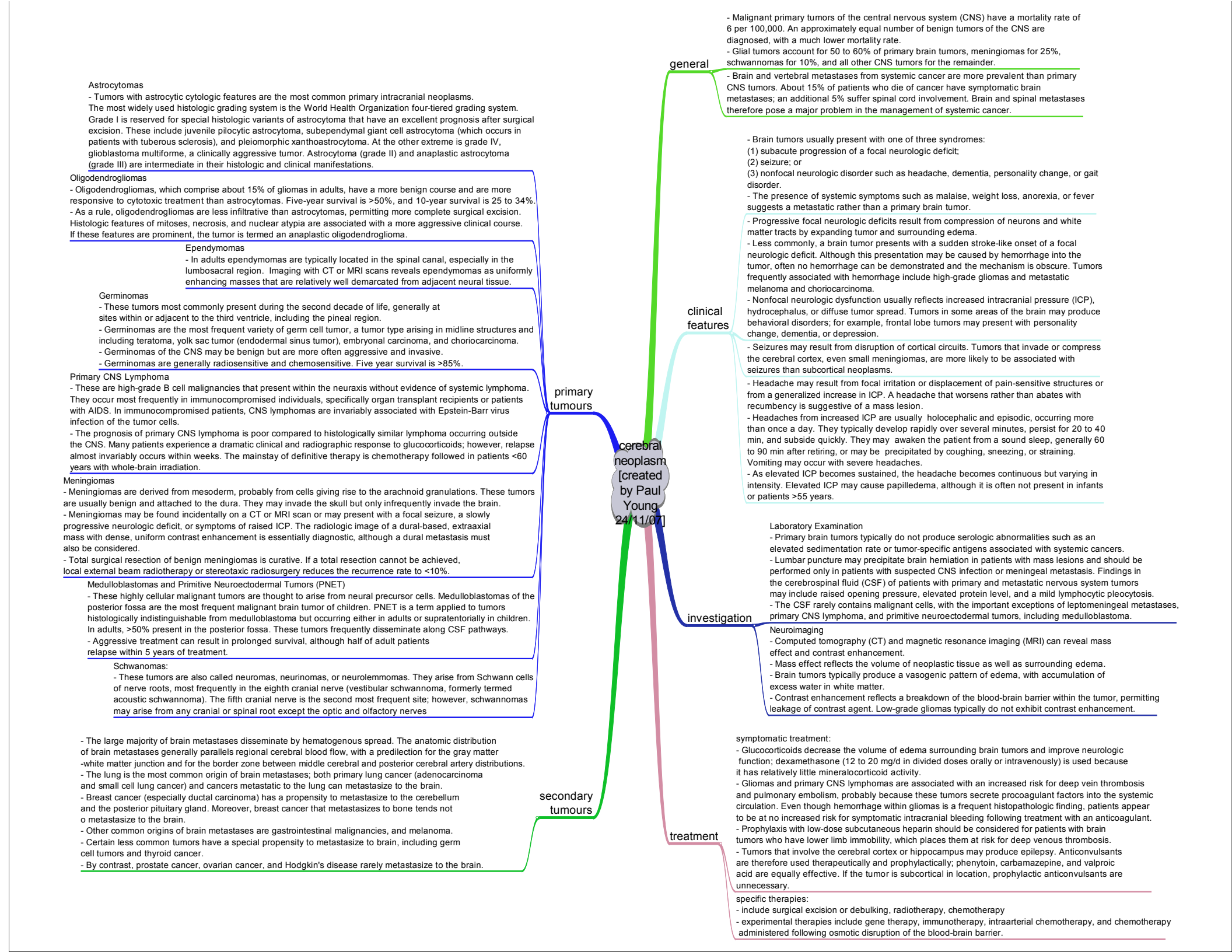
cerebral neoplasm [created by Paul Young 24/11/07]
general
- Malignant primary tumors of the central nervous system (CNS) have a mortality rate of 6 per 100,000. An approximately equal number of benign tumors of the CNS are diagnosed, with a much lower mortality rate.
- Glial tumors account for 50 to 60% of primary brain tumors, meningiomas for 25%, schwannomas for 10%, and all other CNS tumors for the remainder.
- Brain and vertebral metastases from systemic cancer are more prevalent than primary CNS tumors. About 15% of patients who die of cancer have symptomatic brain metastases; an additional 5% suffer spinal cord involvement. Brain and spinal metastases therefore pose a major problem in the management of systemic cancer.
clinical features
- Brain tumors usually present with one of three syndromes:
(1) subacute progression of a focal neurologic deficit;
(2) seizure; or
(3) nonfocal neurologic disorder such as headache, dementia, personality change, or gait disorder.
