A 67-year-old male patient was admitted to the Department of Neurology, Medical University of Silesia, because of progressive weakness of the left limbs persisting for several days. The patient’s relatives reported memory disturbances that had started a few months earlier. The patient had a history of untreated cardiac arrhythmia and a neurological event of unknown origin (12 months earlier, he experienced right arm weakness that resolved spontaneously 3 days after the onset). Moreover, he experienced disorientation, and neurological examination on admission revealed left limb paresis and ataxia. A neuropsychological examination revealed cognitive impairment in memory and visual spatial orientation. A transcranial Doppler ultrasound showed reduced cerebrovascular reactivity to changes in partial CO$_2$ pressure. Magnetic resonance imaging (MRI) of the head showed heterogeneous lesions corresponding to intracranial hemorrhages in the course of amyloid angiopathy (FIGURE 1). Other diagnostic procedures did not reveal any neurological symptoms or abnormalities. A preliminary diagnosis of cerebral amyloid angiopathy (CAA) according to the Boston criteria was established, and the patient was transferred to the Department of Internal Medicine for further diagnostic workup. A biopsy of oral and rectum mucosa was nonspecific; however, a biopsy of the abdominal fat tissue suggested amyloid deposits. Serum immunofixation revealed an abnormal ratio of lambda and kappa light chains. Scintigraphy with $^{123}$I-labeled serum amyloid P component (SAP) showed abnormal SAP levels in the kidney. The patient was diagnosed with amyloidosis and treated with bortezomib.

The term “amyloidosis” is used to describe hyaline eosinophilic extracellular deposits in various tissues. The clinical presentation is nonspecific and may vary depending on the site of deposition. Neurological involvement is observed in about 20% of the cases, and the median duration of neuropathic symptoms before the diagnosis of amyloidosis is 2 years.$^2$ CAA is a cerebrovascular
disease in which amyloid is deposited in the leptomeningeal, cortical, and subcortical arteries as well as in the capillaries. Most individuals with CAA are asymptomatic but they can present cognitive impairment and other neurological deficits caused by vascular wall amyloid deposition leading to hemorrhage.\(^3\) Amyloidosis results in intracranial hemorrhages in 15% to 20% of the patients aged over 60 years.\(^3\) An MRI scan of the hemorrhages in patients with CAA needs to be differentiated with hemorrhages due to hypertension, brain changes in posterior reversible encephalopathy syndrome, or, rarely, hemorrhagic metastases.

In our case, which is quite unique, amyloidosis initially occurred in the central nervous system without systemic manifestation. Cognitive deterioration, left limb weakness, and previously undiagnosed neurological symptoms were caused by amyloid pathology in the central nervous system. MRI of the head suggested CAA as the main etiology of the hemorrhage, which prompted further investigation. Of note, we did not observe neuropathy, which is the most characteristic neurological symptom of amyloidosis. The final diagnosis was based on abdominal fat biopsy and SAP scintigraphy, which allowed to initiate treatment before systemic manifestations of amyloidosis occurred.

REFERENCES