Brain tumors: tumor types

Tumor types
There are more than 120 types of brain tumors. Today, most medical institutions use the World Health Organization (WHO) classification system to identify brain tumors. The WHO classifies brain tumors by cell origin and how the cells behave, from the least aggressive (benign) to the most aggressive (malignant). Some tumor types are assigned a grade, ranging from Grade I (least malignant) to Grade IV (most malignant), which signifies the rate of growth. There are variations in grading systems, depending on the tumor type. The classification and grade of an individual tumor help predict its likely behavior. This section describes the most frequently diagnosed types.

Acoustic neuroma
An acoustic neuroma is also known as a schwannoma, vestibular schwannoma, or neurilemmoma.

Characteristics
• Arises from cells that form a protective sheath around nerve fibers
• Typically grows around the eighth cranial nerve, but can be found around other cranial or spinal nerves

Symptoms
• Hearing loss in one ear
• Dizziness or vertigo
• Tinnitus (ringing in the ear)
• Tingling or numbness in the face
• Walking and balance problems
• Lack of coordination

Treatment. An acoustic neuroma may be observed in order to monitor its growth, or surgery may be performed. The goal of surgery is the complete removal of the tumor without harming the seventh cranial nerve (which controls facial movement) or causing hearing loss. Radiosurgery can be a viable option for many patients. This focused, high-energy radiation prevents the growth of acoustic neuromas, but actual shrinkage of the tumor may never occur or may take several months.

Astrocytomas
Grade I - Pilocytic astrocytoma
Also called Juvenile Pilocytic Astrocytoma (JPA)

Characteristics
• Slow growing, with relatively well-defined borders
• Grows in the cerebrum, optic nerve pathways, brain stem and cerebellum
• Occurs most often in children and teens
• Accounts for two percent of all brain tumors

Treatment. Surgery is the standard treatment. If the tumor cannot be completely resected, radiation or chemotherapy may be given. Chemotherapy may be given to very young children instead of radiation therapy to avoid damage to the developing brain. Some of these tumors can progress to a higher grade, so it is important to be diligent about following up with the medical team after treatment.

Grade II - low-grade astrocytoma
An astrocytoma is a type of glioma that develops from star-shaped cells (astrocytes) that support nerve cells. The WHO classifies a low-grade astrocytoma as a grade II tumor.

Characteristics

• Slow growing
• Rarely spreads to other parts of the CNS
• Borders not well defined
• Common among men and women in their 20s-50s

Treatment. Treatment depends on the size and location of the tumor. The doctor will most likely perform a biopsy or surgery to remove the tumor. Partial resections or inoperable tumors may be treated with radiation. Recurring tumors may require additional surgery, radiation and/or chemotherapy.

Grade III - anaplastic astrocytoma
An astrocytoma is a glioma that develops from star-shaped glial cells (astrocytes) that support nerve cells. An anaplastic astrocytoma is classified as a grade III tumor.

Characteristics
• Grows faster and more aggressively than grade II astrocytomas
• Tumor cells are not uniform in appearance
• Invades neighboring tissue
• Common among men and women in their 30s-50s
• More common in men than women
• Accounts for two percent of all brain tumors

Treatment. Treatment depends on the location of the tumor and how far it has progressed. Surgery and radiation therapy, with chemotherapy during or following radiation, are the standard treatments. If surgery is not an option, then the doctor may recommend radiation and/or chemotherapy. Many clinical trials (experimental treatments) using radiation, chemotherapy, or a combination are available for initial and recurrent anaplastic astrocytomas.

Grade IV - Glioblastoma multiforme (GBM)
Glioblastoma multiforme (GBM) is the most common and deadliest of malignant primary brain tumors in adults and is one of a group of tumors referred to as gliomas.

Classified as a Grade IV (most serious) astrocytoma, GBM develops from the lineage of star-shaped glial cells, called astrocytes, that support nerve cells.

