results but still has memory and behavioral issues. Renal function was managed with intermittent hemodialysis through a right jugular tunneled catheter.

Discussion

Sporadic cerebral amyloid angiopathy (CAA) is a common, age-related microangiopathy characterized by the deposition of amyloid β-peptide in the walls of leptomeningeal and cortical vessels. These amyloid deposits weaken the vascular walls, predisposing them to rupture and resulting in lobar intracranial hemorrhages (ICH) that primarily affect the cerebral cortex and juxtacortical white matter. CAA is a significant contributor to dependency, dementia, and mortality in the elderly population.

CAA accounts for 5% to 20% of intracerebral hemorrhages in individuals over the age of 70. Its prevalence increases with age, rising from 2.3% in patients aged 65–74 years to nearly 100% in those over 80 years. Notably, the disease exhibits no sex-related predisposition [1]. While arterial hypertension is not a direct risk factor for the development of CAA, it is strongly associated with an increased risk of CAA-related intracranial hemorrhage and may contribute to cases of mixed cerebral microangiopathies [2].

In the presented case, the patient's history of long-standing, poorly controlled hypertension was notable. However, the lobar distribution of her intracranial hemorrhage strongly supports the diagnosis of CAA. Other recognized risk factors for CAA include the apolipoprotein E (ApoE) ε2 and ε4 alleles, as well as the use of anticoagulants and antiplatelet

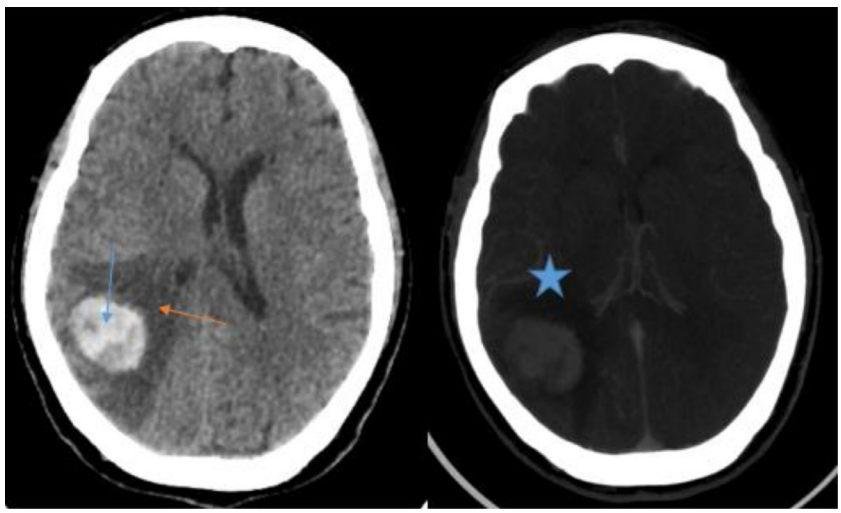

Fig. 1 – Cerabral scan with contrast injection showing a right parietal parenchymal hematoma (blue arrow) with perilesional vasogenic edema (red arrow) all responsible for a mass effect on nearby vascular structures (star).

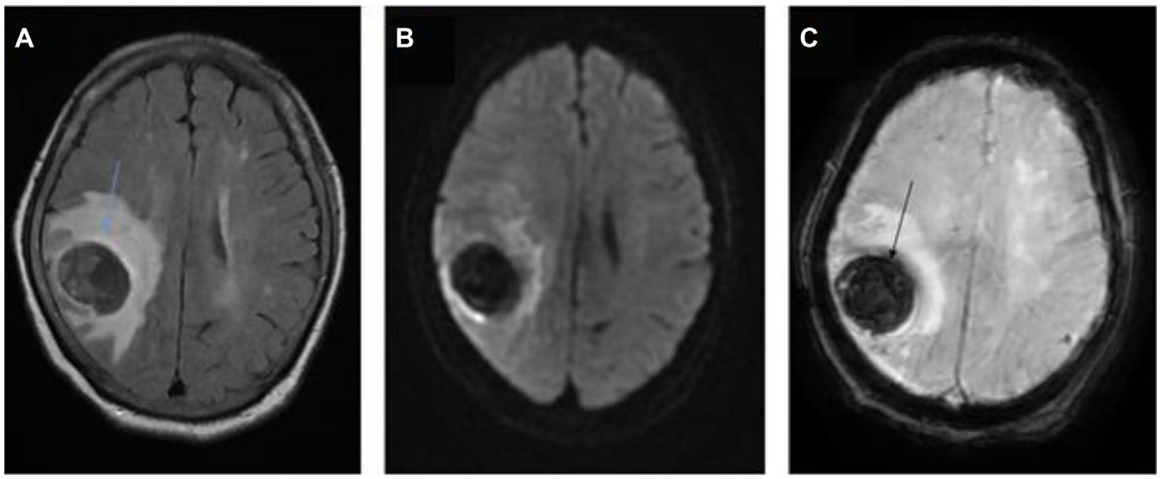

Fig. 2 – Cerebral MRI showing an intraparenchymal right parietal cerebral hematoma (black arrow) with perilesional vasogenic edema (blue arrow).

