Recurrent craniospinal subarachnoid hemorrhage in cerebral amyloid angiopathy

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Abstract

Cerebral amyloid angiopathy (CAA) usually manifests as cerebral hemorrhage, especially as nontraumatic hemorrhages in normotensive elderly patients. Other manifestations are subarachnoid (SAH), subdural, intraventricular hemorrhage (IVH) and superficial hemosiderosis. A 52-year-old hypertensive woman presented with recurrent neurological deficits over a period of 2 years. Her serial brain magnetic resonance imaging and computed tomography scans showed recurrent SAH hemorrhage, and also intracerebral, IVH and spinal hemorrhage, with superficial siderosis. Cerebral angiograms were normal. Right frontal lobe biopsy showed features of CAA. CAA can present with unexplained recurrent SAH hemorrhage, and may be the initial and prominent finding in the course of disease in addition to superficial cortical siderosis and intracerebal and spinal hemorrhages.

Key Words

Amyloid angiopathy, convexity bleeds, subarachnoid hemorrhage, superficial siderosis

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Introduction

Cerebral amyloid angiopathy (CAA) is characterized by beta-amyloid deposition in the medium/small cortical and leptomeningeal arterial walls. It usually presents as intracerebral hemorrhage (ICH), representing 2% of ICHs and 38–74% of nontraumatic hemorrhages in normotensive elderly patients.[1] CAA-related ICHs are multiple, lobar and generally sparing the deep white matter, basal ganglia and brainstem. Other manifestations are subarachnoid (SAH), subdural (SDH), intraventricular haemorrhage (IVH) and superficial hemosiderosis.[1] Here, we describe a case of probable CAA with supporting pathology, with SAH as the initial presentation, later having had recurrent convexity SAHs and spinal hemorrhage.

Case Report

A 48-year-old female presented with sudden onset of loss of consciousness and left hemiplegia in July 2007. Computed tomography (CT) of the brain showed right frontotemporal ICH with SAH in the right sylvian fissure and temporal region [Figure 1a]. The digital subtraction angiogram (DSA) was normal. Her sensorium continued to fluctuate. CT brain done after 10 days showed SAH in the parietal, frontal and temporal regions [Figure 1b]. Magnetic resonance imaging (MRI) of the brain showed subacute right frontal hemorrhage extending from the sylvian fissure [Figure 1c] and sulcal SAH at the vertex bilaterally. Superficial cortical hemosiderosis was seen in the parasagittal region on both sides [Figure 1c]. She gradually improved with residual deficits (word finding difficulty and left facial paresis). Repeat DSA after 6 weeks was unremarkable. In October 2007, she had sudden onset numbness/weakness in the right faciobrachial region lasting 15 min. MRI brain showed subacute hemorrhages in the right posterior frontal and left anterior frontal regions. In December 2007, she had an episode of giddiness lasting 10 min. CT brain showed persisting subacute blood in the old lesions and ventricular dilation. Since then, there was progressive slowing of psychomotor activity and cognitive decline. In December 2008, she presented with altered sensorium and parapresis. MRI spine showed subacute hemorrhages, intrathecal at the S1-2 level, along the anterior surface of the cord at the L1-L2 level and along the filum terminale at the L4-S1 level [Figure 1d]. MRI brain showed bifrontal and right temporal gliotic areas with superficial hemosiderosis [Figure 1e]. Nodular enhancement was seen along the surface of the brain stem, cerebellum and spinal cord. In May 2009, she came with progressive altered sensorium. MRI brain showed ventricular

