

Atraumatic bloodless removal of intramedullary hemangioblastomas of the spinal cord

LEONARD I. MALIS, M.D.

Department of Neurosurgery, The Mount Sinai School of Medicine, New York, New York

Object. The goal of this study was to summarize the author's personal experience in the surgical treatment of 19 patients with intramedullary spinal cord hemangioblastomas.

Methods. All cases were from the author's private practice and were treated between 1967 and 1990. In all cases the intramedullary hemangioblastomas were totally removed by using a unique microsurgical technique of bipolar coagulation, which is fully described in this paper. A bipolar forceps was used to shrink each tumor and detach it from its feeding and draining vessels. Tumor resection was successfully accomplished in all patients. Blood loss was minimal, averaging less than 100 ml, and what little bleeding occurred did so during laminotomy or laminectomy. No bleeding occurred during tumor removal, and no transfusions were given. All patients were symptomatic preoperatively, and all recovered or improved following surgery.

Conclusions. The technique described in this paper makes tumor removal safe, effective, and relatively easy.

KEY WORDS • hemangioblastoma • intramedullary lesion • spinal lesion

SOLITARY hemangioblastomas of the spinal cord are indeed rare, accounting for only 19 (13%) of the 146 intramedullary spinal cord tumors that I have surgically treated. Even this number is skewed by the fact that 14 of these 19 patients previously had undergone one or more laminectomies before their lesions were deemed probably inoperable and the patients were referred to me. Samii and Klekamp¹⁵ treated 10 hemangioblastomas in their series of 100 intramedullary tumors. Five of my patients harbored thoracic tumors and 14 harbored cervical ones. Patients with von Hippel–Lindau syndrome were excluded from this report. Patients with foramen magnum or medullary hemangioblastomas were also excluded and are the focus of a separate report.

Despite the rarity of spinal hemangioblastomas, their characteristics have been extensively reported. Most are solitary tumors. Nearly all are intramedullary, although unique extramedullary and extradural lesions have been reported. These lesions appear brilliant red because they are filled with rapidly flowing arterial blood supplied by a network of enlarged thin-walled arteries that provide dangerous vessels of passage. Spinal hemangioblastomas drain into arterialized veins that are sometimes difficult to distinguish from feeding arteries. This venous shunting is responsible for the characteristic edematous enlargement of the spinal cord. Small syringomyelic cavities are occasionally present at either end of the tumor, but these are generally too small to provide assistance in tumor removal. The spinal lesion is histologically identical to the cystic hemangioblastoma of the cerebellum. Unlike its cystic counterpart, in which the tumor nodule is characteristically attached to the inner wall of a much larger cyst, the spinal tumor is solid and adherent all around to the cord, except

for the pial surface of the tumor and the small wall separating it from the syrinx, if one is present. I have never seen a solitary intramedullary spinal cord hemangioblastoma that did not appear to lie just below the pia mater at some point. This is usually a rather large area, strangely and fortunately located on the posterior surface of the spinal cord, dorsal to the dentate ligaments. This finding is always sufficient to confirm the diagnosis and to permit the resection to begin without requiring a myelotomy. This type of tumor is not invasive and is fully encapsulated. The capsule is extremely thin and delicate. It can very easily rupture, producing a major hemorrhage that is very difficult to control.

Rarely, patients in whom tumors present as a solitary lesion on careful study will return for consultation when additional lesions develop years later. A small group of patients have von Hippel–Lindau syndrome, harboring multiple tumors that may be scattered (Fig. 1). In their series of 18 cases, Murota and Symon¹⁰ found six such cases. Some lesions may be accompanied by renal and other malignancies. In fact, only 40 years ago the multiple tumors associated with this disease were occasionally misdiagnosed as metastatic hypernephromas. Some patients with multiple lesions do not harbor the ocular lesions described by von Hippel. The pathological characteristics of solitary tumors and the multiple ones associated with von Hippel–Lindau syndrome are apparently identical.^{9,13} In new advanced studies authors are currently demonstrating the genomic pattern of von Hippel–Lindau syndrome, a fascinating entity both historically and scientifically.

