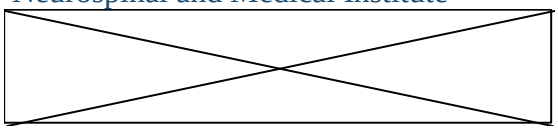
Neurospinal and Medical Institute



Brain and Spinal Tumors

The Neurospinal and Cancer Research Institute combines multidisciplinary clinical care and a strong research program in an effort to facilitate rapid transfer of basic scientific findings into clinical protocols for patients with tumors of the brain, skull base and spine. The Institute has innovative clinical initiatives organized under the auspices of the Neuro-Oncology Program; the NCRI Pediatric Brain Tumor Program, the NCRI Skull Base Surgery Program, the NCRI Tumor Radiosurgery Program, the NCRI Spinal Tumor Program and the NCRI Neuro-Endocrinology Program.

Diseases treated in the Center include both benign and malignant tumors of the brain, spinal cord, skull base, and spine in adults and children. Targeted pathologies include primary parenchymal tumors (including gliomas, craniopharyngiomas, germ cell tumors, and lymphomas), metastatic tumors (including parenchymal, leptomenngeal, and osseous lesions) and tumors of the skull base (including meningiomas, osseous and cartilagenous tumors, pituitary adenomas, acoustic neuromas and glomus tumors).

Minimally invasive therapies, such as **microneurosurgery**, **stereotactic neurosurgery**, **endoscopic neurosurgery**, **radiosurgery**, are a particular strength. Studies of innovative treatment currently underway include the following:

Radiosurgery of Brain tumors having size less than 30mm, deep seated / inapproachable by surgery, highly vascular, old age patients, or having high morbidity are treated by single session radiosurgery on **Gamma knife** as day care (one day) treatment.

Other indications of Gamma Knife include arteriovenous malformations, pituitary adenomas, acoustic neurinomas, trigeminal schewanomas, pinealomas, neurofibromas, juglare body tumors, brain stem gliomas, trigeminal neuralgias, germinomas and metastasis.

Fractionated radiosurgery for all the tumors more than

30mm in size including gliomas are treated on X-Knife

Robotic Synergy-S having micro-radiosurgery facilities called **IGRT, IGMRT soft wares like Ergo, and VMAT**. All tumors of brain; spine and whole body are being

treated on X-knife either in single or hypo-fractions 2-5.

This facility is part of Pakistan Gamma knife stereotactic radiosurgery center – a center of excellence for radiosurgery.

Chemotherapy:

- Timozodamide for adult malignat gliomas
- Combination chemotherapy for pediatric tumors

Brain tumors, or neurological complications of other cancers, often have profound implications for patients and their families. Caring for patients with neuro-oncological disease is a complex task that requires input from many specialists.

To meet these challenges, the NCRI, Neuro-Oncology Program has developed an interdisciplinary team whose goals are the following:

- To provide a specific and accurate diagnosis
- To communicate to the patient and the patient's family the nature, treatment options, and possible outcomes of the disease
- To offer both conventional and experimental treatments
- To optimize the quality of each patient's life
- To assist the patient and the patient's family in coping with the disease

The NCRI maintains an active pediatric tumor practice and runs a separate clinic twice a week. Here, advanced multimodality therapy for a wide variety of brain tumors in children is offered.

Specialties

Acoustic neuroma

Acoustic neuroma (sometimes termed a neurolemmoma or schwannoma) is a benign (non-cancerous) tissue growth that arises on the eighth cranial nerve leading from the brain to the inner ear. This nerve has two distinct parts, one part associated with transmitting sound and the other sending balance information to the brain from the inner ear. These pathways, along with the facial nerve, lie adjacent to each other as they pass through a bony canal called the internal auditory canal. This canal is approximately 2 cm (0.8 inches) long and it is here that acoustic neuromas originate from the sheath surrounding the eighth nerve. The facial nerve provides motion of the muscles of facial

expression.

Acoustic neuromas usually grow slowly over a period of years. They expand in size at their site of origin and when large can displace normal brain tissue. The brain is not invaded by the tumor, but the tumor pushes the brain as it enlarges. The slowly enlarging tumor protrudes from the internal auditory canal into an area behind the temporal bone called the cerebellopontine angle. The tumor now assumes a pear shape with the small end in the internal auditory canal. Larger tumors can press on another nerve in the area (the trigeminal nerve) which is the nerve of facial sensation. Vital functions to sustain life can be threatened when large tumors cause severe pressure on the brainstem and cerebellum part of the brain.

Tumors are typically described as small (less than 1.5 cm), medium (1.5 cm to 2.5 cm) or large (more than 2.5 cm).

