Diffuse Leptomeningeal Gliomatosis with Osteoblastic Metastases and No Evidence of Intraaxial Lesions

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Summary: An autopsy-proved case of cerebral and medullary leptomeningeal gliomatosis and diffuse osteoblastic metastases without evidence of intraaxial tumor is described. MR findings included diffuse thickening of the cerebral and medullary leptomeninges on T1-weighted, proton density–weighted, and T2-weighted images and abnormal enhancement of the sulci and cisterns of the cerebrum, brain stem, cerebellum, and medulla on postcontrast T1-weighted images. MR also showed several areas of replacement of the normal bone marrow of the skull. No intraaxial lesion was seen.

Index terms: Meninges, neoplasms; Brain, neoplasms; Gliomatosis

Diffuse leptomeningeal gliomatosis is a rare condition usually caused by a primary intraaxial lesion (1), although cases without underlying glial tumor have been reported (2–5). Glial tumors can metastasize outside the central nervous system (6, 7). This is rare and occurs most often after surgical procedures.

We report a case of cerebral and medullary leptomeningeal gliomatosis and diffuse osteoblastic metastases without evidence of intraaxial tumor. No primary intraaxial lesion was found at autopsy.

Case Report

A 19-year-old man was admitted to the hospital after 4 weeks of headache, nausea, and vomiting. The fundi of the eyes revealed bilateral papilledema; neurologic examination was otherwise normal. Examination of the cerebrospinal fluid showed no evidence of neoplastic cells. Computed tomography of the brain before admission was reported to be normal.

Magnetic resonance (MR) imaging showed diffuse thickening and abnormal enhancement of the leptomeninges on postcontrast T1-weighted images of the brain, cerebellum, brain stem, and medulla (Fig 1A–D). No intraaxial lesion was seen. The examination also showed multiple areas of replacement of normal bone marrow of the skull (Fig 1B) that appeared as osteoblastic lesions on skull radiographs (Fig 1E).

A bone survey demonstrated focal areas of sclerosis involving vertebral bodies, ribs, pelvis, and limbs. Bone scintigraphy showed multiple foci of hyperactivity, corresponding to the osteoblastic areas seen on plain films. All of the radiologic studies performed to look for a possible primary neoplasm in other parts of the body were normal. Blood tests and bone and bone marrow biopsies also were normal.

Forty days after admission, the neurologic state of the patient rapidly deteriorated, with signs of diencephalic and brain-stem impairment. He died after 50 days of hospitalization. At autopsy, diffuse thickening and opacification of the leptomeninges of the cerebrum, cerebellum, brain stem, and medulla were found. Leptomeningeal thickening was most pronounced about the basal cisterns and the medulla. A whitish gray material obliterated the cisterns and subarachnoid space around the medulla. No intraaxial tumor was found. Findings on general gross autopsy were unremarkable.

On microscopic examination, the leptomeninges were diffusely infiltrated by small, mainly monomorphic astroglial cells (malignant astrocytoma). These were confined to the leptomeninges, without extending into the parenchyma. Immunohistochemical studies (glial fibrillary acidic protein) demonstrated the glial nature of the leptomeningeal tumor and bone metastases (Fig 2A and B).

Discussion

Diffuse glial leptomeningeal involvement is a rare condition that usually results from the diffusion of a primary intraaxial lesion (1). In most of the reported cases, the primary tumor could be diagnosed during life or was discovered at autopsy (6). In reports describing diffuse leptomeningeal gliomatosis without underlying pa-

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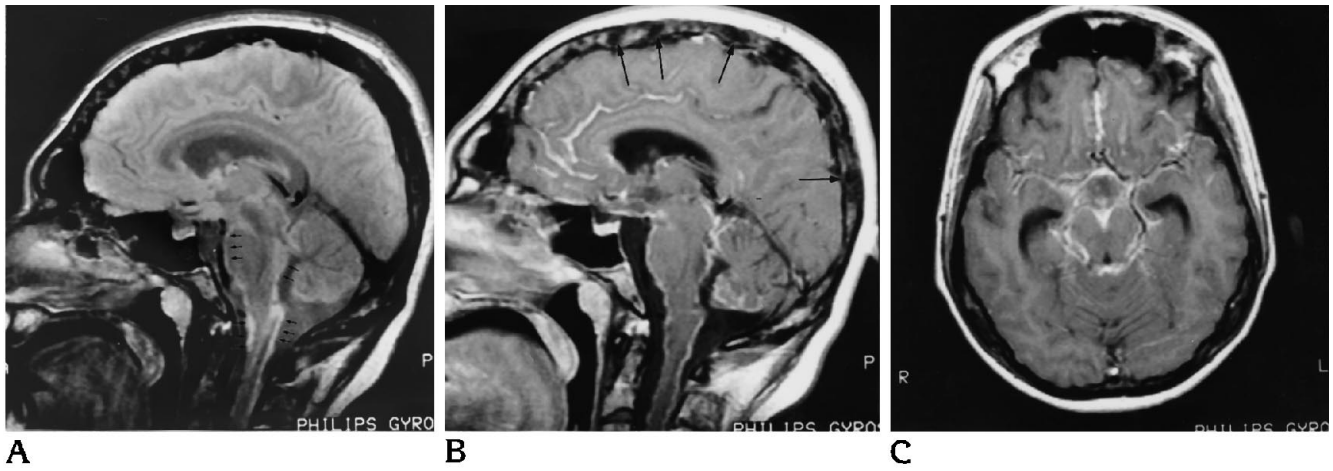