GBM develops primarily in the cerebral hemispheres but can develop in other parts of the brain, brainstem, or spinal cord.

Because of its lethalness, GBM was selected as the first brain tumor to be sequenced as part of The Cancer Genome Atlas (TCGA Website), a national effort to map the genomes of the many types of cancer. In this effort, researchers discovered that GBM has four distinct genetic subtypes that respond differently to aggressive therapies, making treatment extremely difficult and challenging. Parallel research Parallel research at Johns Hopkins University also contributed to the expansion of genomic information on GBM.

Characteristics
• Can be composed of several different cell types
• Can develop directly or evolve from lower grade astrocytoma or oligodendroglioma
• Most common in older individuals and more common in men than women
• Less common in children
• Median survival rate of ~15 months; 5-year survival rate of ~4%
• The cause is unknown, but increasingly research is pointing toward genetic mutations

Incidence. The incidence, or the number of new diagnoses made annually is 2 to 3 per 100,000 people in the United States and Europe. GBM accounts for 12% to 15% of all intracranial tumors and 50% to 60% of astrocytic tumors.

Treatment. Standard treatment is surgery, followed by radiation therapy or combined radiation therapy and chemotherapy. If inoperable, then radiation or radiation/chemotherapy can be administered.

Treatment requires effective teamwork from neurosurgeons, neuro-oncologists, radiation oncologists, physician assistants, social workers, psychologists, and nurses. A supportive family environment is also helpful

Chordoma

Characteristics

• Rare and low grade
• Occurs at the sacrum, near the lower tip of the spine, or at the base of the skull
• Originates from cells left over from early fetal development
• Invades the bone and soft tissues but rarely the brain tissue
• Can block the ventricles, causing hydrocephalus (water on the brain)
• Can metastasize (spread) or recur

Symptoms

• Double vision
• Headaches

Treatment. Surgery and radiation therapy are the common forms of treatment. Chordomas at the base of the skull can be difficult to remove. Surgical resection may be possible if the tumor is located in the spine.

CNS Lymphoma

CNS Lymphoma is a type of cancer that develops in the lymphatic system. The lymphatic system is a network of small organs called lymph nodes and vessels (similar to blood vessels) that carry a clear, watery fluid called lymph throughout the body. This fluid supplies cells called lymphocytes that fight disease and infection. To correctly diagnose primary CNS Lymphoma, staging must be done. Staging is the process of using CT scanning to examine many parts of the body. Staging helps to confirm where the cancer originated and how far it has spread.

Characteristics

• Very aggressive
• Usually involves multiple tumors throughout the central nervous system (CNS)
• More common in people whose immune systems are compromised
• Often develops in the brain, commonly in the areas adjacent to the ventricles
• Can be primary (originating in the brain) or secondary
• Most common among men and women in their 60s-80s, but incidence is increasing in young adults
• More common in men than women
• Accounts for about two percent of all brain tumors

Symptoms

• Headaches
• Partial paralysis on one side of the body
• Seizures
• Cognitive or speech disorders
• Vision problems

Treatment. Radiation therapy, chemotherapy, and steroids are the most common forms of treatment. Surgery is rarely an option because there are usually multiple lesions. However, a biopsy at the start of steroid treatment can be critical to ensure the correct diagnosis.

Craniopharyngioma

Characteristics

• Most common in the parasellar region, an area at the base of the brain and near the optic nerves
• Also grows in the regions of the optic nerves and the hypothalamus, near the pituitary gland
• Tends to be low grade
• Often accompanied by a cyst
• Originates in cells left over from early fetal development
• Occurs in children and men and women in their 50s and 60s

Symptoms

• Headaches
• Visual changes
• Weight gain

• Delayed development in children

Treatment. Surgery is the most common treatment. Radiation therapy may be used.

