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ABSTRACT

Management of Central Nervous System Hemangioblastomas in von Hippel-Lindau Disease

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Central nervous system (CNS) hemangioblastoma (HB) is the most frequent tumor in von Hippel- Lindau disease (VHL). The tumor occurs in cerebellum, brain stem, and spinal cord. However, management of CNS HBs in VHL has been controversial. Here, we discuss the management of CNS HBs in VHL.Surgically treated 62 CNS HBs (cerebellar 41, brain stem 3, spinal 18) in 34 VHL patients from 1992 to 2010 were retrospectively examined in diagnosis and surgical treatment. Diagnosis of CNS HBs depended on symptoms, neurological examination, laboratory data, CT, MRI, and gene diagnosis. Symptoms in cerebellar HB were faintness, gait disturbance, nausea, and headache while those in spinal ones were pain, numbness, and weakness in extremities. Neurological examination revealed cerebellar dysfunction for cerebellar HB and regional hypesthesia or muscular weakness for spinal one. Laboratory data often reveal polycythemia due to secretion of erythropoietin from the tumor. Gene diagnosis for VHL gene mostly revealed ger-mline mutation in VHL gene. Treatment for CNS HBs in cerebellum as well as spinal cord after symptomatic was surgical resection in the first choice. Surgical outcomes was mostly excellent or good except for large tumors (cerebellar HB >4cm, spinal HB>2 cm). In the second choice, stereotactic irradiation did not always control the tumor. VHL patients bearing CNS hemangioblastoma mostly developed other CNS ones in a different site. In conclusion, surgical resection is the first-choice treatment for CNS HB in VHL, and it should be done at appropriate timing without neurological deterioration.