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Primary Amyloidoma of the Brain: CT and MR Presentation

Jeffrey Lee, George Krol, and Marc Rosenblum

Summary: We present CT and MR imaging studies of a histologically proved case of a primary intraaxial amyloidoma of the brain. Noncontrast CT and T1-weighted MR images revealed increased density and hyperintensity respectively of amyloid tissue. There was mixed intensity on proton-density and T2-weighted sequences, and marked contrast enhancement was demonstrated on both modalities.

Index terms: Amyloidosis; Brain, inflammation

The amyloidoses comprise a diverse group of diseases characterized by the deposition of insoluble fibrillar proteins in tissues. Involvement of the central nervous system (CNS) can take many forms, rarest of which is a localized mass or amyloidoma.

Case Report

A 61-year-old woman presented with a 1-year history of deteriorating mental function and a recent history of tonicoclonic seizures. A computed tomographic (CT) scan of the head at a local medical facility revealed an enhancing, partially calcified mass in the left parietal lobe. A brain tumor was diagnosed, and she was given 30 Gy whole-brain radiation. A CT scan of the head performed 6 months later again demonstrated a large, dense, enhancing left parietal mass without surrounding edema (Fig 1). Biopsy from the core of the mass revealed replacement of the neuropil by irregular clumps of amorphous and largely acellular eosinophilic material exhibiting microscopic foci of calcification and metaplastic ossification. At its periphery, these deposits clearly were centered on blood vessels, expanding and replacing their walls. This material stained a brick color in Congo red preparations and exhibited the dichroism and apple-green birefringence characteristic of amyloid on polarization. Transmission electron microscopy demonstrated that the material was fibrillar in nature, having an average diameter of approximately 8.5 nm. The final pathologic diagnosis was cerebral amyloidoma (Fig 2). Follow-up magnetic resonance (MR) revealed an irregular area of slightly increased intensity on T1-weighted sequences (Fig 3A) and mixed intensity on T2-weighted sequences (Fig 3B). Postcontrast images showed marked



Fig 1. CT scan without contrast. There is a high-attenuation mass around the atrium of the left lateral ventricle (arrow).

enhancement with peripheral extensions (Fig 3C). There was conspicuous lack of adjacent edema (Fig 3A–C). The patient remains in good general condition 5 years later and takes phenytoin for seizure control.

Discussion

The amyloidoses are a diverse group of diseases characterized by the deposition of insoluble fibrillar proteins (glycoproteins) in tissues. Clinically, two main forms are distinguished: systemic and localized. The amyloids deposited in both types share similar physiochemical properties, including a characteristic fibrillar appearance on electron microscopy, apple-green birefringence in Congo red-stained sections viewed under polarized light, beta-pleated sheet secondary structure, a high degree of insolubility, resistance to proteolysis, and poor immunogenicity (1).

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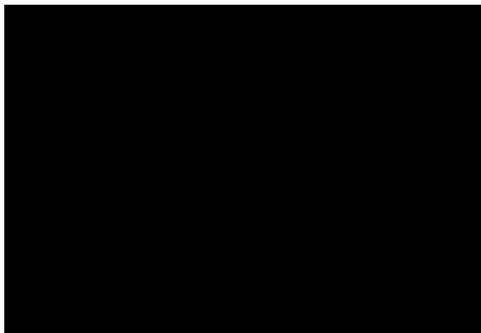


Fig 2. This histologic section of the cerebral mass, stained with Congo red and viewed under polarized light, demonstrates the dichromic blue-green birefringence characteristic of amyloid.

The most common manifestation of CNS involvement by amyloidosis is congophilic angiopathy, in which amyloid is deposited in blood vessels and occasionally causes cerebral hemorrhage. Deposits of amyloid also are found in senile or neuritic plaques associated with advanced age and Alzheimer disease and in the spongiform encephalitides of Kuru, Gerstmann-Strausler syndrome, and Jacob-Creutzfeldt disease (2). Localized masses of amyloid, called *amyloidomas*, are the rarest form of cerebral amyloidosis. Within the CNS, amyloidomas have been reported in the spinal canal, gasserian ganglion, and pituitary gland as well as in the brain.

Primary amyloidomas of the brain parenchyma are rare (3–8). Previously reported ce-

rebral amyloidomas were located in the cerebral white matter, with one case also having lesions in the posterior fossa (8). Single or multiple, they range in size from several millimeters to 8 cm. The CT presentation is that of masses of increased attenuation, with postcontrast enhancement. In 1935, Saltykow (3) reported several hazelnut-size masses in the cerebral cortex of a psychiatric patient. Harris and Rayport ("Primary 'Cerebral Amyloidoma'," *J Neuropathol Exp Neurol* (abstract), 1979;38:318) presented a case of a 28-year-old woman who had experienced headaches and focal seizures since 16 years of age. CT scan revealed a large mass of patchy increased density in the right frontal region. Spaar et al (4) described a walnut-size mass of amyloid in the occipital cortex and subcortical white matter in a 46-year-old woman with progressive visual loss. On CT, the mass was hypodense with contrast enhancement. Two cases of cerebral amyloidoma were described by Townsend et al (5). A 47-year-old woman with progressive cognitive decline had two amyloidomas, one in the left centrum semiovale and the other in the right optic radiation. The second case was a 50-year-old man with right homonymous hemianopsia and a high-density 0.8-cm enhancing mass in the region of the left trigone on CT scan. Hori et al (6) reported a 60-year-old man who died of lung cancer but had no evidence of systemic amy-

