Ependymoma

Definition
Ependymomas are neuro-epithelial tumors of the central nervous system. They originate from ependymal cells in the choroid plexus, cerebral ventricles, the spinal canal, or ependymal clusters in the filum terminale\(^1,2\).

Epidemiology
In the United States, ependymomas account for 1.9% of all primary brain tumors and 3.1% of all gliomas\(^3\). Incidences of ependymomas have a bimodal age distribution. The first peak is in the early childhood (< 4 years) and the second peak occurs during the 4\(^{th}\) to 5\(^{th}\) decade\(^4\).

Pathophysiology
Ependymal cells are cells that form the epithelium of the ventricles in the brain, central canal in the spinal cord, and the choroid plexus. It is a type of neuroglia. The ependymal cells secrete cerebrospinal fluid (CSF) and the cilia on the cell surface facilitate CSF circulation around the central nervous system\(^5,6\).

Locations of ependymomas vary between age groups. In children, most ependymomas arise in the posterior fossa especially within the 4\(^{th}\) ventricle\(^2,7,8\). In adults, ependymomas are more common in the supertentorial compartment and spinal cord. Spinal ependymomas are most often in the cervical region, conus medullaris, and filum terminale\(^9,9\).

Ependymomas are well demarcated and slow growing but have potential for local invasion\(^2,4\). The World Health Organization classifies ependymomas into three grades. Grade I includes subependymomas and myxopapillary ependymomas. Grade II includes cellular, clear cell, tanyctic, and papillary ependymomas. Grade III includes anaplastic ependymomas\(^10,11\). The survival rate for 5 years is approximately 66\(^%\)\(^12\).

Manifestations
Patients with ependymomas may have a wide variety of presentations depending on the size and location of the tumor.

Ependymomas located in the 4\(^{th}\) ventricle may affect flow of CSF and lead to obstructive hydrocephalus and increased intracranial pressure (ICP) (Ghosal et al., 2010; Jun et al., 2012). Increased ICP may display signs and symptoms of cerebellar and/or cranial nerve dysfunction such as cerebellar ataxia, hemiparesis, and blurred vision\(^10,13,14\). Frontal parietal lobe tumors may present with motor hemiparesis and motor weakness\(^2\). Focal neurological deficits and seizures are common in extraventricular tumors. Cortical ependymomas may present with seizures\(^15,16,17\).

Ependymomas at the conus medullaris and filum terminale may present with backache, paraparesis, paresthesia, and bladder and bowel dysfunction due to medullary compression\(^18,19\). Other symptoms include headache, nausea, vomiting, ataxia, vertigo, and papilledema\(^12,14\).

Diagnostic Tests
Computerized tomography (CT) scan and magnetic resonance imaging (MRI) are the most common diagnostic tests for patients with brain tumors. CT scans usually reveal the location and size of the tumor, and possible hydrocephalus. T1 and T2 weighted MRIs usually reveal if there is a hemorrhage, necrosis, and/or calcification of the tumor\(^10\).

Treatment Options
Urgent treatment may be required when patients present with signs of increased ICP such as altered in level of consciousness. These treatments include steroid therapy and an
external ventricular drain. The urgent treatment provides a temporary reduction of ICP until the tumor can be resected\(^{10}\). Treatment options for ependymomas include surgery, radiotherapy, and chemotherapy.

**Surgery**

Primary ependymomas are typically managed with surgical treatment. Tumor removal is usually performed under microscopic magnification\(^ {10}\).

**Radiotherapy**

Radiotherapy is used as a post-surgical adjunct therapy, especially for small sized tumors, or for treating recurrent ependymomas. Children under three years of age are not recommended to receive radiotherapy\(^ {10,20,21}\).

**Chemotherapy**

Chemotherapy is another adjunct therapy for ependymomas. Results from chemotherapy such as temozolomide, carboplatin, cisplatin, or vincristine are not encouraging\(^ {10,20,21}\).

**Nursing Implications**

Monitor patients' neurological status closely for signs of post-operative complications such as altered level of consciousness, limb weakness, or changes to CSM. Early rehabilitation has been proven to be effective in improving motor strength\(^ {1,10}\).

Patients with posterior fossa tumors usually have an occipital craniotomy bone flap\(^ {10}\). Caution should be made not to position patient in the supine position to avoid pressure on the surgical site.

Steroid therapy may be prescribed before surgery. Gradual tapering of steroid therapy may be started immediately after surgery\(^ {10}\).

**Reference**


©2013

Disclaimer: The author of this article neither represents nor guarantees that the practices described herein, if followed, ensure safe and effective patient care. The author further assumes no responsibility or liability in connection with any information or recommendations contained in this article. The recommendations and instructions in this article are based on the knowledge and practice in neuroscience as of the date of publication. These recommendation and instructions are subject to change based on the availability of new scientific information.