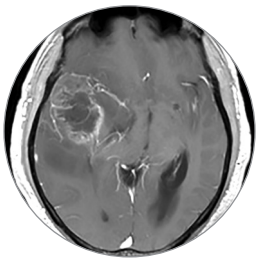

Gliosarcoma: Case Report and Findings by Tomography andConventional and DTI Magnetic Resonance Imaging

Gliosarcoma: presentación de un caso y hallazgos por tomografía y resonancia magnética convencional y tractografíag

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Summary

Gliosarcoma is a rare and highly malignant central nervous system tumor. It is classified by the WHO as a variant of glioblastoma (grade IV) and has a poor prognosis. Histologically it is characterized by having both glial and mesenchymal components. Clinically, it varies depending on the location and size of the tumor, the most frequent symptoms being seizures, headaches and focal neurological deficit. The initial diagnostic approach is computed tomography, which provides suspicionus data; however, magnetic resonance is the diagnostic pillar, providing important data that becomes more significant with the use of functional sequences such as tractography. A clinical case is presented with a literature review and the most significant findings in the imaging studies.

Resumen

El gliosarcoma es un tumor raro del sistema nervioso central y de alto grado de malignidad. La OMS lo clasifica como variante del glioblastoma (grado IV) y es de mal pronóstico. Histológicamente se caracteriza por tener componentes gliales y mesenquimatosos. El cuadro clínico varía dependiendo de su localización y tamaño, los signos y síntomas más frecuentes son convulsiones, cefalea y déficit neurológico focal. El acercamiento diagnóstico inicial es la tomografía computarizada que aporta datos de sospecha; sin embargo, la resonancia magnética constituye el pilar diagnóstico, con importantes elementos de diagnóstico que se vuelven más significativos con el uso de secuencias funcionales como la tractografía. Se presenta un caso clínico con revisión de la literatura y los hallazgos más significativos en los estudios de imagen.

Introduction

Gliosarcoma was described in 1805 by Strobe. It is a rare, highly malignant tumor with a poor prognosis.

It is considered a grade IV tumor by the WHO, its main characteristics are: rapid growth, high mitotic rate, neoplastic blood vessels, as well as areas of necrosis. Histologically, it is characterized by having both glial and mesenchymal components, the latter due to an angiogenic stimulus that generates microvascular hyperplasia, which gives it a biphasic or compound nature (1, 2).

Clinically, the symptomatology is variable depending of the location and size of the tumor. The Most frequent symptoms are: convulsions, headache, focal neurological deficits and those related to increased intracranial pressure.

The initial diagnostic approach is the tomography computerized tomography (CT); magnetic resonance imaging (MRI) constitutes the diagnostic pillar, since it provides data important that become more significant with the use of functional sequences such as tractography that generates information about its relationship with the brain tracts; some kind of affection is observed in the path of the same (3).

Clinical case

65-year-old male patient with genetic load for diabetes mellitus and high blood pressure from his parents. Positive smoking rate of 18 packets per year, systemic arterial hypertension of 7 years of evolution in medical treatment and obstructive prostatic hyperplasia with surgical treatment. He started suffering three months before his admission with a picture characterized by frontal headache of oppressive type of moderate intensity, 5/10 in analogical visual scale (EVA) that remits spontaneously, with progression to incapacitating headache associated to nausea, for

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which he was taken twice to his general hospital of area and received symptomatic treatment, without improvement.

Two days prior to his admission, he presented a picture of paresis and paresthesia of the left hemicorphin.

He was evaluated by the neurosurgery service, who in the neurological exploration found preserved mental functions, with lateralization of the march to the right and decrease of the muscular force of the left hemicorpus, for which a cranial CT scan was requested as an initial diagnostic approach (figure 1). In view of the findings, MRI was performed (Figure 2) as a diagnostic complement and for surgical planning with additional tractography sequence (Figure 3).

Surgical treatment with extensive complete resection is decided upon and a surgical piece is sent to the pathological anatomy service with the final result of grade IV gliosarcoma according to the WHO (Figure 4).

The patient's progress is favorable and he is discharged.

A control MRI was performed (Figure 5) 47 days after the surgery, which showed significant tumor remnant.

