Comparison of anterior and posterior surgical approaches in the treatment of ventral spinal hemangioblastomas in patients with von Hippel–Lindau disease

RYSZARD M. PLUTA, M.D., PH.D., BRAHAN IULIANO, M.D., HETTY L. DEVROOM, R.N., TUNG NGUYEN, M.D., AND EDWARD H. OLDFIELD, M.D.

Surgical Neurology Branch, National Institute of Neurological Disorders and Stroke, National Institutes of Health, Bethesda, Maryland

Object. Von Hippel–Lindau (VHL) disease is an autosomal-dominant neoplastic syndrome with manifestations in multiple organs, which is evoked by the deletion or mutation of a tumor suppressor gene on chromosome 3p25. Spinal hemangioblastomas (40% of VHL disease–associated lesions of the central nervous system) arise predominantly in the posterior aspect of the spinal cord and are often associated with an intraspinal cyst. Rarely, the tumor develops in the anterior aspect of the spinal cord. Ventral spinal hemangioblastomas are a surgical challenge because of difficult access and because vessels feeding the tumor originate from the anterior spinal artery.

The goal of this study was to clarify whether an anterior or posterior surgical approach is better for management of hemangioblastomas of the ventral spinal cord.

Methods. The authors performed a retrospective analysis of clinical outcomes and findings on magnetic resonance (MR) imaging studies in eight patients (two women and six men with a mean age of 34 ± 15 years) who underwent resection of ventral spinal hemangioblastomas (nine tumors: five cervical and four thoracic). Two surgical approaches were used to resect these tumors. A posterior approach was selected to treat five patients (laminationomy and posterior or myelotomy in four patients and the posterolateral approach in one patient); an anterior approach (corpectomy and arthrodesis) was selected to treat the remaining three patients.

Immediately after surgery, the ability to ambulate remained unchanged in patients in whom an anterior approach had been performed, but deteriorated significantly in patients in whom a posterior approach had been used, because of motor weakness (four of five patients) and/or proprioceptive sensory loss (three of five patients). This difference in ambulation, despite significant improvements over time among patients in the posterior access group, remained significant 6 months after surgery. In all cases, MR images revealed complete resection of the tumor and in five patients significant or complete resolution of the intramedullary cyst was demonstrated (present in six of eight patients).

Conclusions. The outcomes of these eight patients with hemangioblastomas of the ventral spinal cord indicate that both immediate and long-term results are better when an anterior approach is selected for resection.

KEY WORDS • von Hippel–Lindau disease • spine • hemangioblastoma
of the thin tumor “capsule” from the surrounding spinal cord.\textsuperscript{11,28,29} This can be accomplished only after proper visualization of the tumor, which in most cases can be achieved using a posterior approach to the spinal cord, even for resection of intramedullary, ventrally located hemangioblastomas. The posterior approach has been considered to be the least traumatic for the patient, especially when the tumor reaches the posterior or posterolateral surface of the spinal cord or when posterior myelotomy is facilitated by the presence of an intramedullary cyst in the vicinity of the tumor.\textsuperscript{6,11,12,20,26,28,29} Nevertheless, even delicate retraction and careful dissection may lead to clinical deterioration immediately after resection of an intramedullary tumor.\textsuperscript{6,18} Thus, Martin, et al.\textsuperscript{20} recently proposed a posterolateral approach to ventral spinal lesions to limit injury to the spinal cord. In this approach, however, the spinal cord must be rotated for proper visualization of the ventrally or ventrolaterally located tumor.\textsuperscript{12,20} Thus, from an anatomical point of view, when the tumor is present on the ventral aspect of the spinal cord the anterior approach should be the least traumatic. In this paper we compare the clinical outcomes of a group of patients in whom ventral spinal hemangioblastomas were resected using either an anterior or a posterolateral approach.

