# Neurosurgical Management of Brainstem Hemangioblastomas: A Single-Institution Experience with 116 Patients

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OBJECTIVE: **Brainstem hemangioblastomas (HBs) are considered one of the most challenging lesions in surgical procedures. We present our institutional experience with 116 patients over a period of 20 years in the treatment of HBs.**

METHODS: **We evaluated the results of microsurgical treatment and highlighted the management strategies. There were 60 male and 56 female patients including 13 cases with clinical evidence of von Hippel-Lindau disease. Tumors were solid in 99 cases and cystic in 17 cases. Tumors were small (≤2 cm) in 43 cases, large (2–4 cm) in 45 cases, and giant (≥4 cm) in 28 cases.**

RESULTS: **Radical removal was achieved in 111 patients (95.7%), and incomplete removal was achieved in 5 cases (4.3%). The immediate postoperative mortality and morbidity were 7.8% and 17.2%, respectively. Detailed analyses of outcomes showed that surgical complications were related to some tumor characteristics. Follow-up study was available in 83 patients by Karnofsky performance scale scores. Most patients maintained their preoperative neurologic status. There were 17 patients with surgical disability who demonstrated a clear improvement with rehabilitation treatment. Worsening of neurologic deficits occurred in 2 patients. Ectopic recurrent lesions developed in 2 patients.**

CONCLUSIONS: **Based on our experience, microsurgery is safe and effective, and excellent outcomes can be obtained for cystic or small tumors. We advocate early surgical intervention for sporadic HBs; giant solid HBs remain a challenge, and meticulous microsurgical technique and perioperative management are vital. Long-term monitoring also is recommended.**

## INTRODUCTION

Hemangioblastomas (HBs) are highly vascular benign neoplasms and account for 1.8% of all central nervous system tumors in our hospital. Brainstem HB is defined as a tumor originating from brainstem, and is responsible for 5% to 15% of all intracranial HBs (12, 35, 42, 43). Patients with brainstem HBs often present with headache and dizziness, pyramidal sign, sensory abnormalities, ataxia, vomiting and cranial nerve deficit, etc (36, 38). Brainstem lesions may be of intrabrainstem type or extrabrainstem type (42). The most common single location of brainstem HBs is frequently located in the medullary region (8, 36, 38, 41). Pathologically, brainstem HBs are identical to HBs that originate elsewhere in the central nerve system (12, 14, 24, 32). Resecting solid brainstem HBs have always been considered a great challenge to most neurosurgeons, although some excellent surgical results have been achieved. The high rates of the mortality and morbidity due to the proximal critical structures and rich blood supply restrict the popularization of the surgical technology. In this study, we retrospectively analyzed our experience in a consecutive series of 116 patients with brainstem HBs and reviewed the advancement in the related field, such as nature history and pathology, in order to approach an optimal (eligible) modality and highlight the management of advance for these particular cases.

## MATERIALS AND METHODS

### Patients

From January 1994 to December 2013, a total of 116 consecutive solitary brainstem HB patients underwent surgery by the senior authors in the Huashan Hospital (part of the cases had been previously reported (39, 42)). Clinical information was obtained from hospital charts, clinic notes, and operative reports.

**Key words**
- Brainstem
- Hemangioblastomas
- Neurosurgical management
- Surgical treatment

**Abbreviations and Acronyms**
HB: Hemangioblastoma
MRI: Magnetic resonance imaging
VHL: von Hippel-Lindau

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This study was approved by the ethics committee of Huashan Hospital, and written informed consent from all patients or their legal representatives.

## NEUROIMAGING ASSESSMENT

All patients were evaluated preoperatively with neuroimaging including computed tomography or magnetic resonance imaging (MRI), and some patients also underwent computed tomography angiography, magnetic resonance angiography, or digital subtraction angiography. All the neuroimaging information was utilized to demonstrate the location, size and blood supply of the tumors as well as the surrounding brainstem tissue involvement.

