

Hemangioblastoma epidemiology

Intracranially, they occur almost exclusively in the posterior fossa ([posterior fossa hemangioblastomas](#) are the most common primary intra-axial [posterior fossa tumor](#) in adults). May occur in the [cerebellar hemisphere](#), [vermis](#), or [brainstem](#).

May also occur in the [spinal cord](#) (1.5–2.5% of spinal cord tumors). Also difficult to distinguish histologically from a [renal cell carcinoma](#) (which is common in VHL adds to the difficulty of this differential).

70% of HGBs occur sporadically, but 30% occur as part of [von Hippel-Lindau disease](#) ¹⁾.

[Retinal hemangioblastoma](#) and/or angiomas occur in 6% of patients with [cerebellar hemangioblastomas](#).

[Hemangioblastoma](#) is the rarest [central nervous system tumor](#), accounting for less than 2% ²⁾, and 7-12% of posterior fossa lesions in adult patients.

They usually occur in adults, yet tumors may appear in VHL syndrome at much younger ages. Men and women are approximately at the same risk.

In [Von Hippel-Lindau disease](#) the incidence of development of hemangioblastoma in cerebellum is 44-72%, in brain stem is 10-25%, and in spinal cord is 13-50% ³⁾.

70% of HGBs occur sporadically, but 30% occur as part of von Hippel-Lindau (VHL) disease.

Retinal HGB and/or angiomas occur in 6% of patients with cerebellar HGBs.

About one-fourth of hemangioblastomas occur in patients with Von Hippel-Lindau disease (VHL); the remainders are sporadic. Although extremely rare, solid hemangioblastomas outside the CNS have been reported, involving peripheral nerve ⁴⁾ retroperitoneum ⁵⁾, soft tissue and bone ^{6) 7) 8)} and visceral organs including the pancreas ⁹⁾, adrenals ^{10) 11)}, liver ¹²⁾, and lung ^{13) 14)}.

Most of the above cases of peripheral hemangioblastoma occurred in patients who also had CNS tumor with few exceptions ¹⁵⁾.

Metastatic tumor involving the lung is exceedingly rare with only four cases previously reported

Two were autopsy studies in patients who died of complications of the CNS hemangioblastomas in 1943 and 1981, and the third was mentioned in a case report addendum providing follow-up information on hepatic hemangioblastoma in 1991. Lu et al. report a case of a 48-year-old man who presented with multiple lung nodules treated by surgical excision. Pathological study revealed features classic for hemangioblastoma. The patient had a remote history of hemangioblastomas having been excised from the posterior fossa 7 and 20 years previously. It is the first report on surgically resected hemangioblastomas from the lung of a living patient with histological and immunohistochemical characterization. ¹⁶⁾

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