

Case 1: CNS

History and Physical Examination

See Radiation Oncology Consult

Imaging

9/14 CT of the brain: 3.5 x 1.8 cm enhancing left hemisphere mass with significant surrounding edema.

9/14 CT of the chest, abdomen, and pelvis: no abnormalities detected.

9/14 PET scan of brain: no abnormalities detected.

9/17 MRI, brain and brain stem: There is a posterior left temporal ring-enhancing mass and significant surrounding edema involving much of the left temporal lobe and hemisphere. The size and configuration of the ventricular system is within normal limits. Right hemispheric cortical sulci as well as central subarachnoid cisterns are symmetric and unremarkable. No additional lesions are noted.

Impression: Left posterior temporal lobe lesion. Primary considerations would be for primary or metastatic brain tumor.

Procedures

9/21 Craniotomy with partial resection and Gliadel wafer implantation.

Pathology

9/21 Glioblastoma multiforme, giant cell type.

Radiation Oncology Consult

9/27 History of Present Illness: This pleasant 58-year-old male was in his usual state of good health until recently. He had been experiencing subtle difficulties with word-finding, misspelling words, concentration, and reading over the last month or so. He underwent initial evaluation at another facility. Work-up included a CT scan of the brain, which showed a left hemisphere mass. He was taken to the operating room on 9/21 and underwent what sounds like a partial resection with Gliadel wafer implantation.

9/27 Physical Exam: This is a pleasant healthy-appearing male resting comfortably in no acute distress. His head is shaved. He has a bandage in place over the left side of his scalp. His pupils were equal, round and reactive to light. Extraocular movements were intact. There was no scleral icterus. Cranial nerves II–XII were grossly intact bilaterally. Sensation was grossly intact in all four extremities. Negative Romberg. Finger-to-nose testing was normal. Rapid alternating movements were normal. He was able to spell “world” backwards without any difficulty. Short-term memory, however, was 0/3. He answered all questions appropriately and speech was fluent. He was able to give the names of multiple objects without difficulty.

Impression: 58-year-old male with a new diagnosis of glioblastoma multiforme. Karnofsky Performance Score is estimated at 90. He underwent a partial resection with placement of Gliadel wafers. We recommend postoperative external beam radiation therapy for a total of approximately six weeks. Additionally, the patient has discussed chemotherapy with the medical oncologist and it sounds as if he was planning for concurrent Temozolomide followed by additional adjuvant Temozolomide

Radiation Oncology Treatment Summary

12/8 Overall, the patient tolerated his treatments well. The patient did complain of having some fatigue as well as headaches in the morning. The patient did develop seizures during the course of his treatment, however, his seizure medication was adjusted and he has not had any seizure since.

Date Treatment Started: 10/18

Date Treatment Ended: 12/8

National Program of Cancer Registries Education and Training Series
How to Collect High Quality Cancer Surveillance Data

Answers

Case 1 CNS	Answer	Rationale
Date of Dx	9/17	MRI; <i>FORDS</i> , p. 89
Primary Site	C71.2	MRI; <i>FORDS</i> , p. 91
Laterality	2	MRI; <i>FORDS</i> , p. 92
Sequence Number-Central	00	Autopsy; <i>SEER Program Coding and Staging Manual (PCSM) 2004</i> , p. 69
Histology	9441/39	Partial resection path; <i>SEER Program Coding and Staging Manual (PCSM) 2004</i> , histology coding rules for single tumors #6, p. 87
CS Extension	10	MRI; <i>Collaborative Staging (CS) Manual</i> , p. 603
CS Lymph Nodes	88	<i>CS Manual</i> , p. 605
CS Mets at Dx	00	<i>CS Manual</i> , p. 605
Surg Primary Site	40	Partial resection, oncology consult; <i>FORDS</i> , p. 281
Scope Reg LN Surg	9	<i>FORDS</i> , p. 138
Surg Proc/Other Site	0	<i>FORDS</i> , p. 142
Rad Reg Treatment Mod	20	<i>FORDS</i> , p. 155
Chemotherapy	02	<i>FORDS</i> , p. 171; single agent administered by implantation of wafer
Hormone Therapy	00	<i>FORDS</i> , p. 175
Immunotherapy	00	<i>FORDS</i> , p. 179
Hem Tsplt & End Proc	00	<i>FORDS</i> , p. 183
Other Treatment	0	<i>FORDS</i> , p. 186

Case 2: CNS

History and Physical Examination

4/29 History of Present Illness: This 62-year-old female has visual problems and had a visual exam done last week. She notes that her vision has deteriorated over the last month and is worse in the right eye. She reports having an occasional headache but these respond to Ibuprofen. Her son has noted some personality changes over the last one to two months. Her husband notes decreased initiative in looking for employment and an increase in fatigue level. She denies any other neurological abnormalities.

4/29 Physical Exam: She is alert, oriented times three and in no acute distress. Her speech is clear. She has a flat affect. Vision in the right eye is 20/200. Vision in the left eye is 20/50. She has a temporal field deficit in the right eye to finger count confrontation. Otherwise cranial nerves II through XII are intact. Muscle strength is strong. Sensation is intact. Deep tendon reflexes are 1–2 and symmetrical. Her gait is stable. All other systems are normal.