Diagnosis is not difficult. Clinically, there is localized pain and some evidence of Brown–Séquard syndrome or sensory cape defect, which leads the astute neurologist to order spinal imaging, rather than a cranial study. On contrast-enhanced CT scans, no other tumor enhances as vividly as hemangioblastoma. Unenhanced MR imaging depicts an isodense mass, which enhances brightly after addition of

Abbreviations used in this paper: CT = computerized tomography; MR = magnetic resonance.

contrast agent. Angiography reveals a vascularity of the lesion that is generally matched by no other and appears unmistakable. In my series, a correct diagnosis was made in all 19 hemangioblastomas preoperatively, and surgical removal of the tumor was performed.

Clinical Material and Methods

All patients underwent surgery after endotracheal administration of general anesthesia. Somatosensory evoked potentials were monitored during induction of anesthesia and during positioning of the patient to prevent spinal cord compression during intubation, which usually requires some extension, and to protect the patient from the flexion required for the operative procedure. This monitoring was discontinued after the patient was placed in position because it was found to be of no benefit during the actual surgery and only seemed to increase operative time.⁷

In all patients with cervical lesions, surgery was performed while the patient was in the semisitting position with the back elevated approximately 30°, the knees elevated to the same angle, and the neck flexed. A transvenous atrial catheter was routinely used, as was a precordial or esophageal Doppler ultrasonography probe for detection and care of a possible air embolism. Patients with cervical lesions were also placed in military antishock compression trousers. The head was fixed in the pinned headrest and was never rotated.

Patients with thoracic lesions underwent surgery while in the 45° prone oblique position. (Fig. 2). This position, rarely used by most neurosurgeons, has been my standard choice since 1967 for all thoracic and lumbar laminectomies. Irrigating fluids drain freely, and the muscle layers are much easier to support than when the patient is in the lateral position. The surgeon does not need to lean over the patient, which is required by the prone position, but instead sits comfortably, looking directly into the wound with or without the aid of the operating microscope. The surgeon's wrists are supported on a padded rest placed alongside the operating table, just below the height of the incision. I would have preferred to perform all these surgeries by using laminotomy,¹² rather than classic laminectomy. Unfortunately, 14 of my 19 patients were referred to me after they had already undergone laminectomy and, thus, laminotomy could only be performed in the five patients who underwent primary surgery.

Results

All tumors in this series were totally removed. In one patient multiple tumors developed 4 years later, providing an updated diagnosis of von Hippel-Lindau syndrome. In another patient who originally presented with a C-5 hemangioblastoma, a T-11 hemangioblastoma developed 12 years later. There was only one complication in this series, the swan-neck deformity that developed in the patient in Case 2 (see *Illustrative Cases*). This complication was corrected by an anterior C4-5 fusion, which was performed by implanting an iliac crest graft. There were no infections or leaks of cerebrospinal fluid in this series, and in no patient was the condition made worse by the procedure. In fact all patients improved from their preoperative status.

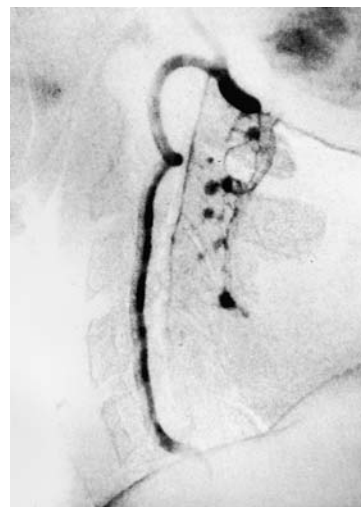


FIG. 1. Vertebral artery angiogram revealing multiple small intramedullary spinal cord hemangioblastomas in a patient with von Hippel-Lindau disease.

Illustrative Cases

Case 1

History. This 21-year-old woman was first seen in 1985. Two years earlier she had begun to experience low-back pain, which was nondescript except that it was worse when she was lying down. By 1985 she had noted right lower-extremity weakness after activity, followed by right lower-extremity numbness and dysesthesia. On examination her gait was normal.

Examination. The patient experienced tenderness at T-12, but no hypalgesia or hyperalgesia in the dermatome. Minimal weakness and dysesthesia were evident throughout the entire right lower limb. There was a slight decrease in the reaction to pinprick in the lower limb on the left side. The patient's reflexes were not significantly altered.

