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MR Imaging of Diffuse Glioma

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Summary: Neuroradiologic diagnosis of "gliomatosis cerebri" is hampered by the diffuse, isodense character of the lesion, and the fact that it may not enhance when intravenous contrast medium is administered. Clinical signs and symptoms are usually nonspecific, nonfocal, and disproportionately mild. We report a case of diffuse glioma in a 30-year-old man, discuss the difficulty in arriving at a precise diagnosis of gliomatosis cerebri, and examine the possible role of MR in its detection and delineation.

Index terms: Gliomatosis; Brain neoplasms, magnetic resonance; Magnetic resonance, contrast enhancement

"Gliomatosis cerebri" is a rare condition characterized by diffuse overgrowth of the central nervous system (CNS) by glial cells in varying stages of differentiation (1). Its neuroradiologic diagnosis using traditional techniques like computed tomography (CT) is difficult because the overgrowth does not usually form a well-defined mass. Recently, the efficacy of magnetic resonance (MR) imaging for the detection of this disease has been reported (2–4); however, at present no imaging modality appears able to show the true extension of neoplastic cells. We report a case of diffuse glioma and discuss the difficulty in making the precise diagnosis of "gliomatosis cerebri."

Case Report

A 30-year-old man was admitted to our hospital because of diplopia and a slowly progressive, painless loss of vision in both eyes. Physical examination at that time was unremarkable, as were the results of routine laboratory studies. The protein content of the cerebrospinal fluid was 26 mg/dL, considered within normal limits. Neurologic examination revealed right abducens nerve palsy. Visual acuity was decreased bilaterally. Perimetric examination revealed bilateral central scotomas and binasal hemianopsia. Funduscopic examination revealed bilateral papilledema. CT of the brain demonstrated the presence of small lateral ventricles and decreased attenuation bilaterally within the white matter of the frontal lobes and in the left temporal lobe without postcontrast enhancement. No enlarged sulci were observed (Fig. 1). The patient underwent MR (General Electric 1.5-Tesla Signa scanner) using T1- and T2-weighted spin-echo pulse sequences. A T1-weighted axial scan was obtained using a repetition time (TR) of 600 msec and an echo time (TE) of 30 msec. This sequence was followed by T2-weighted scans employing a TR of 2500 msec and TEs from 30 to 80 msec. Intravenous (IV) administration of gadolinium diethylenetriaminepentaacetic acid (Gd-DTPA) was carried out for enhanced MR examination. MR using T1-weighted images (T1WI) revealed enlargement of the left temporal lobe with herniation of the mesial aspect of the temporal lobe into the prechiasmatic cistern; it also revealed enlargement of the left frontal lobe with backward deviation of the left middle cerebral artery (Fig. 2). On T2WI, high intensity was observed in the regions noted above, and bilateral involvement of the periventricular and subcortical white matter was noted. Neither the brain stem nor cerebellum were affected (Fig. 3). Following IV administration of Gd-DTPA, T1WI revealed no enhanced tumor mass (Fig. 4). Cerebral angiography was performed, but revealed no tumor stain.

Stereotactic brain biopsy was carried out revealing reactive astrocytes rather than tumor cells; therefore, a left craniotomy was performed to establish a final diagnosis. On macroscopical examination of the brain, the temporal lobe was found to be enlarged and the gyri flattened. Microscopically, the cortex and white matter of the temporal lobe were infiltrated with many neoplastic cells. Mitotic figures were rare, and areas of necrosis were not detected. Almost all of the tumor cells were gemistocytes and a diagnosis of gemistocytic astrocytoma was made (Fig. 5).

Irradiation to the cerebrum (64.8 Gray) and interferon immunotherapy were carried out in the 2 months following surgery. Following this therapy, although the patient lost his visual field, he was discharged with no motor paresis.

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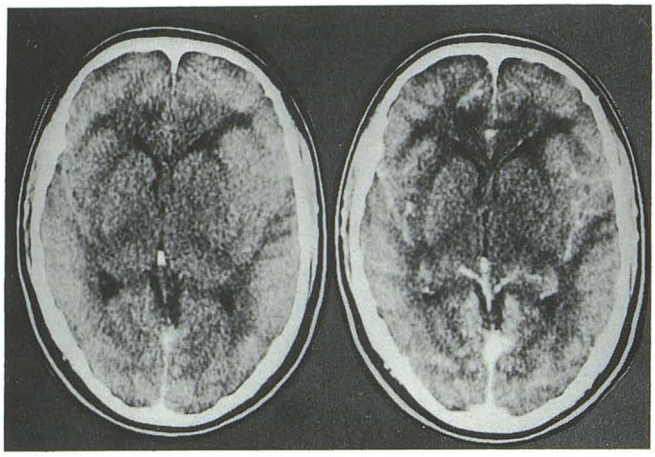


Fig. 1. Plain and postcontrast-enhanced CT on admission reveals small lateral ventricles and decreased attenuation bilaterally within the white matter of the frontal lobes and in the left temporal lobe. No enhancement is seen. No enlarged sulci were observed.

CT at discharge revealed enlarged sulci and slightly enlarged lateral ventricles.

