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This information is current as of May 6, 2024.

AJNR Am J Neuroradiol 1990, 11 (4) 800-801
<http://www.ajnr.org/content/11/4/800.citation>

Gadopentetate-Dimeglumine-Enhanced MR Imaging of Gliomatosis Cerebri: Appearance Mimicking Leptomeningeal Tumor Dissemination

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The term gliomatosis cerebri was first used by Nevin in 1938 [1] and represents a neoplastic process of glial origin characterized by its diffuse pattern of infiltration along anatomic pathways [1, 2]. Radiologic methods of detecting gliomatosis cerebri have relied heavily on CT [3–6]. Recently, the efficacy of MR imaging in detecting this disease has been described [7, 8]. We report a case of histologically documented gliomatosis cerebri that on gadopentetate-dimeglumine-enhanced MR images exhibited an enhancement pattern that mimicked one that has been seen in children with leptomeningeal spread of primary intracranial neoplasms [9, 10]. In our case, examination of the CNS at autopsy demonstrated an absence of leptomeningeal tumor but provided a morphologic explanation for the appearance of the contrast-enhanced MR examination.

Case Report

The patient was a 16-year-old boy who initially presented with a right sixth nerve palsy. MR revealed a lesion within the right medial temporal lobe and thalamus. Stereotactic biopsy yielded a diagnosis of infiltrating glioma. The patient was treated with 5400 rads of local radiotherapy and corticosteroids. MR (1 year later) showed focal enhancement within the right temporoparietal lesion after IV administration of gadopentetate dimeglumine (0.2 ml/kg) (Fig. 1A). In addition, linear enhancement about the cerebellar and right posterior temporal sulci/gyri was identified that suggested the presence of leptomeningeal tumor seeding (Figs. 1B and 1C). CSF studies yielded the following results: glucose = 67 mg/dl, protein = 68 mg/dl, cytology was negative. The patient had a protracted downward clinical course and died.

At autopsy, the brain revealed gross enlargement of the white matter with blurring of the corticomedullary junction within the right temporal lobe. Microscopic examination showed a diffuse infiltration of both cerebral hemispheres with malignant glial cells. The right temporoparietal region, basal ganglia, and thalamus were most se-

verely involved. Diffuse bilateral tumor infiltrates were identified in all portions of the brainstem and cerebellum. The cerebellar involvement exhibited prominent subpial extension, associated gyral expansion, and marked focal edema within the molecular layer of the cortex (Fig. 1D). Intervening sulci were obliterated. The subarachnoid spaces and leptomeninges showed no tumor. Similar findings in the right posterior temporal lobe were seen. No areas of necrosis, evidence of radiation change, or chemotoxicity of the brain were present.

Discussion

Gliomatosis cerebri is a relatively rare glial neoplasm of presumed astrocytic origin [2]. All age groups can be affected; however, the lesion is more common in the second and fifth decades [7]. Preoperative diagnosis of gliomatosis cerebri by traditional radiologic methods, including contrast-enhanced CT, is difficult. These lesions are diffuse, commonly isodense, and usually do not enhance with administration of IV contrast medium [7]. Enhancement may, however, be seen in the late stages of this disease [3]. Only rarely are focal cerebral masses formed [3, 5]. The CT appearance of gliomatosis cerebri may simulate demyelinating diseases [4] or pseudotumor cerebri [3]. Although ventricular compression is most common, ventricular dilatation has been reported as well [6].

Previous reports [7, 8] suggest a marked disparity in the ability of CT and MR to image the pathologic manifestations of gliomatosis cerebri. Lesions that are subtle or inapparent on contrast-enhanced CT may be recognized with MR [7, 8]. The addition of gadopentetate dimeglumine for enhanced MR in this case resulted in a focus of enhancement within the right temporoparietal region (Fig. 1A). Pathologically, this corresponded to dense tumor infiltration and edema. To our knowledge, the appearance of gliomatosis cerebri on gadopentetate-dimeglumine-enhanced MR has not been described.

Received September 11, 1989; revision requested November 2, 1989; revision received December 20, 1989; accepted December 20, 1989.

This work was supported in part by NINDS institutional training grant no. T32-NS07304.