Lumbar myelography revealed an enlarged conus medullaris with a high-grade partial block at T-12 and enlargement extending up the thoracic cord. A T₂-weighted MR image (Fig. 3) revealed the enlarged conus and cord extending up to T-9. An intensely enhanced intramedullary tumor, 2.1 cm long and 1.5 cm in diameter, was located at T-12. The upper end of the lesion was capped by a small syrinx, and a smaller cyst was found at the lower end of the lesion.

These findings were diagnostic of an intramedullary hemangioblastoma. A search for other lesions indicative of possible von Hippel-Lindau syndrome yielded no new findings, and the patient was prepared for surgery. Early in my experience I warned patients of the possibility of major blood loss and possible cord injury; by the time this woman was treated (1985), patients were simply told that transfusion would not be needed and that minor symptoms would probably resolve quickly.

Operation. After the patient had been placed in the prone oblique position, the spinous processes and laminae of T-11, T-12, and L-1 were exposed, and the three-level laminotomy was undertaken by making bone incisions just medial to the facets with the aid of a 1-mm high-speed burr.



FIG. 2. Preoperative photograph showing a patient in the prone oblique position.

Sharp dissection of the ligament permitted these three levels to be removed en bloc. The dura mater was opened and sutured to muscles laterally over the bone margins. The arachnoid was opened and also sewn laterally.

The posterior surface of the spinal cord was covered with a network of large arteries and arterialized veins. At the T-12 level, at approximately the dorsal root entry zone on the right side, the surface of the hemangioblastoma could be seen (Fig. 4 upper). The tumor was bright red, and its surface measured approximately 1 cm in diameter. The lesion protruded only 1 to 2 mm from the spinal cord. Both cephalad and caudad to the tumor, the spinal cord was markedly enlarged.

Using a broad-tipped bipolar forceps, I repeatedly stroked the surface of the hemangioblastoma. The bipolar coagulator was set at a very low power, and continuous saline irrigation was applied. This procedure converted the surface of the hemangioblastoma from bright red to tan, as the tumor was progressively devascularized (Fig. 4 center). A very small amount of pale xanthochromatic fluid drained from a site cephalad as the tumor began to separate.

After the entire surface of visible tumor had been stroked in this manner, the lesion had shrunk enough to expose its lateral, superior, medial, and inferior sides. These areas were also reduced by using the bipolar forceps in the same manner. The progressive decrease in size exposed vascular attachments specific to the tumor. These were coagulated and divided, and the tumor was removed, leaving an untraumatized resection bed (Fig. 4 lower).

The spinal cord was still widened at this area and laterally remained adherent to the arachnoid, which adhered to the dura mater on the right side. This area was separated to permit free movement of the cord within the arachnoid and movement of the arachnoid within the dura. The dura mater was closed and covered with a sheet of Gelfoam. The three laminar arches and spinous processes were replaced in one piece and sewn into place through drill holes made around the bone margins. The usual layered closure was performed. Blood loss was less than 100 ml and occurred entirely during opening and closure.

Postoperative Course. The patient walked the next day and, at discharge a few days later, her neurological signs had almost completely resolved. She remains well 14 years later and is now the mother of two children.



FIG. 3. Case 1. Early (1985) T₂-weighted MR image demonstrating a markedly widened lower thoracic cord with an enhancing intramedullary hemangioblastoma at T-12 and a small syrinx at T-11.

Case 2

History. This 44-year-old man experienced numbness and weakness of his left hand, which had progressed over a 4-year period. Following the appearance of mild weakness of the legs, which was greater on the left side, and some hypalgesia, which was more marked on the right side, he was referred to a neurosurgeon. Myelography revealed marked enlargement of the entire cervical cord, extending down into the upper thoracic segments with a partial block at C-5. These findings were confirmed by MR imaging. The neurosurgeon performed a C3-6 laminectomy. Intraoperative findings included an enlarged cord and a highly vascular, intramedullary tumor surfacing at C-5 on the left side. A biopsy procedure was attempted through a small arachnoid incision, but bleeding was virtually uncontrollable. When the bleeding finally stopped, the wound was closed without further manipulation.

The decompression achieved during this operation resulted in some improvement, but progression of the patient's symptoms during the next year led to his being referred to me.