Discussion

Tumors of the central nervous system represent 2% of all neoplasms. They constitute a heterogeneous group of neoplasms that include from well differentiated and benign lesions, to highly invasive and undifferentiated, such as glioblastoma Recently, researchers from The Cancer Genome Atlas (TCGA) established the existence of four subtypes of glioblastoma (1).

The World Health Organization, in its classification of tumors of the central nervous system, integrates gliosarcoma as a Glioblastoma variant, grade IV (high degree of malignancy). The Characteristics of tumors of this grade are: rapid growth, high mitotic rate with neoplastic vessels and areas of necrosis, which which gives it a bad prognosis (2).

They are rare tumors, representing 2 to 8% of glioblastomas and about 0.48% of all intracranial tumors.

The natural history, clinical presentation and radiological profile are similar to those of the primary glioblastoma.

Due to their infiltrating character, they recur in up to 90%. Within the histological aspects that define gliosarcoma, a bimorphism of glial and mesenchymal elements is recognized. The glial areas usually correspond to glioblastoma and very rarely may have areas of squamous or glandular differentiation with trabecular patterns, adenoid and even papillary structures. As for the component mesenchymal, this is phenotypically that of a fibrosarcoma (3).

The symptoms caused by a primary tumor of the central nervous system are divided into two large groups: focal symptoms and widespread symptoms. The most frequent in the presentation of a Rapidly growing tumors such as gliosarcoma, are headache, nausea or vomiting, as well as seizures.

Gliosarcoma is a tumor frequently located in the temporal lobe, because it has a greater relationship in the clinical picture; other locations are the frontal, parietal and occipital lobes (4).

The diagnostic approach, after a clinical suspicion, requires a forced evaluation with an image study. Neuroimaging studies provide important information that reduce the diagnostic possibilities and point to a probable etiology, although the definitive diagnosis is given by the histopathological study. Another important issue regarding imaging studies is the usefulness for surgical planning (5).

CT is useful as an initial diagnostic approach; it usually identifies the lesion, its location, its morphology, and evaluates if there is bone infiltration. It is especially helpful when there is an urgent need to obtain the image or when the patient has a contraindication for MRI.

MRI is composed of different sequences that provide information capable of reducing diagnostic options, and is the study of choice for a central nervous system tumor.

Generally, high-grade gliomas typically show up with low signal in T1-weighted sequences and high signal in T2-weighted sequences. in T2, heterogeneously and predominantly peripheral with the paramagnetic contrast medium; they produce mass effect and significant perilesional edema; usually have restricted diffusion because of their high cellularity (6).

Spectroscopy has been used in the last years; it allows to improve the differentiation of infiltrating tumors through the analysis of the tumor metabolites; N-acetylaspartate (NAA) is a marker of the neuronal cellular integrity that is diminished in high-grade gliomas, including gliosarcoma; the hill, which is a component of the cell membranes, increases and lactate rises when there is necrosis (7).

Tractography is an image-based sequence of the diffusion tensor (DTI) that uses the same principle of diffusion imaging. In patients with brain tumors it is a fundamental sequence for distinguishing the surrounding white matter tracts, and it allows distinguishing the spatial relationship between the tumor boundary and the white matter by visualizing the fibers.

It is a very useful sequence for preoperative planning of the tumor resection, it evaluates its relationship with fibers related to vital brain functions, such as motor, sensory, auditory, visual or language. In this way it has become an integral part of the preoperative diagnosis in many neurosurgical centers (8, 9).

The relationship between the tracts and tumors is classified into three types: type I or simple displacement, type II displacement with interruption and type III simple interruption; it allows the visualization of the relationship of the tumors with eloquent tracts, and in the case presented it was beneficial in the neurosurgical planning and postoperative evaluation (8).

Currently there are many options for the treatment of high-grade glial neoplasms that revolve around a combination of surgery, radiotherapy and chemotherapeutic agents.

T1-weighted MRI images with contrast medium are the mainstay for the diagnosis of residual/recurrent brain tumors.

They are easy to perform sequences and accurately evaluate the margins of most primary and secondary brain tumor lesions (10).

Surgical therapy evaluation often begins with an imaging scan to determine the extent of resection which should ideally be performed within the first 24 hours.

Although a mild reactive enhancement may be seen after the surgical event, the nodular or similar enhancement to the initial lesion within 24 hours usually represents a residual tumor; however, distinguishing the residual tumor from a reactive change is often difficult.