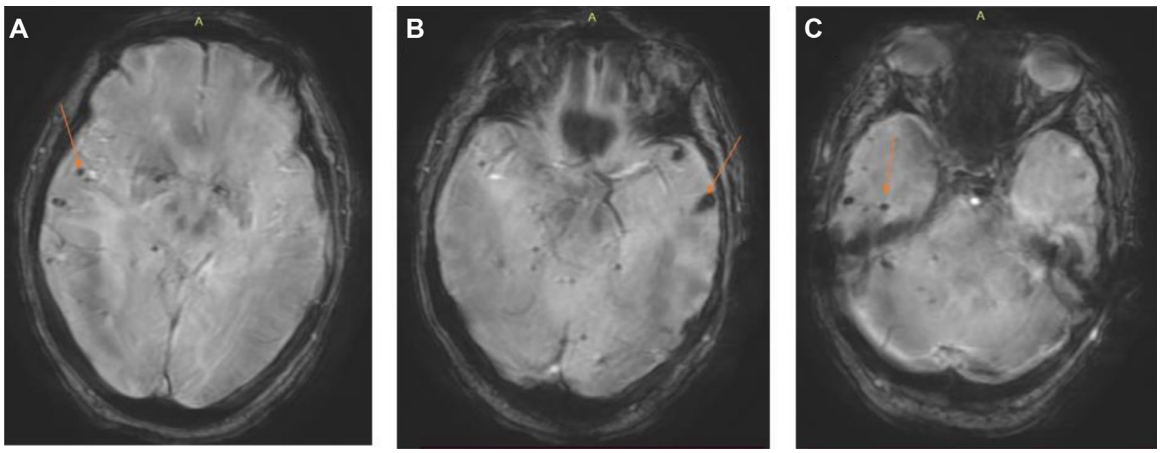

Fig. 3 – Cerebral MRI showing scattered signal-free spots (red arrows) associated with amyloid angiopathy.

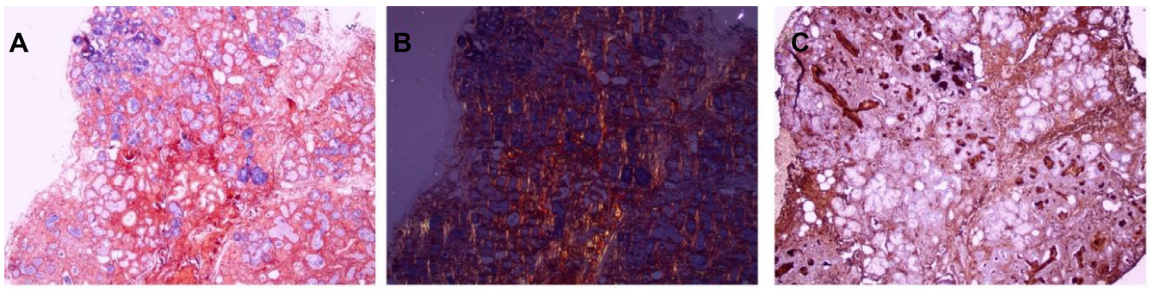

Fig. 4 – Histological examination of our patient's accessory salivary gland biopsy showing amyloid deposits at the periacinal interstitial level, stained with Congo red (A), with yellow-green birefringence under polarized light (B). Immunohistochemistry showed the deposits to be positive for anti-Kappa light chain antibodies (C).

agents, both of which are associated with an elevated risk of recurrent hemorrhage [3,4].

CAA is associated with a high mortality rate, ranging from 10% to 40%. Survivors often face chronic disability and cognitive decline, with recurrence rates estimated between 4.4% and 14.1% per year [5]. This highlights the need for early diagnosis and careful management to minimize long-term complications.

