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Long-term stability after multilevel cervical laminectomy for spinal cord tumor resection in von Hippel-Lindau disease

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Abstract

Object—Despite the frequent multiplicity and development of new spinal cord hemangioblastomas that require multiple resections in patients with von Hippel-Lindau (VHL) disease, the long-term effects of spinal surgery on spinal column stability in this neoplasia disorder are not known. To determine the effect of multilevel cervical laminectomy for spinal cord tumor resection in VHL, the authors analyzed long-term clinical and radiographic outcomes.

Methods—The authors included consecutive patients enrolled in a prospective VHL disease natural history study who underwent cervical laminectomy(s) for spinal cord hemangioblastoma resection. Serial clinical examinations, neck disability indices, and radiographs (static and dynamic), as well as operative records, were analyzed.

Results—Twenty-five adult patients (16 female, 9 male) with VHL disease underwent 34 operations (mean 1.4 ± 0.7 [$\pm$ SD]/patient) for the resection of cervical spinal cord hemangioblastomas (mean number of lamina removed/surgery 3.0 ± 1.3). The mean age at surgery was 33.9 ± 11.9 years (range 18–61 years), and the mean follow-up duration was 9.1 ± 5.6 years. At last follow-up, radiographic criteria indicated that 9 patients (36%) had spinal column instability, 13 patients (52%) developed a cervical spinal deformity, 4 patients (16%) developed moderate to severe neck disability, and 3 patients (12%) met the criteria for clinical instability. Removal of the C-2 lamina was associated with the development of clinical instability ($p = 0.02$, Fisher exact test); older age at surgery was associated with the development of cervical deformity ($p = 0.05$, logistic regression); and a greater number of operations (suboccipital–T4) were associated with increased neck disability indices ($p = 0.01$, linear regression).

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Disclaimer

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Conclusions—Whereas patients with VHL disease will often require multiple laminectomies for cervical spinal cord hemangioblastoma resection, a limited number of patients (12%) will develop clinical instability. Because prophylactic cervical instrumentation confers limited benefit at the time of spinal cord tumor resection for most patients, and because these patients need life-long MR imaging of the spinal cord, the quality of which may be affected by instrumentation, longitudinal clinical and radiological evaluation may be used to determine which patients will require stabilization.

Keywords

cervical laminectomy; instability; neck disability index; spinal cord tumor; spinal deformity; von Hippel-Lindau disease

Von Hippel-Lindau disease is a heritable tumor syndrome caused by a germline mutation of the *VHL* tumor suppressor gene located on chromosome 3.¹³ Patients with VHL disease are predisposed to tumors of the CNS and viscera. Hemangioblastomas are the most common CNS manifestation in VHL disease and occur in 60%–80% of patients.¹⁴ Approximately 50% of craniospinal hemangioblastomas are spinal tumors, the majority (90%) of which are equally distributed between the cervical and thoracic spine.¹⁵ In the spinal canal, most spinal cord hemangioblastomas (94%) are located dorsal to the dentate ligament and are most frequently found within the dorsal root entry zone (66%).^{15,19}

Because spinal cord hemangioblastomas are most frequently located within or on the dorsal aspect of the spinal cord, they are resected via a posterior approach that includes laminectomies. Although the features associated with cervical instability after laminectomy in patients undergoing resection of sporadic spinal cord tumors are becoming better defined, little is known about long-term spinal stability in patients with multiple neoplasia syndrome (including VHL disease and neurofibromatosis) who have unique management features. Specifically, patients with VHL disease and other neoplasia syndromes will frequently require multiple spinal operations, will harbor multiple tumors that will not require resection, and may have fixed neurological deficits that could increase the likelihood of spinal instability after laminectomy.

To more accurately determine the incidence of clinically significant instability and/or deformity after multilevel cervical laminectomy performed for spinal cord tumor resection in a multiple neoplasia syndrome, we analyzed the long-term radiographic follow-up and clinical outcomes in patients with VHL disease in whom cervical hemangioblastomas were excised.