Courtesy of Acoustic Neuroma Association

Anaplastic astrocytoma

Anaplastic astrocytomas or grade II fibrillary astrocytomas are intermediate in pathological characteristics and rapidity of growth between astrocytomas and glioblastomas. Anaplastic astrocytomas occur predominantly in the cerebral hemispheres of middle aged adults. Headaches, depressed mentation, and focal neurologic deficits are more common and seizures somewhat less common than with non-anaplastic astrocytomas. Most of these tumors enhance on contrast MRI scans. The preferred treatment is the surgical removal of as much tumor as is safely possible, followed by a six-week course of radiation therapy. Chemotherapy, usually with a nitrosourea, is often indicated, particularly for tumors that recur. Radiosurgery is often considered for residual or recurrent tumor that appears focally contained. Despite such therapies, these tumors frequently recur in a life-threatening manner between two and five years after initial treatment.

Arachnoid cyst

Arachnoid cysts are collections of CSF cerebrospinal fluid contained within the arachnoidal lining of the brain. They represent approximately one percent of brain mass lesions. Most common in infants, they may also present in adulthood. In infants, they cause head enlargement or signs of obstructive hydrocephalus such as headache, lethargy, double vision, and developmental delay. In adults, they are more likely to cause seizures, particularly those of the middle cranial fossa. Arachnoid cysts are readily seen on CT and

MRI scans as non-enhancing cystic masses of near CSF density/intensity. Treatment options include fenestration of the cyst into normal CSF pathways, either endoscopically or by open craniotomy, or shunting of the cyst and/or ventricles into the peritoneal cavity. Successful cystic opening or fluid diversion is usually effective treatment.

Astrocytoma brain tumor

Astrocytomas include a large group of gliomas whose predominant cell type resembles an astrocyte. Over half of all brain gliomas are astrocytic. They arise from and expand within the brain and almost never spread to other parts of the body. Astrocytomas are graded from grade I to IV depending on the presence of features implying rapid growth such as irregular cell structures, cells actively dividing, and abnormally formed blood vessels. Astrocytomas usually become clinically evident by virtue of seizure, headache, or focal neurologic deficits such as speech difficulty, motor weakness, and visual loss. Magnetic resonance imaging shows them to be a mass that replaces normal brain. Lower grade tumors are less likely to enhance (turn white) with intravenous contrast agents than are higher-grade tumors. Surgery is often indicated: to establish the diagnosis pathologically; to alleviate mass effects causing neurologic symptoms and deficits; to remove as much tumor as is safely possible in order to decrease the chance of lower grade tumors progressing to more malignant forms. Surgery is also performed to improve the safety and efficacy of other treatments such as radiation therapy, radiosurgery, and chemotherapy.

Choroid plexus carcinoma

The choroid plexus is tissue located in the spaces inside the brain called ventricles. The choroid plexus makes the fluid that fills the ventricles and surrounds the brain and spinal cord. Tumors of the choroid plexus can grow slowly (choroid plexus papilloma) or grow more rapidly (anaplastic choroid plexus papilloma). The rapidly growing tumors are more likely to spread to other places in the brain and to the spinal cord.

Choroid plexus papilloma

Choriod plexus papillomas and choriod plexus carcinomas are tumors that arise within the ventricles from the choriod plexus which secretes cerebrospinal fluid. Almost half present in infants and three-quarters within children less than ten years old. About two-thirds are more benign papillomas and one-third are malignant carcinomas. Both types of tumors usually present with signs of increased intracranial pressure usually from a combination of overproduction of cerebrospinal fluid and obstructive hydrocephalus (blockage of the fluid pathways). Symptoms include headache, vomiting, lethargy, double vision, and unstable walking. MRI shows a tumor within the ventricles that enhances brightly with

contrast. Surgery is indicated in an attempt to remove the tumor completely and reopen the obstructed fluid pathways. Surgical removal of more malignant choriod plexus carcinomas may be facilitated by preoperative chemotherapy. Radiation therapy following complete resection of a choroid plexus papilloma, particularly in the young child, is not indicated. Incompletely resected papillomas and all carcinomas warrant radiation therapy in patients of sufficient age.

Colloid cyst

Colloid cysts are epithelial lined collections of mucoid fluid that usually occur in the anterior third ventricle. Here they cause symptoms by obstructing CSF flow. Headache, lethargy, and blurred vision can result. Symptoms from colloid cysts can be intermittent secondary to position-related ball valve obstruction of the foramen of Munro. Sudden, transient leg weakness causing the patient to collapse is relatively rare but frequently suggestive of an obstructive colloid cyst. Sudden death is rare. Dementia and seizures can also occur. MRI shows a non-enhancing cyst at the front of the third ventricle and expansion of the lateral ventricles if obstruction is present. Treatment options include observation of small cysts not causing obstruction, placement of a ventriculo-peritoneal shunt when obstruction is present and removal of the cyst by either craniotomy or stereotactic aspiration. If obstructive hydrocephalus is effectively prevented or treated, the lesion is not life threatening.

