Neoplasms of the BRAIN and CNS

2015-2016 FCDS Educational Webcast Series
Steven Peace, BS, CTR
October 15, 2015

2015 Focus
• Anatomy
• SSS 2000
• MPH Rules
• AJCC TNM

Presentation Outline
• Overview
• Reportable CNS Neoplasms
• Anatomy of the Human Brain & CNS
• WHO Grade for Brain and CNS Neoplasms
• Multiple Primary and Histology Coding Rules
• AJCC TNM Stage and SEER Summary Stage 2000
• Site Specific Factors
Overview

Brain tumors are:

- **Primary “brain” tumors** - those that begin in the brain or central nervous system (or its supporting tissues) and tend to stay in the brain - occur in people of all ages, but they are statistically more frequent in children and older adults.

- **Metastatic “brain” tumors** – those that begin as a cancer elsewhere in the body and spread to the brain – are more common in adults than in children.

Source: American Brain Tumor Association Facts and Statistics http://abta.org
Brain tumors are:

- Usually described as intracranial neoplasms with varying behaviors (benign, borderline, malignant – ref. ICD-O-3)
- Include most identifiable structures within the cranium including the brain itself, small hormone-secreting ducts like the pineal and pituitary glands, the cranial nerves (primarily the optic nerve, olfactory nerve, acoustic nerve), the outer protective lining of the brain (meninges), and the spinal cord.
- Metastatic neoplasms are excluded
- Certain benign “tumors” are excluded
- No malignant neoplasms are excluded
- Benign bone tumors are excluded

Brain Tumor Characteristics

- Start in the brain and grow steadily there.
- Very rarely spread to other organs through the bloodstream.
- Are named for the anatomic location of tumor and/or the cells from which they arise, each having a certain function essential to normal physiological functioning of the brain.
- For example:
  - Brain Stem Gliomas arise in the lowest part of the brain.
  - Meningiomas arise in the meninges.
  - Gliomas arise from glial cells that support the CNS.
  - Astrocytomas arise from astrocytes
  - Ependymomas arise from ependymal cells which line the ventricles
  - Oligodendrogliomas arise from oligodendrocyte cells which make up the fatty substance called myelin that covers nerves like electrical insulation.
Characteristics of Brain Tumors

Range of Tumors and Symptoms

- There are over 120 different types of brain/CNS tumors.

- CNS tumors are associated with a range of symptoms and complications such as edema, seizures, endocrinopathy, fatigue, psychiatric disorder, venous thromboembolism that can seriously impact quality of life.

- Symptoms depend very much on the size and location of the tumor. General symptoms include persistent headaches which tend to be worse with activity, at night or early in the morning, convulsions, vomiting, subtle changes in personality, memory, mental ability, drowsiness, lethargy.
Brain Anatomy and Function

Tumor Location and Symptoms

- Symptoms are often tumor location specific or provide clues.
- Symptoms on the right side of the body may occur if the tumor is located on the left side of the brain and vice-versa.
  - The speech center in most people is on the left side of the brain. Symptoms of a tumor located here may include difficulty saying correct words while still capable of understanding what is being said.
  - If the tumor is located in the frontal lobe which controls intellectual function, thought process, behavior and memory, those activities may be affected.
- Similarity to closed head injury victims (motorcycle crash).
ALL Brain Tumors are Reportable

- Public Law 107-260, the Benign Brain Tumor Cancer Registries Amendment Act, [PDF-185KB] requires programs participating in the National Program for Cancer Registries (NPCR) to collect data on benign and borderline tumors of the central nervous system in addition to the previously required data on malignant tumors.

- In addition to NPCR, the National Cancer Institute's (NCI) Surveillance, Epidemiology and End Results (SEER) program and the American College of Surgeons' (ACoS) Commission on Cancer began requiring that these tumors be reported, starting with cases diagnosed on January 1, 2004.