Methods

Patient Population

Of 250 patients enrolled in a prospective natural history study of CNS lesions in VHL disease (NIH protocol 00-N-0140), 30 underwent resection of a cervical spinal cord hemangioblastoma via laminectomy at the NIH, and this population was included in this study. In all patients, VHL disease was diagnosed by clinical and genetic criteria.¹⁴ Informed consent was obtained in all patients. Patients who underwent cervical

hemangioblastoma resection but in whom anterior cervical approaches (for resection of ventral tumors) necessitated instrumented fusion (2 cases) and those unwilling to undergo dynamic evaluation of the spine (3 cases) were excluded.

Patient Evaluation

Clinical Evaluation—All patients underwent detailed serial neurological examinations, first conducted at the initial screening, immediately before and after an operation, and at approximately 6-month intervals thereafter. The NDI, a questionnaire designed to quantify neck pain and assess its affect on conducting activities of daily living, was administered via phone and confirmed with review of subjective complaints cataloged from subsequent patient visits.⁴⁰ Sections of the NDI that were not pertinent to individual persons (reading ability in blind patients, inability to work/drive due to comorbidity) were scored according to the mean value obtained from assessable sections.³² Patients were classified according to established NDI scaling intervals (0–4, no disability; 5–14, mild disability; 15–24, moderate disability; 25–34, severe disability; and 35, complete disability). Patients with NDI scores greater than 14 were considered negatively impacted by sequelae of VHL disease or operative intervention. Patients were also contacted by telephone at the conclusion of the study period to confirm and supplement the data.

Imaging Evaluation—Preoperative and postoperative MR imaging was performed at 6-month intervals in conjunction with clinical examinations. Postcontrast T1- and T2-weighted MR imaging sequences were used to determine the presence of hemangioblastomas and a spinal cord syrinx. At last follow-up, plain-film radiographs of the cervical spine in the neutral upright, maximal flexion, and extension positions were obtained.

Factors Affecting Development of Deformity, Radiographic Stability, and Increased NDI

Variables that have been previously hypothesized to promote postlaminectomy deformity, radiographic instability, and neck pain (age, number of laminae removed, presence of a C-2 laminectomy, presence of a C-7 laminectomy, amount of facet disruption, preexisting kyphotic deformity, radiotherapy, and presence of a syrinx at the time of operation)^{1,5,6,11,22,35} were recorded. Additional variables unique to patients with multiple neoplasia syndromes (development of new cervical tumors or syringes during the course of follow-up, number of operations performed in the cervical spinal cord [C1–7] and adjacent segments [suboccipital craniectomy, T1–4] for removal of other tumors)^{3,18,21,36,37} were recorded.

Cervical Spine Measurements

All measurements were performed using a PACS Carestream Client (Eastman Kodak Co.). The curvature of the cervical spine was determined by measuring the angle formed by the lines extending from the posterior borders of C-2 and C-7. Spines were classified based on C2–7 curvature as kyphotic ($> 0^\circ$), lordotic ($< -10^\circ$), straight/hypolordotic (0° to -10°), or hyperlordotic ($< -45^\circ$).^{12,28} Cervical spinal deformity was categorized either based on the presence of a focal kyphosis measuring greater than 10° or the presence of a hyperlordotic, straight, hypolordotic, or kyphotic C2–7 curvature (Fig. 1).

Cervical ROM was calculated by the absolute difference between the C2–7 curvature measured in maximal upright flexion and extension. Two measures of intervertebral motion, sagittal rotation and horizontal displacement, were calculated for each level from C-2 to C-7. Sagittal rotation was defined as the change between flexion and extension of the angle created by the lines extended from the inferior endplates of adjacent levels. Horizontal displacement (shear) was defined as the displacement between flexion and extension of the posterior inferior corner of the superior vertebra in the direction defined by the superior posterior endplate of the inferior vertebra. Greater than 20° of intervertebral rotation or 2.7 mm of horizontal displacement at any level was used to identify radiographic cervical spine instability (Fig. 2).^{41,42}

Clinical Instability

Clinical stability was defined as the ability of the spine to limit its patterns of motion under physiological loads, so as not to damage or irritate the spinal cord or the nerve roots.^{25,26,42} Because overall spinal column dysfunction that results in clinical instability may be indicated by symptoms such as muscle fatigue, chronic neck pain, or headaches that disrupt patients' ability to execute ADLs rather than overt neurological deficit, we used the presence of moderate to severe NDI scores or overt neurological sequelae in patients with known radiographic instability, as a marker of overall cervical spine stabilizing system dysfunction.