Craniopharyngioma

Craniopharyngiomas are mixed solid and cystic tumors that arise at the base of the brain near the pituitary gland and hypothalamus. They represent approximately 3% of all intracranial tumors. Approximately half present in children and half in adults. Symptoms include endocrinological disturbances such as growth delay and loss of sexual function, visual loss, headache, memory loss, and depression. CT and MRI scans can demonstrate solid and cystic portions. Calcification and contrast enhancement of the solid portion are characteristic. Surgery is directed both at relief of obstructive hydrocephalus and removal of the tumor. Craniotomy is often the first treatment of choice. Tumor attachment to major arteries, optic nerves and chiasm, and the hypothalamus can preclude complete removal. Postoperative radiotherapy is very valuable in preventing tumor recurrence. Hormone replacement is frequently indicated.

Dermoid tumor

Dermoid cysts contain elements of skin such as hair and sebaceous glands in addition to desquamated keratin. They constitute approximately 0.3% of intracranial tumors. Dermoid

cysts have a preference for the midline of the posterior fossa. Because of their more rapid growth and midline location, they become symptomatic, usually from obstructive hydrocephalus, at an earlier age than do epidermoid cysts. Dermal cysts may have a dermal sinus track connecting them to the skin, which is a source of infection. Bacterial meningitis may result. Dermoid cysts are best treated by surgical resection. This usually is curative.

DNET

DNET or dysembryoplastic neuro-epithelial tumors are benign, in that the tumor mass is composed of malformed neuronal and glial cells. Most of these appear in children and young adults with seizures. Except for their irregular shape and occasional enhancement with contrast, DNET's are difficult to distinguish from surrounding normal brain. Surgical removal is the preferred treatment, is often curable, and frequently results in seizure control.

Ependymoma brain tumor

Ependymomas are tumors of the brain and spinal cord that arise from ependymal cells lining the central fluid spaces (ventricles) of the brain and the central canal of the spinal cord. They occur both in children and adults and constitute between five and ten percent of central nervous system tumors. More aggressively growing anaplastic ependymomas can be distinguished from more benign ependymomas by virtue of pathologic features such as irregular cellular shapes, greater cell density, and mitoses (cells in division). Ependymomas usually present with signs of increased intracranial pressure either from the size of the tumor itself or hydrocephalus (blockage of fluid flow) that results from the intraventricular location. Headache, lethargy, vomiting, and double vision are common. Seizure, facial numbness or weakness and difficulty swallowing can also occur. MRI usually shows a mass within the ventricles that turns bright with contrast. Given the tendency of ependymomas to spread through the brain and spinal fluid spaces, spinal MRI preoperatively and spinal fluid sampling (lumbar puncture) postoperatively are important in staging the spread of tumor. Surgical removal is the preferred treatment. The extent of resection possible is often determined by the pattern of tumor growth through the CSF spaces. Postoperatively, most patients receive fractionated radiotherapy. Whether the spinal cord as well as the brain is radiated depends on the pathology of the tumor and the results of staging. Chemotherapy is indicated in infants too young to undergo radiation therapy and in patients with recurrent growth of their tumor. The likelihood of tumor regrowth depends on the initial grade and extent of tumor, the extent of surgical resection,

and the patient's age.

Epidermoid cyst

Epidermoid cysts are epithelial lined collections of skin debris (desquamated keratin). Epidermoid cysts constitute approximately 1.5% of intracranial tumors. They commonly occur in the parasellar region and cerebellar pontine angle. Visual loss and endocrine dysfunction occur with parasellar cysts. Those in the posterior fossa cause cranial nerve deficits such as double vision, facial numbness or weakness, hearing loss, and swallowing difficulty. In either location, epidermoid cysts can present with signs of increased intracranial pressure associated with obstructive hydrocephalus. Seizures and hydrocephalus can result from aseptic meningitis caused by leakage of the debris into the subarachnoid spaces. Treatment is surgical removal. It is usually curative. Recollection of the cysts can also be treated by surgery.

Epidermoid tumor

An epidermoid tumor is a rare, benign neoplasm of the brain. It is congenital and is formed by skin cells that are in the wrong place during fetal development. It consists of an outer covering (the capsule part of it) and an inner mixture of skin cells, and fatty acids. The capsule is about the thickness of wet tissue paper, and the inside contents are the consistency of cottage cheese. These tumors are slow growing, and generally are diagnosed in the middle twenties to the late fifties. The incidence of intracranial epidermoids is between 0.2% to 1.8% of all brain tumors. Epidermoid tumors grow at a slow rate, and if not removed entirely, tend to grow back.

