Gliomatosis cerebri

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Section: Neuroradiology
Imaging Technique: MR
Case Type: Clinical Cases
Authors: K. Khalatbari, H. Yilmaz, A. Ibrahim, P. Dardel, JC. Froment
Patient: 34 years, male

Clinical History:

The patient presented after three episodes of left partial motor epileptic seizures.

Imaging Findings:

The patient was referred after three episodes of left partial motor epileptic seizures. A brain MRI was performed, which demonstrated findings compatible with a diagnosis of gliomatosis cerebri. This diagnosis was confirmed histologically via a brain biopsy specimen.

Discussion:

Gliomatosis cerebri (GC) is a rare form of the commonest brain tumour, namely the glioma. The term was first used by Nevin in 1938. The WHO classification of brain tumours recognises GC as a distinct clinicopathological entity among the neuroepithelial tumours of uncertain origin; the criterion for diagnosis is considered to be involvement of at least two lobes of the brain by small elongated cells without a cellular, centrally necrotic centre.

The incidence of GC is greatest in the third to the fifth decades (although there is one case report of GC in a newborn). The disease is usually slowly progressive, although its clinical duration can vary from a few weeks to more than 20 years. Nonspecific personality and mental changes are the most frequent clinical manifestations, and clinical findings are disproportionate to the extent of the brain involvement, which may relate to the diffuse but generally nondestructive nature of the disease process. GC has a poor prognosis, and in the longterm, no treatment has been proved effective.

MRI is significantly more sensitive than CT in both the diagnosis of GC and in determining its true extent. Pathological specimens derived from MRI-guided stereotaxic biopsies, correlated with clinical and imaging findings, may be of help in reaching a correct antemortem diagnosis; however, even with multiple biopsy specimens, the antemortem diagnosis of GC still remains difficult.

CT studies in these patients generally show, iso- to hypodense lesions, with a more or less diffuse mass effect, and minimal or no contrast enhancement; a definable mass is not present. Lesions that appear subtle or are not apparent on CT may be identified by MRI, as poorly defined iso- or hypointense areas on T1-weighted images, and hyperintense areas on T2-weighted images. Abnormalities are frequently observed in the basal ganglia, thalamus and the hypothalamus, usually with infiltration of the process along the anatomic white matter pathways. Commissural structures, such as the corpus callosum, are frequently affected and expanded. Subpial and cortical extension of the disease may also be observed. Focal areas of contrast enhancement may be noted, presumably in areas of more
dense infiltration of tumour cells. Although diffuse infiltrative tumoral processes that are associated primarily with focal centres or masses (most likely the origins of the lesions) are better termed diffuse gliomas, focal malignant transformation of a GC lesion can occur, along with the appearance of focal neurological signs. Development of mass, necrosis and contrast enhancement may parallel the clinical deterioration in such cases.

The following entities are included in the radiological differential diagnosis of this disorder: multiple sclerosis, leukodystrophy, ischaemic lesions, infiltrative conventional gliomas and inflammatory or infectious processes.

**Differential Diagnosis List:** Gliomatosis cerebri

**Final Diagnosis:** Gliomatosis cerebri

**References:**


**Description:** Axial T1-weighted SE image (without contrast) obtained at the level of the basal ganglia, demonstrates an infiltrative process predominantly involving the right temporal lobe; the ipsilateral basal ganglia and internal capsule are also affected. The process extends into the splenium of the corpus callosum, and exerts a mild mass effect (demonstrated as obliteration of the ipsilateral sulci and compression of the right lateral ventricle), on the adjacent structures. **Origin:**
Description: Axial T1-weighted SE image (with contrast) obtained at the level of the basal ganglia, demonstrates an infiltrative process predominantly involving the right temporal lobe; the ipsilateral basal ganglia and internal capsule are also affected. The process extends into the splenium of the corpus callosum, and exerts a mild mass effect (demonstrated as obliteration of the ipsilateral sulci and compression of the right lateral ventricle), on the adjacent structures. No enhancement is observed on this post-contrast image. Origin:
Description: Axial T2-weighted TSE image obtained at the level of the basal ganglia, demonstrates an infiltrative process (hyperintense on this T2-weighted image) predominantly involving the right temporal lobe; the ipsilateral basal ganglia and internal capsule are also affected. The process extends into the splenium of the corpus callosum, and exerts a mild mass effect (demonstrated as obliteration of the ipsilateral sulci and compression of the right lateral ventricle), on the adjacent structures. Origin:
**Description:** Axial FLAIR MR image obtained at the level of the basal ganglia, demonstrates an infiltrative process (hyperintense on this FLAIR image) predominantly involving the right temporal lobe; the ipsilateral basal ganglia and internal capsule are also affected. The process extends into the splenium of the corpus callosum, and exerts a mild mass effect (demonstrated as obliteration of the ipsilateral sulci and compression of the right lateral ventricle), on the adjacent structures. **Origin:**