Operative Technique

Through a dorsal incision, sharp dissection and monopolar cautery were used to follow the midline raphe to the level of the spinous processes as described previously.¹⁵ Briefly, the posterior cervical musculature was dissected from its attachment to the spinous processes and lamina in the subperiosteal plane. Laminectomies were performed using rongeurs, bone punches, and/or the high-speed drill. Bone was removed to obtain an exposure that allowed 1–2 cm rostral and caudal visualization of the tumor from the upper and lower tumor margins. Care was taken to preserve the capsular ligaments and ensure that less than one-fourth of the medial aspect of each facet was removed. Subsequent resection of the hemangioblastoma and standard closure were performed as previously described.¹⁵

Statistical Evaluation

Patient characteristics, imaging findings, and surgical procedures were analyzed for their predictive capability in determining outcomes, specifically in the development of increased neck disability, cervical spinal deformity, radiographic instability, and clinical instability. The Fisher exact test was used to assess the relationship between categorical patient characteristics and operative procedures and specific outcome measures. Univariate logistic regression was performed when continuous predictors were considered, and linear regression analysis was performed for continuous outcome measures. The 2-sample t-test was used to compare means of sets of tumor-level continuous variables. Analyses were performed on data obtained at last follow-up. Data were analyzed using the software R (R version 2.7.2; The R Foundation for Statistical Computing). Statistical significance was determined by a 2-sided p value of 0.05. Mean values are presented $\pm$ SD.

Results

Patient Characteristics

Twenty-five patients (16 women, 9 men) who underwent posterior cervical laminectomy for microsurgical resection of 34 cervical spinal cord hemangioblastomas (mean 1.4 tumors; range 1–3 tumors) were included in this study (Table 1). All patients were alive at last follow-up (mean follow-up after initial surgery 9.1 ± 5.6 years; median, 7.75 years; range 2–19 years). The mean age at initial surgery was 33.9 ± 11.9 years (range 18–61 years). The mean number of levels of cervical laminectomies was 3.0 ± 1.3 . A C-2 laminectomy was performed in 7 patients (28%) and a C-7 laminectomy in 9 patients (36%). No patients underwent instrumented or noninstrumented fusion. An additional 32 operations (mean 1.3 ± 1.8 procedures) were performed at the craniocervical junction and immediately adjacent spinal segments (T1–4) for resection of other spinal cord, brainstem, and cerebellar hemangioblastomas in 13 patients (52%).

Clinical Assessment

Patients underwent surgery for the treatment of symptomatic hemangioblastomas (31 tumors in 23 patients) or for large tumors that displayed rapid growth over a short interval (3 tumors in 3 patients) in asymptomatic patients. (One patient had both an asymptomatic and a symptomatic spinal cord tumor [at different times].) In all patients, complete resection of hemangioblastomas was performed. One patient (4%) had undergone stereotactic radiosurgery to a cervical hemangioblastoma that subsequently required surgical resection. No patient experienced long-term worsening of motor function related to surgery, but 1 patient (4%) suffered increased neuropathic pain and sensory dysesthesia of the neck and upper extremity.