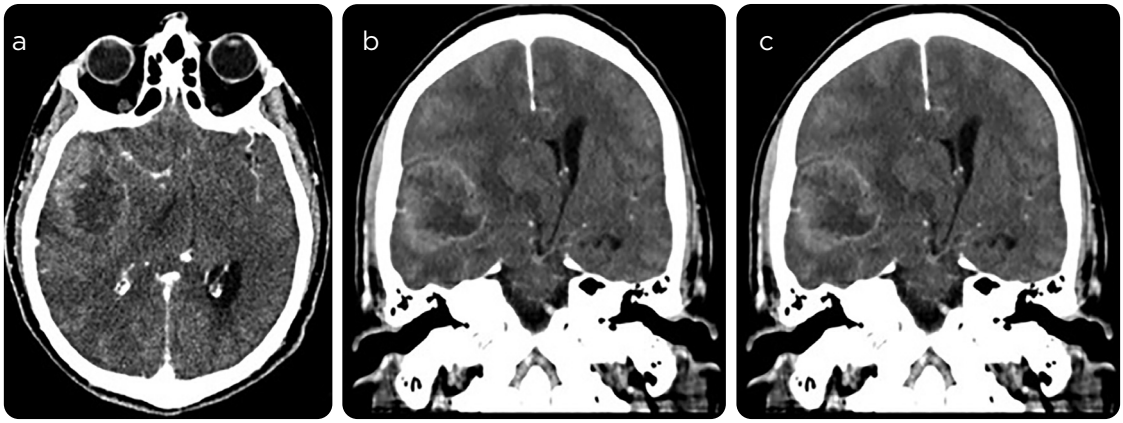


Figure 1. Skull CT with contrast medium. a) Axial section with reconstructions b) coronal and c) sagittal. Right temporal intraaxial lesion, amorphous, of low density in relation to the cerebral parenchyma with dimensions of 35 × 50 × 35 mm in its longitudinal, anteroposterior and transverse axes. It shows peripheral enhancement and mass effect on the frontal lobe, with decrease in width and depth of the grooves and clefts, the frontal shaft and body of the lateral ventricle, the basal ganglia and the ipsilateral thalamus, with deviation of the midline of up to 15 mm.