BUT – Do Not Report Benign/Borderline Tumors that were Diagnosed BEFORE 1/1/2004 – NOT EVEN HISTORICAL

- This creates an ERROR for FCDS at Call for Data
- FCDS then has to Delete the Case and Adjust Sequences
ICD-O Topography Codes (Anatomic Site)

- Brain (C71.0 - C71.9)
- Cerebrum (C71.0)
- Frontal lobe (C71.1)
- Temporal lobe (C71.2)
- Parietal lobe (C71.3)
- Occipital lobe (C71.4)
- Ventricle (C71.5)
- Cerebellum (C71.6)
- Brain stem (C71.7)
- Overlapping lesion of the brain (C71.8)
- Brain NOS (C71.9)

- Meninges (C70.0 - C70.9)
  - Cerebral meninges (C70.0)
  - Spinal meninges (C70.1)
  - Meninges NOS (C70.9)
- Spinal cord (C72.0)
- Cauda equina (C72.1)
- Cranial nerves (C72.2 - C72.5)
  - Olfactory nerve (C72.2)
  - Optic nerve (C72.3)
  - Acoustic nerve (C72.4)
  - Cranial nerve NOS (C72.5)
- Other CNS (C72.6, C72.9)
- Pituitary gland (C75.1)
- Cricopharyngeal duct (C75.2)
- Pineal gland (C75.3)

Brain Tumor Characteristics

- Benign – Borderline – Malignant
- Patient Age
- Tumor Location
- Tumor Histologic Type
- WHO Grade of Primary Tumor

American Brain Tumor Association — http://www.abta.org
Tumor Behavior

- **Benign Tumors** • Slow growing • Distinct borders • Rarely spread

- **Malignant Tumors** • Usually rapid growing • Invasive • Life threatening

- **Borderline Malignant Tumors** • Rare • Likely to Recur following Surgical Resection • May become Life threatening

Common Histologic Types

- Astrocytoma, Grade I (Juvenile Pilocytic Astrocytoma)
- Astrocytoma, Grade II (Low Grade Pilocytic Astrocytoma)
- Astrocytoma, Grade III (Anaplastic Astrocytoma)
- Glioblastoma (Glioblastoma Multiforme/Astrocytoma, Grade IV)
- Ependymoma
- Medulloblastoma
- Meningioma
- Oligodendroglioma
- Oligoastrocytoma
- Pituitary Adenoma
WHO Classification Groups

- Tumors of Neuroepithelial Tissue
- Tumors of Cranial and Paraspinal Nerves
- Tumors of Meninges
- Tumors of Uncertain histogenesis
- Lymphomas and Hematopoietic Malignancies
- Germ Cell Tumors
- Cysts and Tumor-Like Lesions
- Tumors of the Sellar Region
- Metastatic Tumors
http://neuro-oncology.oxfordjournals.org/content/16/suppl_4.toc

ALL Tumors

Primary Site and Behavior

Malignant Only

http://neuro-oncology.oxfordjournals.org/content/16/suppl_4.toc
Distribution Primary Brain & CNS Tumors 2007-2011

Behavior and Histologic Type

Malignant Only

Non-Malignant Only

http://neuro-oncology.oxfordjournals.org/content/16/suppl_4.toc
Childhood Brain Tumors

Tentorium - extension of the dura mater separating the cerebellum from the occipital lobes

- 50% of childhood brain and CNS tumors are infratentorial, originating below the tentorium
- 20+% of childhood CNS tumors are located in the sellar or suprasellar region around the sella turcica (the bone that contains the pituitary gland)
- Remainder of tumors occur in spinal cord, brain stem, cranial nerves, etc.

Childhood Brain Tumors

<table>
<thead>
<tr>
<th>Supratentorial - childhood</th>
<th>Infratentorial - childhood</th>
</tr>
</thead>
<tbody>
<tr>
<td>Craniohypopharyngiomas.</td>
<td>Cerebellar astrocytomas (usually high-grade).</td>
</tr>
<tr>
<td>Diencephalic and hypothalamic gliomas.</td>
<td>Medulloblastomas (primitive neuroectodermal tumors).</td>
</tr>
<tr>
<td>Germ cell tumors.</td>
<td>Ependymomas (low-grade or anaplastic).</td>
</tr>
<tr>
<td>Low-grade astrocytomas.</td>
<td>Brain stem gliomas (high-grade or low-grade).</td>
</tr>
<tr>
<td>Anaplastic astrocytomas.</td>
<td>Atypical teratoid tumors</td>
</tr>
<tr>
<td>Glioblastoma multiforme.</td>
<td></td>
</tr>
<tr>
<td>Mixed gliomas.</td>
<td></td>
</tr>
<tr>
<td>Oligodendrogliomas.</td>
<td></td>
</tr>
<tr>
<td>Primitive neuroectodermal tumors.</td>
<td></td>
</tr>
<tr>
<td>Low-grade or anaplastic ependymomas.</td>
<td></td>
</tr>
<tr>
<td>Meningiomas.</td>
<td></td>
</tr>
<tr>
<td>Choroid plexus tumors.</td>
<td></td>
</tr>
</tbody>
</table>
WHO Grade for Brain/CNS Tumors

