Cerebral amyloid angiopathy presenting as multiple intracranial lesions on magnetic resonance imaging

Case report

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Cerebral amyloid angiopathy is recognized as an important cause of spontaneous intracerebral hemorrhage in the elderly normotensive patient. Magnetic resonance (MR) imaging characteristics of this disease entity are rarely mentioned in the literature. The MR imaging findings of an elderly normotensive patient presenting with an acute spontaneous intracerebral hemorrhage secondary to amyloid angiopathy are reported and a brief review of amyloidosis is presented.

Key Words • amyloidosis • amyloid angiopathy • intracerebral hemorrhage • magnetic resonance imaging

Examination. On arrival, the patient continued to be confused and agitated. She had no focal neurological deficits; however, neurological examination on the following day revealed mild motor dysphasia, finger agnosia on the right, and left-right confusion. She was less agitated and more lucid than on admission. The multiplicity of neurological findings not corresponding to the area of the hemorrhage led us to obtain a repeat CT scan with contrast enhancement and eventually an MR image with and without contrast enhancement.

Precontrast CT of the head demonstrated a 1.5 × 1-cm intraparenchymal hemorrhage in the right occipital region adjacent to the atrium of the right lateral ventricle (Fig. 1). The MR appearance was very unusual and not characteristic of any disease process (Fig. 2). These images demonstrated multiple intracranial lesions. As documented in the CT scan, there was an oval well-circumscribed area in the right anterior occipital region adjacent to the atrium of the right lateral ventricle with signal characteristics of a recent hemorrhage (Fig. 2). Innumerable subcortical punctate areas of hypointensity were seen on the T2-weighted images, which were not observed on the T1-weighted images or the CT scan. Following intravenous administration of contrast medium, there was no contrast enhancement. The signal from the posterior fossa structures was unremarkable. The diagnostic possibilities of these images included...
Cerebral amyloid angiopathy

A 64-year-old man presented with an acute onset of left-sided weakness, aphasia, and right-sided headache. A noncontrast computerized tomography (CT) scan demonstrated a 1.5 x 1-cm intraparenchymal hemorrhage in the right anterior occipital region (black arrowhead). The CT scans, MR images, results of lumbar puncture, and electroencephalograms were suggestive of amyloidosis, a disease complex with the unifying feature of deposition in tissues of nonbranching fibrillar proteins having the common crystallographic characteristics of a beta-pleated sheet. This beta-pleated sheet conformation is responsible for its characteristic staining by Congo red dye and subsequent green birefringence when viewed through a polarized microscope.

Operation. Based on the above differential diagnosis and lack of specificity of the diagnostic studies (CT scans, MR images, results of lumbar puncture, and electroencephalograms), the patient was taken to surgery and a right frontal craniotomy with open cortical biopsy was performed. Upon opening of the dura, three small brownish oval punctate lesions were seen on the cortical surface (Fig. 3). One of these lesions was biopsied in the routine manner. Sections of the cortical brain biopsy revealed a small area of hemorrhage just beneath the pia in the cortex. Within the surrounding neuropil, the small blood vessels demonstrated variable homogeneous eosinophilic stains suggestive of amyloid which was confirmed by staining with Congo red dye and birefringence (Fig. 4).

Postoperative Course. The postoperative course was uneventful. On discharge, the patient was alert and oriented with significant improvement in her dysphasia. She had no focal motor or sensory loss upon discharge.

