Ependymoma

Ependymomas are tumors that arise from the ependymal cells that line the ventricles in the CNS. These occur more commonly in children in the fourth ventricle, but also occur in adults, most often in the spinal canal in the cauda equina region. Since ependymal cells line the ventricles , proliferation of these cells can cause obstruction to CSF flow and result in hydrocephalus. Histologically, the tumor cells can present as aggregates of cells around a blood vessel, known as perivascular pseudorosettes. Rod-shaped blepharoplasts can also be seen. These are basal ciliary bodies, and are found in fully differentiated ependymal cells. Since ependymomas are glial cell tumors, they can usually be identified by positive staining for GFAP (glial fibrillary acidic protein). Ependymomas have a poor prognosis due to its ability to disseminate throughout the CSF. Ependymomas can also be seen with those with Neurofibromatosis type II, a genetic disorder marked by the predisposition to develop a variety of tumors of the central and peripheral nervous systems.



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Occur in Children
Baby in Diaper
Ependymomas are common in children and present in the 4th ventricle.

Fourth Ventricle
(4) Fork Vent
The most common location for ependymomas in children is in the fourth ventricle. Because of this positioning, they can obstruct CSF outflow.

Hydrocephalus
Hydras
Ependymomas can present with hydrocephalus due to obstruction of CSF outflow, especially if the tumors are in the posterior fossa. This can present clinically as headache, nausea and vomiting.

Perivascular Pseudorosettes
Vascular Sumo-rosettes
This describes the rosette arrangement of cells around a blood vessel rather than a lumen. Perivascular pseudorosettes can be found in ependymomas.

Rod Shaped Blepharoplasts
Blue-pharaoh holding Rod
Rod shaped blepharoplasts are also known as basal ciliary bodies. This is a feature of fully differentiated ependymal cells. Therefore, rod shaped blepharoplasts can be found in ependymomas.

Cauda Equina in Adults
Cod Horse-tail and Adult panda
While the fourth ventricle is the most common location for ependymomas in children, adults commonly present with ependymomas in the cauda equina region of the spinal canal.

Poor Prognosis
Gravestone
Ependymomas can have a poor prognosis due to its ability to disseminate throughout the CSF.

Neurofibromatosis Type II
Neuron-Fabios in (2) Tutus
Ependymomas can be seen in those with Neurofibromatosis type II. NFII is also known as MISME syndrome (Multiple Inherited Schwannomas, Meningiomas, and Ependymomas). It is an inherited disorder manifesting as non-malignant brain tumors.

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