**Clinical Material and Methods**

**Patient Evaluation**

All patients were screened at the National Institutes of Health, under an institutional review board–approved protocol, for the presence of mutations and deletions of the VHL gene, and received the diagnosis of VHL disease.\textsuperscript{1} To compare these patients’ neurological outcomes after an anterior or posterior surgical approach had been used to treat hemangioblastomas of the ventral spinal cord, we retrospectively identified eight patients in whom tumors were located in the ventral aspect of the cord (that is, anterior to the dentate ligament) among 73 patients with VHL who were treated for spinal hemangioblastomas between January 1, 1988, and July 31, 2001. All patients were evaluated with serial MR imaging performed using a 0.5 or 1.5-tesla magnet (General Electric Medical Systems, Milwaukee, WI). An independent neuroradiologist assessed tumor size and location.

Neurological examinations were conducted at admission to the hospital, immediately before and after surgery, at approximately 6 months postoperatively, and thereafter at yearly intervals. The neurological status of each patient was assessed at each of these times and a category of neurological function was assigned to each individual based on a retrospective assessment of findings of neurological examinations in the patient’s record (Table 1).\textsuperscript{2}

**Surgical Techniques**

**Posterior Approach for Ventrally Located Tumors.** Patients were placed prone and laminectomies were performed to provide adequate access to the tumor. Ultrasoundography was used to assess the position of the tumor and contiguous cyst.\textsuperscript{3,18,22,24} After dural incision, a small midline myelotomy was made in the dorsal median raphe, and the incision was carried deeper in layers to expose the cyst or the posterior surface of the hemangioblastoma. The tumor was dissected using microneurosurgical techniques and precise identification, as well as bipolar coagulation and division of individual vessels entering and leaving the tumor capsule. In cases in which there were very large tumors, partial removal of the lesion was performed by incremental coagulation followed by sharp excision or removal of the tumor with an ultrasonic aspirator. This allowed visualization of the deep margin of the tumor, including its margin from the anterior spinal artery in some patients, and successful microscopic dissection. After tumor resection and careful hemostasis, the dura mater and superficial layers were closed in a standard fashion.

**Posterolateral Approach for Ventrolaterally Located Tumors.** Combined midline and transverse skin and muscle incisions were used. The laminae, a portion of the rib, the unilateral facet, and a portion of one or more pedicles were removed above and below the tumor. After dural incision, the posterior rootlet of the thoracic nerve root and the dentate ligament were interrupted with the aid of an operating microscope. The veins covering the tumor were coagulated and dissected to expose its margin. After tumor resection was performed using microneurosurgical techniques
Ventral spinal hemangioblastoma

and careful hemostasis, the dura mater and the superficial layers were closed in a standard fashion.

Anterior Access. After insertion of a lumbar drain, the patient was placed supine with the neck slightly extended. Cervical distraction was accomplished with 5 lb of traction. Intraoperative x-ray films were obtained to confirm neck position and to localize the level of interest. A skin incision was made on the right side over the vertebral body parallel to a skin crease. The vertebral body was exposed by a transverse neck incision and dissection, and the proper vertebral level was identified using intraoperative x-ray films. Cervectomy was performed using a high-speed drill equipped with a diamond-tipped burr. After exposure of the dura mater a Doppler ultrasound probe was used to assess tumor position and to confirm adequate exposure. After a vertical incision of the dura, bipolar coagulation, and division of the arterialized veins crossing the margin of the tumor, the pia mater was incised precisely at the edge of the tumor by using a diamond knife to develop a plane of dissection between the tumor capsule and the spinal cord. The tumor was then dissected using microscopic techniques and precise identification, bipolar coagulation, and division of individual vessels entering and leaving the tumor capsule. Following resection of the hemangioblastoma and careful hemostasis, dural closure was achieved with the aid of Gel film and fibrin glue. Arthrodesis was performed using a titanium cage filled with a cancellous bone allograft (National Institutes of Health bone bank) and anterior cervical plating. Lumbar drainage of cerebrospinal fluid was used postoperatively for 72 hours to minimize the risk of a postoperative cerebrospinal fluid leak.

Results

All mean data presented in this paper represent means ± standard deviations.