Other gliomas
Brain stem glioma
Characteristics
• Named for its location at the base of the brain
• Can range from low grade to high grade
• Occurs most often in children between three and ten years of age, but can occur in adults

Symptoms
• Headaches
• Nausea
• Speech or balance abnormalities
• Difficulty swallowing
• Weakness or numbness of the arms and/or legs
• Facial weakness
• Double vision

Symptoms can develop slowly and subtly and may go unnoticed for months. In other cases, the symptoms may arise abruptly. A sudden onset of symptoms tends to occur with rapidly growing, high-grade tumors.

Treatment. Surgery may not be an option because the brain stem controls vital life functions and can easily be damaged. Radiation therapy can reduce symptoms and help slow the tumor's growth. Low-grade brain stem gliomas can have very long periods of remission.

Ependymoma
Ependymal tumors begin in the ependyma, cells that line the passageways in the brain where cerebrospinal fluid (CSF) is produced and stored. Ependymomas are classified as either supratentorial (in the cerebral hemispheres) or infratentorial (in the back of the brain). Variations of this tumor type include subependymoma, subependymal giant-cell astrocytoma, and malignant ependymoma. Ependymoblastoma, which occurs in infants and children under three years, is no longer considered a subtype of ependymoma. For ependymoblastoma, see primitive neuroectodermal tumor (PNET) in the Non-glial Tumors section.

Characteristics
• Usually localized to one area of the brain
• Develops from cells that line the hollow cavities at the bottom of the brain and the canal containing the spinal cord
• Can be slow growing or fast growing
• May be located in the ventricles (cavities in the center of the brain)
• May block the ventricles, causing hydrocephalus (water on the brain)
• Sometimes extends to the spinal cord
• Common in children, and among men and women in their 40s and 50s
• Occurrence peaks at age five and again at age 34
• Accounts for two percent of all brain tumors

Symptoms
• Severe headaches
• Nausea and vomiting
• Difficulty walking
• Fatigue and sleepiness
• Problems with coordination
• Neck pain or stiffness
• Visual problems

Treatment. The doctor will perform tests to determine if it has spread to the spinal cord. Surgery followed by radiation therapy is the usual course of treatment. A shunt may be needed to treat hydrocephalus caused by blockage

of the ventricles.

Mixed glioma
A mixed glioma is often a combination of an astrocytoma and an oligodendroglioma (see oligodendroglioma for more).

Characteristics
• Composed of two or more types of glioma cells
• Graded according to the most aggressive type of tumor cells
• Common among men and women in their 20s-50s
• Accounts for one percent of all brain tumors

Symptoms
• Headaches
• Seizures
• Weakness or paralysis
• Nausea and vomiting
• Visual problems
• Behavioral and cognitive changes

Treatment. Mixed gliomas are generally treated for the most anaplastic (cancerous) type of cell found in the tumor. For example, in the case of a tumor composed of an anaplastic astrocytoma and a low-grade oligodendroglioma, the treatment would be based on the anaplastic astrocytoma – the more aggressive of the two cell types.

Optic nerve glioma
Characteristics
• Named for its location on or near the nerve pathways between the eyes and the brain
• Can range from low grade to high grade
• Occurs most often in infants and children, but can occur in adults
• Symptoms
• Headaches
• Progressive loss of vision
• Double vision

Treatment. Surgery is standard treatment, usually followed by radiation therapy or chemotherapy. Chemotherapy may be given to very young children instead of radiation therapy to avoid damage to the developing brain.

Subependymoma
This tumor forms from ependymal cells, and is a variation of an ependymoma. Ependymal cells are cells line the passageway in the brain where cerebral spinal fluid (CSF) is produced and stored. Ependymal tumors are classified as either supratentorial (in the cerebral hemispheres) or infratentorial (in the back of the brain). See "Ependymoma" for more information.

Characteristics
• Slow growing
• Usually located in the fourth and lateral ventricles
• More common in men than in women

Symptoms
• Headaches
• Nausea
• Loss of balance
• Sometimes no symptoms occur and tumor is detected incidentally

Treatment. Surgery will be performed when possible. Radiation therapy may be used if the tumor progresses or recurs. A shunt may be needed to treat hydrocephalus (water on the brain).

