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AJNR Am J Neuroradiol 1988, 9 (1) 200-202

<http://www.ajnr.org/content/9/1/200.citation>

This information is current as
of August 5, 2025.

Gliomatosis Cerebri Presenting with Hydrocephalus and Dementia

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We report a case of gliomatosis cerebri with massive enlargement of the white matter commissural tracts (including the corpus callosum, the anterior commissure, and the fornices) that may have been directly responsible for obstruction of CSF pathways and subsequent hydrocephalus.

Case Report

A 64-year-old man had no significant medical history until 1 year before he died. At this time he showed signs of excessive forgetfulness, confusion, urinary incontinence, and a shuffling gait that progressed over a 2-week period to inability to ambulate without assist-

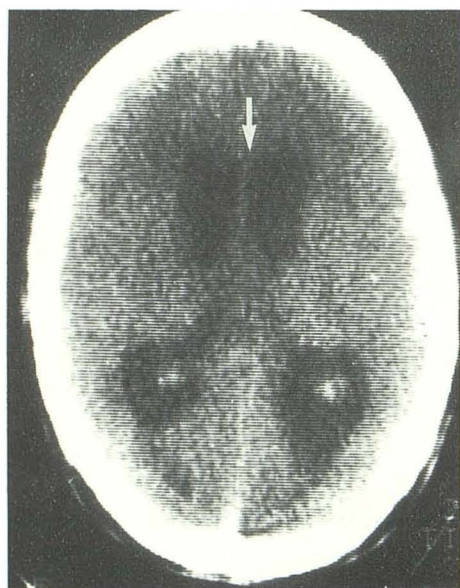


Fig. 1.—Axial CT scan obtained without contrast shows moderate enlargement of lateral ventricles and, in retrospect, thickening of septum pellucidum (arrow).

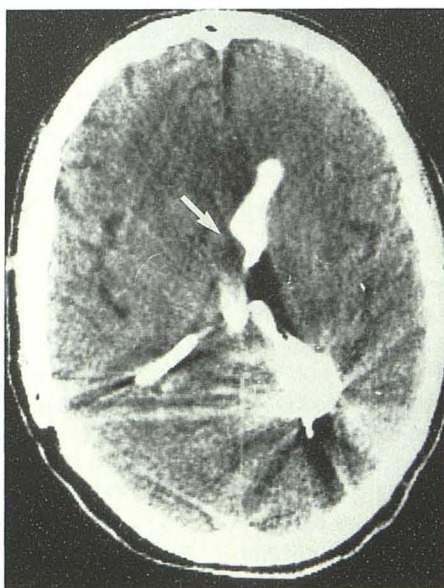


Fig. 2.—CT scan obtained after insertion of catheters into both lateral ventricles. At this time both lateral ventricles are small and cortical sulci have enlarged. When metrizamide was introduced into left shunt, there was free flow of contrast material into third ventricle, but none of it passed the right foramen of Monro, which may have been obstructed by enlarged fornices (arrow).

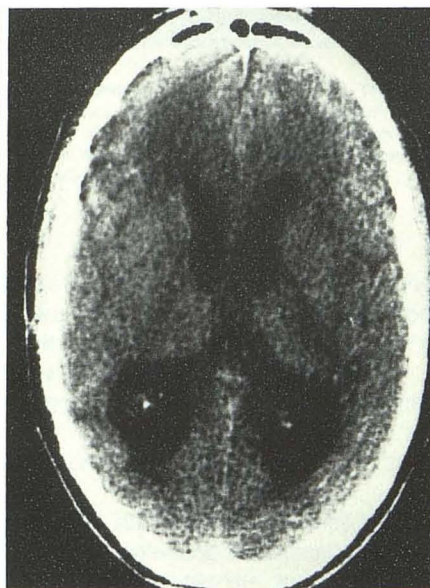


Fig. 3.—Axial CT scan done shortly before the patient died shows recurrence of hydrocephalus and widespread low attenuation of cerebral white matter.

Received October 2, 1985; accepted after revision March 6, 1986.