- WHO Tumor Grades – Grade I, II, III, and IV
- Higher the grade – the more malignant the tumor
- A tumor can contain more than one grade of cell
- Always record and code the highest tumor grade noted
- Record WHO Grade in SSF1 NOT in “Grade/Differentiation”

http://www.abta.org/brain-tumor-information/diagnosis/grading-staging.html

WHO Grade for Brain/CNS Tumors

LOW GRADE Neoplasms

- **Grade I:** least malignant tumors associated with long-term survival. They grow slowly and have an almost normal appearance when viewed through a microscope. Surgery alone may be an effective treatment for this grade tumor.

- **Grade II:** tumors are slow-growing and look slightly abnormal under a microscope. Some can spread into nearby normal tissue and recur, sometimes as a higher grade tumor.

http://www.abta.org/brain-tumor-information/diagnosis/grading-staging.html
WHO Grade for Brain/CNS Tumors

- **Grade III:** These tumors are, by definition, malignant although there is not always a big difference between grade II and grade III tumors. The cells of a grade III tumor are actively reproducing abnormal cells, which grow into nearby normal brain tissue. These tumors tend to recur, often as a grade IV.

- **Grade IV:** The most malignant tumors. Tumors reproduce rapidly, can have a bizarre appearance when viewed under the microscope, and easily grow into nearby normal brain tissue. These tumors form new blood vessels so they can maintain their rapid growth. They also have areas of dead cells in their centers.

http://www.abta.org/brain-tumor-information/diagnosis/grading-staging.html
<table>
<thead>
<tr>
<th>Name</th>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pilocytic astrocytoma</td>
<td>I</td>
<td>Common in children and young adults and in people with neurofibromatosis type 1 and rarely causes death</td>
</tr>
<tr>
<td>Diffuse astrocytoma</td>
<td>II</td>
<td>Also known as low-grade diffuse astrocytoma, it is common in young adults and in people with Li-Fraumeni syndrome. It commonly forms in the cerebrum but can form in any part of the brain. It grows slowly, often spreading into nearby tissues. It can progress to become an anaplastic astrocytoma or a glioblastoma</td>
</tr>
<tr>
<td>Anaplastic astrocytoma</td>
<td>III</td>
<td>Also known as malignant astrocytoma or high-grade astrocytoma, this kind of tumor is more often found in younger adults. It forms in the cerebrum. It grows quickly and spreads into nearby tissues. An anaplastic astrocytoma may transform and become a glioblastoma</td>
</tr>
<tr>
<td>Glioblastoma</td>
<td>IV</td>
<td>Also known as glioblastoma multiforme. About 50% of the gliomas are glioblastomas. It grows and spreads rapidly and has the highest degree of malignancy but poor prognosis</td>
</tr>
</tbody>
</table>

Adapted from American Brain Tumor Association (www.abta.org)
<table>
<thead>
<tr>
<th>Type of Tumor</th>
<th>5-Year Relative Survival Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Age</td>
</tr>
<tr>
<td></td>
<td>20-44</td>
</tr>
<tr>
<td>Low-grade (diffuse) astrocytoma</td>
<td>65%</td>
</tr>
<tr>
<td>Anaplastic astrocytoma</td>
<td>45%</td>
</tr>
<tr>
<td>Glioblastoma</td>
<td>17%</td>
</tr>
<tr>
<td>Oligodendroglioma</td>
<td>86%</td>
</tr>
<tr>
<td>Anaplastic oligodendroglioma</td>
<td>67%</td>
</tr>
<tr>
<td>Ependymoma/anaplastic ependymoma</td>
<td>51%</td>
</tr>
<tr>
<td>Meningioma</td>
<td>62%</td>
</tr>
</tbody>
</table>