Imaging

4/27 MRI, brain: There is an extra-axial well-defined homogeneously enhancing mass, measuring 7.2 x 7.2 x 5.2 cm, attached to the anterior cranial fossa. The lesion is consistent with olfactory groove meningioma. It compresses the anterior horn of the lateral ventricles bilaterally and stretches the optic chiasm and optic nerves inferiorly. The pituitary stalk and lamina terminalis are also stretched posteriorly. No hydrocephalus is noted.

Impression: Olfactory groove meningioma.

Procedures

5/20 Bifrontal craniotomy for resection of olfactory groove meningioma. As the frontal lobes were retracted superiorly and laterally, the tumor easily came into view. It was dissected free from the surrounding brain using bilateral electrocautery and patties. After several hours of meticulous dissection of the tumor sparing the capsule and performing several internal debulkings, the tumor was removed in its entirety. There were no areas of significant damage to the surrounding brain. The tumor was the size of a tangerine. To facilitate removal the ultrasonic aspirator had to be used.

Pathology

5/20 Gross: A. Multiple friable tan irregularly shaped soft tissue fragments measuring 1.8 x 1.4 x 0.4 cm in aggregate. B. Multiple irregularly shaped friable soft tan-red and brown tissue fragments measuring 8.5 x 7.8 x 2.5 cm in aggregate.

Microscopic: A, B. The tumor is composed of nests, vague lobules and fascicles of cells with mildly spindled plump nuclei with bland cytologic features. Cell borders are indistinct. The chromatin is finely dispersed. No mitoses are identified. The lesion is cellular, but no necrosis, increased NC ratio, prominent nucleoli, or sheet-like growth patterns are identified. No brain invasion is seen.

Diagnosis: A, B. Brain, bifrontal tumor, excision: meningioma.

National Program of Cancer Registries Education and Training Series
How to Collect High Quality Cancer Surveillance Data

Answers

Case 2 CNS	Answer	Rationale
Date of Dx	4/27	MRI; <i>FORDS</i> , p. 89
Primary Site	C70.0	MRI; <i>FORDS</i> , p. 91
Laterality	9	MRI, olfactory groove meningioma arises in the midline; <i>FORDS</i> , p. 92
Sequence Number-Central	60	Autopsy; <i>SEER Program Coding and Staging Manual (PCSM) 2004</i> , p. 69
Histology	9530/09	Resection path; <i>SEER Program Coding and Staging Manual (PCSM) 2004</i> , histology coding rules for single tumors #1, p. 86
CS Extension	05	Resection path; <i>Collaborative Staging (CS) Manual</i> , p. 603
CS Lymph Nodes	88	<i>CS Manual</i> , p. 605
CS Mets at Dx	00	<i>CS Manual</i> , p. 605
Surg Primary Site	55	Resection op report, tumor was removed in its entirety; <i>FORDS</i> , p. 281
Scope Reg LN Surg	9	<i>FORDS</i> , p. 138
Surg Proc/Other Site	0	<i>FORDS</i> , p. 142
Rad Reg Treatment Mod	00	<i>FORDS</i> , p. 155
Chemotherapy	00	<i>FORDS</i> , p. 171
Hormone Therapy	00	<i>FORDS</i> , p. 175
Immunotherapy	00	<i>FORDS</i> , p. 179
Hem Tsplt & End Proc	00	<i>FORDS</i> , p. 183
Other Treatment	0	<i>FORDS</i> , p. 186

Case 3: CNS

History

4/4/XX This 21-year-old female had a midline pineal mixed germ cell tumor excised seven years ago. At that time, embryonal carcinomatous, seminomatous, and teratomatous components were identified. At that time, she also had associated hydrocephalus on CT scan, and a ventriculo-peritoneal shunt was inserted. She now has lesions of the right parieto-parasagittal area and left temporal area and undergoes stereotactic biopsy of the parieto-parasagittal area.

Procedures

4/4/XX Parieto-parasagittal stereotactic biopsy

Pathology

4/4/XX Gross: The first specimen is designated "tumor-center" and consists of multiple fragments of brain tissue measuring 1.5 cm in aggregate diameter. They are submitted in toto as A. The second specimen is designated "tumor at edge". The third specimen is designated as "tumor at edge" with no further distinguishing designations from the previous. The fourth specimen is designated "tumor at edge" with no distinguishing designation from the previous specimens. These three specimens are submitted to the Laboratory for Millipore filtration.

Microscopic: Sections reveal portions of blood and portions of brain parenchyma with increased numbers of glial cells. These astrocytic cells have a protoplasmic appearance in most of the biopsy; however there are focal fibrillary features. There is no evidence of anaplasia or necrosis. Immunoperoxidase staining is positive for vimentin, glial fibrillary acidic protein, and S-100 protein, and negative for cytokeratin, alpha fetoprotein, beta human chorionic gonadotropin, and placental alkaline phosphatase.

Final Diagnosis: Brain, right parieto-parasagittal region, stereotactic biopsy; low-grade astrocytoma.

Autopsy

Date: 04/17/XX

Clinical History

The patient was a 21-year-old white female who was admitted on April 14, XX in a coma. This young woman was found to have a mixed germ cell tumor located in the pineal region at age 14. The neoplasm was successfully treated with partial resection, irradiation, and chemotherapy. At age 15 she was found to have right lung metastases which were treated with irradiation, chemotherapy, and resection. She then did until late March XX when she developed aphasia. She returned to the hospital for evaluation and was found to have right parasagittal and a left temporal frontal mass. Stereotactic biopsy was performed on April 4, XX, which was diagnosed as a low-grade astrocytoma. The patient was scheduled for left craniotomy with tumor biopsy, debulking, and partial lobectomy on April 18. However the patient became increasingly more lethargic over the day of admission. When she was found to be unarousable by her parents, she was brought to the hospital where she was admitted directly to the Intensive Care Unit.