Fig 1. *A*, Sagittal proton density–weighted MR image (2179/30/1 [repetition time/echo time/excitations]) on admission. Leptomeningeal thickening is seen as diffuse hyperintensity surrounding the upper cervical cord and the brain stem (*arrows*).

B, Sagittal contrast-enhanced MR image (600/20/2) shows diffuse enhancement of the sulci and cisterns of the cerebrum, brain stem, cerebellum, and medullary surface. Multiple hypointense areas replacing the normal bone marrow of the skull are also evident (*arrows*).

C, Axial contrast-enhanced MR image (600/20). Sulcal and cisternal enhancement is particularly evident about the basal cisterns.

D, Axial T2-weighted MR image (2179/90/1) at the same level as *C* shows no evidence of intraaxial lesion. *E*, Plain radiograph, lateral view, shows multiple osteoblastic areas of the skull (*arrows*).

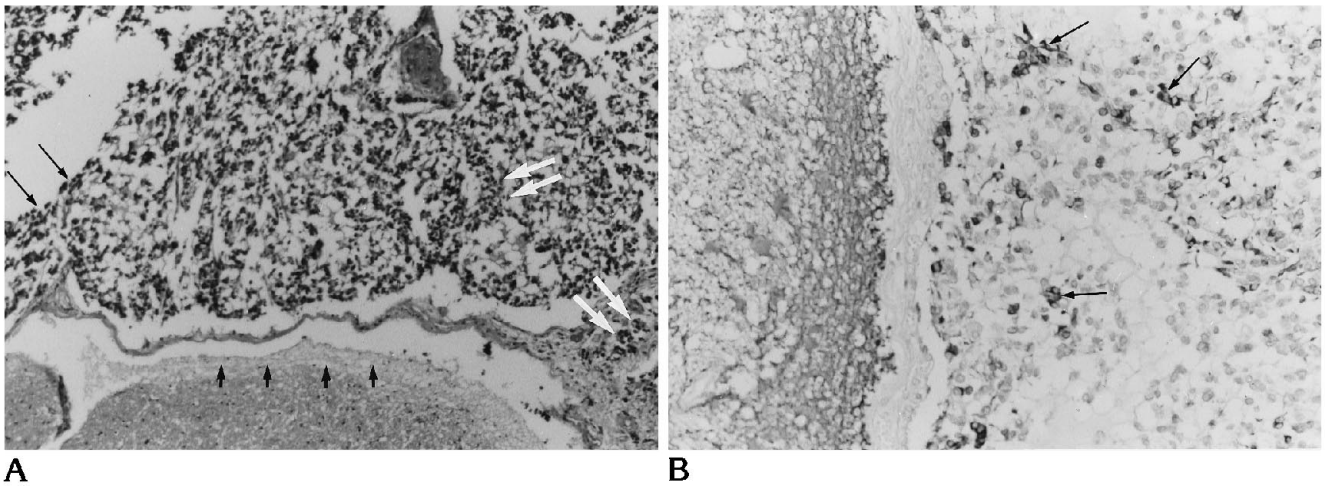

Fig 2. Photomicrographs (hematoxylin-eosin stain, *A*; glial fibrillary acidic protein, *B*) show that the leptomeninges are infiltrated by small, mainly monomorphic astroglial cells (*long arrows*) sparing the cerebral cortex (*short arrows*).

renchymal tumor (2–5), it has been postulated that the presence of heterotopic glial tissue might be the source for leptomeningeal gliomatosis (4, 8). It is well recognized that glial tumors can metastasize to bone. Extraneural metastases have been described after surgical procedures (9), which are thought to increase the risk of dissemination. Bone metastases are usually limited to one or a few sites (10) and are most often osteolytic (9). Diffuse osteoblastic metastases are rare but have been described (9, 10).

In our case, the presence of osteoblastic lesions suggested the neoplastic nature of the leptomeningeal thickening that was otherwise difficult to characterize on clinical grounds alone. As in the report of Whelan et al (5), we found no neoplastic cells in the cerebrospinal fluid. The absence of other primary tumors in the body raised the suspicion of leptomeningeal gliomatosis, but, as in other cases, the clinical course was rapid and we made the diagnosis only at autopsy.

This case serves as a reminder that diffuse leptomeningeal thickening of a glial nature can occur in the absence of a primary intraaxial tumor.

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