ANATOMY OF THE HUMAN BRAIN

Source: National Geographic, courtesy of Fred Houder/Getty Images

Source: University of Illinois
Ventricular System of the Brain


Source: solarnavigator.net/human_brain
Midline Shift and Mass Effect

• The bony cranium protects the brain from outside impacts to the head. When swelling occurs in the brain, there isn’t much “give”.

• The swelling results in intracranial pressure and can cause a number of effects that begin to impact quality of life and comfort for the patient.

• The easiest way to describe midline shift is to bring to mind sitting in a movie theater. As soon as the person to one side of you puts his elbow onto the shared armrest between you, you tend to shift away.

Midline Shift and Mass Effect

• Midline is a central boundary separating the left and right hemispheres.

• Midline Shift – Tumor crosses the brain to shift across the center line

• Mass Effect is – Edema or swelling causes the brain to shift across center line

• Both create new symptoms at cross-over
  • Depends on the size and location of the tumor and level of spread
  • Edema caused by many things
  • Either cause pushes midline out of alignment

Source: Medscape
Meninges and Brain Stem

- The dura mater is the tough outer membrane.
- The arachnoid is the middle web-like membrane.
- The pia mater is the delicate, highly vascular innermost membrane.

Cranial Nerves

1. Olfactory (CN I)
2. Optic (CN II)
3. Oculomotor (CN III)
4. Trochlear (CN IV)
5. Trigeminal (CN V)
6. Abducens (CN VI)
7. Facial (CN VII)
8. Vestibulocochlear (CN VIII)
9. Glossopharyngeal (CN IX)
10. Vagus (CN X)
11. Spinal Accessory (CN XI)
12. Hypoglossal (CN XII)
### Cranial Nerve Functions

<table>
<thead>
<tr>
<th>Cranial Nerve:</th>
<th>Major Functions:</th>
</tr>
</thead>
<tbody>
<tr>
<td>I Olfactory</td>
<td>smell</td>
</tr>
<tr>
<td>II Optic</td>
<td>vision</td>
</tr>
<tr>
<td>III Oculomotor</td>
<td>eyelid and eyeball movement</td>
</tr>
<tr>
<td>IV Trochlear</td>
<td>turns eye downward and laterally, controls superior oblique muscles</td>
</tr>
<tr>
<td>V Trigeminal</td>
<td>chewing, face &amp; mouth touch &amp; pain</td>
</tr>
<tr>
<td>VI Abducens</td>
<td>turns eye laterally</td>
</tr>
<tr>
<td>VII Facial</td>
<td>facial expressions, taste, tears, saliva</td>
</tr>
<tr>
<td>VIII Vestibulocochlear</td>
<td>Also referred to as Auditory Nerve: hearing, equilibrium sensation</td>
</tr>
<tr>
<td>IX Glossopharyngeal</td>
<td>Taste, senses carotid blood pressure</td>
</tr>
<tr>
<td>X Vagus</td>
<td>aortic blood pressure, heart rate, stimulates digestive organs, taste</td>
</tr>
<tr>
<td>XI Spinal Accessory</td>
<td>controls trapezius &amp; sternocleidomastoid muscles, controls swallowing</td>
</tr>
<tr>
<td>XII Hypoglossal</td>
<td>controls tongue movements</td>
</tr>
</tbody>
</table>

### Sinus, Olfactory, Base of Skull Tumors

- Cancer Registries treat many of these as Head & Neck Neoplasms
- Some are intra-cranial but many are not intra-cranial or CNS
- Primary Site of Tumor Critical for Stage, Treatment, and Prognosis
- Histology is used to identify which are abstracted as Brain/CNS
- Highly specialized surgical procedures – location of tumor
- Some Prognostic Factors Overlap
Histologic Type - Glioma

