

Hemangioblastoma

Hemangioblastoma is a benign vascular tumor that occurs most commonly in the CNS and retina. It is most often found in the cerebellum and can present with ataxia or disordered gait. This tumor can occur sporadically but it is also commonly associated with Von Hippel Lindau disease. Von Hippel Lindau disease is a familial syndrome of multiple tumors and cysts caused by mutation of the VHL tumor suppressor gene on chromosome 3. This tumor is highly vascular and is known to produce VEGF as well as erythropoietin, which can result in secondary polycythemia. Histologically, hemangioblastoma demonstrates networks of endothelial cells around capillaries, amidst large foamy cells containing lipid.



PLAY PICMONIC

Most Often Cerebellar

[Silver-cerebellum-bell](#)

This tumor is most commonly found in the cerebellum and can present with symptoms of ataxia and gait disturbance.

Associated with Von Hippel-Lindau

[Van-hippo Landing-owl](#)

This tumor is associated with Von Hippel Lindau syndrome, a mutation of the VHL tumor suppressor gene. A mutation in this gene produces multiple tumors and cysts throughout the body and is associated with hemangioblastomas.

Retinal Angiomas

[Red-tin-eye Angel](#)

Retinal angiomas are benign tumors that can occur sporadically or in association with Von Hippel Lindau disease. High vascularization can cause vascular leakage and, eventually, retinal detachment in the eye.

Can Produce EPO

[Biker-injecting with EPO](#)

Hemangioblastomas are highly vascular and are known to produce erythropoietin.

Secondary Polycythemia

[Poly-cyclist with red-blood-cell-helmet](#)

Due to the ability to produce erythropoietin, this tumor can cause secondary polycythemia, which is an increase in the RBC count independent of stem cells.

Foamy Cells

[Foam](#)

Histologically this tumor contains foamy cells, which are large cells containing lipid droplets.

High Vascularity

[Blood-vessels](#)

This tumor is highly vascular and can be seen in its gross and histological appearance, demonstrating large amounts of capillary networks.