Physical examination on admission was remarkable for unresponsiveness except to pain and posturing in response to pain. Vital signs were stable at normal levels. The left pupil was fixed and dilated while the right was less dilated and reactive. There were no purposeful movements or localization.

The clinical impression at the time of admission was that the patient suffered left uncal herniation secondary to the known left temporal frontal mass. She was intubated and treated with hyperventilation, Mannitol, and Decadron. By the following morning the patient had awakened, responded to her family,

and was able to take liquids. However, her level of consciousness subsequently diminished. No further aggressive measures were undertaken and death occurred on April 17, XX at 12:45 PM.

Summary and Interpretation

The autopsy confirmed the presence of residual mixed germ cell tumor in the pineal region with fibrosis and differentiation to ganglioglioma. Malignant gliomatosis was also present with diffuse infiltration of the cerebral hemispheres consistent with gliomatosis cerebri. This malignant gliomatosis developed after several symptom free years following irradiation and chemotherapy for the mixed germ cell tumor of the pineal region. The cause of death in this case is attributed to gliomatosis cerebri which caused bilateral uncus herniation, cerebellar tonsillar herniation, herniation of the left cingulate gyrus and right hippocampal gyrus, and compression of the mid brain and anterior lobe of the cerebellum.

Neuropathology Report

Gross Examination: The cerebral hemispheres are diffusely swollen, more so on the left hemisphere than the right. The gyri over the convex surfaces of the hemispheres are diffusely flattened. The sulci are in general obliterated. The right superior frontal gyrus, anteriorly, the gyri over the left temporo-parieto-occipital region, and the left superior temporal gyrus throughout its extent are particularly broadened or swollen. There is a small defect at the anterior part of the right superior frontal gyrus, which represents the site of previous biopsy. As the base of the brain, the left uncus shows marked herniation, while the right uncus shows mild herniation. The posterior portion of the right hippocampal gyrus also shows mild herniation into the incisura of the tentorium; the herniated portion has compressed and the medial surface of the right anterior lobe of the cerebellum. The tegmentum of the midbrain on that side is also markedly compressed. The arteries at the base of the brain are unremarkable. The optic nerves appear to be somewhat swollen. Coronal sections through the frontal gyrus. It measures approximately 2 cm anteroposteriorly, 2 cm transversely and 3–4 cm dorsoventrally. It involves the cortex of the superior frontal gyrus extending to the central white matter and has reached the white matter of the cingulate gyrus and corpus callosum. The right cerebral hemisphere is generally compressed due to a marked expansion of the left cerebral hemisphere. The left cingulate gyrus is herniated beneath the cerebral falx and the midline structures are shifted to the right as a result of marked swelling of the left hemisphere. In the left cerebral hemisphere, there is a massive greenish-gray, almost semi-translucent, soft lesion which has occupied most of the posterior temporal and parietal white matter extending to the occipital white matter. The lesion extends medially to involve the entire lateral wall of the atrium, then central white matter of the temporal lobe up to the plane of the subthalamic nuclei. Anterior to this plane, the temporal white matter over the roof of the temporal horn, mesial temporal region and the cortex and white matter of the superior temporal gyrus are mainly affected. The lesion extends almost to the temporal pole. The left frontal white matter is diffusely swollen as compared with the right despite that the white matter texture appears to be normal. The corpus callosum is somewhat more voluminous than usual. The splenium of the corpus callosum is completely degenerated. A part of the degenerated splenium is replaced by a lobulated grayish-white nodule which measures approximately 1.5 x 1.0 cm involving mainly the right half of the genu; it protrudes to the atrium. Near the nodule, the fornix is somewhat degenerated more so on the right than the left. The internal veins are widely patent but surrounded by some rusty brown-colored tissue, attached to this gray-white mass is another small nodule which is pedunculated and appears to be the pineal gland. It measures approximately 0.8 x 0.5 x 0.5 cm. It is situated between the internal cerebral veins and the tectum of the midbrain where the pineal gland sits. The ventricles are patent, although somewhat narrowed. Transverse sections through the brain stem disclose marked compression of the midbrain from side to side, particularly at the tegmentum and tectum on the right. There is focal necrosis in the tectum. The cerebellum again shows severe compression of the dorsal surface with scaphoid compression and relative elevation of the superior vermis. The cerebellar tonsils are mildly herniated. Transverse sections through the cerebellum and brain stem disclose no significant abnormalities. The spinal cord shows no significant abnormalities.

Microscopic Examination: Sections from the pineal show an irregularly lobulated, small mass between the two internal cerebral veins, which is surrounded by the splenium of the corpus callosum, the posterior extremities of the hippocampi, the pulvinar, and the superior colliculi. The mass has three distinct portions. The first is made up of fibrillary glial tissue with little or no pineal parenchymal cells. The second is dense collagenous lobulated fibrous tissue with an island of fibrocartilage and a few glandular structures. The third is lobulated viable tumor composed of well-differentiated neurons of varying sizes and clusters of poorly differentiated glial (or neural) cells. The latter lobule occupies the most dorsal portion involving the right side of the splenium of the corpus callosum protruding into the atrium. All three portions have multifocal calcifications or calcospherites. The mass is surrounded by leptomeninges. Also noted are the tela choroidea of the third ventricle and the pineal recess lined by the ependymal cells.