- Most common category of primary brain tumors. They begin in glial cells (supporting cells of the CNS) – can be Grade I-IV not just III-IV.
- Often spread into surrounding brain tissue along nerve fibers invading the spaces between nearby normal brain cells. Some invade the surrounding brain more than others.
- Difficulty obtaining complete surgical removal. MRI scans show the largest part of the glioma, but cannot reliably show areas of the brain where tumor cells have invaded. Aggressive efforts to remove small numbers of tumor cells within the brain could cause loss of neurologic function.
- When it is not possible to remove the entire glioma, post-op radiation therapy and chemotherapy may be advised.
- Even with maximum safe resection followed by radiation and chemotherapy, gliomas can grow back.
Glioma – 3 Main Histologic SubTypes

1. **Astrocytoma:** In adults most often arise in the cerebrum. In children they occur in the brain stem, cerebrum and cerebellum. Rarely in brain stem in adults. Felt to be most aggressive of brain tumors.
   - Grade I and II astrocytomas are low-grade astrocytomas.
   - Grade III astrocytoma is an “anaplastic astrocytoma”.
   - Grade IV astrocytoma is a “glioblastoma multiforme”.

2. **Oligodendroglioma:** Rare tumor that usually occurs in the cerebrum, grows slowly and usually does not spread into surrounding brain tissue like astrocytoma does. Most common in middle-aged adults.

3. **Ependymoma:** Most commonly arise in children and young adults. They are also seen with neurofibromatosis Type II. (which we will discuss in a bit)
Glioma – Other Subtypes

There are other subtypes of gliomas, each with their own specific characteristics and modes of growth.

- Brain Stem Glioma
- Juvenile Pilocytic Astrocytoma
- Pleomorphic Xanthoastrocytoma
- Subependymoma
- Ganglioglioma

Glioma Tumor Markers

<table>
<thead>
<tr>
<th>Biomarker</th>
<th>Molecular Compartment</th>
<th>Purpose</th>
<th>Analytic Validity Demonstrated</th>
<th>Level of Evidence</th>
<th>NCCN Category of Evidence</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>1p/19q codeletion (unbalanced translocation)</td>
<td>Tumor DNA</td>
<td>Diagnostic (oligodendroglioma)</td>
<td>FISH, aCGH, LOH, MPLA</td>
<td>IA</td>
<td>1</td>
<td>Smith et al. 96</td>
</tr>
<tr>
<td>IDH mutation (IDH1 c.395 G→A p.R132H (IDH2)</td>
<td>Tumor DNA</td>
<td>Tumor protein</td>
<td>Positive is favorably prognostic; also a diagnostic marker</td>
<td>IHC, DNA sequencing</td>
<td>IIB</td>
<td>Houlilier et al. 98, Dubbink et al. 99</td>
</tr>
<tr>
<td>MGMT methylation</td>
<td>Tumor DNA</td>
<td>Prognostic, predictive (benefit for chemotherapy), pharmacodynamic (pseudorecurrence),</td>
<td>MS-PCR, MS-pyrosequencing, MS-MPLA</td>
<td>IIB</td>
<td></td>
<td>Hegi et al. 101, Gilbert et al. 103</td>
</tr>
</tbody>
</table>

Markers With Emerging Evidence

- BRAF fusion (pilocytic astrocytoma)
- CIMP (CpG island methylator phenotype)
Glioma Chromosome Alterations