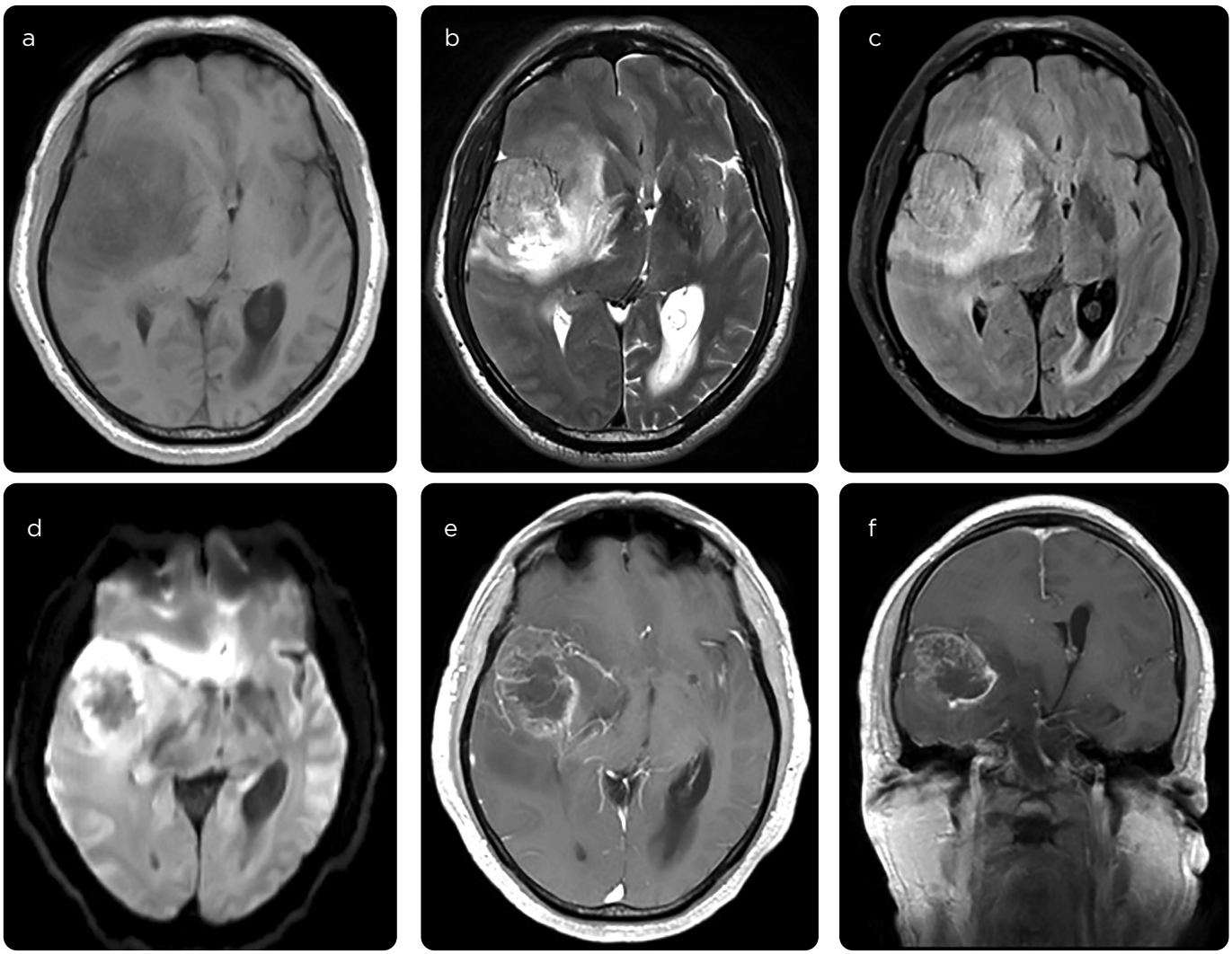


Figure 2. MRI of conventional brain. a) T1-weighted cross sections, b) T2, c) FLAIR sequence, d) diffusion sequence, e) T1-weighted sequence with contrast medium and f) T1-weighted coronal sequence with contrast medium: Supratentorial and intraaxial right temporal lesion, oval, 35 × 50 × 35 mm in its longitudinal, anterior-posterior and transverse axes, heterogeneous predominantly low signal in T1-weighted sequences and high signal in T2/FLAIR-weighted ones. In diffusion sequence: areas of peripheral location restriction and after administration of the peripheral enhancement contrast medium. It is associated with significant perilesional edema and mass effect on the frontal lobe, frontal shaft and body of the lateral ventricle, basal ganglia and thalamus, with contralateral midline deviation of up to 15 mm.