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ance. Examination revealed diffuse hyperreflexia and hypertonia with gegenhalten and bilateral Babinski signs; he was hypophonic and unable to answer simple questions, write his name, or copy words. A CT scan of the head showed large lateral ventricles, normal-sized third and fourth ventricles, and absence of cortical atrophy (Fig. 1). The CSF was under slightly increased pressure, but was otherwise normal. The EEG showed bilateral frontal slowing with slow and sharp waves in the right frontal area. He improved transiently, then became totally mute after a ventriculoperitoneal shunt was placed in the occipital horn of the right lateral ventricle. A repeat CT scan showed a collapsed, shunt-containing right lateral ventricle, while the left side remained dilated. Little clinical improvement followed place-

ment of a second shunt in the left lateral ventricle despite CT evidence that both ventricles had returned to normal size. Truncation of the anterior third ventricle and prominent fornices were noted on a metrizamide shunt study that demonstrated free flow of contrast material from the left lateral ventricle to the third ventricle, but no filling of the right lateral ventricle (Fig. 2). The shunts were removed because of a persistent staphylococcal shunt infection. The patient died at home 10 months after surgical intervention in a state of profound dementia. A repeat CT scan obtained shortly before he died showed recurrent hydrocephalus and extensive radiolucency of the deep cerebral white matter (Fig. 3).

At postmortem examination, the brain was large (1600 grams).

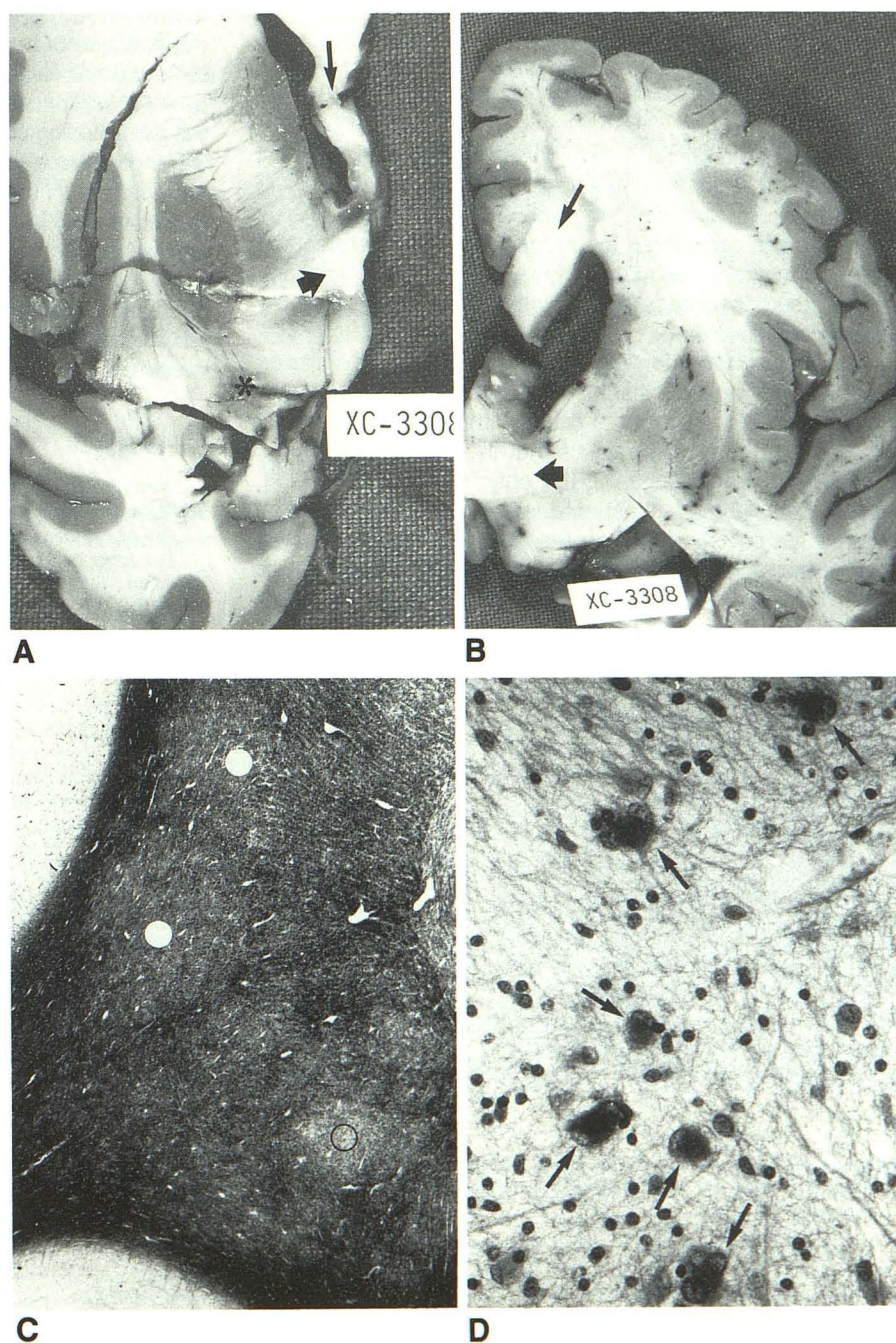


Fig. 4.—A, Coronal section of left hemibrain at level of head of caudate nucleus. Note enlargement and discoloration of the substantia innominata (*asterisk*); anterior commissure (*short arrow*) and septum pellucidum (*long arrow*) are hypertrophied.