Medulloblastoma
Characteristics

• A type of primitive neuroectodermal tumor (PNET) (see below)
• Often located in the cerebellum or near the brain stem
• Can spread to the spinal cord through the cerebrospinal fluid (CSF)
• May obstruct the fourth ventricle, causing hydrocephalus (water on the brain)
• Occurs most often in children under the age of ten, but may occur in adults
• Slightly more common in males than females

Symptoms
• Headaches
• Early morning vomiting
• Lethargy or sleepiness
• Lack of coordination
• Double vision
• Behavioral or personality changes
• Signs of pressure seen behind the eye when examined with an ophthalmoscope

Treatment. Surgery is the standard treatment when possible. Chemotherapy is usually part of the treatment plan. Radiation of the brain and spine is often recommended in adults and children over three years of age. A shunt may be needed to treat hydrocephalus. This tumor may recur years later if not totally resected.

Meningioma
These tumors grow from the meninges, the layers of tissue covering the brain and spinal cord. As they grow, meningiomas compress adjacent brain tissue. Symptoms are often related to this compression of brain tissue, which can also affect cranial nerves and blood vessels. In some cases, meningioma growth can also extend into the bones of the head and face, which may produce visible changes. Most meningiomas are considered nonmalignant or low grade tumors. However, unlike nonmalignant tumors elsewhere in the body, some of these brain tumors can cause disability and may sometimes be life threatening. In many cases, meningiomas grow slowly. Other meningiomas grow more rapidly or have sudden growth spurts. There is no way to predict the rate of growth of a meningioma or to know for certain how long a specific tumor was growing before diagnosis. Meningiomas are graded from low to high. The lower the grade, the lower the risk of recurrence and aggressive growth.

The WHO classification divides meningiomas into three grades:
Grade I: Benign Meningioma
Grade II: Atypical Meningioma
Grade III: Malignant (Anaplastic) Meningioma

Characteristics
• May arise after previous treatment from ionizing radiation or excessive x-ray exposure
• Common among women and men in their 40s-50s, but can occur at any age
• Twice as common in women as in men
• Accounts for 34 percent of all primary brain tumors
• In very rare cases, can invade the skull or metastasize to the skin or lungs
• Women with meningiomas can experience tumor growth during pregnancy
• In rare cases, multiple meningiomas can develop at the same time in different parts of the brain and/or spinal cord

Symptoms
• Seizures
• Headaches
• Nausea and vomiting
• Vision changes
• Behavioral and cognitive changes
• Sometimes no symptoms occur and tumor is detected incidentally

Treatment. If there are no symptoms, the doctor may monitor the tumor with MRIs. Otherwise, surgery is the standard treatment. If the tumor cannot be completely resected or if it recurs, radiation therapy may be given as well. Chemotherapy for unresectable, aggressive, atypical, or recurrent meningiomas is being tested through clinical trials. Follow-up scans are needed indefinitely, because meningiomas can recur years or even decades after treatment.

(This information was written, in part, by Nancy Conn-Levin, M.A. For more detailed information about meningiomas, please contact her at Mngioma634@aol.com)

Metastatic brain tumors (brain metastases)
A metastatic, or secondary, brain tumor is one that begins as cancer in another part of the body. Some of the cancer cells may be carried to the brain by the blood or lymphatic fluid, or may spread from adjacent tissue. The site where the cancerous cells originated is referred to as the primary cancer. Metastatic brain tumors are often referred to as lesions or brain metastases. Metastatic brain tumors are the most common brain tumors. There has been an increase in metastatic lesions as people are surviving primary cancers for longer periods of time.