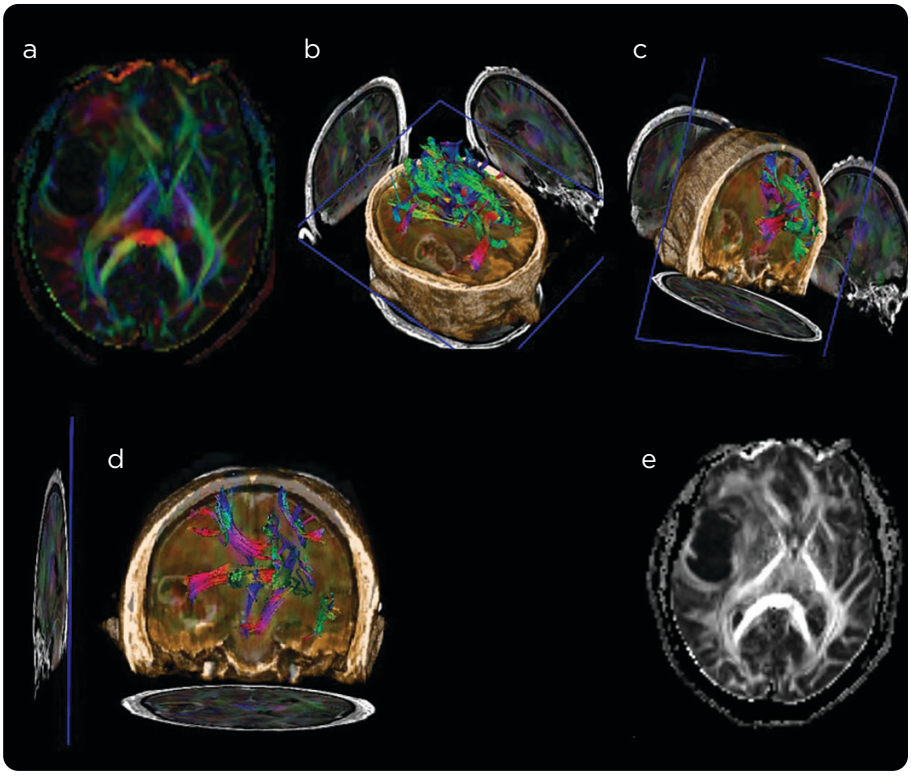


Figure 3. MRI of the brain with tractography. a) In cross section, b) transverse volumetric reconstructions, and c and d) coronal, e) tractography source image.

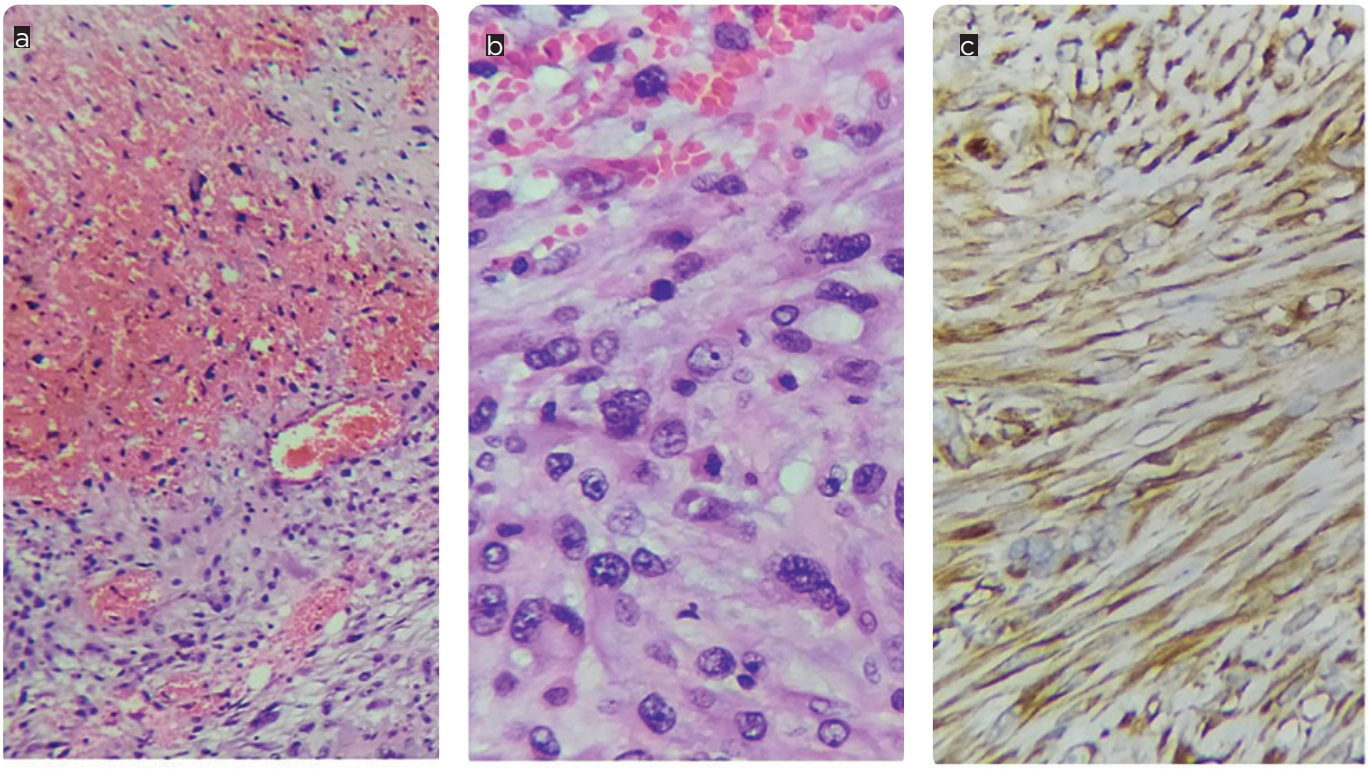
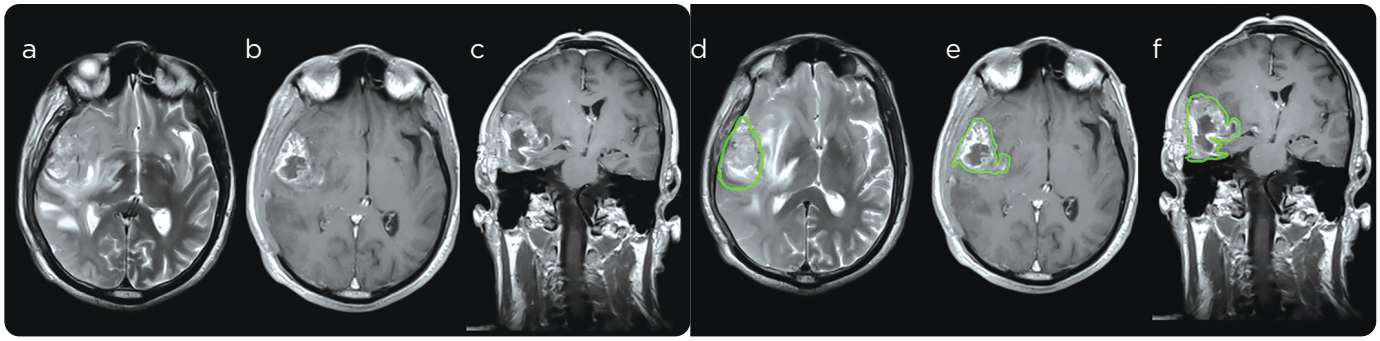