The splenium of the corpus callosum shows multicystic degeneration in its midpoint and marked fibrillary astrogliosis which separates the tumor nodule and the areas of cystic degeneration. The left side of the corpus callosum shows diffuse infiltration by a malignant glioma which is to be described below. The malignant gliomatous infiltration and the nodular tumor described above are clearly separated by areas of cystic degeneration and old gliosis with many fibrillary astrocytes containing hemosiderin granules.

Sections from the right frontal, left temporo-parieto-occipital region, and left superior temporal gyrus, where there was massive gross tumor infiltration, show highly cellular tumor infiltration in which the tumor cells are relatively uniform and small. They have round or elongated, hyperchromatic nuclei and very scanty cytoplasm with or without bipolar processes. In places they tend to aggregate around blood vessels and many areas show cystic degeneration with rare frank necrosis. Acute hemorrhage is noted in the right frontal mass at the site of previous biopsy.

Both the white matter and the cortex are diffusely infiltrated in and near the massive tumor infiltration. The tumor cells in many areas have extended to the ???????? region where they form multiple foci or bands regardless of the apparent tumor infiltration in the underlying cortex.

Many sections from the right and left frontal white matter, corpus callosum and temporal white matter with or without gross swelling show tumor cell infiltration in varying densities. Some areas show mild diffuse infiltration, particularly in the left cerebral hemispheres, while others, the tumor cells are sparsely scattered. Atypical cells having hyperchromatic, bizarre nuclei and scanty cytoplasm are scattered singly or in small clusters. They are observed in the white matter of almost all sections from the left cerebral hemisphere and the right frontal lobe, while the right parietal and occipital lobes appear to be spared. There are clusters of capillaries with severe fibrosis of their walls in the right frontal white matter near the tumor mass. Also noted is focal acute ischemic degeneration of the cortex of the right superior temporal gyrus.

The right basal ganglia and thalamus are also spared, while the left basal ganglia and thalamus are infiltrated by tumor cells in varying degrees.

The tectum of the midbrain shows mild tumor cell infiltration and petechial hemorrhages while the pons, medulla and spinal cord are spared. The cerebellum, on the other hand, shows occasionally scattered atypical glial cells in the white matter suggestive of tumor cell infiltration. The leptomeninges are, however, spared in all areas.

Final Neuropathologic Diagnosis

Mixed germ cell tumor of pineal region

- a. Status seven years post surgical resection
- b. Status post X-irradiation and chemotherapy
- c. Residual mixed germ cell tumor with fibrosis and differentiation to ganglioglioma

Gliomatosis cerebri

- a. Status post three months post right frontal biopsy
- b. Herniation of bilateral unci, right hippocampal gyrus, left cingulate gyrus and cerebellar tonsils
- c. Compression of midbrain and anterior lobe of cerebellum

Notes/Comments: This 21-year-old woman had a mixed germ cell tumor in the pineal region which was partially resected about 7 years before her demise. Subsequently, the patient received X irradiation and chemotherapy. On autopsy examination of the brain, there was a fibrosed mass with gliosed area and a viable tumor nodule in the pineal region representing the residual tumor. The glandular structure and fibrocartilage in the fibrosed mass and dense gliosis indicates the residue of the original germ cell tumor and pineal gland which had undergone X-irradiation changes. The viable portion of the tumor has the features of ganglioglioma which probably represents the differentiation of a primitive neuroectodermal portion of the germ cell tumor or its gangliogliomatous portion resistant to the therapies. In any case, both the residual fibrosed germ cell tumor and ganglioglioma are distinct from the malignant glioma which has diffusely infiltrated the cerebral hemispheres. The poor differentiation of the latter tumor cells with the minority being GFAP positive and the pattern of diffuse infiltration with multiple mass formations are consistent with gliomatosis cerebri. The malignant gliomatosis must have developed after six years of a relatively symptom free period following X-irradiation and chemotherapy for the original mixed germ cell tumor.

National Program of Cancer Registries Education and Training Series
How to Collect High Quality Cancer Surveillance Data

Answers

Case 3 CNS	Answer	Rationale
Date of Dx	4/4	Biopsy path; <i>FORDS</i> , p. 89
Primary Site	C71.8	Autopsy, gross, soft lesion occupies posterior temporal and parietal white matter extending to occipital white matter; <i>FORDS</i> , p. 91
Laterality	2	Autopsy, gross; <i>FORDS</i> , p. 92
Sequence Number-Central	02	Autopsy; <i>SEER Program Coding and Staging Manual (PCSM) 2004</i> , p. 69
Histology	9381/39	Autopsy; <i>SEER Program Coding and Staging Manual (PCSM) 2004</i> , coding instructions #2, p. 85, use path from procedure that resected the majority of the primary tumor
CS Extension	50	Autopsy, involves left cerebral hemisphere, right frontal lobe, left basal ganglia, thalamus, midbrain, and cerebellum; <i>Collaborative Staging (CS) Manual</i> , p. 604
CS Lymph Nodes	88	<i>CS Manual</i> , p. 605
CS Mets at Dx	00	<i>CS Manual</i> , p. 605
Surg Primary Site	90	Autopsy; <i>FORDS</i> , p. 281
Scope Reg LN Surg	9	<i>FORDS</i> , p. 138
Surg Proc/Other Site	0	<i>FORDS</i> , p. 142
Rad Reg Treatment Mod	00	<i>FORDS</i> , p. 155
Chemotherapy	00	<i>FORDS</i> , p. 171
Hormone Therapy	00	<i>FORDS</i> , p. 175
Immunotherapy	00	<i>FORDS</i> , p. 179
Hem Tsplt & End Proc	00	<i>FORDS</i> , p. 183
Other Treatment	0	<i>FORDS</i> , p. 186