Characteristics
• The primary cancer is usually in the lung, breast, colon, kidney, or skin (melanoma), but can originate in any part of the body
• Most are located in the cerebrum, but can also develop in the cerebellum or brain stem
• More than half of people with metastatic tumors have multiple lesions (tumors)
• Common among middle-aged and elderly men and women

Symptoms
• Seizures
• Headaches
• Behavioral and cognitive changes
• Lack of coordination

Treatment. Surgery and radiosurgery are the standard treatments if lesions are limited in number and accessible. Both of these treatments may be followed by whole brain radiation therapy (WBRT). In cases of multiple lesions, WBRT alone may be given. Chemotherapy specific to the brain-located metastatic tumor may be used.

Oligodendroglioma
This tumor type develops from glial cells called oligodendrocytes.

Characteristics
• Occurs frequently in the frontal or temporal lobes
• Can be classified as low grade or high grade
• Common among men and women in their 20s-40s, but can occur in children
• More common in men than women
• Accounts for two percent of all brain tumors
• May be associated with 1p or 19q chromosomal losses

Symptoms
• Seizures
• Headaches
• Behavioral and cognitive changes
• Weakness or paralysis

Treatment. Treatment options depend on the grade of the tumor. If the tumor is low grade and symptoms are not severe, the doctor may decide to perform surgery, then “watch and wait” and evaluate tumor growth through MRIs. There is a malignant form called anaplastic oligodendroglioma and a mixed malignant astrocytoma-oligodendroglioma. The common treatment for these high-grade tumors is surgery followed by radiation therapy and/or chemotherapy. Both low- and high-grade oligodendrogliomas can recur. If a tumor recurs, the doctor will evaluate it for a second surgical procedure, radiation, and/or chemotherapy. Gene expression studies are used to classify gliomas based on certain characteristics, or genetic profiles. Oligodendrogliomas can be identified by deficiencies in certain chromosomes named 1p and 19q. Genetic profiling of oligodendrogliomas provides a more accurate predictor of prognosis and treatment options than does standard pathology.

Pituitary tumors
The pituitary gland produces hormones that affect growth and the functions of other glands in the body. Certain pituitary tumors secrete abnormally high amounts of their respective hormones and cause related symptoms. Other

pituitary tumors do not secrete hormones, but grow and compress brain tissue, causing other symptoms.

Characteristics
• Named for its location on or near the pituitary gland, located at the center of the brain behind and above the nose
• Can range from low grade to high grade
• May cause excessive secretion of hormones
• Common among men and women in their 50s-80s
• Accounts for about 13 percent of all brain tumors

Symptoms
• Headache
• Depression
• Vision loss
• Nausea or vomiting
• Behavioral and cognitive changes
• Cessation of menstrual periods (amenorrhea)
• Leaking of fluid from the breasts (galactorrhea)
• Hair growth in women
• Impotence in men
• Abnormal growth of hands and feet
• Abnormal weight gain

Treatment. If the tumor is large or compressing the optic nerve, standard treatment is surgery. This can be transphenoidal surgery, which gets access to the tumor by entering through the nasal passage. Radiation therapy may also be used. Some pituitary tumors may be treated with medication, and/or observed with MRI scans. Certain drugs can block the pituitary gland from making too many hormones. Follow up with an endocrinologist may be necessary to manage hormonal changes.

Primitive neuroectodermal tumor (PNET)
There are several tumor types in this category. Names of specific PNETs may be based on the tumor location. Examples include pineoblastoma (located in the pineal region, a form of pineal tumor), medulloblastoma (located in the cerebellum), and cerebral cortex PNET (located in the cerebral cortex).

Characteristics
• Highly aggressive and tend to spread throughout the CNS
• Grow from undeveloped brain cells
• Commonly include cysts and calcification (calcium deposits)
• Tend to be large

Symptoms
• Can vary depending on location of tumor
• Weakness or change in sensation on one side of the body
• Morning headache or headache that goes away after vomiting
• Nausea and vomiting
• Seizures
• Unusual sleepiness or lethargy
• Behavioral or personality changes
• Unexplained weight loss or weight gain

Treatment. Surgery is the standard treatment when possible. In adults and children over three years of age, surgery may be followed by radiation therapy to the whole brain and spinal cord, and chemotherapy. In children under three years of age, surgery may be followed by chemotherapy or a clinical trial of chemotherapy to delay or reduce the need for radiation therapy.