Figure 4. Histological sections show a hypercellular neoplasm with atypical proliferation of malignant looking pleomorphic astrocytes. There are areas of focal necrosis, vascular proliferation and mitotic activity. Some areas simulate a typical glioblastoma; however, there are spindle cells and atypical nuclei. With vimentin-positive immunostaining, the mesenchymal component of the neoplasm is confirmed.



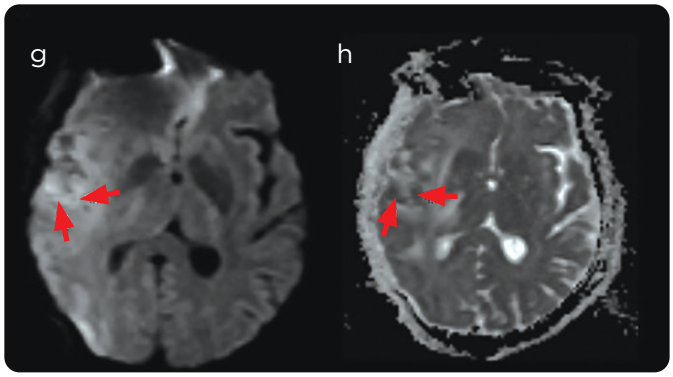


Figure 5. Post-surgical control brain MRI. a) T2-weighted cross sections, b) T1-weighted with contrast medium, c) coronal reconstruction, d) diffusion sequence and e) ADC map: surgical changes by right pterional craniectomy. In the surgical bed in the right temporal lobe, there is a tumor remnant of irregular morphology, predominantly hypointense with respect to the parenchyma in T1-weighted sequences and hyperintense in T2-weighted sequences. It shows reactive peripheral, gyral and meningeal enhancement and vasogenic edema associated with medial displacement of the base ganglia with partial collapse of the lateral ventricle on the same side. In sequence of diffusion and ADC map: restriction to diffusion (high signal in diffusion and low in ADC map) of the edges of the lesion showing enhancement.

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There are other sequences that can help when diagnosing recurrent brain injury. T2/FLAIR-weighted images may be useful in differentiating between unenhanced residual tumor areas and post-surgical reaction. This sequence shows areas of abnormal signal intensity and extension, although in high gliomas grade may not reliably differentiate the infiltrating tumor from vasogenic edema, as both are shown to be high signal.

The diffusion-weighted images provide information on water diffusivity. Many pathophysiological processes result in diffusion restriction, as occurs in high-grade glial neoplasms; therefore, it is important to correlate any new enhancements with diffusion-restricted areas that may correspond to tumor recurrence or progression.

Other useful sequences for the evaluation of recurrence or progression are perfusion and spectroscopy.

Individually, none of these have been shown to be very specific; however, together, these sequences make it possible to diagnose residual tumor and differentiate it from changes secondary to oncological treatment (11).

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