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Gangliocytoma

Gangliocytomas, gangliomas, and anaplastic gangliogliomas are tumors that include neoplastic nerve cells that are relatively well differentiated. Gangliogliomas have inter-mixed neoplastic glial cells that are relatively benign; in anaplastic gangliogliomas, the glial cells are more malignant. Together these tumors represent approximately 1% of brain tumors. Most occur in young adulthood. The cerebral hemispheres, thalamus and brainstem are common sites. Presenting symptoms include seizures, hypothalamic endocrine dysfunction, and weakness. They appear on MRI scans as well circumscribed masses. The more benign tumors show less enhancement, mass effect, and edema. Surgical excision is the treatment of choice. Radiation therapy is reserved for anaplastic

tumors. Survival of patients with gangliocytomas and gangliogliomas is usually long.

Ganglioglioma

These rare, benign tumors arise from ganglia-type cells, which are groups of nerve cells. Gangliocytomas (sometimes called ganglioneuromas) are tumors of mature ganglion cells. Gangliogliomas are tumors of both mature nerve and supportive cells.

Tumors arising from ganglia most frequently occur in children and young adults. They represent 0.4% of all primary brain tumors and about 4% of all pediatric brain tumors.

The most common sites are the temporal lobe of the cerebral hemispheres and the third ventricle, although they might also occur in the spine.

Cyst formation and calcification can be present. Seizures are the most common symptom.

These tumors are small, slow growing, and have distinct margins. Metastasis and malignancy are very rare.

Surgery is the standard treatment.

Courtesy American Brain Tumor Association

Germinoma

Germinomas are the most common type of a group of intracranial tumors derived from germ cells (which includes embryonal carcinoma, choriocarcinoma, endodermal sinus tumor, yolk sac carcinoma, and teratoma. Germinomas may occur either in the pineal region or just above the pituitary gland. They present with signs of increased intracranial pressure from obstructive hydrocephalus, double vision, and unstable gait. MRI scans show a uniformly enhancing tumor in the pineal region, posterior third ventricle, and, occasionally, above the sella. It is usually important to obtain tumor tissue, either by stereotactic or endoscopic biopsy or by open craniotomy, to establish the diagnosis. The spine should be screened for tumor deposits by MRI and, if there is no hydrocephalus, by cytologic analysis of lumbar cerebrospinal fluid. Surgical resection can accomplish both tumor removal and reopening of the obstructed fluid pathways. In that germinomas are highly sensitive to radiation, surgery can often be avoided if tissue diagnosis can be otherwise established and hydrocephalus is either absent or treatable by diversion until radiation can be delivered. Radiotherapy is indicated for patients older than three years. Long term remission is common. Chemotherapy is indicated in children too young to

receive radiotherapy and in older patients with recurrent tumors.

Glioblastoma multiforme

Glioblastomas (glioblastoma multiforme) are highly malignant tumors that constitute approximately half of all gliomas and one-quarter of intracranial tumors in adults. They occur most frequently in the middle aged and elderly. They may present with seizures, focal neurologic deficits such as speech disturbance, weakness and visual loss, or signs or symptoms of increased intracranial pressure such as headache and mental slowing. They often appear as ring contrast enhancing masses on MRI scans. Unfortunately, at that time of discovery, most have extensive infiltration far from the contrast enhancing solid tumor. Nonetheless, the initial treatment of often surgical with an effort to remove this focal solid tumor. Radiation therapy and chemotherapy are used in an effort to forestall the spread of microscopically infiltrative tumor and the return of a tumor mass. Despite current best therapies, these tumors are often life threatening within a year of diagnosis.

Glioma

Gliomas are tumors that arise from glial cells of the brain and spinal cord. There are three main types: astrocytomas, oligodendrogliomas, and ependymomas. Gliomas also differ from one another by virtue of their location within the brain and spinal cord and their grade, which is an index of the rate and invasiveness of growth. Treatment options include surgery, radiation, radiosurgery, and chemotherapy. Choice among these options depends on the characteristics and preferences of the individual patient, the size and location of the tumor, and the tumor's pathology and grade. Prognosis depends on the gliomas' pathology and grade, the patient's age and level of function, and the tumor's response to therapy.

Hemangioblastoma brain tumor

Hemangioblastomas are brain tumors derived from blood vessels. They represent approximately 2% of intracranial tumors and 10% of tumors in the posterior fossa. Hemangioblastomas are twice as frequent in men as in women. Approximately 5% of patients with hemangioblastomas have the dominantly inherited disease of Von Hippel-Lindau. Von Hippel-Lindau Disease is characterized by hemangioblastomas of the retina, cerebellum, and spinal cord, by tumors of the kidneys, and by cysts of the kidney, liver and pancreas. Approximately half of patients with the Von Hippel-Lindau disease gene have hemangioblastomas. Patients with Von Hippel-Lindau disease are more likely to have multiple hemangioblastomas than patients with a sporadic tumor. Both sporadically occurring and inherited tumors may produce erythropoietin, a hormone that causes over-production of red blood cells. Hemangioblastomas most commonly occur in the

cerebellum, floor of the fourth ventricle, and spinal cord. They form as a solid tumor nodule, associated in the majority of cases with a cyst. Clinical symptoms and signs reflect the tumor's location. Posterior fossa tumors can cause headache and double vision from obstructive hydrocephalus, weakness in coordination, and gait instability. Spinal cord tumors can cause peripheral sensory loss, weakness, and gait difficulties. MRI clearly shows the brightly enhancing tumor nodule and the non-enhancing cyst. Surgical excision is the preferred method of treatment. Indications for surgery include relief of symptoms and removal of the risk of hemorrhage. Tumors that can only be partially removed by virtue of location or vascularity can be treated with radiosurgery or fractionated radiation therapy.