Nineteen patients (76%) reported no neck pain at the time of last evaluation, 2 patients (8%) reported mild neck pain, and 4 patients (16%) reported moderate to severe neck pain (NDI Scores 15, 18, 23, and 27, respectively). Three patients (12%) reported they were unable to perform their usual work due to neck pain. Twenty-one patients reported that neck pain did not disturb sleeping (17 patients [68%]) or barely affected sleeping (4 patients [16%]). Four patients reported moderate disturbances in sleep (2–3 hours of sleeplessness) related to neck pain. The most frequently reported symptom attributed to neck pain was the presence of moderate to severe headaches (7 patients [28%]).

Radiographic Assessment

Twenty-one patients had evaluable preoperative MR imaging studies, and all patients had MR images and plain radiographs of the cervical spine in the neutral upright, maximal flexion, and extension positions at last follow-up. The mean preoperative C2–7 angulation was $-23^\circ \pm 10^\circ$ (median -22° , range -6° to -46°). Three patients had preoperative deformity, including 2 with cervical hypolordosis (-6° and -8°) and 1 patient with hyperlordosis (-46°). In 14 patients (64%), new cervical spinal cord hemangioblastomas developed that were not identified on MR images obtained at the first visit. Eleven patients (44%) had at least 1 cervical spinal cord hemangioblastoma identified on MR imaging

performed at last follow-up. Twenty-two patients (88%) had a tumor-associated syrinx preoperatively, and 9 patients (36%) had one at last the follow-up visit.

The mean C2–7 cervical curvature on neutral upright plain radiographs obtained at last follow-up was $-24^{\circ} \pm 14^{\circ}$ (range -82° to 0°) compared with $-27^{\circ} \pm 16^{\circ}$ (range -78° to -11°) on MR images obtained at last follow-up. Dynamic plain flexion/extension radiographs revealed a mean cervical ROM of $52.3^{\circ} \pm 18.1^{\circ}$ (range 10° – 85°). Based on static plain radiographs obtained in the neutral upright position, no patient developed C2–7 postlaminectomy kyphosis, but 13 patients (52%) developed a cervical spine deformity by last follow-up. These included 10 patients (40%) who had focal kyphosis, 7 (28%) who developed hypolordosis/straight neck deformity, and 1 (4%) who developed hyperlordosis.

Dynamic plain flexion/extension radiographs revealed spinal column instability in 9 patients (36%) based on radiographic criteria. In 4 patients (12%), these criteria were met by surpassing the upper limit of physiological shear alone, and in 1 patient (4%), the criteria were met by greater than 20° of intervertebral mobility in at least 1 cervical intervertebral level. Four patients (12%) exceeded both normal limits of physiological shear and had greater than 20° of intervertebral mobility in at least 1 cervical intervertebral level.

Clinical Instability

At last follow-up, 3 cases (12%) met the criteria for the definition of clinical instability based on the presence of moderate to severe neck disability and radiographically documented instability. In these patients, no overt neurological symptoms (long tract signs, myelopathy, radiculopathy, and so on) attributable to spinal cord compression or nerve root impingement from deformity or radiographic instability were present at last follow-up.

Analysis of Factors Affecting Disability, Radiographic Stability, and Clinical Stability

Increasing age at surgery, but no other outcome measures, was correlated with the development of cervical spinal deformity ($p = 0.05$, logistic regression). An increasing number of surgeries (subocciput–T4) was associated with an increase in neck disability indices ($p = 0.01$, linear regression), but not with other outcome measures. Removal of the C-2 lamina (7 patients [28%]) was associated with the development of clinical instability in 3 patients (42.8%; $p = 0.02$, Fisher exact test). There was no significant difference ($p = 0.36$) in the mean number of laminae removed in the group of patients who developed radiographic instability (mean 3.3 ± 1.4 levels; median 3 levels) and those who did not (mean 2.8 ± 1.3 levels; median 3 levels). Preexisting deformity and radiotherapy were not found to correlate with the development of increased neck disability, cervical deformity, radiographic instability, or clinical instability. No other variables were statistically correlated with markers for poorer outcome (Table 2).