Spontaneous (nontraumatic) intracranial hemorrhage is the hallmark lesion of cerebral amyloid angiopathy (CAA),

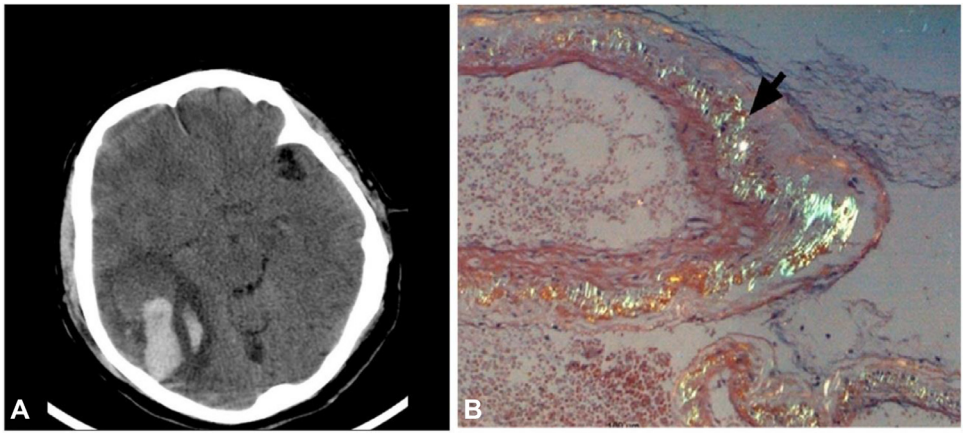

Fig. 5 – A 67-year-old man presented with acute-onset headache and vomiting. Axial CT scan reveals a large right temporoparietal hematoma (A). Histopathology revealed amyloid deposits in the outer tunica media of large-calibre leptomeningeal blood vessels (arrow) with apple-green birefringence (B) [11].

typically involving one or more hemorrhages in the cortex and juxtacortical white matter. The posterior regions of the brain, particularly the frontal and occipital lobes, are most commonly affected [5]. Leptomeningeal arteries tend to exhibit more severe amyloid deposition compared to cortical arteries, while vessels within the white matter are generally spared [6]. Although the presence of microbleeds is a recognized feature of CAA, it is not specific to the condition and can also be observed in hypertensive angiopathy. However, microbleeds localized within territories typically associated with CAA vascular involvement are highly suggestive of the diagnosis [1].

Clinically, intracranial hemorrhage in CAA presents with focal neurological deficits, often accompanied by symptoms such as headache, nausea, seizures, and altered consciousness [5]. Cognitive impairment is frequently noted in patients with CAA, irrespective of the presence of intracranial hemorrhage, and has been linked to the presence of lobar microbleeds [7]. In some cases, CAA may manifest as transient, stereotyped focal neurological episodes, frequently involving sensory symptoms such as paresthesia or pseudo-angina, as reported in the literature [8,9]. Rarely, CAA can present in an inflammatory form, characterized by a constellation of symptoms including headache, altered consciousness, cognitive decline, behavioral changes, focal neurological signs, and seizures [8].

Definitive diagnosis historically relied on postmortem examination of brain or leptomeningeal tissue, demonstrating the presence of eosinophilic hyaline material in the arterial media or adventitia, consistent with amyloid β deposition. Congo red staining, revealing characteristic apple-green birefringence under polarized light, remains a key histopathological feature (Fig. 5).

Advances in neuroimaging now allow for in vivo diagnosis using the Boston criteria, which rely on identifying hemorrhagic markers on brain magnetic resonance imaging (MRI), including macrohemorrhages (>1 cm in diameter) and microhemorrhages (<1 cm in diameter) [10]. Additional diagnostic tools, such as positron emission tomography (PET) imaging or the assessment of Aβ40 and Aβ42 protein levels in cerebrospinal fluid (CSF), may further support the diagnosis of CAA [11].

The primary goal of treatment in cerebral amyloid angiopathy (CAA) is to control systemic factors contributing to brain injury. Reducing the risk of intracranial hemorrhage focuses on the management of comorbid conditions, particularly cardiovascular risk factors, and careful evaluation of anticoagulation therapy. In cases of transient neurological episodes, antiepileptic drugs such as valproic acid, lamotrigine, and topiramate have shown efficacy [8,9]. For inflammatory forms of CAA, immunosuppressive therapies, typically corticosteroids and/or cyclophosphamide, are the mainstay of treatment. These approaches generally yield favorable outcomes, with rapid clinical improvement and a low rate of recurrence [8,12].

Emerging therapeutic strategies for CAA are centered on interventions targeting amyloid β at different stages of its pathophysiological cascade. Ongoing clinical trials are investigating approaches to reduce amyloid β production using antisense oligonucleotides, facilitate its clearance with amyloid-binding agents such as tramiprosate or immunotherapies, and promote its proteolytic degradation via enzymes like neprilysin [8].

Conclusion

Cerebral amyloid angiopathy is a small vessel disease that significantly contributes to intracranial hemorrhage and cognitive impairment in elderly individuals. Early diagnosis is critical for managing associated risks and addressing contributing factors. However, there is currently no effective treatment to halt or reverse disease progression, underscoring the urgent need for further research into novel therapeutic options.

Patient consent

I confirm that written informed consent for the publication of this article has been obtained from the patient, as appropriate. Copies of the consent forms have not been provided to ensure patient anonymity, but they are retained in my records for reference if required.

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