Examination and Operation. Angiography revealed a 2- to 3-cm hyperintense tumor stain at C4-5 (Fig. 5) in a markedly enlarged cervical and upper thoracic spinal cord. After the patient had been placed in the semisitting position, surgery was performed via the old midline scar. The vertebral canal was abnormally wide and the remaining lateral portions of the arches were thin. The surgical exposure was carried laterally to reach the medial edges of the facets. Although the dura mater had not been closed during the previous operation, the intact arachnoid made microdissection of the scar reasonable.

The tumor measured approximately 2.5 × 2 cm at the site where it surfaced on the left side. The tumor surface was bright red and was surrounded by a large vascular network (Fig. 6 upper). I stroked the surface of the tumor with a 2-mm-wide, round-ended bipolar forceps with the bipolar coagulator powered at a setting of 20. Constant irrigation was applied throughout this procedure and the tumor progressively changed color from bright red to tan (Fig. 6 cen-

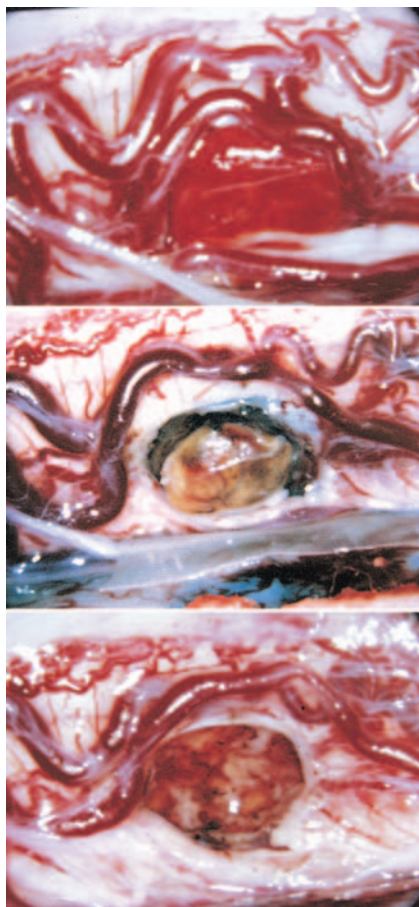


FIG. 4. Case 1. *Upper*: Intraoperative photograph demonstrating the thoracic spinal cord and a bright red hemangioblastoma. *Center*: Same area depicted in the upper panel. After the tumor has been stroked with the bipolar forceps, as described in the text, its color changes to tan and it is decreased in size, creating a gap separating it from the spinal cord. *Lower*: Same area shown in the upper and center panels. The tumor has been removed, leaving a clean resection bed and an untraumatized cord.

ter). A large arterialized venous drain was preserved. Dorsal rootlets of the left C-4 and C-5 vertebrae were engulfed by tumor. Additional use of the bipolar forceps to stroke the lesion's surface reduced its dimensions to 1.5×1 cm. The major draining vessel sedimented and was divided, and the tumor was removed, leaving a normal-appearing resection bed (Fig. 6 *lower*). The very small syrinx located at the lower pole of the lesion drained, but the cord remained much larger than normal both above and below the site where the lesion had been excised.

Postoperative Course. There was immediate improvement in the patient's left arm function, and excellent neurological recovery was attained within 3 months. By that time, however, a swan-neck deformity had begun to develop. The patient was readmitted to the hospital and, following traction reduction, an anterior C4-5 iliac graft was placed, resulting in solid fusion and a normal alignment. A postoperative CT scan demonstrated that the cervical cord was smaller than normal and there was no residual sign of the previous swelling. The patient's only deficit was a min-

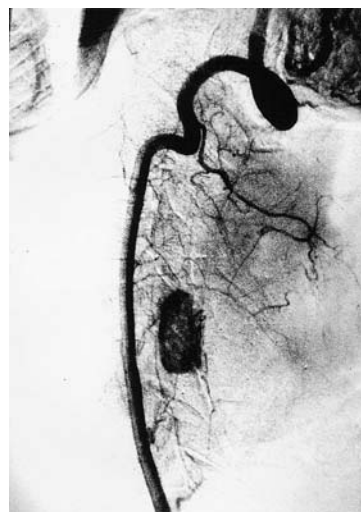


FIG. 5. Case 2. Vertebral artery angiogram demonstrating a dense opacification of C4-5 due to an intramedullary hemangioblastoma.

imal decrease in pin-prick sensation in the first and second digits of his left hand, which was present 2 years later; this remained unchanged when he was examined 12 years later.

