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## CT and MR Imaging of Intracerebral Amyloidoma: Case Report and Review of the Literature

Dheeraj Gandhi, Roberto Wee, and Mayank Goyal

**Summary:** Intracerebral amyloidoma is the least common form of amyloid deposition in the brain. CT and MR imaging features in a case of pathologically proved cerebral amyloidoma are presented, and the available literature is reviewed. Typical imaging features of this entity include solitary or multiple supratentorial white matter masses that are hyperattenuated on nonenhanced CT. They have little or no mass effect on surrounding structures, extend medially up to the lateral ventricle wall, and have fine, irregular, enhancing margins.

The clinical manifestations of amyloidosis affecting the central nervous system vary with the degree and pattern of involvement. In its most common form, the amyloid protein is deposited in the arterial walls and results in cerebral amyloid angiopathy. In the following case report, we describe a patient with an intracerebral mass consisting of an amyloid-protein amyloidoma.

### Case Report

A 54-year-old right-handed woman presented with two left hemispheric seizures involving shaking and weakness of the right arm. She had associated dysphasia during the seizures and difficulty in finding words in the postictal period. She had no history of headache or other neurologic symptoms. Her medical history was noncontributory. Results of a neurologic examination were unremarkable. CT scanning showed a comet-shaped hyperattenuated mass in the left posterior frontal white matter. This mass extended medially to the lateral wall of the left lateral ventricle (Fig 1). The differential diagnosis considered on the basis of CT findings was oligodendroglioma or a calcified glioma. T1- and T2-weighted MR images showed a hypointense mass, which had intense enhancement after the administration of gadopentetate dimeglumine (Fig 2). Fine, irregular, radiating lines were seen at the medial edge of the mass on the gadolinium-enhanced T1-weighted images.

Stereotactic craniotomy was performed with the patient under regional anesthesia and with cortical mapping and excision of this mass. The tumor was situated 1.5 cm from the cortical surface. It had a gritty consistency and was well demarcated from the surrounding white matter. The lesion was defined and progressively debulked. The tail of this mass was followed to the lateral ventricle.

The specimen for pathology was a  $2.5 \times 1.7 \times 1$ -cm soft tumor with a yellow interior and hemorrhagic debris. On his-

tologic analysis, the specimen consisted of brain tissue that was replaced by masses of amorphous eosinophilic material and scattered aggregates of plasma cells and lymphocytes. The eosinophilic material was deposited in most of the vessel walls as well. This eosinophilic material was Congo red-positive and exhibited dichroism with rotation of the Nicol prism. The immunologic stains for kappa and lambda chains were positive, with a lambda predominance. The transthyretin stain was positive, and staining for beta-amyloid was negative.

The pathologic diagnosis was cerebral amyloid tumor. On extensive laboratory workup, no evidence of systemic amyloidosis was found. Serum and urinary protein electrophoresis results were normal.

### Discussion

Amyloidosis is a disease complex that results in the extracellular deposition of insoluble fibrillar protein with a beta-pleated sheet configuration. This particular protein configuration accounts for various biochemical and physical properties of amyloid proteins that permit their identification (1, 2). The characteristic staining properties—resistance to protease digestion and the insolubility of amyloid that promotes its accumulation within organs—are reflections of the configuration of the amyloid protein (1, 2). Amyloid deposition can occur in a systemic or localized fashion. The systemic form includes primary amyloidosis related to plasma cell dyscrasias, reactive or secondary amyloidosis, and the familial forms. Localized amyloidosis includes organ-limited forms, focal amyloid deposits, and senile amyloidosis.

Amyloid deposition within the brain can take many forms, including cerebral amyloid angiopathy, senile plaques of Alzheimer dementia, and deposits seen in the encephalopathy of Kuru and of Creutzfeldt-Jacob disease (1). Tumorlike deposition (amyloidoma) is the least common form of the brain involvement by amyloid proteins (3, 4), with only 13 previously reported cases (2–13).

A review of previously reported cases (2–13) reveals that the average patient age at presentation with cerebral amyloidoma is 47.8 years, and a slight female preponderance, with a female-male ratio of 8:5 (Table). Major presenting features of this lesion are seizures, headache, focal motor signs, and a cognitive decline.