Other brain-related conditions
Cysts. A cyst is a fluid-filled sac that may encapsulate (surround) or be located next to a tumor. Based on its location in the brain, a cyst can cause symptoms such as headache, pain, seizures, or a neurological deficit. Cysts can be

surgically removed or drained. If a cyst is not causing neurological difficulties, the doctor will conduct ongoing MRI and CT scans to observe its growth pattern.

Neurofibromatosis. Neurofibromatosis is a genetic disorder that can cause tumors in various parts of the central nervous system (CNS). There are two types of neurofibromatosis. Type 1, the more common kind, usually occurs outside of the CNS. Type 2 occurs within the CNS. Type 2 neurofibromatosis causes multiple CNS tumors, including neurofibromas, multiple meningiomas, bilateral vestibular schwannomas, optic nerve gliomas, and spinal cord tumors. Symptoms include loss of balance, tinnitus, total hearing loss, facial pain or numbness, and headache. Surgery is the standard treatment.

Pseudotumor cerebri. This condition is not a brain tumor, but its symptoms mimic a brain tumor. Pseudotumor Cerebri most commonly afflicts obese adolescent girls and young women. Symptoms include headaches, blurred vision, dizziness, and a slight numbness of the face. The symptoms are caused by a buildup of cerebrospinal fluid (CSF). Treatment is given to relieve the symptoms, particularly visual impairment. Treatment may include repeated lumbar punctures or medications to decrease CSF. In severe cases, a shunt may be needed.

Tuberous sclerosis. Tuberous Sclerosis is a genetic disorder that causes numerous neurological and physical symptoms, including tumors of the CNS, eyes, and kidneys. Most cases occur in children under 20 years of age. About fifty percent of tuberous sclerosis patients develop brain tumors. Subependymal giant-cell astrocytomas are the most common type, but other tumor types are also associated with this condition. Most patients suffer from seizures. Treatment may involve inserting a shunt to prevent hydrocephalus (water on the brain). Surgery is another treatment option.

Schwannoma
Also known as vestibular schwannoma, neurilemmoma and acoustic neuroma (see acoustic neuroma).

Characteristics
• Arises from cells that form a protective sheath around nerve fibers
• Typically grows around the eighth cranial nerve, but can be found around other cranial or spinal nerves

Symptoms
• Reduced hearing in the ear on the side of the tumor when eighth cranial nerve is involved Tinnitus (ringing in the ear)
• Balance problems
• Deficits depend on the nerve that is affected

Treatment. Surgery and radiotherapy are the most common forms of treatment. If the tumor is not completely removed, recurrence is likely.

The following tumor types are more common in children than in adults:
• Brain Stem Glioma
• Craniopharyngioma
• Ependymoma
• Juvenile Pilocytic Astrocytoma (JPA)
• Medulloblastoma
• Optic Nerve Glioma
• Pineal Tumor
• Primitive Neuroectodermal Tumors (PNET)
• Rhabdoid Tumor

Keep in mind that many tumors have different subtypes; for example, an astrocytoma can be a juvenile pilocytic astrocytoma, an anaplastic astrocytoma or a glioblastoma. In addition, the same tumors sometimes have different names; even pathologists are not always consistent in what they call them. Finally, it is important to note that nonmalignant, or benign, brain tumors can be just as difficult to treat as malignant brain tumors.

This information was developed by the National Brain Tumor Society and is herewith used with permission.

National Brain Tumor Society. Tumor Types. Available at: https://braintumor.org/brain-tumo-

-information/understanding-brain-tumors/tumor-types/. Accessed September 4, 2018.