Case 3

History. This 53-year-old man had a 15-year history of progressive weakness and spasticity in all four extremities. The presence of a C2-3 tumor had previously been determined. Over the years three attempts were made at tumor excision (including a laser technique) in major neurosurgical centers, but each attempt was unsuccessful. An intense tumor blush was still visualized on follow-up CT scans. A full course of radiotherapy was given, but symptom progression continued and the patient was referred to me.

Examination. The patient was unable to walk or support himself. All four extremities were spastic. Position sense was very poor in all extremities, as was vibration sense. Respiration was still adequate. Angiography demonstrated a somewhat atypical hemangioblastoma blush; better filling was achieved only by combining both vertebral artery injections.

Operation. Surgery was performed using the usual protocol, with the patient in the semisitting position. The midline scar was reopened and dissection was carried in to reach the distended scar that partly replaced the dura mater. The dura was reopened and sutured laterally. The posterior surface of the spinal cord from C-2 to C-4 was densely scarred to an intramedullary hemangioblastoma. The surface of the tumor was typically red in some small areas, but gray, thickened, and scarred in others (Fig. 7 *upper*). The arachnoid was opened where separable. Using a 2-mm-wide, blunt-tipped bipolar forceps with the power set at 20, I gently stroked the exposed portion of tumor under continuous irrigation. The lesion changed from bright red to tan and shrank, creating traction on the surrounding scar. Aided by this traction, the surface attachment could be sharply dissected, and further stroking with the bipolar forceps decreased the tumor bulk further. Fortunately, there was no scarring anteriorly within the cord despite the prior surgery

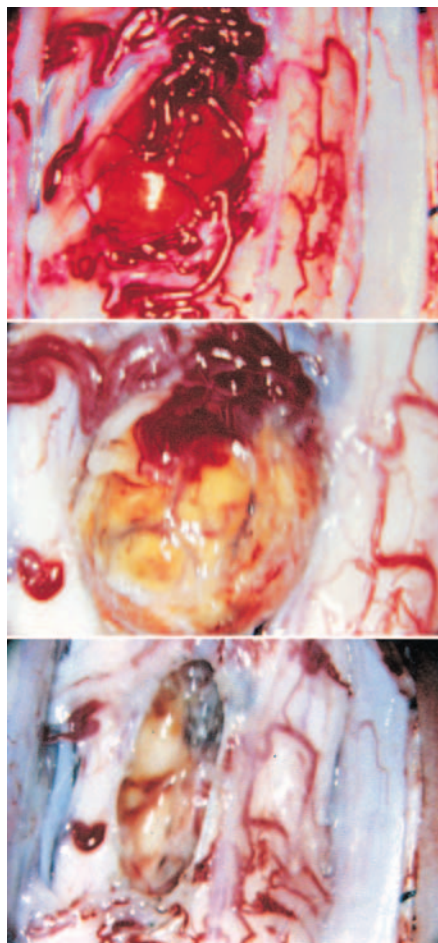


FIG. 6. Case 2. *Upper:* Intraoperative photograph demonstrating exposure of C4-5 and a bright red intramedullary hemangioblastoma. *Center:* Same area shown in the upper panel. The tumor has been stroked with the bipolar forceps and its color is changing to tan. *Lower:* Same area shown in the upper and center panels. The tumor has been removed.

and radiotherapy, and the tumor virtually delivered itself (Fig. 7 lower). This made it easy to manage the vascular pedicle while preserving the branches of the anterior spinal artery, which coursed laterally around the tumor to the cord, as well as the radicular arteries of C-2. After total en bloc resection of the tumor, the usual layered closure was performed, using a strip of Gelfoam to reinforce the scarred dura. Blood loss was negligible throughout the operation.

Postoperative Course. The patient's postoperative course was remarkably benign, although somewhat prolonged by the need to withdraw the high-dose steroid medications that he had received for many months preoperatively. The patient regained sufficient neurological function to walk and care for himself and was finally able to return to work as a salesman.