Cerebral lobar white matter is most commonly affected, with occasional involvement of the cortex (5, 11). The mass is supratentorial, with only one reported case involving simultaneous involvement of the pons (10). Single and multiple lesions occur with

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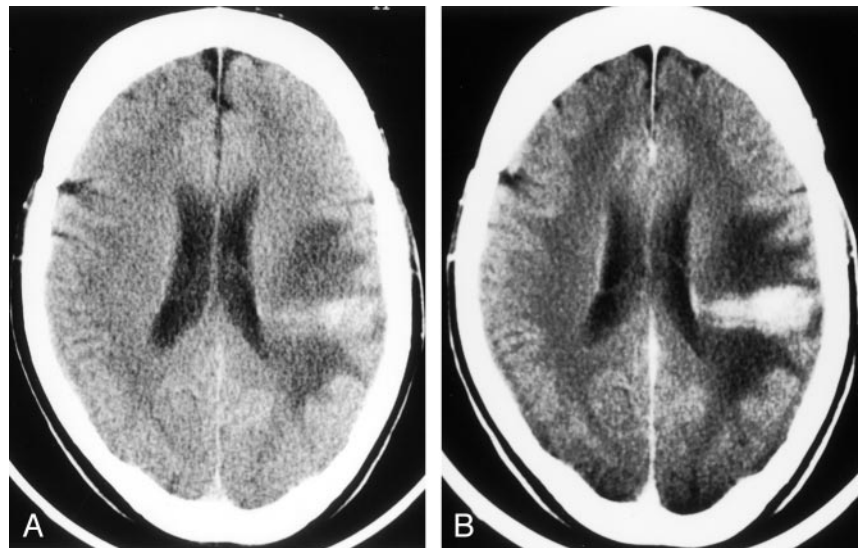
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FIG 1. CT scans obtained in 54-year-old woman with intracerebral amyloidoma.

A, Axial non-contrast enhanced CT scan of the brain reveals a hyperattenuated comet-shaped mass in the left posterior frontal white matter. Significant vasogenic edema is also present around the mass.

B, The mass enhances after administration of contrast material. Note the medial extension of the mass up to the lateral ventricular ependyma.



similar frequency; seven cases each are reported in the literature (Table).

Imaging findings are available in 12 of the 14 reported cases. On nonenhanced CT scans, the amyloid material appears hyperattenuated (cases 2, 4, 5, 8, 10, 12, 14) and shows enhancement with the use of iodinated contrast material. Spaar et al (7) have described multiple hypoattenuated masses in their patient; these showed enhancement after the administration of contrast agent. With these lesions, little or no mass effect and, frequently, no perilesional edema are observed. On MR images, the appearance is more variable. Amyloidomas can be hypointense (11, present case), isointense (4), or hyperintense (3) on T1-weighted images. Lee et al (3) postulated that increased signal intensity on T1-weighted images could be attributed to dense amyloid protein deposition. The signal intensity on T2-weighted sequences is mixed, with areas of high and low signal

intensity. In our patient, low signal intensity on T2-weighted images was seen, as was hyperintense perilesional edema. On gadolinium-enhanced T1-weighted images, faint or intense enhancement is seen. Cerebral angiographic results are normal, or they reveal vascular displacement due to the tumor. Townsend et al (8) observed absent filling of the veins in the affected region of the brain.

On reviewing the surgical and imaging findings (when available), we found that most reported cases of intracerebral amyloidoma show a medial extension up to the lateral ventricle ependyma (2-4, 7-9, 11-13 and the present case). Thickening of lateral ventricular wall has also been observed (9). In two cases (5, 6), we could not find adequate operative and imaging details. The reason for the medial extension of the tumor edge is unclear. In some cases (3, 4, 7, 11), including our own, interesting features on the images