Low grade glioma

Low-grade gliomas include pilocytic astrocytomas, grade II fibrillary astrocytomas, and oligodendrogliomas. These tumors have less malignant pathological features, are more likely to be circumscribed and thus completely removed by surgery, and more slowly growing. Surgery is the preferred form of treatment. Many, however, may require radiation therapy or chemotherapy. Although therapy initially provides long-term remission, these tumors may show recurrent growth particularly following transformation into a higher-grade tumor.

Malignant glioma

Malignant glioma is a term which refers to astrocytomas and oligodendrogliomas with pathologic features such as irregular cell and nuclear shape, frequent mitotic (cells in division) figures, and widely invasive growth. These are likely to recur within one to three years following treatment. They include anaplastic astrocytomas, anaplastic oligodendrogliomas, mixed anaplastic gliomas or oligoastrocytomas, gliomatoses cerebri, gliosarcoma, and glioblastoma multiforme.

Meningioma brain tumor

Meningiomas are tumors that arise from the dural coverings, the meninges, of the brain or spinal cord. They represent approximately 20% of intracranial tumors. They are twice as common in women as in men. The incidence of meningiomas increases with age; most occur in women in mid to late adulthood. Meningiomas occur at arachnoid granulations, sites of CSF absorption into the venous sinuses. In order of likelihood, these are the falx and parasaggital area, the convexity, the sphenoid wing and middle cranial fossa, the floor of the anterior cranial fossa, and the posterior fossa. Almost 90% of meningiomas are benign, slowly growing masses that produce symptoms by compressing brain or cranial

nerves. Symptoms thus include headache, seizure, and focal neurologic loss of vision, strength, sensation, and speech. Except in the 10 to 15% of meningiomas that are atypical or malignant, growth is slow and progression of symptoms is insidious. Meningiomas have a highly characteristic appearance on MRI scans of a homogeneously enhancing mass with a dural tail. The preferred treatment for a symptomatic, enlarging, surgically accessible meningioma in a healthy person is surgical removal. In some cases, removal of the tumor may be facilitated by angiographic blockade (embolization) of the tumor's blood supply before surgery. Complete surgical removal provides cure or decades of local tumor control in almost all cases. In some cases, because of inextricable attachment to cranial nerves or critical blood vessels or because of the age or general health of the patient, complete surgical resection is not possible. Radiation either by radiosurgery or fractionated radiotherapy is then indicated. Small tumors, particularly in the elderly, are often quite indolent and can be observed radiographically without treatment. Evidence of growth that threatens the development of symptoms warrants treatment. Atypical or malignant features, found in 10 to 15% of cases, are usually considered indications for the addition of radiation even in completely removed tumors. The risk of tumor recurrence depends on the extent of the resection, which, in it's self, reflects tumor location.

Metastatic brain tumor

Metastatic brain tumors are tumors that have spread to the brain from cancers originated in other parts of the body. Lung, colon, melanoma, breast, and prostate are the most common types. Approximately a third of patients with systemic cancer have brain metastases. Approximately half of patients with brain metastases have only a single clinically apparent tumor. Brain metastases occasionally are evident even before the systemic tumor. Common presenting signs and symptoms include headache, weakness, seizure, sensory loss, change in mentation, and unstable gait. Contrast enhanced MRI scan is the most sensitive way of detecting metastatic tumors. The choice of treatment is based upon a number of factors including number, size, location, and pathology of the metastatic tumors, the location, pathology, systemic spread, and treatment of the primary tumor, and the patient's age and general physical condition. Whole brain radiation therapy is usually indicated for patients with multiple brain metastases and progressive systemic disease. However, whole brain radiation therapy produces local control of tumor in less than half of the patients. Thus, in patients with effectively treated systemic disease whose function and survival are limited by one to several brain metastases, focal treatment such as surgery and radiosurgery is usually indicated. In general, surgery followed by whole brain radiation therapy is indicated for a single, surgically accessible metastasis in a highly functioning patient whose systemic disease is well controlled. Some physicians offer focal treatment to patients with two to four lesions as well. When there is only one metastasis, it

is controversial whether whole brain radiation therapy is required following surgical resection or radiosurgical treatment. Even in some patients with several metastases, whole brain radiation therapy may be deferred following effective focal treatment because of the long-term risk of radiation-induced brain injury. In conclusion, the current treatment of choice for one to four cerebral metastases in a patient with well-controlled systemic disease is focal treatment with either surgery or radiosurgery followed by whole brain radiation therapy. For most pathologies, this combination will provide local control or these tumors for at least a year in the large majority of patients.