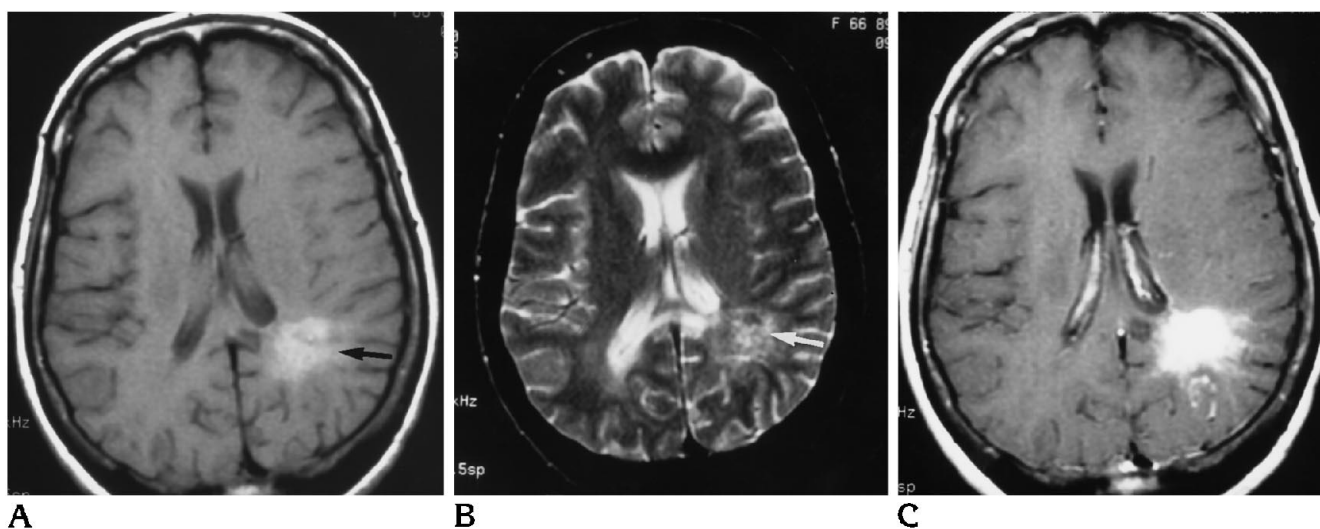


Fig 3. MR examination.

A, Precontrast T1-weighted scan (450/11/2 [repetition time/echo time/excitations]). The lesion has increased signal intensity (arrow).

B, T2-weighted image (2500/80/2). There is mixed intensity (arrow) without significant edema of the surrounding tissues.

C, On T1-weighted postcontrast examination (450/11/2), prominent enhancement with peripheral extensions is noted.

loid and had no neurologic symptoms. Autopsy revealed a 0.7-cm \times 0.4-cm amyloidoma in the occipital white matter and microscopic deposition of amyloid in the basal ganglia. Cohen et al (7) reported a case of a 28-year-old man with headaches, seizures, and optic disk swelling who had multiple large, hyperdense, enhancing white matter lesions on CT scan. They were located in the centrum semiovale, cerebellum, and pons. On MR, the lesions were hypointense on T1-weighted images, had mixed signal intensity on T2-weighted images, and showed faint, inhomogeneous enhancement.

The amyloidoma in our patient also was of increased attenuation on CT. Histologic studies showed the increased density to be primarily from the amyloid protein deposits, because the mass contained only microscopic foci of calcifications. Unlike a previously reported case in which an MR image was obtained (7), our case showed the amyloidoma to be hyperintense on the T1-weighted sequences; this was felt to be attributable to the dense amyloid deposits. On the T2-weighted images, the amyloidoma had patchy areas of high signal intensity centrally and intermediate signal intensity at its periphery. The exact reason for the different signals in the mass is unclear, but it likely represents non-uniform deposition of amyloid protein, the areas of more dense amyloid deposit having brighter signal. The postcontrast images showed intense enhancement in the center and an irregular, radial type at the periphery of the amyloidoma.

These peripheral extensions (Fig 3C) correspond to the deposits of amyloid in the blood vessel walls as seen on histologic sections. Contrast enhancement was seen on MR in both cases but was more intense and homogenous in our case.

The clinical presentation of amyloidomas is similar to that of slowly growing neoplasms, and differentiation on the basis of radiologic studies may not be possible. Primary amyloidomas, although rare, should be included in the differential diagnosis of intraaxial masses, which are hyperintense on T1-weighted MR and which show postcontrast enhancement.

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