Amyloid Beta-Related Angiitis Presenting as Visual Hallucination and Rapid Cognitive Decline

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A 72-year-old woman presented with a 1-month history of visual hallucinations and rapidly progressive cognitive decline. Neurological examination revealed disorientation, frontal release signs, ideomotor and ideational apraxia, and generalized hypertonia with hyperreflexia. Cerebrospinal fluid analysis showed normal opening and closing pressure, one white blood cell, and normal protein levels, and was negative for infectious, autoimmune, and syphilitic etiologies. Brain MRI demonstrated numerous microbleeds in the bilateral temporo-parieto-occipital regions, predominantly on the right, along with T2/FLAIR hyperintensities surrounding the microbleeds and focal leptomeningeal enhancement involving the bilateral parieto-occipital regions. Alternative causes of rapidly progressive cognitive decline were excluded. A diagnosis of probable amyloid beta-related angiitis (ABRA) was established. High-dose intravenous methylprednisolone (1 g/day for 3 days) was administered, followed by oral prednisone tapering off. At 2 months, the patient's psychotic symptoms had resolved, functional status in basic activities of daily living had improved, and her Thai Mental State Examination score increased from 2/30 at baseline to 15/30. Follow-up brain MRI demonstrated marked radiologic improvement. By 6 months, executive dysfunction had significantly improved, and no residual leptomeningeal enhancement was observed.

Figure Gadolinium-enhanced brain MRI of a 72-year-old woman presenting with a 1-month history of rapidly progressive cognitive decline. (A, B) T2/FLAIR images demonstrate asymmetric hyperintensities in the bilateral temporoparietal regions. (C)

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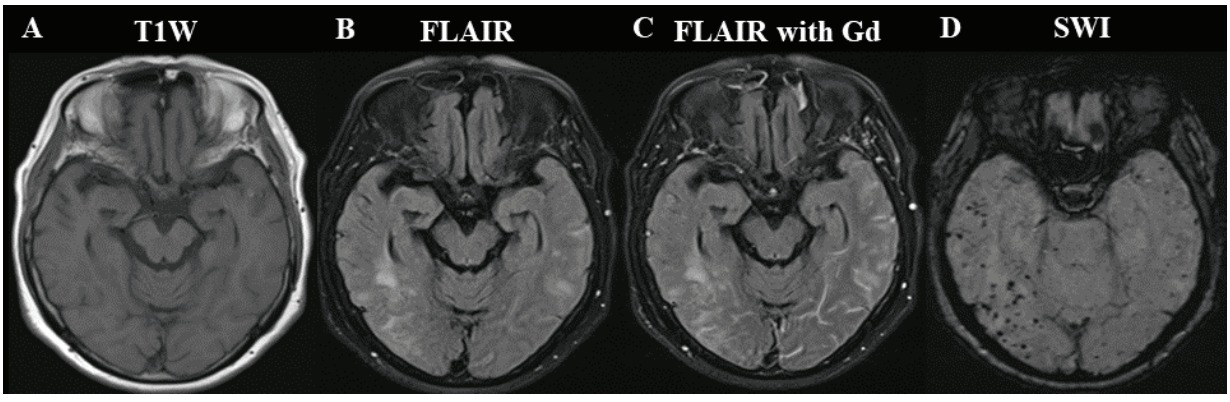
Leptomeningeal enhancement is observed in the parieto-occipital regions. (D) Susceptibility-weighted imaging reveals multiple cortical microbleeds extending into the adjacent subcortical white matter.



(Front cover)

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