Seventy-three patients underwent 102 surgeries for resection of 141 spinal hemangioblastomas at the Surgical Neurology Branch of the National Institute of Neurological Disorders and Stroke between January 1, 1988, and July 31, 2001. Eight of these patients (two women and six men with a mean age of 34 ± 15 years [median 32.5 years, range 21–51 years]) harbored a total of nine ventral spinal hemangioblastomas (five cervical and four thoracic). These eight patients comprised the study population and the nine tumors they harbored constitute 6% of all spinal hemangioblastomas treated at our institution during the study period.

Two surgical approaches were used to resect these tumors. A posterior approach was selected to treat five patients (laminectomy and posterior myelotomy in four patients and the posterolateral approach in one patient); an anterior approach (corpectomy and arthrodesis) was selected to treat the remaining three patients (Table 2).

We attempted to resect a ventral spinal hemangioblastoma via the posterolateral approach in three patients (Cases 1, 7, and 8).14,20 In one patient (Case 1), however, the surgery was abandoned because we could not gain access to the tumor through this approach and the tumor was subsequently resected via an anterior approach. In the second patient (Case 8), the posterolateral approach was abandoned because a scar caused by prior surgery in the vicinity of the tumor prevented adequate rotation of the spinal cord; this tumor was resected via a posterior myelotomy.

In all patients, postoperative MR images revealed complete resection of the tumor(s) and a collapse, decrease in size, or complete resolution of the cyst within 6 months after surgery in five of six patients with a syrinx (Table 3). Clinical outcomes were analyzed in all eight patients and assessed according to the neurological scale proposed by Aminoff and Loque (Table 1).2 Immediately after surgery, the ability to ambulate remained unchanged in patients who had undergone an anterior approach, but it deteriorated significantly in patients who had undergone a posterior approach due or weakness (four of five patients) and/or sensory loss (three of five patients). This difference in ambulation, despite improvement over time among patients in the posterior approach group, remained significant 6 months after surgery (Table 3) and at the last follow-up examination (mean 28 ± 9.2 months, median 30 months for the anterior approach group; and mean 79.6 ± 38.6 months, median 78 months for the posterior approach group).

### Table 3
Clinical outcomes: a comparison of anterior and posterior approaches*

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Immediate Postop Neurological Status</th>
<th>Preop</th>
<th>At Discharge</th>
<th>At 6–12 Mos Follow Up</th>
<th>MR Imaging Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anterior approach</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>50, M</td>
<td></td>
<td></td>
<td>worsening lt hemiparesis</td>
<td>mild difficulty (II)</td>
<td>required walker (IV)</td>
<td>normal w/ ankle brace (II)</td>
<td>no tumor</td>
</tr>
<tr>
<td>21, F</td>
<td></td>
<td></td>
<td>mild lt arm &amp; leg dysesthesia</td>
<td>normal (I)</td>
<td>normal (I)</td>
<td>normal (I)</td>
<td>no tumor; syrinx persisted</td>
</tr>
<tr>
<td>27, M</td>
<td></td>
<td></td>
<td>no deficits</td>
<td>normal (I)</td>
<td>normal (I)</td>
<td>normal (I)</td>
<td>no tumor; syrinx collapsed</td>
</tr>
<tr>
<td>Posterior approach</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>26, M</td>
<td></td>
<td></td>
<td>lt hand &amp; rt leg weakness, loss of proprioception &amp; vibratory sense in both legs</td>
<td>normal (I)</td>
<td>required cane (III)</td>
<td>normal (I)</td>
<td>no tumor; syrinx resolved in 6 mos</td>
</tr>
<tr>
<td>31, M</td>
<td></td>
<td></td>
<td>proprioceptive loss in lt leg; weakness in lt anterior tibialis, rt EHL &amp; quadriceps</td>
<td>normal (II)</td>
<td>required walker (IV)</td>
<td>normal (I)</td>
<td>no tumor; syrinx diminished</td>
</tr>
<tr>
<td>34, M</td>
<td></td>
<td></td>
<td>severe rt lower-extremity dysesthesia, weakness in rt lower extremity</td>
<td>mild difficulty (II)</td>
<td>required walker (IV)</td>
<td>required cane (III)</td>
<td>no tumor</td>
</tr>
<tr>
<td>43, M</td>
<td></td>
<td></td>
<td>significant weakness in lower extremities</td>
<td>moderate difficulty (II)</td>
<td>required walker (IV)</td>
<td>required cane (III)</td>
<td>no tumor; syrinx collapsed</td>
</tr>
<tr>
<td>41, F</td>
<td></td>
<td></td>
<td>significant paraparesis, headache, pain</td>
<td>normal (I)</td>
<td></td>
<td>required walker (IV)</td>
<td>no tumor; syrinx persisted</td>
</tr>
</tbody>
</table>