- The presence of systemic symptoms such as malaise, weight loss, anorexia, or fever suggests a metastatic rather than a primary brain tumor.
- Progressive focal neurologic deficits result from compression of neurons and white matter tracts by expanding tumor and surrounding edema.
- Less commonly, a brain tumor presents with a sudden stroke-like onset of a focal neurologic deficit. Although this presentation may be caused by hemorrhage into the tumor, often no hemorrhage can be demonstrated and the mechanism is obscure. Tumors frequently associated with hemorrhage include high-grade gliomas and metastatic melanoma and choriocarcinoma.
- Nonfocal neurologic dysfunction usually reflects increased intracranial pressure (ICP), hydrocephalus, or diffuse tumor spread. Tumors in some areas of the brain may produce behavioral disorders; for example, frontal lobe tumors may present with personality change, dementia, or depression.
- Seizures may result from disruption of cortical circuits. Tumors that invade or compress the cerebral cortex, even small meningiomas, are more likely to be associated with seizures than subcortical neoplasms.
- Headache may result from focal irritation or displacement of pain-sensitive structures or from a generalized increase in ICP. A headache that worsens rather than abates with recumbency is suggestive of a mass lesion.
- Headaches from increased ICP are usually holocephalic and episodic, occurring more than once a day. They typically develop rapidly over several minutes, persist for 20 to 40 min, and subside quickly. They may awaken the patient from a sound sleep, generally 60 to 90 min after retiring, or may be precipitated by coughing, sneezing, or straining. Vomiting may occur with severe headaches.
- As elevated ICP becomes sustained, the headache becomes continuous but varying in intensity. Elevated ICP may cause papilledema, although it is often not present in infants or patients >55 years.
investigation
Laboratory Examination
- Primary brain tumors typically do not produce serologic abnormalities such as an elevated sedimentation rate or tumor-specific antigens associated with systemic cancers.
- Lumbar puncture may precipitate brain herniation in patients with mass lesions and should be performed only in patients with suspected CNS infection or meningeal metastasis. Findings in the cerebrospinal fluid (CSF) of patients with primary and metastatic nervous system tumors may include raised opening pressure, elevated protein level, and a mild lymphocytic pleocytosis.
- The CSF rarely contains malignant cells, with the important exceptions of leptomeningeal metastases, primary CNS lymphoma, and primitive neuroectodermal tumors, including medulloblastoma.
Neuroimaging
- Computed tomography (CT) and magnetic resonance imaging (MRI) can reveal mass effect and contrast enhancement.
- Mass effect reflects the volume of neoplastic tissue as well as surrounding edema.
- Brain tumors typically produce a vasogenic pattern of edema, with accumulation of excess water in white matter.
- Contrast enhancement reflects a breakdown of the blood-brain barrier within the tumor, permitting leakage of contrast agent. Low-grade gliomas typically do not exhibit contrast enhancement.
treatment
symptomatic treatment:
- Glucocorticoids decrease the volume of edema surrounding brain tumors and improve neurologic function; dexamethasone (12 to 20 mg/d in divided doses orally or intravenously) is used because it has relatively little mineralocorticoid activity.
- Gliomas and primary CNS lymphomas are associated with an increased risk for deep vein thrombosis and pulmonary embolism, probably because these tumors secrete procoagulant factors into the systemic circulation. Even though hemorrhage within gliomas is a frequent histopathologic finding, patients appear to be at no increased risk for symptomatic intracranial bleeding following treatment with an anticoagulant.
- Prophylaxis with low-dose subcutaneous heparin should be considered for patients with brain tumors who have lower limb immobility, which places them at risk for deep venous thrombosis.
- Tumors that involve the cerebral cortex or hippocampus may produce epilepsy. Anticonvulsants are therefore used therapeutically and prophylactically; phenytoin, carbamazepine, and valproic acid are equally effective. If the tumor is subcortical in location, prophylactic anticonvulsants are unnecessary.
specific therapies:
- include surgical excision or debulking, radiotherapy, chemotherapy
- experimental therapies include gene therapy, immunotherapy, intraarterial chemotherapy, and chemotherapy administered following osmotic disruption of the blood-brain barrier.
secondary tumours
- The large majority of brain metastases disseminate by hematogenous spread. The anatomic distribution of brain metastases generally parallels regional cerebral blood flow, with a predilection for the gray matter -white matter junction and for the border zone between middle cerebral and posterior cerebral artery distributions.
- The lung is the most common origin of brain metastases; both primary lung cancer (adenocarcinoma and small cell lung cancer) and cancers metastatic to the lung can metastasize to the brain.
