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Glioblastoma and Other Primary Brain Malignancies in Adults: A Review

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Abstract

Importance: Malignant primary brain tumors cause more than 15 000 deaths per year in the United States. The annual incidence of primary malignant brain tumors is approximately 7 per 100 000 individuals and increases with age. Five-year survival is approximately 36%.

Observations: Approximately 49% of malignant brain tumors are glioblastomas, and 30% are diffusely infiltrating lower-grade gliomas. Other malignant brain tumors include primary central nervous system (CNS) lymphoma (7%) and malignant forms of ependymomas (3%) and meningiomas (2%). Symptoms of malignant brain tumors include headache (50%), seizures (20%-50%), neurocognitive impairment (30%-40%), and focal neurologic deficits (10%-40%). Magnetic resonance imaging before and after a gadolinium-based contrast agent is the preferred imaging modality for evaluating brain tumors. Diagnosis requires tumor biopsy with consideration of histopathological and molecular characteristics. Treatment varies by tumor type and often includes a combination of surgery, chemotherapy, and radiation. For patients with glioblastoma, the combination of temozolomide with radiotherapy improved survival when compared with radiotherapy alone (2-year survival, 27.2% vs 10.9%; 5-year survival, 9.8% vs 1.9%; hazard ratio [HR], 0.6 [95% CI, 0.5-0.7]; $P < .001$). In patients with anaplastic oligodendroglial tumors with 1p/19q codeletion, probable 20-year overall survival following radiotherapy without vs with the combination of procarbazine, lomustine, and vincristine was 13.6% vs 37.1% (80 patients; HR, 0.60 [95% CI, 0.35-1.03]; $P = .06$) in the EORTC 26951 trial and 14.9% vs 37% in the RTOG 9402 trial (125 patients; HR, 0.61 [95% CI, 0.40-0.94]; $P = .02$). Treatment of primary CNS lymphoma includes high-dose methotrexate-containing regimens, followed by consolidation therapy with myeloablative chemotherapy and autologous stem cell rescue,

nonmyeloablative chemotherapy regimens, or whole brain radiation.

Conclusions and relevance: The incidence of primary malignant brain tumors is approximately 7 per 100 000 individuals, and approximately 49% of primary malignant brain tumors are glioblastomas. Most patients die from progressive disease. First-line therapy for glioblastoma is surgery followed by radiation and the alkylating chemotherapeutic agent temozolomide.

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