* Grades were assigned according to Aminoff and Loque. Abbreviation: EHL = extensor hallucis longus.
Illustrative Cases

Patient characteristics and postoperative outcomes are summarized in Tables 2 and 3.

Posterior Approach

Case 5. This 31-year-old right-handed man with VHL disease had undergone resection of a cerebellar hemangioblastoma 6 years before the present admission. He also harbored multiple cervical and thoracic hemangioblastomas with cervicothoracic syringomyelia. He complained of progressive weakness in the right lower extremity and a burning sensation in the left lower extremity, which had lasted approximately 3 months. These symptoms were produced by an enlarging T-2 ventral hemangioblastoma and its associated syrinx (Fig. 1).

Operation. Laminectomy of T-1 and T-2 was performed and intraoperative ultrasonography was used to identify the echogenic ventrally located tumor. This large intramedullary tumor did not reach the posterior surface of the spinal cord (Fig. 1B). A midline myelotomy was made and a 20-mm-diameter tumor and associated cystic cavity were identified. The ventral portion of the tumor was separated from the anterior spinal artery and excised. The wound was closed in layers in the usual manner.

Postoperative Course. The patient’s sensory and motor deficits in his lower extremities increased appreciably after surgery. He had decreased strength and diminished sensation of vibration and proprioception in the lower extremities bilaterally, and he required the assistance of a walker on discharge. At follow-up visits he walked with a cane and the proprioceptive deficits had significantly improved.

Posterolateral Approach

Case 7. This 43-year-old man had received the diagnosis of VHL disease 2 years before admission. He experienced progressive spastic paraparesis in the lower extremities for approximately 5 years. The patient was ambulatory; however, his strength in both legs was 4/5 with increased tone, hyperreactive reflexes, and bilateral Babinski sign with clonus. Sensation to light touch and pinprick was decreased in both lower extremities below L-1; this decreased sensation was greater on the left side than on the right. These symptoms and signs were produced by a large thoracic cyst associated with two hemangioblastomas (Fig. 2).

Operation. Laminectomies of T2–6 were performed via a posterolateral approach, with partial resection of the fourth

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Fig. 1. Case 5. Magnetic resonance images demonstrating a T-2 hemangioblastoma in a 31-year-old man with VHL disease. The **black arrows** indicate the tumor and the **white arrows** indicate the associated cyst. A: Sagittal view. B: Axial T1-weighted image with contrast enhancement.

Fig. 2. Case 7. Sagittal contrast-enhanced T1-weighted MR image obtained in a 43-year-old man with VHL disease, demonstrating a large thoracic cyst (**white arrows**) with two hemangioblastomas (**black arrows**).
rib and the transverse process. Intraoperative ultrasonography revealed a syringomyelic cavity and defined the location of two vascular tumors that were located ventral and to the right of the spinal cord. The small extramedullary portion of the rostral tumor was identified where it lay beneath the dentate ligament. The sensory rootlets of the fourth thoracic nerve root were divided and the tumor was removed. The second tumor, which was mostly (70%) intramedullary and was located lower and also ventrally, was resected.

After surgery, the patient’s motor deficits significantly increased, although his strength gradually improved over the next several days, and he ambulated with a walker at discharge. One year later, after this patient received a heart transplant, he became wheelchair bound, a condition initially attributed by his physicians to his cardiac difficulties. Two years later, two more posterior thoracic spinal cord hemangioblastomas were resected and his weakness improved; however, he remains confined to a wheelchair 4 years after the initial spinal surgery.

**Anterior Approach**

**Case 1.** This 50-year-old man received the diagnosis of VHL disease 8 years before the present admission and had previously undergone several surgical procedures for peripheral tumors (retinal angiomas, renal carcinomas, and adrenal pheochromocytoma) and resection of a cerebellar hemangioblastoma. Increasing weakness in his left arm and leg was attributed to a ventral hemangioblastoma at C-6 (Fig. 3A).

