Long-term outcome after resection of brainstem hemangioblastomas in von Hippel-Lindau disease.

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Abstract

OBJECT: Brainstem hemangioblastomas are frequently encountered in patients with von Hippel-Lindau (VHL) disease. These tumors can cause significant morbidity, and their optimal management has not been defined. To better define the outcome and management of these tumors, the authors analyzed the long-term results in patients who underwent resection of brainstem hemangioblastomas.

METHODS: Consecutive patients with VHL disease who underwent resection of brainstem hemangioblastomas with a follow-up of 12 months or more were included in this study. Serial functional assessments, radiographic examinations, and operative records were analyzed.

RESULTS: Forty-four patients (17 male and 27 female) underwent 51 operations for resection of 71 brainstem hemangioblastomas. The most common presenting symptoms were headache, swallowing difficulties, singultus, gait difficulties, and sensory abnormalities. The mean follow-up was 5.9 ± 5.0 years (range 1.0-20.8 years). Immediately after 34 operations (66.7%), the patients remained at their preoperative functional status; they improved after 8 operations (15.7%) and worsened after 9 operations (17.6%) as measured by the McCormick scale. Eight (88.9%) of the 9 patients who were worse immediately after resection returned to their preoperative status within 6 months. Two patients experienced functional decline during long-term follow-up (beginning at 2.5 and 5 years postoperatively) caused by extensive VHL disease-associated CNS disease.

CONCLUSIONS: Generally, resection of symptomatic brainstem hemangioblastomas is a safe and effective management strategy in patients with VHL disease. Most patients maintain their preoperative functional status, although long-term decline in functional status may occur due to VHL disease-associated progression.

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