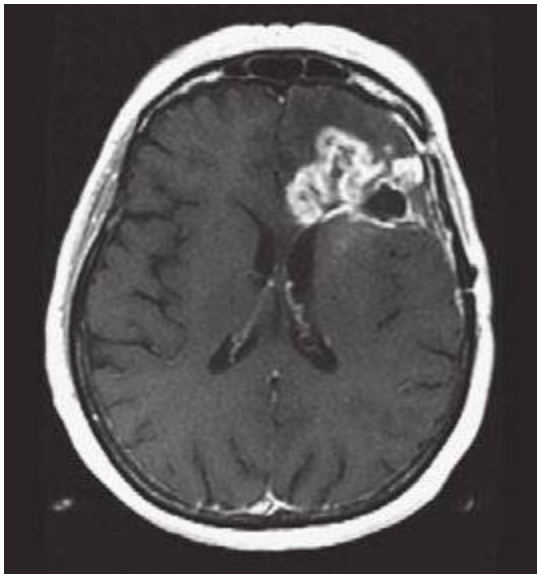
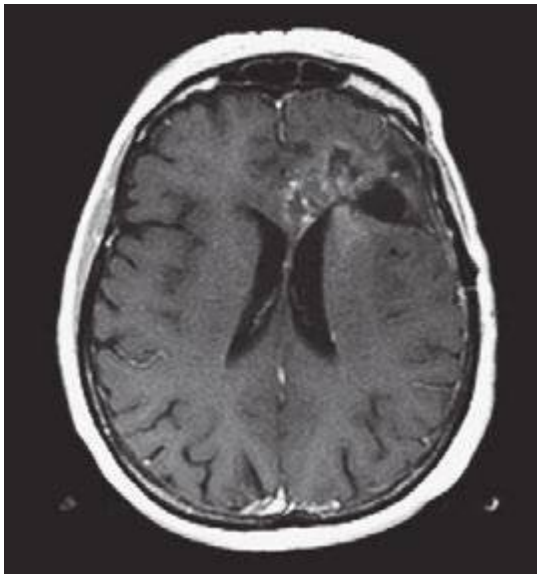


A



B

FIGURE 118-3 Postgadolinium T1 MRI of a recurrent glioblastoma before (***A***) and after (***B***) administration of bevacizumab. Note the decreased enhancement and mass effect.

features such as perinuclear clearing—giving rise to a "fried-egg" appearance—and a reticular pattern of blood vessel growth. Some tumors have both an oligodendroglial as well as an astrocytic component. These mixed tumors, or oligoastrocytomas (OAs), are also classified into well-differentiated OA (grade II) or anaplastic oligoastrocytomas (AOAs) (grade III).

Grade II oligodendrogliomas and OAs are generally more responsive to therapy and have a better prognosis than pure astrocytic tumors. These tumors present similarly to grade II astrocytomas in young adults. The tumors are nonenhancing and often partially calcified. They should be treated with surgery and, if necessary, RT and chemotherapy. Patients with oligodendrogliomas have a median survival in excess of 10 years.

AOs and AOAs present in the fourth and fifth decades as variably enhancing tumors. They are more responsive to therapy than grade III astrocytomas. Co-deletion of chromosomes 1p and 19q, mediated by an unbalanced translocation of 19p to 1q, occurs in 61–89% of patients with AO and 14–20% of patients with AOA. Tumors with the 1p and 19q co-deletion are particularly sensitive to chemotherapy with procarbazine, lomustine (cyclohexylchloroethylnitrosourea [CCNU]), and vincristine (PCV) or temozolomide, as well as to RT. Median survival of patients with AO or AOA is approximately 3–6 years, but those with co-deleted tumors can have a median survival of 10–14 years if treated with RT and chemotherapy.

EPENDYMOMAS

Ependymomas are tumors derived from ependymal cells that line the ventricular surface. They account for approximately 5% of childhood tumors and frequently arise from the wall of the fourth ventricle in the posterior fossa. Although adults can have intracranial ependymomas, they occur more commonly in the spine, especially in the filum terminale of the spinal cord where they have a myxopapillary histology. Ependymomas that can be completely resected are potentially curable. Partially resected ependymomas will recur and require irradiation. The less common anaplastic ependymoma is more aggressive and is treated with resection and RT; chemotherapy has limited efficacy. Subependymomas are slow-growing benign lesions arising in the wall of ventricles that often do not require treatment.

OTHER LESS COMMON GLIOMAS

Gangliogliomas and pleomorphic xanthoastrocytomas occur in young adults. They behave as more indolent forms of grade II gliomas and are treated in the same way. Brainstem gliomas usually occur in children or young adults. Despite treatment with RT and chemotherapy, the prognosis is poor, with a median survival of only 1 year. Gliosarcomas contain both an astrocytic as well as a sarcomatous component and are treated in the same way as glioblastomas.

PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMA

Primary central nervous system lymphoma (PCNSL) is a rare non-Hodgkin lymphoma accounting for less than 3% of primary brain tumors. For unclear reasons, its incidence is increasing, particularly in immunocompetent individuals.

PCNSL in immunocompetent patients usually consists of a diffuse large B cell lymphoma. PCNSL may also occur in immunocompromised patients, usually those infected with the human immunodeficiency virus (HIV) or organ transplant recipients on immunosuppressive therapy. PCNSL in immunocompromised patients is typically large cell with immunoblastic and more aggressive features. These patients are usually severely immunocompromised, with CD4 counts of less than 50/mL. The Epstein-Barr virus (EBV) frequently plays an important role in the pathogenesis of HIV-related PCNSL.

Immunocompetent patients with PCNSL are older (median 60 years) compared to patients with HIV-related PCNSL (median 31 years). PCNSL usually presents as a mass lesion, with neuropsychiatric symptoms, symptoms of increased intracranial pressure, lateralizing signs, or seizures.

On contrast-enhanced MRI, PCNSL usually appears as a densely enhancing tumor (Fig. 118-4). Immunocompetent patients have solitary lesions more often than immunosuppressed patients. Frequently there is involvement of the basal ganglia, corpus callosum, or periventricular region. Although the imaging features are often characteristic, PCNSL can sometimes be difficult to differentiate from high-grade gliomas, infections, or demyelination. Stereotactic biopsy is necessary to obtain a histologic diagnosis. Whenever possible, glucocorticoids should be withheld until after the biopsy has been obtained because they have a cytolytic effect on lymphoma cells and may lead to nondiagnostic tissue. In addition, patients should be tested for HIV and the extent of disease should be assessed by performing PET or CT of the body, MRI of the spine, CSF analysis, and slit-lamp examination of the eye. Bone marrow biopsy and testicular ultrasound are occasionally performed.

TREATMENT PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMA

PCNSL is more sensitive to glucocorticoids, chemotherapy, and RT than other primary brain tumors. Durable complete responses and long-term survival are possible with these treatments. High-dose methotrexate, a folate antagonist that interrupts DNA synthesis, produces response rates ranging from 35–80% and median