**Operation.** Laminectomies were performed at C-5–6 and, after rotation and retraction of the spinal cord, exploration revealed the presence of a hemangioblastoma that had no accessible surface presentation (Fig. 3B); only the most lateral and posterior tip of the tumor was visualized. The approach was abandoned, and 1 week later the patient underwent surgery via an anterior approach. Corpectomies were performed at C-6 and C-7. Intraoperative ultrasonography and dural and arachnoid incision revealed the hemangioblastoma to be located to the left of the midline and to the left of the anterior spinal artery (Fig. 3C). A 1-cm-diameter hemangioblastoma was resected (Fig. 3D). The dura was closed and a C5–T1 cervical arthrodesis was accomplished using a Harm cage and anterior cervical plates.

**Postoperative Course.** After surgery, the patient’s left-sided weakness progressively decreased. Magnetic resonance images confirmed total resection of the ventral spinal hemangioblastoma. The patient underwent radiation treatment.
(5040 cGy) for a large hemangioblastoma of the brainstem. Follow-up assessment (23 months after surgery; Table 3) revealed clinical Grade II neurological status with no resid-
ual symptoms of the spinal cord tumor.

Case 2.

This 21-year-old, right-handed woman had VHL disease and Chiari I malformation with syringomyelia. She underwent a resection of two cerebellar hemangioblasto-
mas 4 years before admission, which resulted in the dis-
appearance of the syringomyelia,17 and a dorsal C-6 hemangioblastoma was resected 1 year before admission. At presentation the patient complained of occasional head-
aches, neck stiffness, decreased endurance, and long-lasting decreased sensation in the right C7–8 distribution, which were associated with recurrent syringomyelia and a ventral C-7 hemangioblastoma that was progressively increasing in size (Fig. 4A and B).

Operation. A corpectomy was performed at C-7. After removal of the posterior longitudinal ligament, a predomi-
nantly exophytic (85% extramedullary) hemangioblastoma was easily identified (Fig. 4C), displacing the anterior spi-
nal artery to the left side (Fig. 4D). The hemangioblastoma was resected (Fig. 4E and F) and the dura mater was sutured and covered with Gel film, Gelfoam, and tissue glue. A tita-
nium Harm cage was fitted into the C-7 vertebral space and arthrodesis was performed at C6–T1. The soft tissue was closed in the usual fashion.

Postoperative Course. This patient experienced no new neurological deficits, except transient hoarseness. Six months after surgery, she underwent resection of two he-
angioblastomas at C6–7 and T-2 because of a persistent syringomyelia in the cervicothoracic spinal cord.

FIG. 4. A and B: Sagittal contrast-enhanced T1-weighted MR images obtained in a 21-year-old woman with VHL dis-
ease and a Chiari I malformation with syringomyelia, demonstrating a ventral C-7 hemangioblastoma (black arrows) and the syrinx (white arrows). A C-7 corpectomy was performed. C–F: Intraoperative photographs. After removal of the pos-
terior longitudinal ligament, the hemangioblastoma is easily visualized through the intact dura (C: rostral to the right). The surface presentation of a predominantly exophytic (85% extramedullary) hemangioblastoma displacing the anterior spinal artery to the left side is revealed (D). The hemangioblastoma is clearly seen during resection. The inferior pole has been displaced rostrally, exposing the tumor bed in the spinal cord (E). The bed is viewed after tumor resection (F).

Discussion

Hemangioblastomas of the central nervous system will always be associated with the name of Dr. A. Lindau,16 who in 1926 described the presence of a highly vascular tumor in the cerebellum, which was later named “hemangioblas-
toma” by Dr. Harvey Cushing.7 The first spinal hemangio-
blastoma was described earlier, however, by Dr. F. Schultze in 1912.23 Since then many surgeons have attempted resec-
tion of these tumors. The first trials prompted Krishnan and Smith15 to state in 1961 that “operation is contraindicated if presence of intramedullary hemangioblastoma is con-
firmed.” Despite this, in 1967 Guidetti and Fortuna14 sum-
marized their surgical experience with these lesions, sug-
gestin’that “the decision concerning total resection of these tumors must rest upon the presence . . . of a clear line of cleavage” between the tumor and spinal cord. Their obser-
vation on the successful dissection of the intramedullary portion of the tumor proved to be valuable and led to a re-
assessment of intraspinal hemangioblastomas as curable by resection.

