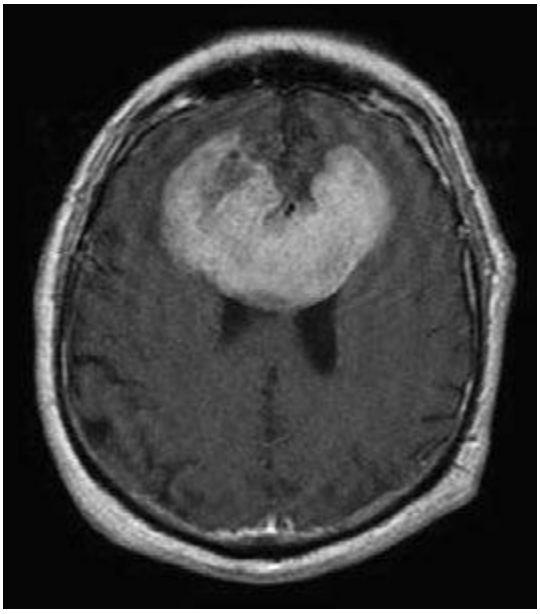

FIGURE 118-4 Postgadolinium T1 MRI demonstrating a large bifrontal primary central nervous system lymphoma (PCNSL). The periventricular location and diffuse enhancement pattern are characteristic of lymphoma.

survival of up to 50 months. The combination of methotrexate with other chemotherapeutic agents such as cytarabine increases the response rate to 70–100%. The addition of whole-brain RT to methotrexate-based chemotherapy prolongs progression-free survival but not overall survival. Furthermore, RT is associated with delayed neurotoxicity, especially in patients over the age of 60 years. As a result, full-dose RT is frequently omitted, but there may be a role for reduced-dose RT. The anti-CD20 monoclonal antibody rituximab has activity in PCNSL and is often incorporated into the chemotherapy regimen. For some patients, high-dose chemotherapy with autologous stem cell rescue may offer the best chance of preventing relapse. At least 50% of patients will eventually develop recurrent disease. Treatment options include RT for patients who have not had prior irradiation, re-treatment with methotrexate, as well as other agents such as temozolomide, rituximab, procarbazine, topotecan, and pemetrexed. High-dose chemotherapy with autologous stem cell rescue may have a role in selected patients with relapsed disease.

PCNSL IN IMMUNOCOMPROMISED PATIENTS

PCNSL in immunocompromised patients often produces multiple ring-enhancing lesions that can be difficult to differentiate from metastases and infections such as toxoplasmosis. The diagnosis is usually established by examination of the CSF for cytology and EBV DNA, toxoplasmosis serologic testing, brain PET imaging for hypermetabolism of the lesions consistent with tumor instead of infection, and, if necessary, brain biopsy. Since the advent of highly active antiretroviral drugs, the incidence of HIV-related PCNSL has declined. These patients may be treated with whole-brain RT, high-dose methotrexate, and initiation of highly active antiretroviral therapy. In organ transplant recipients, reduction of immunosuppression may improve outcome.

MEDULLOBLASTOMAS

Medulloblastomas are the most common malignant brain tumor of childhood, accounting for approximately 20% of all primary CNS tumors among children. They arise from granule cell progenitors or from multipotent progenitors from the ventricular zone. Approximately 5% of children have inherited disorders with germline mutations of genes that predispose to the development of medulloblastoma. Gorlin syndrome, the most common of these inherited disorders, is due to mutations in the patched-1 (*PTCH-1*) gene, a key component in the sonic hedgehog pathway. Turcot syndrome, caused by mutations in the adenomatous polyposis coli (*APC*) gene and familial adenomatous polyposis, has also been associated with an increased incidence of medulloblastoma. Histologically, medulloblastomas are highly cellular tumors with abundant dark staining, round nuclei, and rosette formation (Homer-Wright rosettes). They present with headache, ataxia, and signs of brainstem involvement. On MRI they appear as densely enhancing tumors in the posterior fossa, sometimes associated with hydrocephalus. Seeding of the CSF is common. Treatment involves maximal surgical resection, craniospinal irradiation, and chemotherapy with agents such as cisplatin, lomustine, cyclophosphamide, and vincristine. Approximately 70% of patients have long-term survival but usually at the cost of significant neurocognitive impairment. A major goal of current research is to improve survival while minimizing long-term complications.

PINEAL REGION TUMORS

A large number of tumors can arise in the region of the pineal gland. These typically present with headache, visual symptoms, and hydrocephalus. Patients may have Parinaud syndrome characterized by impaired upgaze and accommodation. Some pineal tumors such as pineocytomas and benign teratomas can be treated simply by surgical resection. Germinomas respond to irradiation, whereas pineoblastomas and malignant germ cell tumors require craniospinal radiation and chemotherapy.

EXTRINSIC "BENIGN" TUMORS

MENINGIOMAS

Meningiomas are diagnosed with increasing frequency as more people undergo neuroimaging for various indications. They are now the most common primary brain tumor, accounting for approximately 35% of the total. Their incidence increases with age. They tend to be more common in women and in patients with neurofibromatosis type 2. They also occur more commonly in patients with a past history of cranial irradiation.

Meningiomas arise from the dura mater and are composed of neoplastic meningothelial (arachnoidal cap) cells. They are most commonly located over the cerebral convexities, especially adjacent to the sagittal sinus, but can also occur in the skull base and along the dorsum of the spinal cord. Meningiomas are classified by the WHO into three histologic grades of increasing aggressiveness: grade I (benign), grade II (atypical), and grade III (malignant).

Many meningiomas are found incidentally following neuroimaging for unrelated reasons. They can also present with headaches, seizures, or focal neurologic deficits. On imaging studies they have a characteristic appearance usually consisting of a partially calcified, densely enhancing extraaxial tumor arising from the dura (Fig. 118-5). Occasionally they may have a dural tail, consisting of thickened, enhanced dura extending like a tail from the mass. The main differential diagnosis of meningioma is a dural metastasis.

If the meningioma is small and asymptomatic, no intervention is necessary and the lesion can be observed with serial MRI studies. Larger, symptomatic lesions should be resected. If complete resection is achieved, the patient is cured. Incompletely resected tumors tend to recur, although the rate of recurrence can be very slow with grade I tumors. Tumors that cannot be resected, or can only be partially removed, may benefit from treatment with external-beam RT or stereotactic radiosurgery (SRS). These treatments may also be helpful in patients whose tumor has recurred after surgery. Hormonal therapy and chemotherapy are currently unproven.

Rarer tumors that resemble meningiomas include hemangiopericytomas and solitary fibrous tumors. These are treated with surgery and RT but have a higher propensity to recur locally or metastasize systemically.

SCHWANNOMAS

These are generally benign tumors arising from the Schwann cells of cranial and spinal nerve roots. The most common schwannomas, termed *vestibular schwannomas* or *acoustic neuromas*, arise from the