

HOSPITAL PHYSICIAN®

NEUROLOGY BOARD REVIEW MANUAL

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The *Hospital Physician Neurology Board Review Manual* is a peer-reviewed study guide for residents and practicing physicians preparing for board examinations in neurology. Each manual reviews a topic essential to the current practice of neurology.

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Cancer Neurology: Primary and Metastatic Brain Tumors

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Cancer Neurology: Primary and Metastatic Brain Tumors

Andrew D. Norden, MD, and Santosh Kesari, MD, PhD

INTRODUCTION

Cancer neurology is a diverse field that involves primary tumors of the nervous system, metastatic cancer, a variety of complications of primary and secondary cancer, and paraneoplastic disease. Neuroimaging and molecular-based technologies have revolutionized the diagnostic evaluation of neoplastic disease in the nervous system, but tissue diagnosis remains critical for choosing an optimal treatment regimen. Multimodal treatment is expected to play an increasingly important role in future management of these disorders.

Although uncommon, primary brain tumors are responsible for a disproportionate share of morbidity and mortality and are increasing in incidence. After stroke, brain tumor is the most common cause of death due to intracranial disease. An estimated 18,500 new diagnoses of primary brain tumors were expected in the United States in 2005 along with 12,760 deaths, and the estimated 5-year survival rate between 1995 and 2000 was 33%.¹ Metastatic brain tumors are thought to be at least twice as prevalent as primary tumors and are associated with an even worse prognosis. Median survival ranges from 2.3 to 13.5 months.²

The only well-validated risk factor for development of a primary brain tumor is exposure to ionizing radiation, generally in the form of therapeutic irradiation for cancer.³ Other proposed risk factors include occupational exposures to environmental carcinogens, high dietary intake of *N*-nitroso compounds such as those found in cured meats, tobacco smoking, exposure to electromagnetic fields, cellular phone use, head injuries, various allergic conditions, and viral infections. None of these exposures has been conclusively linked to brain tumor risk.⁴ Several rare familial syndromes confer increased risk (**Table 1**) but account for fewer than 5% of brain tumors.

A World Health Organization consensus panel developed a comprehensive classification scheme for brain tumors in 1993 and updated it in 2000.^{5,6} Relevant entities are summarized in **Table 2**. Gliomas account for about one third of primary brain tumors, and two thirds of these are high grade (mostly anaplastic

astrocytomas and glioblastomas), which are associated with an extremely poor prognosis. Meningiomas represent nearly 25%, pituitary tumors 8%, and primary central nervous system (CNS) lymphoma (PCNSL) 4.1%, followed by all other tumor subtypes.⁷

This review focuses on primary and metastatic brain tumors and presents 3 cases that evolve over the course of the discussion to encompass evaluation and management of a solitary brain mass, multiple brain masses, and leptomeningeal metastases.

PRIMARY BRAIN TUMORS

CASE 1 PRESENTATION

A 54-year-old man presents to the emergency department (ED) after a minor motor vehicle accident. He cannot recall the specific circumstances that led to the accident, but a bystander reported that the patient was “shaking all over” just before the collision. The patient had been well until several weeks earlier. Since then, he has become lost while driving in a familiar area, left home wearing only his right shoe, and on more than one occasion put only his right arm into his jacket. His gait also has seemed unsteady, and he has repeatedly bumped into objects on his left side. He has increasingly complained of a bilateral frontal headache in the morning.

Examination in the ED reveals mild drowsiness. Basic cognitive functions are normal. When the patient is asked to draw a clock, he includes only numbers 12 through 6. Cranial nerve examination is notable for left inferior quadrantanopsia. Strength is normal and symmetric. There is decreased sensation to all modalities on the left side of the face, arm, and leg. Extinction to double simultaneous visual and tactile stimulation is apparent on the left. The patient can identify an object by touch when placed in the right hand but not when placed in the left hand. Coordination is normal. Gait is moderately unsteady, with a tendency to fall to the left. Reflexes are normal and symmetric. There is a Babinski sign on the left.

- What elements of this patient’s history and examination are suggestive of a mass lesion?