Case 4: CNS

History and Physical Examination

The patient is an 11-year-old male with a myriad of neurologic symptoms. His parents reported changes in personality and clumsiness in a young man who previously was a good athlete. Some speech difficulties were noted.

Imaging

10/3 CT scan of brain: Enhancement in frontal lobe.

Procedures

10/10 Craniotomy with brain biopsy. Mass, 3 cm removed, but residual tumor visible.

Pathology

10/10 Gross: Right frontal mass, 3.2 x 3.3 x 1.9 cm.

Microscopic: Malignant glial tumor that infiltrates the leptomeninges and dura. It contains both astrocytic and oligodendroglial differentiation as well as areas of necrosis, vascular proliferation, and calcification.

Diagnosis: Right frontal lobe, mixed glioma, astrocytoma and oligodendroglioma, grade III.

Oncology

10/17 Patient began treatment with Procarbazine, CCNU, and Vincristine today.

National Program of Cancer Registries Education and Training Series
How to Collect High Quality Cancer Surveillance Data

Answers

Case 4 CNS	Answer	Rationale
Date of Dx	10/10	Biopsy; <i>FORDS</i> , p. 89
Primary Site	C71.1	CT scan; <i>FORDS</i> , p. 91
Laterality	1	Biopsy; <i>FORDS</i> , p. 9
Sequence Number-Central	00	Autopsy; <i>SEER Program Coding and Staging Manual (PCSM) 2004</i> , p. 69
Histology	9382/33	Biopsy path; <i>SEER Program Coding and Staging Manual (PCSM) 2004</i> , histology coding rules for single tumors #3, p. 86
CS Extension	60	Biopsy path, tumor invades leptomeninges and dura; <i>Collaborative Staging (CS) Manual</i> , p. 604
CS Lymph Nodes	88	<i>CS Manual</i> , p. 605
CS Mets at Dx	00	<i>CS Manual</i> , p. 605
Surg Primary Site	20	Biopsy, residual tumor visible; <i>FORDS</i> , p. 281
Scope Reg LN Surg	9	<i>FORDS</i> , p. 138
Surg Proc/Other Site	0	<i>FORDS</i> , p. 142
Rad Reg Treatment Mod	00	<i>FORDS</i> , p. 155
Chemotherapy	03	<i>FORDS</i> , p. 171
Hormone Therapy	00	<i>FORDS</i> , p. 175
Immunotherapy	00	<i>FORDS</i> , p. 179
Hem Tsplt & End Proc	00	<i>FORDS</i> , p. 183
Other Treatment	0	<i>FORDS</i> , p. 186

Pathology Report (Case 5: CNS)

Collected: 11/10/XX

Diagnosis: Cerebellopontine angle, left, "tumor", excision. Schwannoma.

History: 30-year-old with acoustic neuroma.

Tissue Submitted: A. Left CPA tumor

Gross: Received in formalin is a 1.5 x 1.0 x 0.5 cm aggregate of yellow pink soft tissue. Bagged all in A1.

Microscopic: Sections show the mass to be comprised of hypercellular bands of spindled cells oriented in various directions. No significant cytologic atypia or mitoses are identified. The majority of the specimen represents the Antoni A patterns of schwannoma. Verocay bodies are identified.

Radiology Report (Case 5: CNS)

MRI

Date: 5/17/XX

History: Persistent severe headache, fever, thrombocytopenia. Evaluate for meningitis, intracranial bleed.

Technique: MRI of the brain and brainstem without relevant comparison study. Sequences include sagittal T1 pre and post contrast, axial T1, axial T1 fat-sat post contrast, coronal T1 post contrast, axial FLAIR, axial T2, axial gradient, axial diffusion and ADC, axial CISS. 10 cc gadolinium IV contrast administered.

Findings: There is an incidental left cerebellopontine angle comma shaped enhancing mass centered at the CP angle involving the proximal half of the internal auditory canal with mild expansion. The distal internal auditory canal, inner ear structures are not involved. The signal characteristics are isointense on T1 and T2 without additional abnormal signal, measuring 1.0 x 1.0 x 0.7 cm. The remainder of the study is unremarkable. The supratentorial, posterior fossa, brainstem, sella parenchyma are unremarkable. No large or small vessel ischemic change, areas of abnormal parenchymal or leptomeningeal enhancement, or parenchymal masses are identified. No intracranial hemorrhage. The calvarium, skull base, temporal bones with mild right mastoid inflammatory change, intratemporal fossa, and orbits are unremarkable. There is moderate left maxillary sinus mucosal thickening. There is mild rightward deviation of the nasal septum.