<table>
<thead>
<tr>
<th>Chromosomal region</th>
<th>Type of alteration</th>
<th>Candidate glioma genes</th>
</tr>
</thead>
<tbody>
<tr>
<td>1p36.31-pter</td>
<td>Gains and deletions</td>
<td>Not known</td>
</tr>
<tr>
<td>1p36.22-q36.31</td>
<td>Gains and deletions</td>
<td>Not known</td>
</tr>
<tr>
<td>1p34.3-1p36.1</td>
<td>Gains and deletions</td>
<td>Not known</td>
</tr>
<tr>
<td>1q32</td>
<td>Gains</td>
<td>RIPK5, MDM4, PH3C2S and others</td>
</tr>
<tr>
<td>1q</td>
<td>Deletions</td>
<td>NEK1, NIMA</td>
</tr>
<tr>
<td>7p11.2-p12</td>
<td>Amplifications or gains</td>
<td>EGFR</td>
</tr>
<tr>
<td>9p21-9p24</td>
<td>Deletions</td>
<td>CDKN2</td>
</tr>
<tr>
<td>10q23</td>
<td>Deletions</td>
<td>PTEN</td>
</tr>
<tr>
<td>10q25-q26</td>
<td>Deletions</td>
<td>MGMT</td>
</tr>
<tr>
<td>11p</td>
<td>Deletions</td>
<td>Between CDKN1C and RB582</td>
</tr>
<tr>
<td>12q13-q15</td>
<td>Amplifications</td>
<td>MDM2, CDK4 and others</td>
</tr>
<tr>
<td>13q11-q13 and 13q14-q24</td>
<td>Loss</td>
<td>RB1, GLTSCR1, GLTSCR2, LGI1, P3CD2 and many others</td>
</tr>
<tr>
<td>19q13</td>
<td>Loss</td>
<td>GLTSCR1, GLTSCR2, LGI1, P3CD2 and many others</td>
</tr>
<tr>
<td>22q11.21-q12.2</td>
<td>Loss</td>
<td>28 genes, including INI1</td>
</tr>
<tr>
<td>22q13.1-q13.3</td>
<td>Loss</td>
<td>Not known</td>
</tr>
</tbody>
</table>

Non-Glial Tumors

- **Medulloblastoma:** Usually arises in the cerebrum, is the most common brain tumor in children, and is sometimes called a “primitive neuroectodermal tumor” or PNET.

- **Meningioma:** Arises from the meninges which are the outside coverings of the brain between the skull and the brain itself. It usually presses on the brain, but does not invade it and often grows slowly.
Meningioma

- Meningiomas are typically diagnosed by CT or MRI imaging.
- Biopsy may be considered for confirmation.
- Options stratified by presence/absence of symptoms and tumor size.
- Most asymptomatic patients with small tumors (<30mm) may just be observed. If neurological impairment is imminent, surgery (if accessible) or radiotherapy (EBRT OR SRS) is feasible.
- Asymptomatic tumors >30mm can be either resected or observed.

Non-Glial Tumors

- **Schwannoma**: Arises from Schwann cells present in certain nerves, including those that control balance and hearing. May be called “neuroma”.

  - A common site is the vestibular nerve which carries signals from the inner ear to the brain stem.

  - Tumors in this location are called “acoustic neuromas” (a.k.a. vestibular schwannoma), and occur most often in adults.
Non-Glial Tumors

- **Craniopharyngioma**: Grows at the base of the brain, arises from the tissue connecting the brain and the pituitary gland and occurs in both adults and children.

- **Pituitary Adenoma**: Arises from the pituitary gland and may cause compression of the optic nerves causing vision problems. Some produce excessive amounts of hormones that can disrupt the body’s metabolism.

Neurofibromatosis

- The neurofibromatoses (NF) are a group of genetic disorders which cause tumors to grow along nerves and can also affect the development of non-nervous tissues such as bones and skin.

- **Neurofibromatosis Type I (NF-I)**, also known as Peripheral NF and historically as von Recklinghausen Disease
  - Occurs in 1:4,000 births
  - Multiple cafe-au-lait spots (not reportable)
  - Many, many neurofibromas on or under the skin (not reportable)
  - Enlargement and deformation of bones and curvature of the spine
  - Tumors may develop in brain, on cranial nerves, or the spinal cord
NF Type I: First documented photo 1871

Source Credit: Dr. Stanley B. Burns
http://www.cbsnews.com/2300-204_162-10007019-6.html#ixzz1elFzAehl

Other Manifestations of NF Type I

Lisch nodules on the eye
- Melanocytic hemartomas

Café-au-lait spots on skin
- Discolored birth marks

Medscape Source: Dermnet.com; Dermatologic Manifestations of NF Type I
Neurofibromatosis Type II

- **Neurofibromatosis Type II (NFII)**, also known as Multiple Inherited Schwannomas, Meningiomas and Ependymomas (MISME) or Bilateral Acoustic Neurofibromatosis (BAN).

