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MODESTUM

Case Report **OPEN ACCESS**

An extremely rare case with diagnoses of cerebral amyloid angiopathy, idiopathic normal pressure hydrocephalus, and dementia with Lewy bodies

Suleyman Emre Kocyigit [1*], Kerime Basaran [2], Derya Kaya [3], Hasan Bozkurt [4], Erdogan Bulbul [5], Ahmet Turan Isik [3]

[1] Department of Geriatrics, Balikesir University, Balikesir, TURKEY
[2] Department of Internal Medicine, Balikesir University, Balikesir, TURKEY
[3] Department of Geriatric Medicine, Dokuz Eylul University, Izmir, TURKEY
[4] Division of Nuclear Medicine, Ataturk City Hospital, Balikesir, TURKEY
[5] Department of Radiology, Balikesir University, Balikesir, TURKEY
***Corresponding Author:** suleymanemrekocyigit@gmail.com

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ABSTRACT

Introduction: Inflammatory cerebral amyloid angiopathy (iCAA) is an inflammatory condition seen in cerebral amyloid angiopathy (CAA) which is a disease pathologically characterized by β-amyloid deposition in the walls of cortical and leptomeningeal arteries. It contributes independently to cognitive impairment in older adults and is most commonly associated with Alzheimer's disease (AD).

Case reports: We describe a 73 year old male patient presenting with rapidly progressive cognitive decline with clinical dementia syndromes other than AD, namely idiopathic normal pressure hydrocephalus and dementia with Lewy bodies at the same time, who is not widely thought to be related to CAA and thus may have been overlooked.

Conclusion: This case is striking to illustrate that inflmmatory CAA may also present in the context of multiple neurodegenerative diseases and interact with them leading to be an independent variable for rapidly progressive decline and immunosupressive treatment may help slow down the cognitive decline and cause improvement in Daily living activities in the life of older adults. It is of importance to diagnose and manage iCAA for the accurate patient care and for research settings.

Keywords: cerebral amyloid angiopathy, idiopathic normal pressure hydrocephalus, dementia with Lewy bodies, rapidly cognitive decline, cognitive impairment

INTRODUCTION

Dementia refers to a range of progressive neurodegenerative diseases affecting the brain. While Alzheimer's disease (AD) is the most common form of dementia, various other types are also observed in clinical practice. One of them, cerebral amyloid angiopathy (CAA), a vasculopathy, is characterized by beta-amyloid accumulation in the arterioles and venules of the leptomeninges and cortex [1]. In some cases, an inflammatory response against Aβ in the leptomeninges and cerebral parenchyma may lead to an inflammatory condition, inflammatory cerebral amyloid angiopathy (iCAA), which may require corticosteroids and immunosuppressive treatment [2]. Symmetric or asymmetric and patchy or confluent T2 hyperintense lesions in the cortex and subcortical white matter are characteristics for iCAA in the MR imaging [3], with cerebral macrohemorrhage, microhemorrhage or microinfarcts [2].

Idiopathic normal pressure hydrocephalus (iNPH) is associated with secondary parkinsonism due to several proposed mechanisms, including increased resistance to cerebrospinal fluid (CSF) outflow, resulting in ventricular enlargement, abnormal cerebral blood flow, brain parenchymal pressure, and increased water content in the periventricular region [4]. One of the core criteria for dementia with Lewy bodies (DLB) is Parkinsonism, which shares a common pathophysiology with Parkinson's disease, referred to as alpha synucleinopathy [5]. Although the coexistence of these two Parkinsonism-causing diseases is rare, it is inevitable that they can lead to serious adverse outcomes when they coincide.

CAA frequently accompanies AD; however, cases that may coincide with iNPH or DLB are highly uncommon [6]. To the best of our knowledge, the coexistence of CAA, DLB, and iNPH in the same patient has yet to be documented, which is why this case has been reported.