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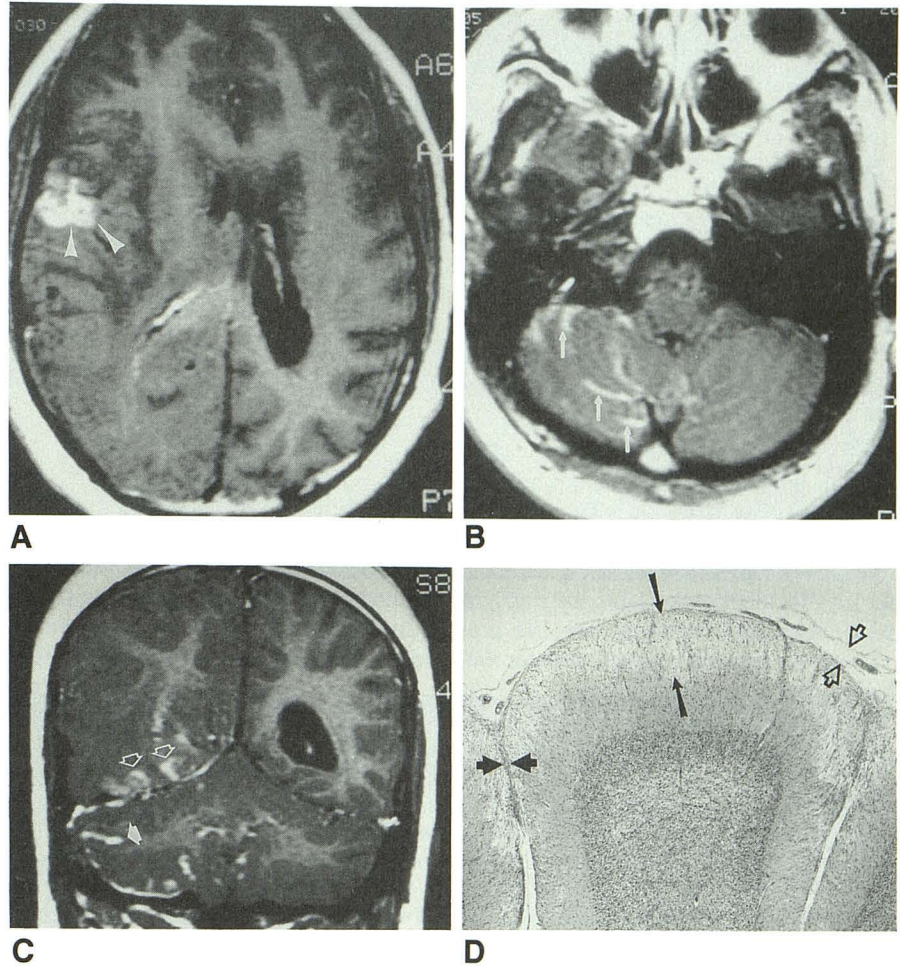
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Fig. 1.—A, Axial contrast-enhanced MR image (500/20) shows focal enhancement near sylvian fissure (arrowheads).

B and C, Axial (B) and coronal (C) contrast-enhanced MR images (500/20). Linear enhancement following course of cerebellar sulci/gyri (closed arrows in B and C) with similar involvement in right posterior temporal lobe (open arrows in C) suggests leptomeningeal spread of tumor.

D, Microscopic examination of cerebellum shows diffuse tumor infiltration and marked edema of cortical molecular layer (long arrows), with resultant focal obliteration of intervening sulci (solid arrowheads). The subarachnoid space is free of tumor (open arrowheads). (H and E $\times 40$)



Linear enhancement within the posterior fossa was also present (Figs. 1B and 1C). This enhancement pattern simulated the appearance of leptomeningeal dissemination of tumor previously described in pediatric patients [9, 10]. Histologic examination, however, revealed no evidence of leptomeningeal disease. Instead, dense tumor infiltration of gyri with marked edema of the cortical molecular layer was noted. Expansion of gyri with obliteration of intervening sulci was also seen (Fig. 1D). Presumably, tumor deposits elicited a localized edema and blood-brain barrier permeability change resulting in the enhancement pattern present on contrast MR studies. Yet, not all areas of tumor infiltration enhanced with contrast.

In summary, MR imaging is well suited for evaluation of patients with gliomatosis cerebri. The addition of gadopentate dimeglumine may reveal focal areas of enhancement. In our case, diffuse cortical infiltration and gyral expansion was present. Linear enhancement on contrast MR images simulated leptomeningeal tumor dissemination.

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