Mixed anaplastic oligoastrocytoma

An anaplastic astrocytoma is a grade III tumor. Astrocytomas often contain a mix of cells and cell grades, but brain tumors are graded by the highest grade (most abnormal) cell seen in the tumor. These tumors tend to have tentacle-like projections that grow into surrounding tissue, making them difficult to completely remove during surgery. The word "anaplastic" means malignant, and because of this, treatment for an anaplastic astrocytoma may be more aggressive than treatment for a lower grade tumor.

The treatment options your doctor outlines will be based on the size and location of the tumor, what it looked like under the microscope, if and how far the tumor has spread, any previous treatment, and your general health. Generally, the first step in the treatment of anaplastic astrocytomas is surgery. The goals of surgery are to obtain tumor tissue for diagnosis and treatment planning, to remove as much tumor as possible, and to reduce the symptoms caused by the presence of the tumor. There are some circumstances, such as certain medical conditions or concerns about the location of the tumor, in which a biopsy may be done in place of surgery. The tissue obtained during the biopsy is then used to confirm the diagnosis.

Mixed glioma

Mixed gliomas or oligoastrocytomas are tumors that have both astrocytic and oligodendrocytic features. Their presentation is similar to that of their composite lesions. Imaging treatment and response to treatment reflect the element with the more anaplastic features.

Neurocytoma

This rare grade II tumor typically occurs in a lateral ventricle in the region of the foramen of Monro, and occasionally extends into the third ventricle as well. It is supplied by many blood vessels. The central neurocytoma shows mature cells, similar to normal neurons of

the gray matter, although their cell of origin is unknown. It is most common in young adult males. Symptoms are those associated with increased intracranial pressure: headache, nausea and vomiting, drowsiness, vision problems and mental changes.

Standard treatment is surgery, which is often successful. Excessive bleeding can limit the extent of tumor removal however. The routine use of radiosurgery/radiotherapy as an adjuvant therapy is best choice.

Neurofibroma brain tumor

Neurofibromas are a symptom of neurofibromatoses, which are genetic disorders of the nervous system that primarily affect the development and growth of neural (nerve) cell tissues. These disorders cause tumors to grow on nerves and produce other abnormalities such as skin changes and bone deformities. The neurofibromatoses occur in both sexes and in all races and ethnic groups. Scientists have classified the disorders as neurofibromatosis type 1 (NF1) and neurofibromatosis type 2 (NF2). Other or variant types of the neurofibromatoses may exist, but are not yet identified.

Courtesy National Institutes of Health, National Institute of Neurological Disorders and Stroke

Oligodendroglioma

Oligodendrogliomas are infiltrative glial tumors whose main cell type resembles the glial cell, oligodendrocyte. They account for about four percent of intracranial gliomas and are most commonly found in young and middle aged adults. Most are localized in the white matter tracts of the hemispheres, along which they spread. About half of patients present with seizure. Headache, mental change, and weakness are also common. Oligodendrogliomas are usually bright white on T2 weighted MRI scans but rarely enhance significantly after contrast is given. Surgical removal is the treatment of choice. If the tumor is identified while relatively superficial or confined to a polar region, gross total removal can be achieved. When diffuse spread precludes this, residual tumor may require radiation therapy. Larger, more aggressive tumors may also require chemotherapy. Although the tumor may remain quiescent for years after treatment, recurrence is the rule. This is treated with additional surgery or radiosurgery where possible and with chemotherapy.

PNET

PNET or primitive neuro-ectodermal tumors, are tumors of neuroglial precursors that

predominate in children. They may cause a focal neurologic deficit or signs of increased intracranial pressure. They enhance brightly with contrast MRI. They tend to spread throughout the brain. Treatment includes maximal resection, irradiation and chemotherapy. Response to therapy is often poor. Many consider PNET's to be the same tumor as meduloblastoma, just occurring intracranially at sites other than the posterior fossa.

Radiation necrosis

Radiation necrosis is a mass of dead tumor tissue and injured surrounding brain that can occur following intensive radiation of a brain tumor. It is sometimes difficult to distinguish from recurrent tumor growth. It most commonly follows focal radiation of a malignant glioma by brachytherapy or radiosurgery. Similar to the original tumor, it can cause headaches, seizures, and neurologic deficits such as weakness and speech loss. The initial treatment is cortical steroids. Should the steroids prove ineffective and the neurologic deficits progress, surgical removal of the mass is sometimes indicated.