Discussion

Hemangioblastomas of the spinal cord and brainstem have been considered to pose difficult and dangerous neurosurgical problems because of the risk of massive bleeding. Unlike cerebellar hemangioblastomas, which are near-

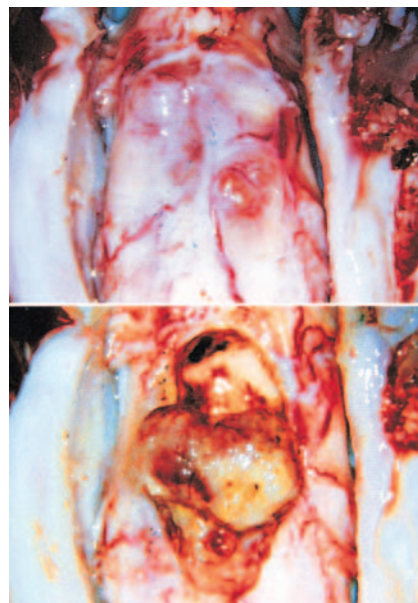


FIG. 7. Case 3. *Upper:* Intraoperative photograph demonstrating the spinal cord at C-2, which has been scarred by prior surgery and radiotherapy. The hemangioblastoma can barely be identified through the scar. *Lower:* Same area shown in the upper panel. After the bipolar forceps has been used to stroke the lesion, it rolls downward out of the spinal cord.

ly always located within large cysts and can easily be resected within the space provided by opening the cyst, these spinal lesions are solid tumors. Accompanying cysts, if present, provide little assistance in the resection. Of course, no adjacent spinal cord tissue may be excised. Most authors who have reported successful series have emphasized the need for a precise angiographic study to delineate the feeding arteries and draining veins associated with the lesion, so that the supply can be isolated and secured before attempting dissection. Even with the most careful microsurgical dissection, this prior knowledge appears to be necessary. It should be noted that the authors of most series reporting good results have been based in major microneurosurgical centers, and the operations have been performed by surgeons with superior qualifications.^{1,8,10,11,14,15} Under these conditions, good results were achieved even in the early days of microsurgery.^{2,6,16}

Embolization performed using a selective microcatheter technique has been recommended, although it poses its own hazards to spinal cord circulation.⁴ I do not believe that embolization should be used. In my technique of tumor removal, angiography has not been necessary to demonstrate the vascular arrangement associated with this lesion, although it was undertaken in a number of patients. (It was routinely performed in patients treated before 1976, when CT scanning became available.) The technique that I have outlined makes the operative procedure relatively easy and safe. As bipolar coagulation empties the tumor of much of its blood and coagulates its tissue, the lesion progressively shrinks and vessels are easily visualized.

Cysts or syringomyelic cavities, ranging from small to large, have been reported to be regularly present in cases of hemangioblastomas. In the series of 10 hemangioblastomas

reported by Samii and Klekamp,¹⁵ there were eight cases in which a syrinx was present. In my series there were only three cases in which there was a significant syrinx. On the other hand, there was frequent enlargement of the spinal cord across a number of segments, both cephalad and caudal to the actual tumor. This enlargement represents edema, which is caused by a marked increase in venous pressure secondary to the shunt through the tumor.⁸ This edema resolves after removal of the tumor. The edematous expansion of the cord has frequently been reported to be a cyst or syringomyelia based on imaging scan findings.

As already noted, these tumors are solid bright red and reach the pia mater at some point. They are encapsulated, but the surface of the capsule is much finer than arachnoid. If they are incised, these lesions bleed almost uncontrollably. Obviously, they should not be subjected to needle or biopsy sampling, and should be removed in one piece. Laser resection has been used successfully in a limited series,³ but my experience in using the laser has made me afraid even to attempt it because it violates the major principle of one-piece resection without opening the capsule.

The technique I began to use in 1967 makes the removal of the tumor both easy and safe. It is based on the fact that most of the tumor volume is composed of circulating blood. After opening the arachnoid, I simply stroke the pial surface of the tumor very gently with a smooth, broad, blunt-tipped bipolar forceps; the bipolar generator⁵ is set at 20, and the procedure is accompanied by continuous saline irrigation. The surface of the tumor shrinks and its color changes to a yellowish tan. It is noteworthy that stroking the surface of the tumor in this manner toughens the capsule, making it less likely to be inadvertently perforated. There is no measurable heat transfer and no spread of current to the spinal cord. The tumor is reduced in size and the plane of cleavage to the cord opens. The adherent pia mater is exposed so well that it may be readily divided. This permits the area to be stroked to extend farther around the tumor, and additional shrinkage exposes the arterial feeders without retraction. These may then be coagulated and divided, sparing arteries of the cord while coagulating branches to the tumor. There is usually at least one major draining vein that can easily be preserved until it is virtually the only remaining attachment to the tumor. After the vein has been coagulated and divided, the tumor may readily be removed. By now it should have been reduced in size by at least one third. Myelotomy above and below the tumor has been recommended.¹⁵ In this series, no myelotomy was required because tumor shrinkage made it unnecessary.