Impression

1. Incidental left internal auditory canal schwannoma (acoustic neuroma), with otherwise unremarkable exam. No findings to account for the clinical concerns.
2. Mild inflammatory change of the left maxillary sinus and right mastoid air cells.

Radiology Report (Case 5: CNS)

Exam: CT, Brain, W/O Contrast

Date: 5/2/XX

History: 30-year-old with thrombocytopenia and coagulopathy now with headaches.

Technique: Multiple transaxial images of the brain, from skull base to vertex, were obtained without intravenous contrast administration. No similar prior studies are available for comparison.

Findings: There are no intra or extra-axial masses or fluid collections. There is no mass-effect, or midline shift. The ventricular system is of normal size and configuration. The posterior fossa is grossly unremarkable. The bony calvarium is unremarkable. Scans through the orbits and paranasal sinuses are unremarkable.

Impression: Unremarkable study of the brain.

Radiology Report (Case 5: CNS)

Exam: Chest – PA and Lateral

Date: 5/12/XX

History: AML on induction chemotherapy and pancytopenia, rule out infiltrate.

Findings/Impression: PA and lateral views of the chest are presented with prior comparison not available. There are no active infiltrates or effusions. Cardiomeastinal silhouette is within normal limits with no pulmonary vascular congestion. Left PICC line tip overlies the superior vena cava.

Operative Report – Otolaryngology (Case 5: CNS)

Surgery Date: 11/10/XX

Preoperative Diagnosis: Left acoustic neuroma.

Operation/Procedure: Left middle fossa craniotomy for excision of tumor and temporalis fascia graft; intraoperative ABR monitoring and direct cochlear monitoring; facial nerve monitoring; use of operating microscope.

Postoperative Diagnosis: Left inferior vestibular nerve schwannoma (1 cm).

Description of Operation/Procedure: The patient was taken to the operating room on 11/10/XX. Following the induction of general anesthesia, she was intubated by the Anesthesia Service staff without difficulty. Subsequently, the table was turned 180 degrees. The hair above the left ear was shaved. A reverse C-incision was marked out above the left ear measuring 6 x 6 cm. This was injected with 4 cc of 1% lidocaine with epinephrine 1:100,000. A facial nerve monitor was placed to monitor the left face. An ABR monitor was also set up. Both of these monitors gave good signaling prior to the procedure. She was then prepped and draped in the sterile fashion.

We began by making a marked incision using a 15 blade. The skin flap was raised posteriorly using electrocautery. A 4 x 5 cm piece of temporalis fascia was harvested and saved on the back table in sterile saline. We raised an anteriorly based temporalis muscle flap. The root of the zygoma was identified and the muscle was elevated from it. A 4 x 5 cm craniotomy flap was marked out and drilled using #4 cutter and #4 diamond burrs. The bone flap was raised from the dura. The dura was elevated posteriorly, anteriorly, and superiorly from the craniotomy site. All bleeding was controlled using bipolar cautery and Oxycel. The dura was then elevated from posterior to anterior on the floor of the middle fossa. Extra bone was drilled from the edges of the craniotomy site.

The operating microscope was advanced into the field and used for the remainder of the procedure. The remainder of the dura was elevated from the floor of the middle fossa, revealing the arcuate eminence, petrosal ridge and greater superficial petrosal nerve. The House-Urban retractor was placed beneath the petrous ridge at the approximate site of the internal auditory canal. We identified the superior semicircular canal and blue-lined it. We then drilled the bone medially, identifying the internal auditory canal. This was traced distally, identifying Bill's bar and the labyrinthine branch of the facial nerve. All of the bone was removed posteriorly and anteriorly to the internal auditory canal, revealing the dura in these areas.

At that point, the ABR monitor gave good signaling. We opened the dura using a 5910 Beaver blade. The superior vestibular nerve was separated from the facial nerve and truncated distally. The tumor of the inferior vestibular nerve was identified and separated from the facial nerve. We identified the cochlear nerve and separated the tumor from that. The tumor was debulked at times to facilitate dissection of the nerves from its surface. Finally, the entire tumor was dissected away from the nerves and removed. At that point, our direct cochlear nerve monitor had good function, as did an ABR. The facial nerve stimulated 0.1 milliamperes, giving both facial nerve movement as well as a good signal from the facial nerve monitor.

The area was thoroughly irrigated with sterile saline. A piece of temporalis muscle was harvested and placed into the dural defect, sealing it. This was coated with a layer of DuraSeal. The temporalis fascia was laid over the floor of the middle fossa. This was coated again with a layer of DuraSeal. The bone flap was laid back into the craniotomy site. The temporalis muscle was closed using 3-0 Vicryl. The skin flap was closed with a deep layer of 3-0 Vicryl and the top layer with interrupted horizontal mattress sutures of 3-0 nylon. The patient was awakened from general anesthesia and extubated while in the operating room.

Findings: Left inferior vestibular nerve schwannoma (1cm).

Discharge Summary – Otolaryngology (Case 5: CNS)

Admission Date: 11/10/XX

Discharge Date: 11/14/XX

Reason for Admission: Treatment of left sided acoustic neuroma.

Primary Diagnosis: Left acoustic neuroma.

Other Pertinent Diagnoses: Acute promyelocytic leukemia.