MRI with T1/T2-weighted imaging and gadolinium-enhanced sequences is the primary method for screening tumors and for surgical planning. Contrast-enhanced MRI permits the identification of small tumor nodules. In addition, MRI is helpful in separating cystic and solid components of the tumor from edema. On MRI, brainstem HBs can appear as solid or cystic forms (Figure 1). In the 116 patients, we found that, most of the HBs were solid including small cysts within the tumors; whereas 17 tumors were cystic forms. The typical cystic HBs had large cyst with small nodules that were markedly enhanced after contrast injection (Figure 1). According to the relationship between the tumor and the brainstem parenchyma, these tumors can be divided into two types (Figure 1): intrabrainstem and extrabrainstem. Generally, those tumors with huge volume and rich blood supply belonged to the extrabrainstem type. Further, we divided the tumors into three groups according to tumor size. Group I: small-size tumors ($\leq$2 cm in diameter for solid tumor; diameter refers to maximal diameter of the nodule in cystic tumors; 43 cases), Group II: medium-size tumors (2—4 cm; 45 cases), Group III: large-size tumors ($\geq$4 cm; the maximal diameter was 5.3 cm; 28 cases).

On angiographic images, the feeding arteries and draining veins were observed with a heavy flush of solid tumors. 32 patients in the series (mainly large or giant solid HBs) had undergone preoperative embolization since 1996. Using superselective catheterization technique, the feeding arteries of the tumor were large or partially embolized with semipermanent materials (gelatin sponge, polyvinyl alcohol particles, or Onyx [ev3 Endovascular, Inc., Plymouth, Minnesota, USA) in the 1-3 days before operation. The purpose of preoperative embolization was to control inaccessible arteries during surgery and reduce tumor vascularity. All patients tolerated the embolization without permanent neurologic deficits.

### Microsurgical Techniques

The technique used for brainstem HBs resection has previously been described[3]. Briefly, for predominantly cystic HBs, complete excision of the mural nodule is not too difficult and the cyst wall does not need to be resected. The mural nodule is excised microsurgically within the cyst by dissecting along the gliotic margin away from the parenchyma of the brainstem when concomitant with meticulous coagulating and cutting the tiny vessels. Noticeably, in some situation, confusion will be met as the tumor nodule embeds into the inner wall and is covered by a xanthochromic fibroproteinaceous membrane, making the surface color of the nodule the same as that of the inner wall of the cyst. Careful review of 3-dimensional MRI findings can assist in finding the nodule to avoid unnecessary and excessive disturbance of the proximal brainstem structures. Otherwise, the tumor nodule will be missing.

Successful removal of solid HBs should abide by the principle of arteriovenous malformation dissection, especially in large or giant tumors. Usually, HBs have an integral capsule and there is a thin rim of soft gliotic tissue at the junction between the capsule and the adjacent brainstem. Along the tumor surface confining at the tumor—neural tissue interface, tumor was microdissected circumferentially and proceeded to the deeper regions until total removal en bloc (Figure 2). Intraoperative electrophysiologic monitoring, including somatosensory evoked potentials and brainstem auditory evoked potentials, was applied in recent 10 years.

### Perioperative Management

The assessment of preoperative conditions, especially cardiopulmonary functions, was strictly enforced. The elaborate postoperative management is of great importance during the early postoperative days at the neurosurgical intensive care unit. Usually, intubation was continued for at least 12 hours on the second postoperative day. If the patients' respiratory function was weak, ventilatory support and assisted respiration by breathing machine was required for some duration until full restoration of normal respiratory function. The endotracheal tube was removed if cough and gag reflexes had recovered. For patients who were at risk of pneumonia or aspiration dyspnea, tracheotomy and nasogastric feeding were required to alleviate respiratory complications and provide nutritional support.

### Surgical Evaluation and Follow-Up Study

Preoperative and postoperative neurological functions of the patients were graded according to the scale described by McCormick et al. (22) before surgery and at discharge from hospital. A Karnofsky Performance Scale score was also accessed for each patient based on clinical evaluations, Contrast MRI scan was usually taken for every patient 3 months after the operation and then annually. Follow-up ranged from 1 to 20 years with a mean of 7 years. At the final examination, all survivals of these patients were followed up in the Outpatient Department or by telephone call.

### Statistical Analysis

Fisher exact test was used to compare the categorical variables by tumor characteristics. All statistical analyses were performed using SAS version 9.3 (SAS Institute Inc., Cary, North Carolina, USA).

## RESULTS

### Patients and Tumor Characteristics

The patient information and tumor characteristics were summarized in Table 1. The patients were on average 28 years old, with a range from 12 to 71 years. Among them 60 were males and 56 females. All the 116 patients had solitary brain HB. 13 patients had clinic evidence of VHL disease: one had a family history with polycystic disease of visceral organ, 2 had 2 central nervous system HBs, 3 had symptomatic tumor recurrence after a remote