Conclusions

Solitary intramedullary hemangioblastomas of the spinal cord are slow-growing, benign lesions that can cause major spinal cord damage. Radiation treatment is unnecessary, dangerous, and relatively ineffective. These tumors have a reputation for being very difficult to remove because perforation of their extremely thin capsule will produce major bleeding. They must be removed en bloc. The usual micro-

surgical approach is delicate, meticulous, and time consuming, and it often requires spinal cord retraction. Using the aforementioned technique of bipolar coagulation, removal of these tumors is remarkably easy. There is no need for myelotomy or retraction, and there is no bleeding or cord trauma. There has been no tumor recurrence in this series of 19 patients, in whom the minimum follow-up period has been 8 years.

Disclosure

Dr. Malis is a principal shareholder and director of Valley Forge Scientific Corp., manufacturer of the Malis bipolar generators. He is also a consultant to the Codman Corp. and receives royalties from Codman (Johnson & Johnson Professional) for the sale of his instruments.

References

1. Fischer G, Brotchi J (eds): **Intramedullary Spinal Cord Tumors**. Stuttgart: Thieme, 1996, pp 1–115
2. Guidetti B, Fortuna A: Surgical treatment of intramedullary hemangioblastoma of the spinal cord. Report of 6 cases. **J Neurosurg** **27**:530–540, 1967
3. Herrmann HD, Neuss M, Winkler D: Intramedullary spinal tumors resected the CO₂ laser microsurgical technique: recent experience in fifteen patients. **Neurosurgery** **22**:518–522, 1988
4. Hoff DJ, Tampieri D, Just N: Imaging of spinal cord hemangioblastomas. **Can Radiol J** **44**:377–383, 1993
5. Malis LI: Electrosurgery. Technical note. **J Neurosurg** **85**: 970–975, 1996
6. Malis LI: Intramedullary spinal cord tumors. **Clin Neurosurg** **25**:512–539, 1978
7. Malis LI: Monitoring is not essential. **Clin Neurosurg** **42**: 203–213, 1995
8. Malis LI: Spinal cord tumors, in Davidoff R (ed): **Handbook of the Spinal Cord**. Basel: Marcel Dekker, 1986, pp 319–369
9. Miller DJ, McCutcheon IE: Hemangioblastomas and other uncommon intramedullary tumors. **J Neurooncol** **47**:253–270, 2000
10. Murota T, Symon L: Surgical management of hemangioblastoma of the spinal cord: a report of 18 cases. **Neurosurgery** **25**: 699–708, 1989
11. Pietila TA, Stendel R, Schilling A, et al: Surgical treatment of spinal hemangioblastomas. **Acta Neurochir (Wein)** **142**: 879–886, 2000
12. Raimondi AJ, Guitierrez FA, DiRocco CD: Laminotomy and total reconstruction of the posterior spinal arch for spinal canal surgery in childhood. **J Neurosurg** **45**:555–560, 1976
13. Rho YM, Sachdev VP, Malis LI: Von Hippel–Lindau's disease: case report and oncogenic consideration. **Mt Sinai J Med** **42**: 245–251, 1975
14. Roonprapunt C, Silvera VM, Setton A, et al: Surgical management of isolated hemangioblastomas of the cord. **Neurosurgery** **49**:321–327, 2001
15. Samii M, Klekamp J: Surgical results of 100 intramedullary tumors in relation to accompanying syringomyelia. **Neurosurgery** **35**:865–873, 1994
16. Yaşargil G, Antic L, Laciga R, et al: The microsurgical removal of intramedullary spinal hemangioblastomas. Report of twelve cases and review of the literature. **Surg Neurol** **3**: 141–148 1976

Manuscript received January 28, 2002.

Accepted in final form May 10, 2002.

Address reprint requests to: Leonard I. Malis, M.D., 219-44 Peck