Tumors of the Nervous System (Dr. Merchut)

1. Characteristics of brain tumors

Primary brain tumors arise from within the central nervous system (CNS), whether from glia, neurons or the meninges. In general, most primary brain tumors are infratentorial in children, such as a cerebellar medulloblastoma or astrocytoma, and supratentorial in adults. Metastatic brain tumors spread from other organs or tissue outside the CNS, such as breast or lung cancer. Metastases to the bone of the vertebral column or skull can spread by direct extension into the brain or spinal cord, or tumor cells from a remote systemic cancer can reach the brain by way of blood vessels. Conversely, the spread of primary brain tumors to other body organs virtually never occurs. Malignant brain tumors are microscopically less differentiated and invade normal surrounding tissue. Malignant gliomas and brain metastases typically grow faster and spread more readily, often leading to death in a matter of months. Benign brain tumors are microscopically more differentiated, and encapsulated or isolated from surrounding normal tissue, with slower growth and less or no tendency to spread. Examples include meningiomas, pituitary adenomas and neurinomas (schwannomas). However, the location of a brain tumor may be more important than its growth characteristics, since a benign tumor in a surgically inaccessible, vital area, like the thalamus or brain stem, may rapidly become fatal.

2. Symptoms of brain tumors

A tumor involving motor or sensory cortex may create a focal neurological deficit, typically at a slower rate than an ischemic infarction or hemorrhage arising at the same area. Sometimes the onset of symptoms is more abrupt or acute, as when a hemorrhage occurs within the tumor itself. Seizures may occur when cortex or adjacent subcortical areas are involved. Whether a focal neurological deficit is obvious or not, a tumor can produce increased intracranial pressure by obstructing cerebrospinal fluid (CSF) pathways or from tumor edema shifting the brain. This is suggested by headache, nausea and vomiting, somnolence or obtundation. The presence of a brain tumor may also be heralded by vague symptoms such as forgetfulness, confusion, irritability, moodiness or behavioral changes which are otherwise unexplained.

3. Malignant primary brain tumors

Gliomas, the most common type of primary brain tumor, include astrocytomas, ependymomas, medulloblastomas and oligodendrogliomas. Astrocytomas, which arise from astrocytes, have differing microscopic degrees of malignancy, the most malignant of which is the glioblastoma multiforme. The prognosis is poorer if the grade of malignancy is worse. Ependymomas arise from the ependymal cells lining the ventricles, and most often occur in the posterior fossa in children, commonly obstructing the fourth ventricle. Ependymomas often develop in the filum terminale of the spinal cord in adults. Medulloblastomas arise from the neuroepithelial roof of the fourth ventricle in
children. Oligodendrogiomas, originate from oligodendrocytes, and often involve the frontal lobes in adults, sometimes slowly progressing over years.

The CT or MRI brain scan of a more malignant primary brain tumor typically shows a more irregular, inhomogeneous, solitary mass which variably enhances (Figure 1). More benign primary brain tumors may appear more spherical and homogeneous, showing uniform enhancement. Treatment is based on the type, location and extent of the tumor, in conjunction with the overall health and age of the patient. In the least, a biopsy is ideally needed to best determine further treatment, obtained either by a surgical procedure or CT-guided stereotactic approach. If complete surgical excision is possible, the prognosis is often best, as with resection of a childhood cerebellar astrocytoma. Combined therapy with radiation or chemotherapy is often undertaken, although the blood-brain barrier impairs the delivery of chemotherapy to the tumor site. A therapeutic success often is prolonged survival rather than a permanent cure. Anticonvulsants are needed to control any seizures which occur.

Figure 1. CT brain scan of a malignant, primary brain tumor in the frontal lobe. Contrast was given for the image on the right, showing irregular, non-uniform tumor enhancement.
4. Metastatic brain tumors

In several series of intracranial neoplasms, metastatic brain tumors outnumber primary brain tumors. Systemic tumor cells in the bloodstream typically are deposited where the cortical arterial supply ends, at the gray-white matter junction. The typical CT or MRI scan appearance of metastatic brain tumors is that of multiple, spherical tumors at the gray-white matter junction of the cerebral cortex, often with surrounding edema (Figure 2). Intravenous contrast may be needed to better delineate the tumor from its edema. A solitary metastatic brain tumor may be difficult to distinguish from a primary brain tumor unless a biopsy is done, although a metastasis is more likely in a patient with a previously diagnosed systemic cancer. Metastases most commonly occur from systemic cancers of the lung, breast, colon, kidney, or melanoma. A systemic lymphoma can spread to the brain, while in immunocompromised patients, particularly those with AIDS, a lymphoma can originate within the brain itself.

Figure 2. MRI brain scans of cerebral metastases. A, Note the peritumoral edema (high MRI signal, bright white area). B, Note homogeneous tumor enhancement with contrast.
Survival is improved when a single brain metastasis is surgically removed and followed by irradiation, with a poorer prognosis for multiple metastases treated with brain radiation alone. Corticosteroids, such as dexamethasone, may decrease the edema around many types of brain tumors, and reduce neurological symptoms. Although chemotherapy may reduce systemic cancer, it usually does not effectively penetrate the blood-brain barrier to affect brain metastases. Anticonvulsants are needed to control any seizures which occur.

Meningeal carcinomatosis is a syndrome where cancer cells spread throughout the CSF, depositing over different areas of cerebral cortex, cranial nerve or spinal nerve roots. The diagnosis is suggested by mental status changes, headache, cranial nerve palsies or radiculopathies, especially in a patient with a known systemic malignancy. Cytological analysis of the CSF confirms the diagnosis. Intrathecal chemotherapy may offer some therapeutic benefit.

5. Benign primary brain tumors

Meningiomas are the most common benign brain tumor, arising from the arachnoid over the parasagittal falx, cerebral convexities, sphenoidal ridge and olfactory groove. They may remain asymptomatic or take years to slowly grow. Whenever complete excision is possible, it is the best prognosis of all brain tumors. Pituitary adenomas may affect the optic chiasm, causing visual deficits, or create symptoms of hormonal excess or deficiency. Often their resection is possible through a trans-sphenoidal approach through the skull base. Acoustic neurinomas (neuromas or schwannomas) arise from the sheath of the vestibular nerve, usually impairing hearing ipsilaterally. Imaging shows a mass at the cerebellopontine angle, which often can be surgically removed.

6. Spinal cord tumors

CNS tumors affecting the spinal cord are often meningiomas or ependymomas. Signs and symptoms are dependent on the level of the spinal cord involved. Treatment options include surgery or irradiation. Metastatic spinal cord compression often presents acutely and progresses rapidly, and must be treated emergently. Systemic cancer which has spread to the bony vertebrae causes local pain early, and tumor expansion thereafter compresses the spinal cord in its canal. Again, signs and symptoms depend on the level of spinal cord affected, but the longer any paralysis persists, the poorer the neurological outcome. MRI is the optimal imaging modality of the spinal cord, and may reveal subclinical, asymptomatic tumor at other spinal cord levels. If the patient has no diagnosed systemic cancer, surgical decompression will provide tissue for tumor identification as well as relieve symptoms. Intravenous corticosteroids, such as dexamethasone, may dramatically reduce peritumoral edema and improve symptoms as well. Irradiation of the tumor, in conjunction with steroid therapy, may achieve the same therapeutic result as surgical resection, provided that the tumor is radiosensitive.