B, Coronal section of cerebral hemisphere at level of lentiform nucleus shows massive enlargement of corpus callosum (*long arrow*) and anterior commissure (*short arrow*).

C, Heidenhain's myelin stain of deep cerebral white matter shows patchy areas of staining pallor (*circles*).

D, Immunoperoxidase staining of tissue sections for glial fibrillary acidic protein demonstrates that infiltrating cells are astrocytes, since their cytoplasm is stained (*arrows*).

Coronal sections of the cerebrum revealed slightly dilated lateral and third ventricles with thickening of the corpus callosum, the septum pellucidum, the anterior commissure, and the fornices (Fig. 4). The substantia innominata was expanded and discolored, while the corpus striatum and the thalamus had pale mottled areas. The deep white matter was expanded and firm. The aqueduct of Sylvius at the level of the superior colliculus was reduced to a pinpoint. Microscopically, the deep white matter displayed attenuated staining with myelin stains and extensive proliferation of atypical glial cells, which were immunocytochemically stained by antisera to glial fibrillary acidic protein (Fig. 4). The cells imperceptibly infiltrated the adjoining tissues, displacing and expanding, rather than destroying, normal structures. The substantia innominata was obliterated by the infiltrate, and the periaqueductal tissue was hypercellular. The cerebral cortex had only scattered neuritic plaques and no evidence of neurofibrillary degeneration.

Discussion

Gliomatosis cerebri is a term coined by Nevin in 1938 [1] to describe a condition characterized by diffuse proliferation of pleomorphic glial cells throughout the central nervous system. Nevin considered the process to be a blastomatous proliferation analogous to the diffuse proliferation of Schwann cells in neurofibromatosis. Most observers believe that the process is more akin to a diffuse glial neoplasm [2].

In gliomatosis cerebri the abnormal glia tend to infiltrate and expand the white matter while preserving normal anatomic landmarks. A review of cases with adequate anatomic descriptions of the brain reveals that the corpus callosum is frequently infiltrated [1, 3-7]; the optic chiasm [1, 3, 6, 7], septum pellucidum [4, 6, 7], and fornix [1, 5] are less often involved. Examination of the present case with CT showed encroachment upon the foramen of Monro by thickened pillars of the fornices and a mass in the anterior third ventricle that most likely corresponded to the massively enlarged anterior commissure.

Although the radiologic features of diffuse and multicentric brain tumors are often nondiagnostic [8], the finding of diffuse enlargement of commissural structures, particularly the corpus callosum, may be a useful indicator of gliomatosis cerebri in future cases.*

It should be pointed out that ventricular compression [1, 3, 4, 7], rather than ventricular dilatation [3, 6], is the more common finding in gliomatosis. Dilatation of the lateral ventricles in this case may have been due to obstruction of CSF

flow at the foramen of Monro. The failure of metrizamide to pass freely through the right foramen of Monro tends to support this hypothesis. Hydrocephalus may have been aggravated by stenosis of the aqueduct of Sylvius due to excessive glial proliferation in the midbrain.

The clinical symptoms of gliomatosis cerebri are relatively nonspecific [6]; global intellectual and personality disturbances tend to precede focal neurologic signs. This patient had gait disturbance, dementia, urinary incontinence, and a CT scan that showed dilated ventricles and no cortical atrophy. This clinical picture is highly suggestive of normal pressure hydrocephalus [9]. The radiologic features of normal pressure hydrocephalus are varied, but 30% of patients have normal-sized fourth ventricles [10], as in this case. Furthermore, failure of ventricular decompression to alleviate symptoms did not exclude normal pressure hydrocephalus, since only 40% of patients improve after shunting [9]. At autopsy the usual findings of normal pressure hydrocephalus, such as meningeal fibrosis and hemosiderosis [11], were lacking.

Dementia in this case may have been multifactorial. Although there was a small number of neuritic plaques in the cortex, they were within normal limits for the patient's age and could not have accounted for dementia. On the other hand, tumor infiltration of the limbic system and the substantia innominata, as well as hydrocephalus, per se, adequately account for progressive dementia.

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* A contrast-enhanced CT scan of our patient (not shown) revealed no abnormal areas of enhancement, adding to the diagnostic difficulty.