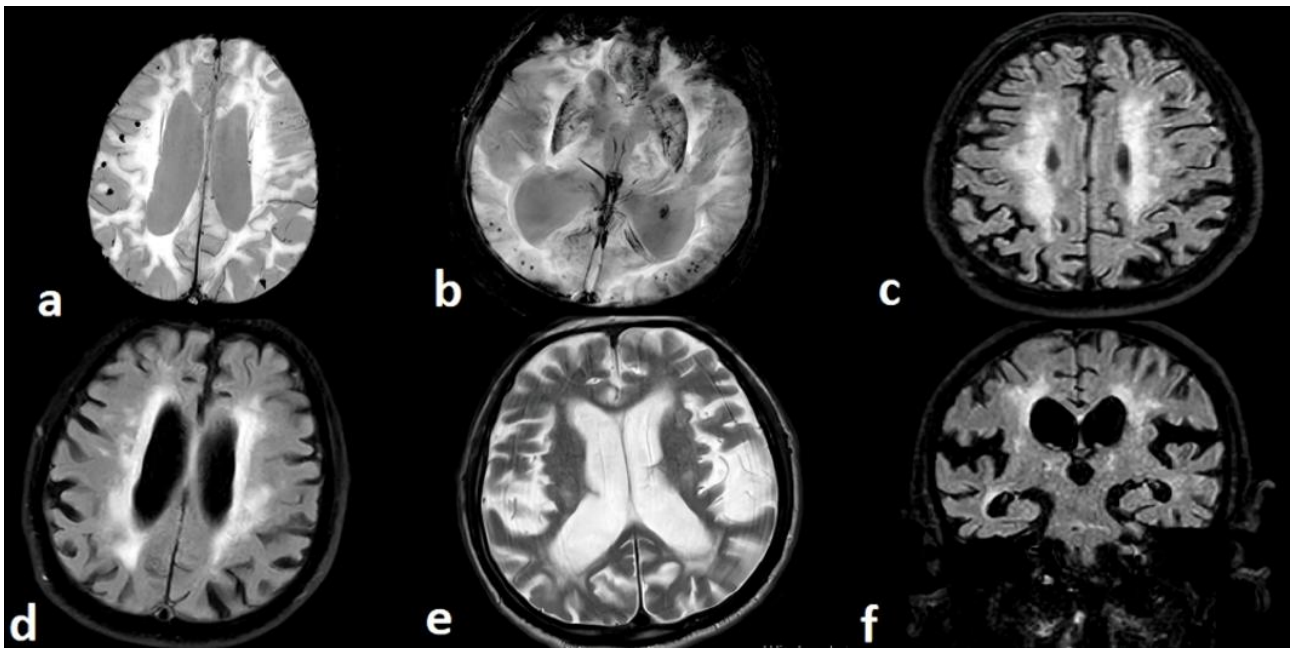

Figure 1. Cranial magnetic resonance imaging of the patient: Microbleedinig on the different area of the cerebral cortex on SWI seguantial imaging (a & b), periventricular hyperintensities extending to the subcortical white matter on different level of axial plan on T2-weighed FLAIR imaging (c & d), lateral ventricle enlargement on the axial plan (e), & callosal angle < 90° on the coronal plan (f) (Department of Radiology, Balikesir University Hospital [patient data anonymized])

CASE PRESENTATION

A 73-year-old male patient with a medical history of hypertension and frequent headaches was admitted to the geriatric outpatient clinic due to forgetfulness and gait-balance disturbances.

His family reported a history of a balance problem for the last two years and a total of six falls in the last year. It was understood that the type of fall was generally forward and sideways. The patient also had urinary incontinence and therefore used diapers. The cognitive history from his wife revealed that he had been experiencing forgetfulness, disorientation in time and place, and difficulty expressing himself for one year. When he woke up at night, he was observed to be confused about the location of the rooms, had visual hallucinations and organized visual delusions. His cognitive symptoms fluctuated during the course of the day. His mini-mental status examination (MMSE) was 14/30, with points missed in multiple sections, including attention, visuospatial function on MMSE was 14/30, with points missed in multiple sections, including attention, visuospatial function, and language. He also had nightmares for some years, with occasional punching or kicking in his sleep and acting out vivid dreams. Over the past 6 months, his forgetfulness progressed and started to negatively affect his instrumental activities of daily living as well as his basic activities of daily living. In the last three months, dressing apraxia, inability to eat his own food (inability to use a fork), pseudobulbar symptoms (occasional laughing or crying attacks), aphasia (decreased word output), and behavioral disorders (swearing and shouting) have been started together with episodes of recurrent falls. He became more dependent on his wife, such that their daughter had to stay with them for the patient care for nearly 2 months before his hospitalization.

In his family history, advanced-stage dementia was noted in his sister. He used only valsartan 80 mg/day as medication. The comprehensive geriatric assessment was performed.

On examination, he was not oriented to time and place. His vital signs were stable, but he had orthostatic hypotension measured by active standing test, with a significant systolic blood pressure drop of 25-27 mmHg after 3 min. He demonstrated mild hypomimia and bilateral postural tremor in the upper extremities as well as bradykinesia and postural instability, but no other parkinsonian features. The further examination was notable for blepharospasm in the right eye, decreased deep tendon reflexes, apraxia, positive frontal releasing signs, visual agnosia, and positive Romberg test. He had a magnetic gait, vertical typing during N-block turns, and deviations during walking. The timed up&go test was observed as 33 seconds.