Discussion

The most common approach to resection of dorsal cervical spinal cord tumors is via a posterior approach after multilevel laminectomy.¹⁵ Although cervical laminoplasty has been proposed to reduce the risk of developing postsurgical deformity,^{9,29} recent reports suggest that no significant reduction in the incidence of spinal deformity occurs, especially in the

setting of intradural spinal tumor resection.^{17,30} Controversy exists over the need for prophylactic management of deformity and instability that may arise during the follow-up period with concurrent instrumented stabilization techniques performed at the time of tumor extirpation.³⁵ Previously, in sporadic tumors, the incidence of progressive deformity and instability following cervical laminectomy was highest in the pediatric population, patients with a malignant intramedullary lesion, those undergoing adjuvant radiotherapy, or patients with preoperative findings of kyphosis and/or instability.^{6,11,44} Other intraoperative factors, including resection of the C-2 lamina, extensive laminectomies (≥ 3 levels removed), and removal of greater than 25% of the articular facet joint, have also been correlated with an increased risk for the subsequent development of deformity, instability, and neurological sequelae.^{10,21,35}

Clinical Outcomes and Neck Disability

Long-term follow-up (mean 9.1 ± 5.6 years) of patients undergoing multilevel cervical laminectomies (mean 3 ± 1.3 levels) revealed that the majority of patients (84%) experienced either no neck pain or mild neck pain after tumor resection. This study shows that patients may have multiple surgeries for cervical hemangioblastomas without the development of disability as a result of neck pain. Of the examined risk factors leading to increased neck disability, patients were most negatively affected by additional surgery at the craniocervical junction or cervicothoracic junction ($p = 0.01$). This confirms the overall importance of muscle insertions at these transition areas in maintaining the posterior tension band and preventing muscular fatigue and neck pain.

Removal of the C-2 lamina resulted in increased NDI scores (mean 10.2 ± 11.4 , median 4.4), but this was not statistically significant compared with the level of neck disability reported from patients not undergoing C-2 laminectomy (mean 6.1 ± 5.4 , median 5) ($p = 0.052$). Analysis of the data did not reveal other patient characteristics, MR imaging characteristics, or surgical factors that pre-dispose to the development of increased neck disability.

Postlaminectomy Deformity

The normal sagittal weight-bearing axis of the cervical spine lies posterior to the vertebral bodies, which, in conjunction with the intact posterior tension band, facilitates the formation of the natural lordotic curvature.²³ Pooled estimates of the mean lordotic alignment of the cervical spine (C2–7) in asymptomatic adults average $-17^\circ \pm 14^\circ$.¹² Radiographs obtained at long-term follow-up revealed that 16 patients (64%) remained within 1 SD (-31° to -3°) of pooled estimates of mean lordotic alignment in normal asymptomatic adults (mean $-17^\circ \pm 14^\circ$),^{12,23} and 24 patients (96%) remained within 2 SDs (-45° to $+11^\circ$) of the mean lordotic alignment of the cervical spine (C2–7) in asymptomatic adults. Although overall C2–7 curvature was maintained within 2 SDs of the normal mean lordotic alignment in most patients, an evaluation of plain radiographs in the upright position reveals that facet-sparing multilevel laminectomy was associated with the development of radiographic deformity in 13 patients (52%). Because previous studies in normal and postlaminectomy models of adults indicate that progression of deformity and displacement gradually stabilize, it is likely

that rates and severity of deformity identified in this group of patients are unlikely to progress or increase with additional follow-up³⁴ (Fig. 3).

The type of deformity that developed varied depending on the gravitational center of the head. In the majority of patients in whom deformity developed (7 [28%]), the center of gravity was located anterior to the C2–7 axis and a straight/hypolordotic neck deformity developed, whereas when the center of gravity was located posteriorly (1 [4%]), a hyperlordotic neck deformity developed.³⁴ Additionally, in 10 patients (40%), focal kyphosis developed in the area of multilevel cervical laminectomy. Despite the deformity that resulted from multilevel cervical laminectomy in these patients, MR imaging and clinical evaluation revealed no evidence of nerve root impingement or spinal cord compression in any patient.