Schwannoma brain tumor

Schwannomas and neurofibromas are tumors of the sheaths that surround cranial and peripheral nerves. The most common nerves involved are the eighth and the fifth cranial nerves producing vestibular schwannomas (acoustic neuromas) and trigeminal schwannomas, respectively. Vestibular schwannomas can occur either sporadically, on one side of a patient with no family history of such tumors, or as part of the dominantly inherited syndrome Neurofibromatosis type 2 (NF2). In the latter case, the patient will often have vestibular schwannomas on both sides and additional brain or spinal cord tumors. Acoustic neuromas present most commonly in young adulthood in NF2 and middle to late adulthood in the spontaneous form. Hearing loss, tinnitus (ringing in the ear), ear fullness, dizziness, vertigo, and unstable gait are all common. Larger tumors can cause headache, double vision, and unsteady gait, as well as facial numbness and difficulty swallowing. Enhanced MRI clearly shows tumor along the eighth nerve complex extending from the brainstem through the internal auditory canal toward the ear. Options for management include observation, surgery and radiation. Observation with close audiologic, neurologic, and MRI follow-up is a reasonable choice in patients with incidentally discovered tumors either with no, mild, or non-progressive symptoms. Enlargement of the tumor or progression of symptoms is an indication for treatment. Radiosurgery (focal radiation involving one to several treatment exposures) is an excellent option for small tumors (less than an inch in diameter) particularly in elderly or medically infirm patients. Surgery is often warranted in patients with large tumors compressing the

brainstem, tumor enlargement following radiosurgery, rapid tumor growth, or high quality hearing. The choice of surgical approach -- suboccipital, translabyrinthine, and middle cranial fossa -- depends on the size of the tumor and the quality of the patient's residual hearing. Almost all of these tumors are benign and one or a combination of these treatments usually achieves long-term tumor control. Trigeminal schwannomas are similar except that their presentation, location, and treatment reflect their origin from the fifth rather than the eighth cranial nerve. They present with facial sensory loss, double vision, headache, weakness and gait instability. If large enough, they, too, may cause hydrocephalus. They may extend into the middle cranial fossa as well as the posterior cranial fossa and even into the cavernous sinus. The same treatment options apply. Generally, surgical removal of this benign tumor is preferred. Long-term control is usually possible. Other cranial nerves such as the vagal, glossopharyngeal, and hypoglossal nerves may also give rise to these tumors. Symptoms reflect the function mediated by the involved nerve and surrounding structures. Surgical treatment is usually preferred as it offers the opportunity for cure. Radiosurgery is often an acceptable alternative.

Teratoma

Teratomas account for 0.5% of intracranial tumors overall but are more common in the pediatric population where they represent 2% of intracranial tumors. Their incidence is even higher in infants. Teratomas usually occur in the pineal region or, less commonly, around the sella. Pathologically, teratomas have elements of all three germ layers. Their growth, particularly in infants, can be quite rapid. Head enlargement and hydrocephalus are common. Surgical treatment is the procedure of choice. In infants, rapid fatal regrowth is common.

Anaplastic oligodendroglioma

Anaplastic oligodendrogliomas are oligodendrogliomas with more malignant pathologic features, more rapid growth, and a greater tendency to recur earlier. They are more likely to enhance on MRI scans and less likely to be completely resectable. Following surgery radiosurgery is often quite effective in forestalling recurrence.

Central neurocytoma

Central neurocytomas are benign tumors of small neuronal cells. They occur within the fluid spaces of the lateral and third ventricles. They may present throughout adulthood with either signs of increased intracranial pressure from the tumor mass or obstructive hydrocephalus or with focal neurologic deficits such as weakness or diplopia secondary to compression of the site of origin. Complete excision is the treatment of choice.

Radiosurgery is reserved for tumors that are clinically aggressive or recurrent. Overall, outcome is quite favorable.

Eosinophilic granuloma

Eosinophilic granulomas are skull lesions caused by abnormal collections of inflammatory cells. They may be single or multiple and usually occur in children. Pain is the most common presentation. Plain films and CT scans show round lytic lesions. Surgical excision of the granuloma is warranted. Low dose radiation should be given to lesions not completely resectable. Some patients will have systemic disease that requires chemotherapy.

Esthesioneuroblastoma

Esthesioneuroblastoma or olfactory neuroblastomas are rare malignant tumors that arise from olfactory cells high in the nasal cavity. They grow invasively either down into the nose to present with nasal obstruction, bleeding, or loss of smell or superiorly through the skull base to compress the brain. The preferred treatment is preoperative chemotherapy and irradiation prior to an attempt at enbloc resection. Recurrence may be either local or metastatic. Most patients with lower grade, less malignant tumors survive more than five years; those with more malignant higher-grade tumors do not.