Discussion

Gliomatosis cerebri is generally considered a glial neoplasm and its precise morphologic definition is difficult. Broad pathologic guidelines for postmortem examination for the diagnosis have been suggested by Scheinker and Evans (5), but the definition of gliomatosis cerebri still remains ambiguous. In general, gliomatosis cerebri is a type of glioma associated with diffuse overgrowth within the CNS of neoplastic, usually astrocytomal, glial elements. Nevin (6) originally proposed the term "gliomatosis cerebri" to describe forms of gliomatous neoplasms in the CNS in which the neoplastic elements are very diffuse and widespread in their distribution. Ultrastructural study of the disease has been reported recently (7), indicating that it is a neoplastic process of small undifferentiated elements, transitional forms of astroglia (to oligodendroglia), and anaplastic cells of astrocytic origin in all stages of development.

The photograph of the pathologic specimen of our case appears to show a rather diffuse astrocytic neoplasm of low to intermediate grade. We arrived at a diagnosis of diffuse glioma in this case based on the imaging studies and results obtained from only one area of biopsy. Establishing the extension of neoplastic infiltration along the neuraxis with only one area of biopsy makes precise diagnosis of gliomatosis cerebri difficult.

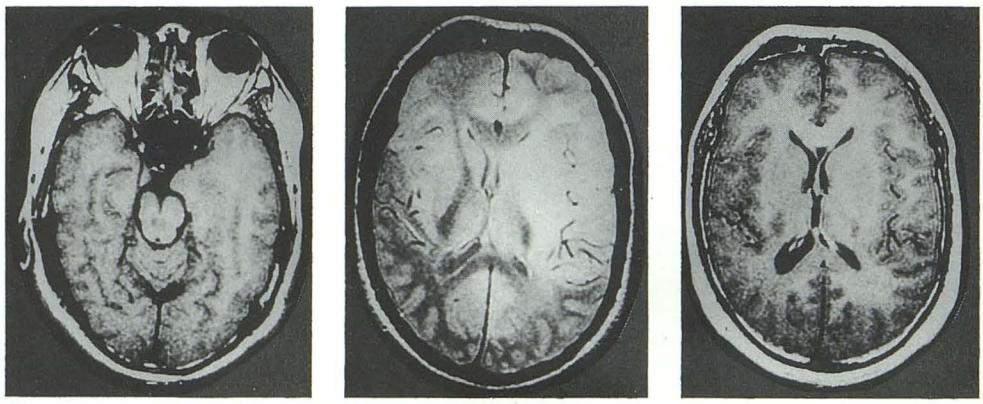
Clinical signs and symptoms of gliomatosis cerebri are usually nonspecific, nonfocal, and disproportionately mild considering the wide extent of parenchymal involvement; these characteristics reflect the diffuse yet generally nondestructive nature of this disease process. The course of disease is most often marked by the development of progressive mental and personality changes, hemiparesis, ataxia, and seizures. Signs of increased intracranial pressure are common late in the course of illness (8). Although all age groups can be affected, this lesion is most commonly observed in the second and fifth decades. It is progressive and its duration can vary from weeks to years (1).

A favorable response to radiation therapy has been reported (9), but long-term control of the disease has not. Radiation therapy was administered to the patient considered here and seemed to be effective, since enlargement of the ventricles was observed on CT following irradiation. This patient was discharged 6 months after onset of illness with moderate disability, but long-term follow-up will be necessary to determine the effectiveness of radiation therapy and immunotherapy.

Fig. 2. Axial MR image (600/30) reveals enlargement of the left temporal lobe with herniation into the prechiasmatic cistern.

Fig. 3 Axial MR image (2500/30) reveals high signal intensity in the left temporal lobe and periventricular and subcortical white matter bilaterally.

Fig. 4 Axial postcontrast-enhanced MR image (600/30) using Gd-DTPA reveals no enhanced tumor mass.



2 3 4

Fig. 5. Microscopic examination of the left temporal lobe revealed a rather diffuse astrocytic neoplasm of low to intermediate grade. Note gemistocytes (*arrows*).

Although the availability of CT has expanded the possibilities of clinical diagnosis, these lesions are diffuse, commonly isodense, and they usually do not enhance following administration of IV contrast medium. Therefore, the diagnosis of gliomatosis cerebri can be only suspected from the presence of clinical signs and an absence of radiologic signs. In the present report, however, we have described a case of diffuse glioma or gliomatosis cerebri in which MR revealed diffuse, contiguous high-intensity areas on T2WI. The findings presumably reflect the prolonged T2 effect of the neoplastic tissue or secondary destruction of myelin fibers because the stereotactic biopsy revealed no tumor cells. Furthermore, changes in the shape of the brain, including enlargement of the temporal lobe, were well-demonstrated on T1WI. The addition of Gd-DTPA failed to demonstrate a focus of enhancement. Rippe (4) reported an enhancement effect after IV administration of gadopentetate dimeglumine for gliomatosis cerebri; however, IV administration of Gd-DTPA proved not to be useful for enhancement of the tumor mass. Brash (10) reported that Gd-DTPA is ideal only in areas where the blood-brain barrier has broken down; therefore, the blood-brain barrier in areas of neoplastic cells may have been preserved in our case.

Our diagnosis in this case is "diffuse glioma," although we suspect it is gliomatosis cerebri. The diagnosis was made with great difficulty, without autopsy, and with biopsy of only one area. Additionally, there was lack of contrast enhancement and clinical symptoms were nonspecific.

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