- Is a genetically inherited disease caused by mutations of the "Merlin" gene, which appears to influence the form and movement of cells.

- Primary manifestation is a development of non-malignant brain tumors in the region of the cranial nerves, frequently bilaterally. The eighth cranial nerve is the auditory-vestibular nerve which transmits sensory information from the inner ear to the brain and is commonly affected.

Source: California Ear Institute

Acoustic Neuroma/Schwannoma

Source: http://thrivingwithneurofibromatosis.blogspot.com
Multiple Primary Rules
Histology Coding Rules

Different Rules for Benign and Malignant
Sequence Numbering for Brain Tumors

- **Malignant primary brain and CNS tumors** are assigned Sequence Codes in the range 00-35
  - Sequence Chronologically 00-35
  - Only count malignant tumors in the sequence
  - If only one malignant tumor occurs, it is coded 00
  - If subsequent (multiple) primary malignant and/or in situ neoplasms, the sequence number for the first tumor begins at 01, the sequence number for the second primary tumor is 02, and so forth.

- **Non-malignant primary brain and CNS tumors** are assigned Sequence Codes in the range 60-87.
  - Sequence Chronologically 60-87
  - Only count benign/borderline or reportable by agreement neoplasms in the sequence
  - If only one non-malignant tumor occurs, it is coded 60.
  - If subsequent (multiple) non-malignant neoplasms are diagnosed, the first tumor should be sequenced as 61, the second 62 and so forth.

Benign and Borderline Tumor Rules
Benign and Borderline Tumor Rules

- When multiple tumors are present registrars should identify and document specific characteristics for MPH Rules Text
  - Date of Diagnosis (Timing is not used to determine number of abstracts or primary neoplasms to abstract)
  - Method and Details of Diagnosis (some are never resected)
  - Location of Tumor
  - Laterality
  - Histologic Type – refer to Chart 1
  - Tumor Behavior
  - Multiple Meningioma’s (meningiomatosis)
  - Neurofibromatosis Characteristics (when applicable)

Malignant Tumor Rules

Chart 1 – Neuroepithelial Malignant Brain and Central Nervous System Tumors

Note: This chart is based on the 1999 Classification of Tumors of the Brain and Central Nervous System. The chart is not a complete listing of histologies that exist within the primary or central nervous system.

Chart instructions: Use this chart to code histology. The tree is arranged in descending order. Each branch is a histologic group starting at the top with the broad specific terms and descending into more specific terms.
Malignant Tumor Rules

- When multiple tumors are present registrars should identify and document specific characteristics for MPH Rules Text
  - Date of Diagnosis (Timing is not used) to determine number of abstracts or primary neoplasms to abstract
  - Method and Details of Diagnosis (most attempt resection)
  - Location of Tumor (not spread or invasion — but bulk of tumor)
  - Histologic Type — refer to Chart 1 and/or Chart 2
  - Tumor Behavior
  - Variations or Combinations of One or More Glial Tumors Over Lifetime — astrocytoma, glioblastoma, ependymoma, or oligodendroglioma
    - Special rules for determining # abstracts
    - Special rules for determining whether or not is mixed glioma
  - Note: Recurrence, progression, or any reappearance of histologies on the same branch in Chart 1 or Chart 2 is always the same disease process.
Do NOT Report Benign Tumors Dx’d < 2004

REMINDER: Sequence numbers for malignant neoplasms and for benign, borderline, and other reportable-by-agreement cases are usually assigned over a lifetime.

• HOWEVER,

WHEN A PATIENT WAS DIAGNOSED WITH A NON-MALIGNANT CNS NEOPLASM BEFORE REPORTING WAS REQUIRED (January 1, 2004),

THE FIRST NEOPLASM IS NOT REPORTABLE TO FCDS EVEN AS A HISTORICAL CASE. THE NEW (SECOND) NEOPLASM SHOULD BE ASSIGNED SEQUENCE NUMBER 60

DO NOT REPORT THE BENIGN NEOPLASM IF DIAGNOSED BEFORE 1/1/2004

THIS HAS BEEN A SOURCE OF CONFUSION — PLEASE REVIEW AND SAVE

FCDS Data Acquisition Manual and CDC Data Collection of Primary Central Nervous System Tumors

Staging Brain and CNS Neoplasms
“Attempts at developing a TNM-based classification and staging system for tumors of the central nervous system have not been successful.” Basic TNM Concepts are not applicable to brain and CNS sites. “It continues to be the recommendation of the ASJCC CNS Tumor Task Force that a formal classification and staging system not be attempted.”