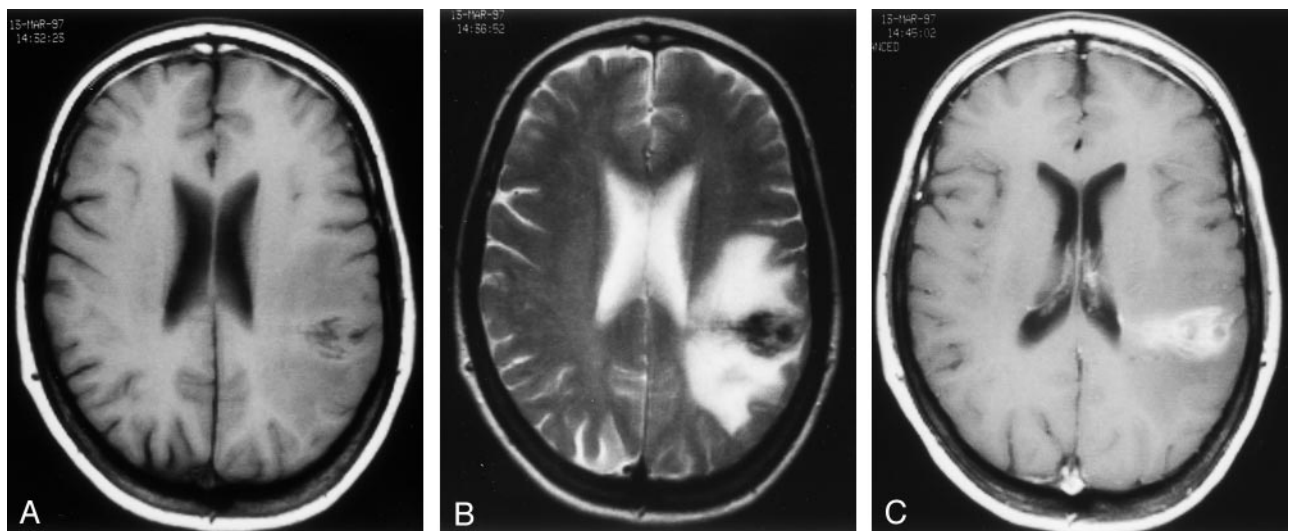


FIG 2. MR images obtained in 54-year-old woman with intracerebral amyloidoma.

A, The mass is hypointense on this axial T1-weighted image (551/5/2 [TR/TE/NEX]).

B, The mass is hypointense on this axial T2-weighted image (4110/110/2).

C, The mass is hyperintense on this T1-weighted postcontrast axial image.