In most cases, the hemangioblastoma is located in the posterior aspect of the spinal cord in the vicinity of pos-
terior nerve roots.14 These tumors are surgically ap-
proached posteriorly through a laminectomy. The posterior approach with a myelotomy has been used for ventrally lo-
cated tumors, especially those associated with a coexisting intraspinal cyst, because it provides proper tumor exposure and avoids important anatomical structures.5,12,13,26 The key factors for choosing this approach have been “ease” and “atraumatic” access to the intraspinal tumor via a posterior myelotomy and the presence of “facilitating” surgical expo-
sure of the intraspinal syrinx. Nevertheless, this technique
often leads to immediate postoperative deterioration, prolonged neurological recovery, and sometimes to permanent neurological deficits, as was confirmed in cases presented here and elsewhere.5,14,18

Other techniques have been developed to resect tumors of the spinal cord that are located ventrally and ventrolaterally. Yasargil, et al.,20 proposed a posterolateral approach through a hemilaminectomy: “by gently retracting the spinal cord and spinal roots, tumors can be removed from the ventral and dorsal aspect of the spinal cord.” This technique, however, has not been used for resection of hemangioblastomas. In 1995 Martin, et al.,20 proposed a more aggressive lateral bone exposure with resection of the whole lamina, as well as the facet joint and pedicle, to expose lesions of the ventral spinal cord. In their series of seven patients who were surgically treated for different types of tumors and vascular malformations, instability requiring stabilization developed in two patients. One patient harbored a ventrally located hemangioblastoma. The result of surgery was determined to be excellent, but the patient was nonambulatory with a dense quadriparesis before surgery and remained neurologically the same after surgery. We used the same technique in one patient (Case 7) and, despite careful and successful dissection of both tumors from the right ventrolateral aspect of the spinal cord, the patient deteriorated by two clinical grades postoperatively (Table 3). Also, this technique may be rejected if the patient has undergone prior spinal surgeries, as in our Case 8.

In their report of a single case, Iwasaki and colleagues13 proposed using the anterior approach for resection of ventral spinal hemangioblastomas because of “the presence of arterial feeders directly from the anterior spinal artery that have to be interrupted in order to complete a gross resection of the tumor.” The authors advocate using this technique only for small, centrally and paracentrally located tumors, however, and conclude that “it is better to approach larger tumors posteriorly or posterolaterally.” Each surgical approach has its advantages and disadvantages, and has been selected for these rare ventrally located tumors mostly on the basis of the surgeon’s preference. In the series of patients described here, three approaches to ventrally located spinal lesions were used. From a surgical, anatomical, and clinical point of view, the anterior surgical approach provided direct access to lesions located on the ventral aspect of the spinal cord, allowed immediate visualization and safe division of the arteries feeding the tumor, and proved to be the least traumatic to the spinal cord.

Although the small number of cases described here do not permit statistical comparisons, the results suggest that for carefully selected patients both the immediate and long-term clinical results of resection of ventral spinal hemangioblastomas were superior when the anterior surgical approach was chosen.

Although we favor an anterior approach for most ventral hemangioblastomas, there are occasions in which a posterior or posterolateral approach will be indicated.

Conclusions

The postoperative clinical outcomes of eight patients with hemangioblastomas of the ventral spinal cord suggests that in selected cases the immediate and long-term results are appreciably better when surgery is performed using an anterior approach rather than a posterior or posterolateral approach. The presence of an intraspinal syrinx should not influence the choice of surgical approach used to remove ventral spinal hemangioblastomas.

References

20. Martin NA, Khanna RK, Batzdorf U: Posterolateral cervical or thoracic approach with spinal cord rotation for vascular malfor-