Discussion

Amyloidosis denotes a disease complex with the unifying feature of deposition in tissues of nonbranching fibrillar proteins having the common crystallographic characteristics of a beta-pleated sheet. This beta-pleated sheet conformation is responsible for its characteristic staining by Congo red dye and subsequent green birefringence when viewed through a polarized microscope.
Classification of Amyloidosis

Human amyloidosis is associated with multiple pathological conditions having diverse etiologies. Amyloidosis can be classified on the basis of clinical presentation and/or biochemical composition of the amyloid protein.2

Clinically, it is useful to divide amyloidosis into systemic or localized disease. Systemic amyloidosis can be further subdivided into: 1) primary amyloidosis (no evidence of pre-existing or coexisting disease); 2) amyloid associated with multiple myeloma; 3) secondary or reactive amyloidosis associated with chronic infectious diseases (such as osteomyelitis, tuberculosis, or leprosy) or chronic inflammatory diseases (such as rheumatoid arthritis or ankylosing spondylitis); 4) hereditary familial amyloidosis associated with familial Mediterranean fever and a variety of neuropathic, renal, cardiovascular, and other syndromes; 5) amyloidosis associated with long-term hemodialysis; 6) familial amyloid polyneuropathy; and 7) systemic senile amyloidosis.

Localized amyloidosis can be subdivided into endocrine and cerebral types. In endocrine-related amyloidosis, deposits consist of prehormonal or preprohormonal precursor proteins. The endocrine type of amyloidosis consists of tumor-like deposits in isolated organs without evidence of systemic involvement. This includes: 1) medullary carcinoma of the thyroid (procalcitonin); 2) pancreatic islet amyloidosis associated with insulinoma and diabetes mellitus; and 3) atrial amyloidosis (atrial natriuretic protein).

Localized cerebral amyloidosis consists of amyloid deposits restricted to the central nervous system (CNS). It includes: 1) Alzheimer’s disease; 2) Down’s syndrome; 3) Parkinson-dementia of Guam; 4) sporadic cerebral amyloid angiopathy; 5) inherited forms of cerebral amyloid angiopathy, both the Dutch type and the Icelandic type; and 6) spongiform encephalopathies (Creutzfeldt-Jakob disease and kuru).

Amyloid Angiopathy

The patient reported here exhibited the MR imaging findings of sporadic cerebral amyloid angiopathy. Three forms of amyloid deposits have been recognized in the CNS. They are: 1) the amyloid core of senile plaque; 2) deposits in the arterial and arteriolar walls of cortical and leptomeningeal vessels (congliphilic angiopathy); and 3) deposits in the capillary and arteriolar walls with extension into the surrounding brain parenchyma (dysphoric angiopathy). The latter two conditions are together called “amyloid angiopathy.”

Amyloid angiopathy is frequently present in the cerebral vessels of elderly patients and its incidence increases with age. There is a close relationship between amyloid angiopathy and intracerebral hematoma. Amyloid angiopathy is one of the causes of spontaneous intraparenchymal hemorrhage, especially in elderly, normotensive, often demented individuals. In contrast to the hypertensive hemorrhages which are predominantly in the basal ganglia, pons, or cerebellum, hemorrhages from amyloid angiopathy tend to be near the cortical surface in the parietal or occipital area. The patient presented here had an acute hemorrhage in the deep occipital lobe with multiple small cortical hemorrhages.

Magnetic Resonance Imaging Appearance

There are no specific MR imaging characteristics of amyloidosis. As a matter of fact, review of the literature does not reveal any thorough investigation of radiological characteristics of amyloid angiopathy. Drayer, in his report on the imaging of the aging brain, discusses cerebral amyloid angiopathy in the context of intracerebral hematoma with an MR image of the lobar hematoma. The present report discusses the MR imaging presentation of cerebral amyloid angiopathy. Although not specific, MR imaging demonstrates that cerebral amyloid angiopathy can present as multiple intracranial lesions. These lesions are especially well visualized on the T2-weighted images. They most likely represent areas of multiple cortical and subcortical punctate hemorrhages in different stages of evolution. Another important point is the lack of enhancement of the multiple intracranial lesions. Multiple cerebral lesions related to an inflammatory or neoplastic process usually demonstrate contrast enhancement. Thus, the above MR imaging findings in an elderly normotensive patient with a spontaneous hemorrhage should raise the possibility of cerebral amyloid angiopathy.

References


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