## Review of Cases of Cerebral Amyloidomas

| Author and Year*                     | Patient (no.)/<br>Age (y)/Sex | Clinical Presentation                        | Location                                                                      | No. of<br>Lesions | CT Findings                                                                                                                           | MR Imaging Findings                                                                                                         | Angiographic Findings                                           |
|--------------------------------------|-------------------------------|----------------------------------------------|-------------------------------------------------------------------------------|-------------------|---------------------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------------------------------------------------------------------|-----------------------------------------------------------------|
| Saltykow, 1935 (5)                   | 1/Unknown                     | Psychiatric disturbances                     | Cortex, WM                                                                    | M                 | NA                                                                                                                                    | NA                                                                                                                          | NA                                                              |
| Harris and Rayport, 1979<br>(6)      | 2/28/F                        | Focal seizures                               | Frontal WM                                                                    | S                 | Hyperattenuating mass on<br>nonenhanced scans                                                                                         | NA                                                                                                                          | NA                                                              |
| Spaar et al, 1981 (7)                | 3/46/F                        | Visual loss, headache,<br>depression         | WM                                                                            | M                 | Hypoattenuating lesions, enhancement<br>on contrast-enhanced scans,<br>extension to the lateral ventricle                             | NA                                                                                                                          | Vascular displacements                                          |
| Townsend et al, 1982 (8)             | 4/47/F                        | Cognitive decline                            | Frontal/optic radiation, WM                                                   | M                 | Hyperattenuating mass on<br>nonenhanced scans                                                                                         | NA                                                                                                                          | Absent venous filling in the<br>affected region                 |
| Townsend et al, 1982 (8)             | 5/50/M                        | Visual field defects                         | Occipital WM                                                                  | S                 | Hyperattenuating mass with<br>enhancement on contrast-enhanced<br>scans                                                               | NA                                                                                                                          | NA                                                              |
| Moreno et al, 1983 (9)               | 6/48/M                        | R hemianopsia, L-arm<br>weakness             | L occipital WM                                                                | S                 | Ring-enhancing lesion, extension to<br>the lateral ventricle, thickened<br>ependyma                                                   | NA                                                                                                                          | Normal                                                          |
| Hori et al, 1988 (10)                | 7/60/M                        | Headache, seizure, dementia                  | L frontal, cerebellum, pons (WM)                                              | M                 | NA                                                                                                                                    | NA                                                                                                                          | NA                                                              |
| Cohen et al, 1992 (11)               | 8/32/M                        | Headache, seizure, decreased<br>cognition    | Bilateral centrum semiovale WM, 1<br>cortical lesion                          | M                 | Hyperattenuating masses, contrast<br>enhancement, extension to the<br>lateral ventricle                                               | Hypointense on T1WI, mixed<br>intensity on T2WI, faint<br>enhancement                                                       | NA                                                              |
| Eriksson et al, 1993 (2)             | 9/76/M                        | Seizure                                      | R parietal WM, extension to lateral<br>ventricle and choroid plexus           | S                 | NA                                                                                                                                    | NA                                                                                                                          | Mass effect on the<br>temporal horn of the<br>lateral ventricle |
| Schroder et al, 1995 (12)            | 10/70/F                       | Ataxia, L-leg weakness,<br>cognitive decline | R subcortical parieto-occipital, 2 smaller<br>lesions in the ventricular wall | M                 | Hyperattenuating, no edema,<br>extension to the lateral ventricle                                                                     | NA                                                                                                                          | Normal                                                          |
| Lee et al, 1995 (3)                  | 11/61/F                       | Seizures, decreased<br>mentation             | L parietal periaxial WM                                                       | S                 | Hyperattenuating, partially calcified,<br>no edema                                                                                    | Hyperintense on T1WI, mixed<br>intensity on T2WI, marked<br>enhancement, no edema,<br>extension to the lateral<br>ventricle | NA                                                              |
| Caerts et al, 1997 (4)               | 12/71/F                       | R pyramidal syndrome                         | L deep periventricular WM                                                     | S                 | Hyperattenuating mass on<br>nonenhanced scans, contrast<br>enhancement, no edema                                                      | Isointense on T1WI, hyperintense<br>on T2WI, strongly enhancing,<br>extension to the lateral<br>ventricle                   | NA                                                              |
| Blatter et al, 2001 (22)             | 13/26/F                       | R-hand paresis, dysarthria                   | L parieto-occipital, smaller R-sided<br>lesions                               | M                 | NA                                                                                                                                    | T1WI and T2WI appearance<br>unknown, contrast<br>enhancement, extension to the<br>lateral ventricle                         | NA                                                              |
| Gandhi et al, 2003<br>(present case) | 14/54/F                       | Seizures                                     | L frontal WM, medial extension to the<br>lateral ventricle                    | S                 | Comet-shaped hyperattenuating mass<br>on nonenhanced scans, contrast<br>enhancement, medial extension up<br>to lateral ventricle wall | Hypointense on T1WI and T2WI,<br>strong contrast enhancement,<br>surrounding edema                                          | NA                                                              |

Note.—L indicates left; M, multiple; NA, not applicable; R, right; S, single; T1WI, T1-weighted image; T2WI, T2-weighted image; and WM, white matter.

\* Numbers in parentheses are reference numbers.

are the finely irregular, radiating lines at the edge of the tumor. These may indicate the deposition of amyloid along the vessels, a finding that has been observed in pathology specimens (2, 3, 6, 12).

The pathogenesis for the tumoral deposition of an amyloid tumor is unclear. Some have argued that the amyloid fibril protein is either derived from components that leak from the vessels or synthesize at the site of deposition (2). Several amyloid accumulations have been shown to have coexistent plasma cells (6, 7, 8, 11). In an immunochemical and biochemical study, Vidal et al (14) have shown monotypic perivascular plasma cells with cytoplasmic staining for the lambda light chain. Their findings suggest that the amyloidoma is related to a benign clonal proliferation of the plasma cells. The origin of intracerebral plasma cells commonly observed in cerebral amyloidomas is unclear. Whether a solitary plasmacytoma can evolve in to an amyloidoma remains to be determined (11).

The clinical course of intracerebral amyloid tumor appears to be benign, although these lesions can show slow growth, and they can also recur after surgical resection (2). Scheroder et al (12) suggest that if the diagnosis is made by means of biopsy, one can wait and control the local findings, especially when no malignant plasma cells are present in the biopsy specimen.

### Conclusion

We have presented a case of intracerebral amyloidoma and reviewed its imaging features. A diagnosis of amyloidoma should be considered in solitary or multiple intracerebral white matter masses with little or no mass effect, hyperattenuation on nonenhanced CT scans, and enhancement after the administration of

contrast material. Medial extension of the mass up to the lateral ventricular ependyma and a fine, irregular, radiating margin on imaging studies could possibly add specificity to this diagnosis.

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