Brief History and Physical: 30-year-old female with a history of acute promyelocytic leukemia diagnosed in April XX. She underwent an MRI during her hospitalization because she had headaches and was found to have a lesion in her left internal auditory canal and cerebellar pontine angle. This lesion was about 1.0 x 1.0 x 0.7 cm in diameter. She was fairly asymptomatic from this lesion. Her hearing was normal with an SRT of 20 in the left and 10 on the right with 96% word understanding. She had intermittent ringing in her left ear and some unsteadiness on quick turn, but did not know if that was due to her other disease process. After it was deemed that she was sufficiently recovered from her chemotherapy treatment, she was consented for left middle cranial fossa approach for excision of acoustic neuroma after the goals, risks, benefits, and possible complications were explained to her.

Hospital Course: The patient was admitted to the Department of Otolaryngology on 11/10/XX. She was taken to the operating room and a middle cranial fossa approach was used to excise a left sided acoustic neuroma. She was observed in the intensive care unit overnight and there were no complications that evening. She was transferred to the floor the following day. Postoperatively, she exhibited facial nerve weakness on the left. Her diet was advanced as expected, and she began ambulating without assistance at an acceptable rate. She was discharged home on 11/14/XX in stable condition, afebrile, tolerating a regular diet, and ambulating without assistance. Patient was discharged after MRI was accomplished.

Condition on Discharge: Stable.

Consultation Note – Neurology (Case 5: CNS)

Date: 4/29/XX

Chief Complaint: Migraine headaches.

History of Present Illness: Patient is a very pleasant 30-year-old woman who was newly diagnosed as AML M3 earlier this month when she noted spontaneous bruising, lower extremity petechiae, and was found to have a platelet count of 23. As a result, the patient was referred to the Hematology Clinic for further evaluation. Bone marrow biopsy was positive for acute leukemia and the cytology was suggestive of AML M3. She was thus started on ATRA, Doxorubicin, Dexamethasone and Cytarabine. Since her admission, her migraines became more frequent requiring more medication for control. Previously she used to have migraines every few months to years and used to go away with Imitrex. She also said that her current headaches have been of the same severity but more frequent than her usual. Currently headaches are constant exacerbating on a daily basis; Imitrex and Phenergan do help but the headache recurs rather quickly.

She reports that previously she was never on a standing medication for prevention yet she was tried on Amitriptyline, which she discontinued because of how it made her feel, as well as Inderal, which dropped her blood pressure significantly. Otherwise only Imitrex was being used.

Past Medical History:

1. Migraines
2. Mild depression
3. Echocardiogram showed patent foramen ovale and mitral valve prolapse. (The echocardiogram was performed when the patient presented with vision loss. The vision loss was eventually attributed to fluid behind the retina.)

Review of Systems:

Constitutional: (-) Eye: (-) HEENT: (-) CV: (-) Lungs: (-) GI: (-)
Musculoskeletal (-) Psychiatric (-) Endocrine (-) Allergic/immunologic: (-)
Lymph/blood (-) Skin (-)

Family History: No known family history of headaches.

Social History: The patient is single and works as a nurse. There is no history of smoking, alcohol, or drug use.

Physical Exam

General Appearance: In no acute distress; Eye: normal sclerae and conjunctivae; ENT: normal mucosa, no thrush, no neck masses; Respiratory: no rales, no wheezing, no dullness to percussion; Cardiovascular: normal S1 S2, no gallop, no murmur, normal distal pulses; GI: no organomegaly, normal bowel sounds, non-tender; no CVA tenderness; MS: no edema or muscle tenderness; Skin: no rash or bruises.

Neurology Exam

General Appearance: In no acute distress. Mental status: oriented to person, place, and time. Level of alertness: normally alert. Affect: normal range of emotions. Appropriateness: normal. Language: normal word choice and pronunciation. Memory: normal recent and remote memory. Attention span and concentration: normal. Fund of knowledge: normal.

National Program of Cancer Registries Education and Training Series
How to Collect High Quality Cancer Surveillance Data

Fundi: Benign, without papilledema or vascular changes. CNII: intact visual fields. CN III, IV, VI: full and conjugate eye movements; pupils normal. CN V: normal facial sensation to pin and touch. CN VII: full and symmetric facial movements. CN VIII: hearing intact bilaterally. CN IX, X: normal palatal movement. CN XI: trapezius, SCM strength intact. CN XII: tongue is midline.

Motor Strength: Full and symmetric throughout. Pronator drift: none. Muscle tone: normal. Sensation: normal to pin and touch. Reflexes: full and reasonably symmetric throughout. Coordination: generally intact. Station and gait: normal.

Heart: Regular rhythm without murmurs. Carotids: no bruits appreciated bilaterally. Jaw movements: smooth and non-painful. Neck and shoulders: no significant tenderness and stiffness; no true trigger points.

Impression: 30-year-old woman with newly diagnosed AML M3 with worsening migraines. CT without contrast was done on 5/2 negative for any bleed.

Plan: We would advise avoiding narcotics for management of headaches as that could potentially exacerbate rebound headaches.

Since the patient is immune compromised we would recommend MRI of the brain to check for meningeal signs of enhancement that may indicate possible meningitis. It is rather worrisome that she is a risk of meningitis, though a spinal tap would have some risk with the patient's low platelet count. If she develops any new symptoms in addition to her headache and fever, then empiric antibiotic coverage would be indicated if a spinal tap could not be performed. Coverage for fungal as well as bacterial meningitis would be considered. As for symptomatic treatment of the patient's headache now, we would recommend starting Benadryl 25 mg IV Q8 hours and increasing the dose as needed to 50 mg. If pain does not resolve in 24–48 hours, then DHE might be considered.