Load transmission occurs along a 3-column model in the cervical spine composed of an anterior column (vertebral body), which transmits 36% of the load, and 2 posterior pillars (articular processes and facet joints), which transmit 32% each.²⁴ Because compressive forces change direction and are redistributed along the laminae of C-2 and C-7, these 2 levels typically have larger laminae and have been considered integral in maintaining normal alignment.^{23,24} We found that in 7 patients who underwent C-2 laminectomy, 4 (42.8%; $p = 0.99$) progressed to develop postlaminectomy deformity and 3 (33%) of 9 patients undergoing C-7 laminectomy developed postlaminectomy deformity ($p = 0.23$).

The concept that older patients may be protected against the development of deformity after multilevel cervical laminectomy due to the development of spondylosis⁴³ has been refuted by others.¹⁰ This study supports the notion that these patients undergoing multilevel cervical laminectomy for the resection of tumors are more likely to develop postlaminectomy deformity.

Postlaminectomy Radiographic Instability

The definition of radiographic instability of the cervical spine remains controversial. White and Panjabi⁴¹ have proposed that flexion and extension of the radiographically stable cervical spine should reveal less than 2.7 mm of subluxation and less than 20° of rotation at any level. These criteria are the most commonly used measurement thresholds for the definition of radiographic instability and were used in this study. Recent publications suggest that the 2.7-mm shear threshold guideline may be too stringent for all but the C6–7 level, leading to the overdiagnosis and overestimation of abnormal shear related instability within our patients.³¹

The typical technique of splitting the nuchal ligament, transection of the supraspinous and interspinous ligaments, and removal of the ligamentum flavum and laminae during the approach to resect spinal cord tumors results in transfer of forces required to resist flexion to the facet and the capsule.^{34,37} Zdeblick et al.⁴⁶ studied the effect of progressive facetectomy on segmental mobility in cadaveric specimens and noted that significant segmental hypermobility of the cervical spine resulted if more than 50% of the facet was removed. Removal of facet joints altered the motion segment so that there was less rotation and more horizontal displacement in flexion. Given these findings, and the results of other clinical

studies reporting on the effects of partial facetectomy, our approach has been to avoid disruption of the facet when performing posterior approaches for resection of spinal hemangioblastomas. Despite our facet-sparing operations in all patients, 9 patients (36%) in this study developed radiographically documented instability, suggesting other stochastic factors may play an important role.

Other risk factors, including age ($p = 0.97$) and removal of the C-7 lamina ($p = 0.67$), were not significantly correlated with an increased risk of developing radiographic instability. There was no significant difference ($p = 0.36$) in the mean number of laminae removed in the group of patients who developed radiographic instability and those who did not. Removal of the C-2 laminae was associated with an increased incidence of radiographic instability (5 patients [56%]) and a trend toward statistical significance ($p = 0.06$).

Clinical Stability

Chronic flexion of the cervical spine, in conjunction with ventral compression that occurs when the spinal cord is draped over the kyphotic deformity, has been described as a cause of anterior-posterior cord compression that leads to selective elongation and compression of cord microvasculature supplying the lateral columns and central gray matter, with relative sparing of the anterior and posterior spinal columns.^{4,38} Preventing resultant myelomalacia and motor myelopathic symptoms has been the primary indication for prophylactic correction of early postlaminectomy deformity.

Although severe manifestations of deformity and radiographic instability are likely to result in neurological symptoms, the degree of postlaminectomy deformity and radiographic instability (13 patients [52%] and 9 patients [36%], respectively) that developed in this cohort of patients after a mean follow-up of 9.1 ± 5.6 years was uniformly neurologically benign. Three patients (12%) developed moderate to severe neck disability associated with deformity and/or radiographic instability, suggesting the NDI questionnaire may be a more sensitive indicator of the overall stabilizing system of the spine. Analysis of risk factors that predispose to clinical instability revealed that patients undergoing C-2 laminectomy are at particular risk ($p = 0.02$). These patients require close follow-up, utilizing the NDI questionnaire, clinical evaluation, and radiographic examination to identify the subgroup of patients in whom clinical instability may develop and in whom instrumented fusion may be of benefit.