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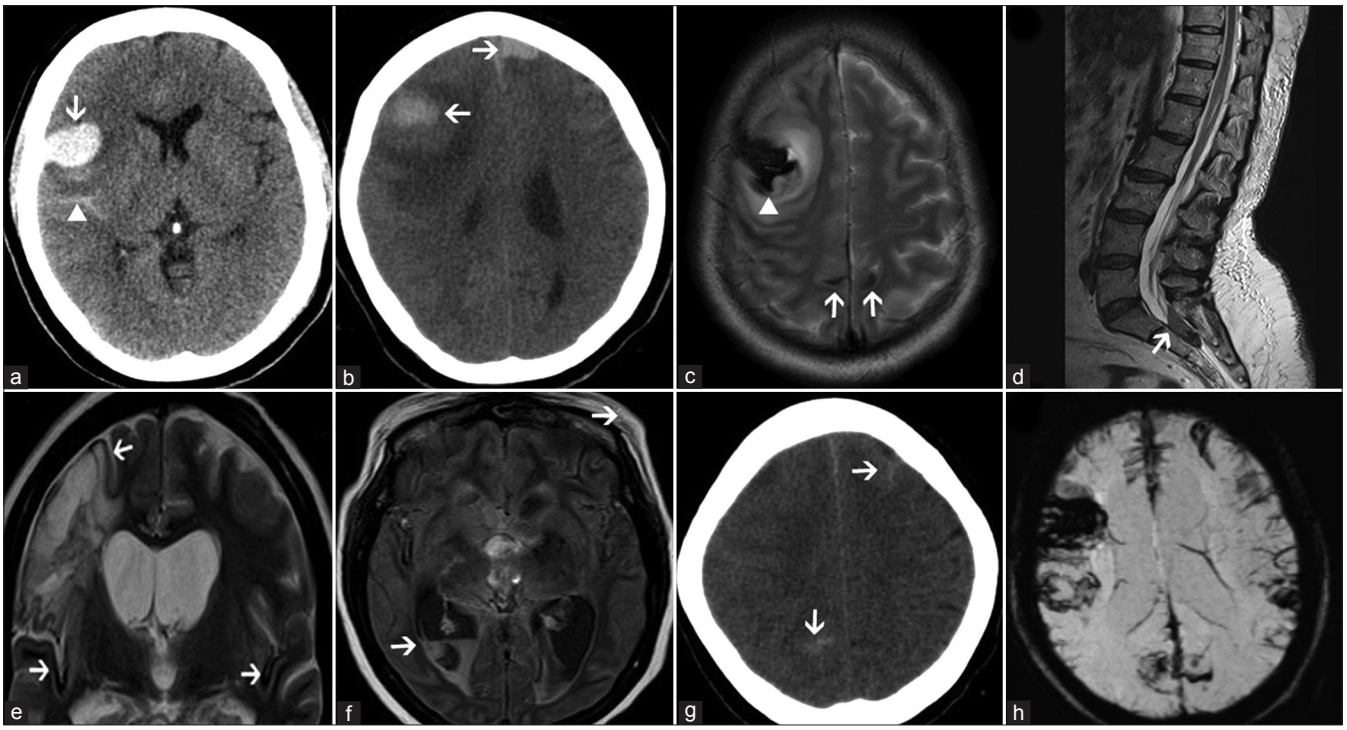

Figure 1: Computed tomography (CT) of the brain, magnetic resonance imaging (MRI) of the brain and spine images. CT showing right frontotemporal hematoma (a, arrow) with subarachnoid (SAH) hematoma (a, arrowhead). CT showing old hematoma and new areas of SAH on the left side (b, arrows). MRI showing subacute hematoma (c, arrowhead) and superficial cortical siderosis (c, arrows). MRI spine showing sacral intrathecal blood (d, arrow). MRI brain showing superficial cortical siderosis (e, arrows) and intraventricular blood (f, arrow). CT showing convexial SAH (g, arrows). SWI image showing siderosis in the supratentorial regions (h)

dilatation with IVH in the occipital horns [Figure 1f]. During the course of her illness, she was evaluated for underlying bleeding diathesis, connective tissue disorders, malignancy and vascular lesions, which were negative. Subsequent brain CT/MRI scans showed evidence of recurrent convexity SAHs in different locations [Figure 1g] with progressive superficial hemosiderosis [Figure 1h]. Right frontal lobe biopsy showed thickened small-/medium-sized meningeal vessels, containing Congo red positive, birefringent, immunofluorescent amyloid material within the tunica media and adventitia [Figure 2]. Many vessels showed concentric separation of their walls (lumen in a lumen appearance). There was reactive gliosis in the white matter with normal parenchymal vessels. Features were consistent with CAA. She continued to worsen with declining sensorium and recurrent infections, and expired in May 2009. The features were of probable CAA with supporting pathology.