Teaching Statement: I have interviewed and examined the patient and confirm the pertinent findings. I have discussed the case with the resident/fellow and agree with the findings and plan as documented.

Staff Physician Comments: I personally interviewed and examined the patient. I have confirmed the above key findings on the history of migraine headaches in a 30-year-old woman who is undergoing chemotherapy for recently diagnosed AML. Her current headache has not responded to PRN analgesics noted above with the headache more severe than usual. Key findings on my current physical examination show a somnolent woman who was recently given analgesics for headache. She was arousable and could follow commands without difficulty but would drift back to sleep when not stimulated. The neurologic exam noted above was confirmed by me. There was no hint of nuchal rigidity. I have discussed the impression of recurrent migraine headaches vs. meningitis in an immune compromised host, which can occur without the classic symptoms of stiff neck, headache, and fever. I have reviewed the current recommendations with the Neurology Consult Resident. Symptomatic treatment with IV Diphenhydramine as described above can be tried at this time; however, I would have a very low threshold to provide antibiotic coverage for this patient, as meningitis is a high risk in this immune compromised patient.

Clinical Notes – Family Care Center (Case 5: CNS)

Date: 11/22/XX

Chief Complaint: Headache.

History of Present Illness: This is a pleasant 30-year-old woman. She has a history of migraine headaches that frequently have an aura. She experiences the pain on the left side of the head of an aching throbbing nature with some mild photophobia and nausea. She generally has obtained effective relief of these in the past with Sumatriptan. She recently had surgical removal of an acoustic neuroma from the left ear. She has some residual neurologic deficits, including a left-sided facial paresis and possibly permanent loss of the hearing function on the left side. She developed a headache soon after the surgery, which did not start with an aura, but was otherwise typical for her migraine type headaches, i.e., was on the left side of the head throbbing in character and accompanied by mild photophobia and nausea. She was seen recently by Ear, Nose and Throat who evaluated her for possible complications of surgery and ruled these out. Per the patient's report, they discussed therapy with Neurology and she was discharged with Percocet, Vistaril, Promethazine, and Ativan. The latter was to attenuate some side effects of the Vistaril, which sound like akathisia. She states that the medications have lowered her head pain from a 10 to a 4–5 and this is acceptable for her, but she is running low of medications. She also wonders if there is something else that can be tried that will block her headache out altogether. She did use some Sumatriptan earlier in the course of the headache and obtained some relief from it; she has not tried both medication regimens together. No other concerns.

Family History: Per the record above, no interval change.

Social History: Per the record above, no interval change.

Past Medical History: per the record above, no interval change.

Review of Systems: Negative except as per HPI.

Physical Exam: This is a pleasant young woman sitting in a darkened room. She is alert, oriented and appropriate, and gives an excellent history. She is afebrile; pulse and blood pressure are unremarkable. She has evidence of surgical incision on the left side of her head, but the surgical wound appears clean and healing well. She has an obvious left-sided facial paresis including the frontalis muscle. The pupils are equal and reactive to light; extraocular movements are full. The fundi are benign with sharp disc margins. Her gait is somewhat unsteady but she can walk without assistance. No further exam today.

Impression: Migraine headache complicated by postsurgical pain.

Plan: We will put her back on her Imitrex, 50 mg as an initial dose followed by two repetitions if relief is not obtained. She is cautioned not to use more than three doses in 24 hours. She is also given renewals for her Phenergan, Vistaril, Ativan, and Percocet, which she will take as she has been using. We will have her follow-up for a routine visit with her regular doctor in the next several weeks. If she is not obtaining satisfactory headache relief, or if the headache changes in severity or quality, she will seek urgent medical attention.

National Program of Cancer Registries Education and Training Series
How to Collect High Quality Cancer Surveillance Data

Answers

Case 5 CNS	Answer	Rationale
Date of Dx	5/17	MRI; <i>FORDS</i> , p. 89
Primary Site	C72.4	MRI; <i>FORDS</i> , p. 91
Laterality	2	MRI; <i>FORDS</i> , p. 92
Sequence Number-Central	60	Autopsy; <i>SEER Program Coding and Staging Manual (PCSM) 2004</i> , p. 69
Histology	9560/09	Excision of tumor path; <i>SEER Program Coding and Staging Manual (PCSM) 2004</i> , histology coding rules for single tumors #1, p. 86
CS Extension	05	Excision of tumor path; <i>Collaborative Staging (CS) Manual</i> , p. 607
CS Lymph Nodes	88	<i>CS Manual</i> , p. 608
CS Mets at Dx	00	<i>CS Manual</i> , p. 609
Surg Primary Site	55	Excision of tumor op report, entire tumor dissected; <i>FORDS</i> , p. 281
Scope Reg LN Surg	9	<i>FORDS</i> , p. 138
Surg Proc/Other Site	0	<i>FORDS</i> , p. 142
Rad Reg Treatment Mod	00	<i>FORDS</i> , p. 155
Chemotherapy	00	<i>FORDS</i> , p. 171
Hormone Therapy	00	<i>FORDS</i> , p. 175
Immunotherapy	00	<i>FORDS</i> , p. 179
Hem Tsplt & End Proc	00	<i>FORDS</i> , p. 183
Other Treatment	0	<i>FORDS</i> , p. 186