Von Hippel-Lindau disease treatment

Resection of individual CNS tumors is usually reserved until symptomatic to decrease the number of operations over a lifetime, since the tumors in VHL are usually multiple, tend to recur, and the growth pattern is saltatory.

Surgery is the treatment of choice for accessible cystic HGBs.

Stereotactic radiosurgery (SRS)

May provide local control rates of > 50% over 5 years. SRS has been recommended for asymptomatic HBG > 5 mm diameter if they are cystic or progressing in size during surveillance.

Cranial treatment plan: using a median dose of 22 Gy (range: 12–40 Gy) prescribed to the median 82% isodose line in 1–4 sessions. In cystic lesions, treatment is confined to the contrast-enhancing mural nodule (the cyst wall is not treated). Spinal treatment plan: a median dose of 21 Gy (range 20–25 Gy) prescribed to the median 77% isodose line in 1–3 sessions. Radiosurgery is usually contraindicated in hemangioblastomas with a cyst.

The guidelines for the diagnosis and treatment of VHL were published in Japan in 2011 [1)].

There is no current way to reverse the presence of the VHL mutation in patients. Nonetheless, early recognition and treatment of specific manifestations of VHL disease can substantially decrease complications and improve quality of life. For this reason, individuals with VHL disease are usually screened routinely for retinal angiomas, CNS hemangioblastomas, clear-cell renal carcinomas and pheochromocytomas.

CNS hemangioblastomas are usually surgically removed if they are symptomatic. Photocoagulation and cryotherapy are usually used for the treatment of symptomatic retinal angiomas, although anti-angiogenic treatments may also be an option. Renal tumours may be removed by a partial nephrectomy or other techniques such as radiofrequency ablation.

1)
Shuin T, editor. Tokyo: Chugai-Igakusha; 2011. Clinical Practice Guideline for the Management of von Hippel-Lindau disease.