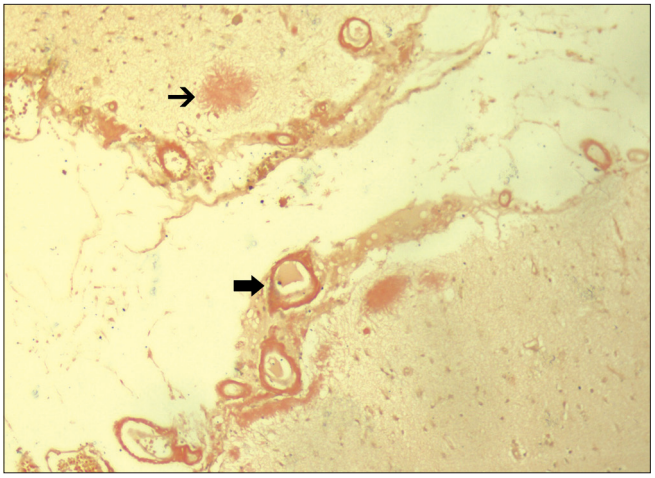

Figure 2: Histopathology image. Leptomeningeal vessels showing thickening of the walls due to amyloid deposits, which stained positive with Congo red and showed apple-green birefringence (thick arrow). Amyloid plaques were also seen (thin arrow). (Images magnification – 10×)

Discussion

CAA is a degenerative vasculopathy. Sporadic CAA-related hemorrhages occur in patients older than 60 years, while familial cases occur at a younger age. CAA is common in women. Our patient was younger than those described in the literature, and without a family history of CAA. CAA can present with acute neurologic deficit, focal seizures and dementia. Headache is usually absent.[1] Similar to this, our patient presented initially with left hemiplegia due to SAH/right frontal hematoma, followed by episodes of transient sensorimotor symptoms during the course of the illness without headache. Imaging findings described in CAA include ICH (macro/microhemorrhages), IVH, SAH or SDH, leukoencephalopathy and atrophy. GRE T2/SWI images show hemorrhagic squelae, subarachnoid (SAS) and superficial cortical hemosiderosis (SCS).[1,2] In our patient, the MR/CT images showed recurrent SAHs, mainly in the sylvian fissure and in the cerebral convexities, few parenchymal macrohemorrhages, IVH, SCS, siderosis along the cerebellar foliae and spinal hemorrhages and nodular enhancement along the meninges. There were no significant microhemorrhages on

SWI. Intracerebral hemorrhages (macro/micro) are common in CAA.[1] This feature was conspicuously not prominent in our case. SAH in CAA may be due extension of the parenchymal hemorrhage (secondary) or disruption of the leptomeningeal vessels (primary).[1] Commonly SAH in CAA is assumed to be secondary.[2] Current evidence suggests involvement of the leptomeningeal arteries in the early stages of CAA, followed by extension to the cortical arterioles.[3] Histopathology in our patient also showed amyloid deposits in the leptomeningeal vessels without any significant changes in the parenchymal vessels. Takeda *et al*. (2003) examined six autopsy cases of subcortical hematoma caused by CAA and concluded that primary hemorrhage occurred in the SAH space of the cerebral sulci due to rupture of the meningeal arteries.[4] These points may suggest that the initial hemorrhage occurs in the SAH space, mainly along the convexities, and then extends into the parenchyma. Our patient presented with sylvian fissure SAH as the initial event, and then had recurrent convexity SAHs, which is being increasingly recognised as a feature of CAA.[3,5,6] Spinal hemorrhage seen in our patient has not been commented upon in the previous reports. Enhancement as seen in our patient was also reported by others, which could be due to hemosiderin-related chemical meningitis.[6,7] Hemosiderin from recurrent SAH penetrates the pia mater and accumulates in the subpial layers of the cortex as linear deposits (SCS). SCS in CAA can be focal/ disseminated, preferentially over the cerebral convexity and often extending beyond the site of acute bleeding, suggesting that previous occult convexity SAH has occurred.[5-7] SCS was seen in our case even on the first MRI, although there were no neurological events in the past indicating subclinical bleed; this progressed to other areas over time. Hemosiderosis was prominently seen in the infratentorial regions also. SCS/ SAS have been reported as the only indicators of bleeding in some patients with CAA,[5,7] with SCS being included in the Modified Boston criteria for diagnosis of CAA.[5]

Conclusion

Our case had probable CAA with supporting pathology, with primary SAH as the first presentation, and later had recurrent convexial SAH and one episode of spinal hemorrhage. Patients with convexity SAH and suspicion of CAA should be followed-up carefully. CT brain shows convexity SAH as only slight sulcal hyperattenuation, which must be carefully looked for in suspected cases. It is important to screen for CAA in those with recurrent sensorimotor symptoms, isolated SCS and convexial SAH by including GRE/SWI sequence, especially in the elderly.

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