Basic laboratories were unremarkable, but erythrocyte sedimentation rate was 89 mm/h, C-reactive protein level was 25 mg/L, vitamin B12 level was 70 pg/mL, and 25-OH vitamin D level was 18.5 ng/mL. Complement level, including C3 was low. CSF studies showed no white blood cells with an elevated IgG index. Electroencephalography demonstrated nonspecific slowing. Further evaluation at the hospital revealed unremarkable laboratory investigations for any other causes of rapidly progressive dementia.

MRI of the brain showed cerebral global atrophy and disproportionately enlarged subarachnoid-space hydrocephalus with an Evans index of 0.35 on T2-weighted images and a callosal angle below 90 degrees at the level of the posterior commissure in coronal sections. In addition, hyperintensity extending to the bilateral periventricular subcortical area was observed on T2-FLAIR imaging, while widespread microinfarctions were observed on diffusion-weighted imaging and scattered microhemorrhages in the frontal and parietal lobes on SWI sequence imaging (**Figure 1**).

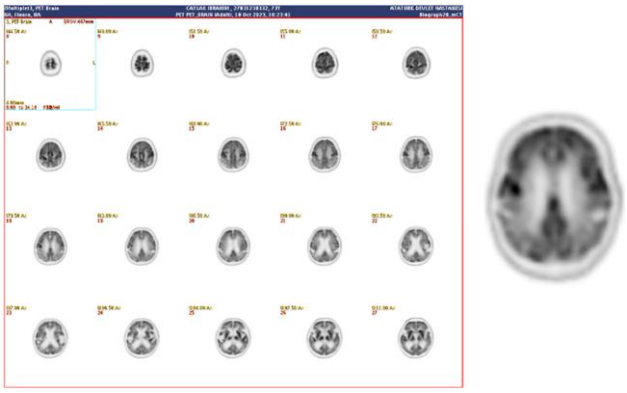

Figure 2. Cranial 18-fluorodeoxyglucose positron emission tomography of the patient (cingulate island sign) (Department of Radiology, Balikesir University Hospital [patient data anonymized])

At this stage, as the patient and his family did not give the consent for lumbar puncture for evaluate CSF biomarkers, a 18-fluorodeoxyglucose positron emission tomography scan of the brain was performed (Siemens biograph MCT S/20)-3R; 2014, USA) for the differential diagnosis, which showed hypometabolism of bilateral occipital, posterior parietal lobe and to a lesser extent, bilateral frontal lobes with relative sparing of the posterior cingulate gyrus (**Figure 2**).

All aforementioned clinical and neuroimaging findings are supportive of a probable diagnosis of DLB, iNPH, as well as a possible CAA-related inflammation (CAA-ri). The patient's vitamin B12 and vitamin D replacements were started immediately after hospitalization. Rivastigmine patch for his dementia and melatonin supplementation for his REM sleep behavior disorder (RBD) were started. For CAA-ri, 1000 mg of intravenous methylprednisolone was administered for 5 days, and the steroid was planned to be tapered off within six months, combined with azathioprine to maintain immunosuppressant treatment. For iNPH, a CSF tap test was performed with a large volume (40 mL) lumbar puncture, with normal CSF opening pressure (12 mmHg). In the first month following the discharge, his balance had improved without experiencing any falls, and there was no progression in the neuroimaging after treatment, and regression was even seen in some lesions (**Figure 3**). His visual hallucinations and RBD improved dramatically, and his pseudobulbar symptoms, headaches, and behavioral problems were also mitigated.

