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GLIOMATOSIS CEREBRI: A CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

Gliomatosis cerebri is a rare and aggressive tumor characterized by diffuse infiltration of the central nervous system, with a highly lethal course. Its clinical presentation is insidious, with gradual neurological decline, intracranial hypertension and focal deficits. The diagnosis of cerebral gliomatosis relies on clinical, radiological and histological findings. The prognosis remains poor due to late diagnosis and extensive brain involvement.

INTRODUCTION

Gliomatosis cerebri is a rare disorder affecting young people. It is defined by diffuse and bilateral infiltration of the brain, sometimes the spinal cord, by glial cells.

The histological diagnosis of gliomatosis cerebri, due to its uncircumscribed nature, is often difficult both for diagnosis and for determining the nature of the tumor cells.

The aim of this study is to illustrate the diagnostic and prognostic aspects, as well as the therapeutic management of this rare pathology through a new observation.

We report the case of an 9 year old boy admitted to the Pediatrics Department of Military Hospital Mohamed V of Rabat for resistants headache and balance disorders.

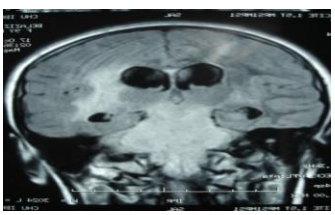
OBSERVATION

The patient is a 9-year-old child with good psychomotor development and no specific medical history had been presenting for one month prior to the examination with occipital headaches resistant to usual analgesic treatment and insidious onset of balance disorders.

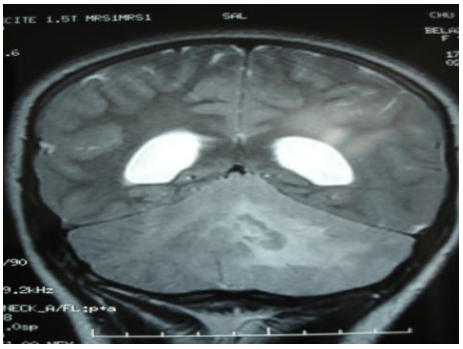
The clinical examination revealed subtle phasic disturbances, decreased muscle strength, normal osteoarticular reflexes, and a cerebellar statokinetic syndrome.

The funduscopy showed bilateral papilledema.

The brain MRI showed diffuse subcortical areas that were hyperintense on T2-weighted images and iso- or hypointense on T1-weighted images, affecting the hemispheric and cerebellar white matter, the splenium, the corpus callosum, and the basal ganglia, with no circumscribed masses and no enhancement after gadolinium injection.



Brain MRI T2 Flair sequence



Brain MRI T2 weighted

The spectrometry demonstrated a moderate increase in the choline/creatine and choline/ N-acetylaspartate ratios (>2.5), creatine and myoinositol, and a decrease in the N-acetylaspartate /Creatine ratio.

The stereotaxic biopsy led to the diagnosis of cerebral gliomatosis in its astrocytic form.

DISCUSSION

The World Health Organization defines gliomatosis cerebri as a high-grade infiltrating glial lesion, affecting at least two lobes.

Clinically, there are no specific signs. The onset of the disorder is gradual, with mental deterioration, personality disorders, and intracranial hypertension. Focal neurological signs appear secondarily. Neurological involvement is not proportional to radiological involvement. Currently, the diagnosis of gliomatosis cerebri is difficult and relies on a combination of morphological, histological, and radiological (MRI spectroscopy) criteria.

Histologically, gliomatosis cerebri is characterized by infiltration of the white matter by often polymorphic cells (astrocytes or, less often, oligodendrocytes), arranged in a "single file" fashion, which can induce demyelination through destruction of axonal myelin. The mitotic index is variable, and vascular proliferation is generally absent. The cytological characterization of gliomatosis cerebri (astrocytic or oligodendrocytic) has a prognostic interest since cerebral gliomatoses of oligodendroglial morphology have a more favorable prognosis than those of the astrocytic or mixed type.

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Morphologically, the tumor infiltration is diffuse and extensive, affecting more than two cerebral lobes, frequently bilateral, with possible infratentorial or even spinal cord extension. The basal ganglia, corpus callosum, thalamus, hypothalamus, and brainstem may be involved.

MRI spectroscopy provides the following additional evidence

– N-acetyl-aspartate (NAA): a marker of neuronal integrity, is decreased.

– creatine (Cr): a reflection of the energy metabolism of normal brain tissue, is increased.

– myoinositol (Ino): a marker of glial activation, is increased.

Creatine and myoinositol peaks are higher in astrocytic infiltrations compared to oligodendroglial infiltrations ($p < 0.01$).

– choline (Cho): linked to membrane activity, is more elevated.

– A Cho/NAA ratio > 2.5 would suggest high grade.

Due to its diffuse and therefore inoperable nature, the gliomatosis cerebri is treated with chemotherapy alone as a first-line treatment (nitrosourea or the combination of procarbazine-CCNU-vincristine or temozolomide, which has the advantage of better tolerance).

A specific genetic profile (loss of chromosomes 1p and 19q) appears to respond better to chemotherapy in oligodendrocyte gliomatosis cerebri.

CONCLUSION

Gliomatosis cerebri is a rare form of glial tumor with an insidious onset. It essentially poses a prognostic problem due to its often late diagnosis and its inoperability.

AKNOWLEDGMENTS

No conflict of interest.

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