AJCC Stage = 88 (Not Applicable)
Includes: ANY Benign or ANY Malignant Neoplasm

Factors Felt to be of Prognostic and/or Clinical Significant Include:
• Tumor Histology
• Location of Tumor
• Unifocal or Multifocal
• WHO Grade of Tumor
• Patient Age at Diagnosis
• Functional Neurologic Status *(KPS)
• Primary or Recurrent Tumor
• Extent of Resection
• Metastatic Spread Proliferative Fraction (Ki-67, M1 B-1)
• Gene Deletions (1p, 19q)
• MGMT Methylation
To obtain a FREE electronic copy of the SS2000 Manual:
Steps to Assign SS2000

Three summary stage groups can be ruled out quickly: in situ, distant, and localized

**FIRST**

In situ

1. **Rule out in situ stage disease.** Carcinomas and melanomas are the only types of cancer that can be classified as in situ. Only carcinomas have a basement membrane. Sarcomas are never described as in situ. A pathologist must examine the primary organ and state that the tumor is in situ. If the cancer is anything except a carcinoma or melanoma, it cannot be in situ.

2. If there is any evidence of invasion (or extension to), nodal involvement or metastatic spread, the case is not in situ even if the pathology report so states. This is a common error in staging cervical cancer where the path report states that the cancer is “in situ with micrometastasis.” Such a case would be staged as localized.

In Situ Stage is Not Applicable for Brain/CNS Tumors

In Situ Stage is Not the Same as Benign or Borderline Behavior

SS2000 for Benign/Borderline Tumors

= 8
Three summary stage groups can be ruled out quickly:
in situ, distant, and localized

SECOND

3. **Rule out distant disease.** If metastases can be documented, there is no need to spend a great deal of time identifying local or regional spread. If distant metastases are recorded on x-ray or needle biopsy, the stage is already determined and the patient does not need to undergo a lot of other tests.

4. Hematopoietic diseases, such as leukemias and multiple myeloma, are considered disseminated or distant at time of diagnosis.

5. Rule out distant spread by reading the operative report for comments about seedling, implants, liver nodules, or other indications of metastases. Read diagnostic reports for references to distant disease.

SECOND

Brain and CNS these are usually CSF Involvement – cells in fluid

On rare occasion you may see “drop metastasis” – code as Distant Stage

THIRD

7. **Rule out that the cancer is “confined to the organ of origin.”** In order for a lesion to be classified as localized, it must not extend beyond the outer limits of the organ and there must be no evidence of metastases anywhere else.

8. Terms such as “blood vessel invasion” or “perineural lymphatic invasion” do not necessarily indicate that the cancer has spread beyond the primary organ. If tumor at the primary site has invaded lymph or blood vessels, there is the potential for malignant cells to be transported throughout the body. Step 1 (invasion), has occurred, but not necessarily steps 2 (transport of cancer cells) and 3 (growth at the secondary site). The case may still be localized.

9. Vascular invasion within the primary is not a determining factor in changing the stage unless there is definite evidence of tumor at distant sites.

Most common Summary Stage – unless tumor crosses midline
Steps to Assign SS2000

Three summary stage groups can be ruled out quickly: in situ, distant, and localized

FINAL (if needed)

10. If in situ, local and distant categories have been ruled out, the stage is regional.
11. For carcinomas, if there are lymph nodes involved with the tumor, the stage is at least regional.
12. For tissues, structures, and lymph nodes, assume ipsilateral unless stated to be contralateral or bilateral.

Additional Resources

- Central Brain Tumor Registry of the United States (CBTRUS), 2015
- NCCN Evidence Based Treatment Guidelines, nccn.org, 2015
- Multiple Primary and Histology Coding Rules, SEER 2007
- SEER Summary Staging Manual 2000
QUESTIONS ??