Spinal Tumors in VHL Disease

Spinal cord hemangioblastomas are common lesions, accounting for more than 40% of VHL disease-associated CNS lesions detected on MR imaging, excluding retinal angiomas.^{7,16,20} Management of patients with VHL disease presents a significant challenge for surgeons because approximately half of tumors (45%) that become symptomatic and require resection are not present at the time of initial imaging evaluation.² In the present study, 14 patients (64%) developed new cervical spinal cord hemangioblastomas not identified on MR imaging at the beginning of the study. Six patients (24%) underwent resection of 15 cervical spinal cord tumors at different times based on MR imaging–documented progression and development of new symptoms. The frequent identification of multiple and new lesions

makes continued and accurate MR imaging of the spinal cord and nerve roots critical. Instrumented fusion, either used to treat or prevent cervical spine–associated instability or deformity, can lead to MR imaging–based artifacts that could prevent identification of new tumors and accurate monitoring of coexisting tumors.^{8,27,33,39,45}

Clinical Implications

Because the management of spinal hemangioblastomas associated with VHL disease requires cautious neurological observation and timely selective removal of symptomatic lesions, it is imperative that high-quality MR imaging be available in the longitudinal assessment of these patients. These results indicate that although a considerable number of patients undergoing multilevel cervical laminectomy for spinal cord tumor removal will develop radiographic instability or cervical deformity over the course of long-term follow-up, only a limited number (12%) will develop clinical instability. Because there may be limited benefit from prophylactic cervical instrumentation at the time of spinal cord tumor resection, close surveillance may help identify the cohort of patients who will require intervention for postlaminectomy changes (surveillance studies include the NDI questionnaire, clinical evaluation, and radiological examination). Particularly close follow-up of patients undergoing C-2 laminectomy or surgery in immediately adjacent regions of the spine is necessary because of the increased risk for developing moderate to severe neck disability and clinical instability. Coupling the need for accurate visualization of the spinal cord and nerves with the low likelihood that patients undergoing multilevel laminectomy will develop significant neck pain and clinical instability, this study suggests that patients can be safely treated without prophylactic concurrent instrumented fusion.

Conclusions

Although patients with VHL disease will often require multiple laminectomies for cervical spinal cord hemangioblastoma resection, a limited number of patients (12%) will develop clinical instability. Because prophylactic cervical instrumentation confers limited benefit at the time of spinal cord tumor resection for most patients, and because these patients need life-long MR imaging of the spinal cord, the quality of which may be affected by instrumentation, longitudinal clinical and radiological evaluation may be used to determine which patients will require stabilization.

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Abbreviations used in this paper

NDI	Neck Disability Index
NIH	National Institutes of Health
ROM	range of motion
VHL	von Hippel-Lindau

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Fig. 1.

Postlaminectomy radiographic changes. **A:** The curvature of the cervical spine is determined by measuring the angle (β) formed by the lines extending from the posterior borders of C-2 and C-7 in the upright neutral position. The spine is classified based on C2–7 curvature as kyphotic ($> 0^\circ$), straight/hypolordotic (-10° to 0°), lordotic ($< -10^\circ$), or hyperlordotic ($< -45^\circ$). Spinal deformity is categorized based on the presence of a kyphotic, straight/hypolordotic (**B**), or hyperlordotic C2–7 curvature (**C**) or the presence of focal kyphosis measuring greater than 10° (α , **D**).

Fig. 2.

Lateral flexion (**A**) and extension (**B**) radiographs were obtained to assess instability. Sagittal rotation is defined as the change between flexion and extension of the angle created by the lines extended from the inferior endplates of adjacent levels ($r_e + r_f$). Horizontal displacement (shear) is defined as the displacement between flexion and extension of the posterior inferior corner of the superior vertebra in the direction defined by the superior posterior endplate of the inferior vertebra. This 21-year-old man underwent resection of 2 cervical spinal hemangioblastomas and the radiographs show greater than 20° of intervertebral rotation and over 2.7 mm of horizontal displacement at the C3–4 level, demonstrating radiographic cervical instability.