Gliomatosis cerebri

Gliomatosis cerebri connotes diffuse proliferation of anaplastic astrocytes throughout the brain. Given the diffuse rather than focal nature of the tumor, generalized signs and symptoms predominate over focal neurologic deficits. These include change in intellect and personality and headache. The widespread nature of the tumor is often best depicted on a T2 weighted MRI scan. Given the diffuse infiltrative nature of the tumor, treatment is usually non-surgical following an initial biopsy to establish the diagnosis. Radiosurgery and chemotherapy are the mainstays of treatment.

Gliosarcoma

Gliosarcomas are relatively rare tumors that combine characteristics of glioblastoma and sarcoma. Often located superficially, they are more likely to invade the dura and skull and to metastasize extracranially. Their treatment and its outcome are similar to those for glioblastomas.

Lipoma

Lipomas are fatty tumors that usually occur in the midline of the brain and are associated with other congenital anomalies, such as myelomeningocele. They rarely cause symptoms. The appearance on CT and MRI scans is that of fat. Occasionally they cause seizures or obstructive hydrocephalus. Surgery, complicated by tenacious attachment of the tumor to surrounding structures, is rarely needed.

Low grade astrocytoma

Astrocytoma, low-grade astrocytoma, grades I and grade II astrocytoma are all names for the less malignant forms of astrocytoma.

Lymphoma

Lymphomas of the brain and spinal cord may be secondary to spread of non-Hodgkin's lymphoma from other parts of the body may arise primarily in the central nervous system. Primary CNS lymphoma (primary central nervous system lymphoma, PCNSL) accounts for approximately one percent of primary brain tumors but its incidence is increasing secondary to the rise of immune deficient states in HIV infection and chemotherapeutic suppression for organ transplantation. CNS lymphomas are predominantly B-cell in type. They may present as a brain or spinal cord mass, as widely disseminated foci in the cerebrospinal fluid, or in the eye. Symptoms include headache, mentation or personality change, double vision, and weakness. The diagnosis is usually made either by lumbar puncture for CSF analysis or MRI scan that shows a homogeneously enhancing mass. In immunosuppressed patients, there may be multiple lesions. Stereotactic biopsy for a pathological specimen is the best way to confirm the diagnosis. Best yield occurs when the patient is not taking steroids, which may disintegrate tumor cells and make diagnosis difficult. Chemotherapy, either with or without radiation therapy, is the mainstay of treatment. Currently methotrexate is the agent of choice. Overall survival can exceed two years.

Medulloblastoma

Medulloblastomas are tumors of the neuro-glial line that most often occur in the posterior fossa of children, usually in the fourth ventricle. Although they can occur in adults, two-thirds to three-quarters occur in children with a mean age of detection of seven years old. Symptoms such as headache, vomiting, and double vision usually occur from obstruction to CSF flow at the level of a fourth ventricular tumor. MRI scans usually show an intraventricular tumor that brightly enhances with contrast. Surgery is usually indicated in

an attempt to remove the entire tumor and reopen the brain fluid pathways. Even when the tumor is removed, patients still may need a shunt for relief of hydrocephalus. Radiation therapy is indicated except in patients less than three years old (in whom chemotherapy is warranted). Because of the propensity of the tumor to spread through the cerebral spinal fluid, the entire brain and spine should be imaged prior to surgery and CSF sampled intraoperatively prior to resection, or several weeks after surgery. Chemotherapy is used extensively following surgery and radiosurgery, particularly for recurrent tumor and is sometimes used even before radiation therapy. Multi-modality treatment with surgery, radiation therapy, and chemotherapy has resulted in marked improvement in outcome over the last twenty years. The five-year disease-free survival rate of older patients with non-disseminated disease and appropriate therapy including complete resection and cranial-spinal irradiation may exceed eighty percent.

Pilocytic astrocytoma

Pilocytic astrocytoma is a variety of astrocytoma that has a particularly favorable prognosis. They tend to occur in young adult life with a mean age of onset of 26 years. They present equally with signs of mass (headache and pappiladema from increased intracranial pressure), seizure, and neurologic deficits. MRI shows the tumor to be circumscribed in most cases. Complete resection is usually possible for superficially located tumors. This is often curative. Growing residual or recurrent tumor warrants radiosurgery

Pineal cell tumor

Pineal cell tumors, pineocytomas and pineoblastomas, are rare tumors of the pineal gland that may be either relatively benign (pineocytoma), of intermediate grade, or malignant (pineoblastoma). This range reflects both pathologic appearance and clinical course. The tumors tend to present with signs of increased intracranial pressure from obstructive hydrocephalus, double vision, and unstable gait. MRI scans show a uniformly enhancing tumor in the pineal region and posterior third ventricle. Surgical resection for tumor removal and reopening of the obstructive fluid pathways is indicated. After surgery, a search of the spine for disseminated tumor with MRI and CSF cytological analysis is important to treatment and outcome. A completely removed pineocytoma may not require radiation therapy. Pineoblastomas warrant craniospinal irradiation and chemotherapy.