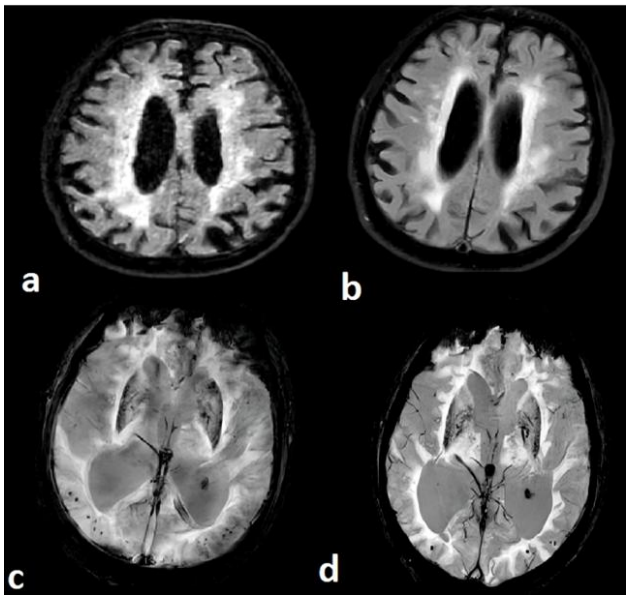

Figure 3. Comparison of pre- and post-treatment magnetic resonance imaging images: Periventricular hyperintensities extending to the subcortical white matter on T2-weighed FLAIR imaging (before pulse methylprednisolone) (a), periventricular hyperintensities on T2-weighed FLAIR imaging (after pulse methylprednisolone) (b), bilaterally microbleeding on the posterior parietal region of the cerebral cortex on SWI seguantial imaging before treatment (c), & reduced microbleeding on the posterior parietal region of the cerebral cortex on SWI seguantial imaging after treatment (d) (Department of Radiology, Balikesir University Hospital [patient data anonymized])

DISCUSSION

The case illustrates that three different types of neurodegeneration, including CAA-ri, iNPH, and DLB, may cause dementia in the same patient. The patient presented with gait-balance disturbance, falls, and progressive cognitive impairment, which worsened significantly in a short period of 3-6 months. It is imperative that one of the different types of dementia may have been missed entirely in such a case without a comprehensive geriatric assessment and a certain clinical suspicion.

CAA is an age-related central nervous system vasculopathy characterized by beta-amyloid deposition in small to medium-sized arteries, arterioles, or capillaries of the leptomeninges and cerebral cortex [2]. Cerebral microbleeds and larger lobar intracerebral hemorrhages are very typical features for the disease, whereas now nonhemorrhagic manifestations such as transient neurological episodes, chronic cognitive decline, dementia, cortical microinfarcts, and leukoencephalopathy are also included in the CAA spectrum [2, 7]. In a subset of CAA, vascular inflammation directed against vascular amyloid accumulation is observed, namely inflammatory CAA, which shows acute or subacute cognitive or functional decline and responses to immunosuppressive agents [8]. As in our patient, CSF inflammation and widespread microhemorrhages in the cerebral cortex in the SWI sequence imaging met criteria for possible CAA-ri, given relatively rapid cognitive decline, headaches, aphasia, and visual symptoms as focal neurological deficits, behavioral and pseudobulbar symptoms, which were incompatible with iNPH or DLB. The subacute cognitive and functional decline over a period of a few months that may be attributed to the immune-mediated vascular dysfunction led us to seek a reason other than DLB or iNPH for his rapidly progressive dementia [9].

Additionally, his symptoms had started to improve at least partially immediately after pulse corticosteroid treatment.

Furthermore, it is utmost important that the typical iNPH symptoms of the patient had benefited significantly from the CSF tap test duration decreased from 33 sec to 21 sec following the procedure). Given the similarity of clinical pictures of iNPH and Parkinsonian syndromes, which may result in a confusing diagnosis, and the common occurrence of iNPH in older individuals with major neurodegenerative disorders, it is crucial to differentiate iNPH from potential coinciding neurodegenerative diseases, such as DLB and AD, in geriatric practice [10]. Additionally, considering the underlying possible

mechanisms of iNPH, such as vascular disease or vascular risk factors, and neuroinflammation [11], it is suggested that CAA-ri may have played a role in the development of iNPH in the case. On the other hand, it is yet unclear how CAA may affect the course of iNPH.

Additionally, the patient also met criteria for DLB, with presence of dementia with cognitive fluctuations, visual hallucinations, parkinsonism, and RBD, as well as the cingulate island sign on his FDG-64 PET-CT imaging [5], as independent from CAA-ri and iNPH. Therefore, it is noteworthy that, in addition to CAA-ri, primary Parkinsonism, DLB, and secondary Parkinsonism, iNPH also coexisted in an older patient, who is one of the first patients, as far as we are concerned. The existing literature has highlighted that vascular factors, complement activation, and inflammation may be related to the development of both iNPH and DLB [12-14].

In conclusion, the present case report emphasized that three different types of neurodegeneration, including CAA-ri, iNPH, and DLB, may cause dementia with atypical clinical manifestations in the same patient. Consequently, healthcare professionals should keep in mind that the co-occurrence of different neurodegenerative conditions causing dementia is not rare and a challenging and intricate situation, which may require a holistic approach, including a comprehensive geriatric assessment and a certain clinical suspicion in dementia practice, especially in patients with atypical presentation.

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Data sharing statement: Data supporting the findings and conclusions are available upon request from the corresponding author.

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