Fig. 3.

Artist's illustrations depicting representative longitudinal changes of the cervical spine in a patient with VHL disease. **A:** At initial symptomatic presentation, a spinal cord hemangioblastoma with large surrounding peritumoral cyst is identified at the C3–4 level and a smaller tumor is identified at the C5–6 level. The tumor was removed via C3–4 laminectomies. **B:** Three years later, a symptomatic tumor at the C6–7 level, not present on initial imaging, is identified. A focal kyphotic deformity at the C3–4 level ($\alpha = +11^\circ$) has developed in the interim. Another hemangioblastoma in the cervical spinal cord (C5–6) remains stable. The symptomatic tumor is removed via C6–7 laminectomies. **C:** Seven years after initial presentation and surgery, the focal kyphotic deformity at the C3–4 level ($\alpha = +11^\circ$) remains stable, and focal changes at the C6–7 level are noted ($\alpha = 0^\circ$). The hemangioblastoma at C5–6 remains essentially unchanged in size for the duration of follow-up.

TABLE 1

Summary of demographic and operative characteristics

Variable	No. of Patients*
<u>age (yrs)</u>	
mean $\pm$ SD	33.9 $\pm$ 11.9
range	18–61
median	33
<u>sex</u>	
female	16
male	9
<u>follow-up</u>	
mean $\pm$ SD	9.1 $\pm$ 5.6
range	1.6–18.8
median	7.75
<u>no. of ops (C1–7)</u>	
mean $\pm$ SD	1.4 $\pm$.7
1	19
2	3
3	3
total	34
<u>no. of ops (subocciput–T4)</u>	
1	8
2	6
3	6
4	5
total	66
<u>no. of laminae removed</u>	
mean $\pm$ SD	3.0 $\pm$ 1.3
1	2
2	8
3	8
4	4
5	1
6	2
<u>syringomyelia (no. of patients)</u>	
preop	22
last examination	9
<u>cervical spinal cord tumor</u>	

Variable	No. of Patients *
developed during follow-up	14
last examination	11

* Values represent numbers of patients unless otherwise specified.

TABLE 2

Correlation of patient characteristics and surgical procedures with outcome measures*

Characteristic	NDI	Moderate or Severe Neck Disability	Radiographic Instability	Radiographic Deformity	Clinical Instability
age	0.188 [†]	0.165 [‡]	0.969 [‡]	0.049 [‡]	0.080 [‡]
duration of follow-up	0.801 [†]	0.655 [‡]	0.578 [‡]	0.444 [‡]	0.901 [‡]
no. of laminae removed	0.745 [†]	0.999 [‡]	0.345 [‡]	0.758 [‡]	0.356 [‡]
3 laminae removed	0.735 [†]	0.999	0.691	0.428	0.999
no. of ops (C1–7)	0.794 [†]	0.659 [‡]	0.646 [‡]	0.852 [‡]	0.425 [‡]
>1 op (C1–7)	0.982 [†]	0.999	0.630	0.999	0.999
no. of ops (C0–T4)	0.010 [†]	0.055	0.241	0.228	0.062
>2 ops (C0–T4)	0.186 [†]	0.260	0.999	0.226	0.544
C-2 laminectomy	0.223 [†]	0.0523	0.058	0.999	0.015
C-7 laminectomy	0.408 [†]	0.260	0.671	0.226	0.280
presence of syrinx	0.832 [†]	0.602	0.999	0.688	0.999
developed tumor during follow-up	0.884 [†]	0.999	0.697	0.695	0.999
tumor present at last follow-up	0.549 [†]	0.999	0.999	0.428	0.565

* Unless otherwise specified, data were calculated using the Fisher exact test. Significant values are highlighted.

Abbreviation: C0 = subocciput.

[†] Linear regression.[‡] Logistic regression.