- Breast cancer (especially ductal carcinoma) has a propensity to metastasize to the cerebellum and the posterior pituitary gland. Moreover, breast cancer that metastasizes to bone tends not o metastasize to the brain.
- Other common origins of brain metastases are gastrointestinal malignancies, and melanoma.
- Certain less common tumors have a special propensity to metastasize to brain, including germ cell tumors and thyroid cancer.
- By contrast, prostate cancer, ovarian cancer, and Hodgkin's disease rarely metastasize to the brain.
primary tumours
Astrocytomas
- Tumors with astrocytic cytologic features are the most common primary intracranial neoplasms.
The most widely used histologic grading system is the World Health Organization four-tiered grading system. Grade I is reserved for special histologic variants of astrocytoma that have an excellent prognosis after surgical excision. These include juvenile pilocytic astrocytoma, subependymal giant cell astrocytoma (which occurs in patients with tuberous sclerosis), and pleiomorphic xanthoastrocytoma. At the other extreme is grade IV, glioblastoma multiforme, a clinically aggressive tumor. Astrocytoma (grade II) and anaplastic astrocytoma (grade III) are intermediate in their histologic and clinical manifestations.
Oligodendrogliomas
- Oligodendrogliomas, which comprise about 15% of gliomas in adults, have a more benign course and are more responsive to cytotoxic treatment than astrocytomas. Five-year survival is >50%, and 10-year survival is 25 to 34%.
- As a rule, oligodendrogliomas are less infiltrative than astrocytomas, permitting more complete surgical excision. Histologic features of mitoses, necrosis, and nuclear atypia are associated with a more aggressive clinical course. If these features are prominent, the tumor is termed an anaplastic oligodendroglioma.
Ependymomas
- In adults ependymomas are typically located in the spinal canal, especially in the lumbosacral region. Imaging with CT or MRI scans reveals ependymomas as uniformly enhancing masses that are relatively well demarcated from adjacent neural tissue.
Germinomas
- These tumors most commonly present during the second decade of life, generally at sites within or adjacent to the third ventricle, including the pineal region.
- Germinomas are the most frequent variety of germ cell tumor, a tumor type arising in midline structures and including teratoma, yolk sac tumor (endodermal sinus tumor), embryonal carcinoma, and choriocarcinoma.
- Germinomas of the CNS may be benign but are more often aggressive and invasive.
- Germinomas are generally radiosensitive and chemosensitive. Five year survival is >85%.
Primary CNS Lymphoma
- These are high-grade B cell malignancies that present within the neuraxis without evidence of systemic lymphoma. They occur most frequently in immunocompromised individuals, specifically organ transplant recipients or patients with AIDS. In immunocompromised patients, CNS lymphomas are invariably associated with Epstein-Barr virus infection of the tumor cells.
- The prognosis of primary CNS lymphoma is poor compared to histologically similar lymphoma occurring outside the CNS. Many patients experience a dramatic clinical and radiographic response to glucocorticoids; however, relapse almost invariably occurs within weeks. The mainstay of definitive therapy is chemotherapy followed in patients <60 years with whole-brain irradiation.
Meningiomas
- Meningiomas are derived from mesoderm, probably from cells giving rise to the arachnoid granulations. These tumors are usually benign and attached to the dura. They may invade the skull but only infrequently invade the brain.
- Meningiomas may be found incidentally on a CT or MRI scan or may present with a focal seizure, a slowly progressive neurologic deficit, or symptoms of raised ICP. The radiologic image of a dural-based, extraaxial mass with dense, uniform contrast enhancement is essentially diagnostic, although a dural metastasis must also be considered.
- Total surgical resection of benign meningiomas is curative. If a total resection cannot be achieved, local external beam radiotherapy or stereotaxic radiosurgery reduces the recurrence rate to <10%.
Medulloblastomas and Primitive Neuroectodermal Tumors (PNET)
- These highly cellular malignant tumors are thought to arise from neural precursor cells. Medulloblastomas of the posterior fossa are the most frequent malignant brain tumor of children. PNET is a term applied to tumors histologically indistinguishable from medulloblastoma but occurring either in adults or supratentorially in children. In adults, >50% present in the posterior fossa. These tumors frequently disseminate along CSF pathways.
- Aggressive treatment can result in prolonged survival, although half of adult patients relapse within 5 years of treatment.
Schwanomas:
- These tumors are also called neuromas, neurinomas, or neurolemmomas. They arise from Schwann cells of nerve roots, most frequently in the eighth cranial nerve (vestibular schwannoma, formerly termed acoustic schwannoma). The fifth cranial nerve is the second most frequent site; however, schwannomas may arise from any cranial or spinal root except the optic and olfactory nerves