They originate from the vascular system.

The clinical presentation typically occurs in the fifth decade for sporadic cases and as early as the third decade for patients with a diagnosis of von Hippel-Lindau (VHL) disease

They may be associated with other diseases such as polycythemia (increased blood cell count), pancreatic cysts and Von Hippel-Lindau syndrome (VHL syndrome). Hemangioblastomas are most commonly composed of stromal cells in small blood vessels and usually occur in the cerebellum, brain stem or spinal cord.