Intramedullary Spinal Cord Tumor

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Tumor Subtypes

Intramedullary spinal cord tumors are rare and challenging entities, comprising 16% to 58% of all primary spinal cord and 2% to 8.5% of all primary central nervous system tumors in the adult and pediatric populations.

Analogous to their extramedullary counterparts, three major histologic subtypes predominate. Together, ependymomas (30-40%), astrocytomas (30-35%) and hemangioblastomas (2-15%) correspond to more than 90% of all intramedullary spinal cord tumors. Other tumors such as gliomas, cavernomas, hamartomas, metastases, and lipomas are rarely encountered.

Several genetic factors have been associated with intramedullary spinal cord tumors. Neurofibromatosis type 1 (NF1), neurofibromatosis type 2 (NF2), and von Hippel-Lindau disease (VHL) are the most common genetic diseases that are prone to cause the development of astrocytomas, ependymomas, and hemangioblastomas, respectively.

Approximately 20% of patients affected by neurofibromatosis type 1 and type 2 will develop an intramedullary spinal cord tumor. Similarly, 20% to 43% of the patients with an intramedullary hemangioblastoma harbor the diagnosis of von Hippel-Lindau disease.

Intramedullary spinal cord tumors are particularly related to syringomyelia (25-58%), especially those tumors located in the lower cervical and upper thoracic spine. The occurrence of a syrinx is seen as a sign of a favorable outcome after resection because it denotes a noninfiltrative tumor and a rapid postoperative recovery after the resolution of the fluid-filled cavity.

Half of the syringes occur above the tumor, whereas 40% are above and below, and only 10% are primarily below the tumor.

For a detailed discussion on extramedullary tumors, please refer to the [Extramedullary Spinal Cord Tumor](#) chapter.

Ependymomas

Intramedullary ependymomas are usually soft, slow-growing, well-circumscribed, gray-reddish masses with moderate vascularity. They typically affect adult patients in their third to fourth decades of life and are rare in children. Neurofibromatosis type 2 can be found in up to 8% of these patients. Intramedullary ependymomas have a male predominance (ratio of 2:1).

Intramedullary ependymoma is the most common histologic subtype associated with syrinx formation. They predominantly affect the cervical and thoracic spines, whereas occurrence in the conus medullaris is considered rare.

Myelopathic symptoms predominate because of the central location of ependymomas. The most frequent presenting symptoms are sensory changes, gait ataxia, motor weakness, and pain. At presentation, most patients are able to walk.

Gross total resection leads to cure for most patients. However, ependymomas are associated with the highest rates of surgical morbidity among all intramedullary tumors.

Astrocytomas

Intramedullary astrocytomas are generally ill-defined, infiltrative masses. They correspond to the second most common intramedullary spinal cord tumor in adults during third to fourth decades of life. High-grade lesions are usually identified in adults, whereas low-grade lesions usually affect younger patients. Malignant degeneration has a relatively common occurrence, affecting one-quarter of the adults with astrocytomas.

Astrocytomas occur mainly in the cervical spine and are associated with a syrinx in 20% of patients. Similarly to intramedullary ependymomas, adult patients commonly experience sensory changes, gait ataxia, and motor weakness in about equal distribution, and approximately one-third of patients experience some sort of pain.

Children have a relatively different presentation; motor weakness is by far the major complaint at admission. Diagnosis in small children is challenging because of the appearance of unusual symptoms, such as abdominal pain, torticollis, and muscle atrophy. Of note, however, 4% of children who suffer from an intramedullary tumor present with progressive scoliosis, mainly associated with pain and other neurologic disturbances.

Holocord presentation, in which most or all of the cervical and thoracic spinal cord is involved, is rare, but mostly related to the diagnosis of a pilocytic astrocytoma.

Hemangioblastomas

Intramedullary hemangioblastomas are small, benign, well defined, richly vascularized masses that commonly affect patients in the fourth decade of life. They are not age discriminant, predominantly affect males, and are mostly sporadic, but occurrence in the spinal cord should raise the suspicion of von Hippel-Lindau disease.

Hemangioblastomas can develop anywhere in the neuraxis, but are most commonly found in the posterior or posterolateral cervical cord. Syringomyelia is not a common occurrence in comparison with the aforementioned histologic subtypes; however, cystic degeneration can be observed in up to 70% of these tumors.

Diagnosis

Most patients demonstrate a typical clinical course starting with a slow progressive myelopathy, without clinical remission. Radicular symptoms can appear if there is lumbosacral involvement. Acute deteriorations are rarely observed, but when present, can occur in the spectrum of

intratumoral hemorrhage or malignant tumors.

Evaluation

Magnetic resonance (MR) imaging with contrast enhancement is the diagnostic study of choice for these tumors. MR imaging can help define the tumor-cord interface, as well as the occurrence of associated cysts, syringomyelia, and hemorrhage.

Intramedullary ependymomas, astrocytomas, and hemangioblastomas demonstrate similar signal characteristics on MR imaging, which makes them difficult to differentiate based on imaging alone. Gliomas (ependymomas and astrocytomas) are commonly hypo- or isointense on T1-weighted and iso- or hyperintense on T2-weighted sequences. Contrast enhancement is similarly heterogeneous.

Hemangioblastomas appear iso- to hyperintense on T1-weighted and hyperintense on T2-weighted images, whereas enhanced images reveal a homogeneous pattern.

Ependymomas are typically midline tumors with symmetrical cord expansion over several vertebral levels. Some may display an eccentric configuration together with an extramedullary component, but these configurations are rare. Astrocytomas, on the other hand, are generally eccentric, infiltrative lesions. Distinction between astrocytomas and inflammatory reactions can be troublesome in the case of no contrast uptake. Finally, hemangioblastomas are commonly solid or cystic masses with a mural nodule and can demonstrate significant perilesional edema.

Intratumoral hemorrhage is relatively common with ependymomas, even though not pathognomonic. The characteristic hemosiderin caps on either tumor poles or within some ependymomas are often present.
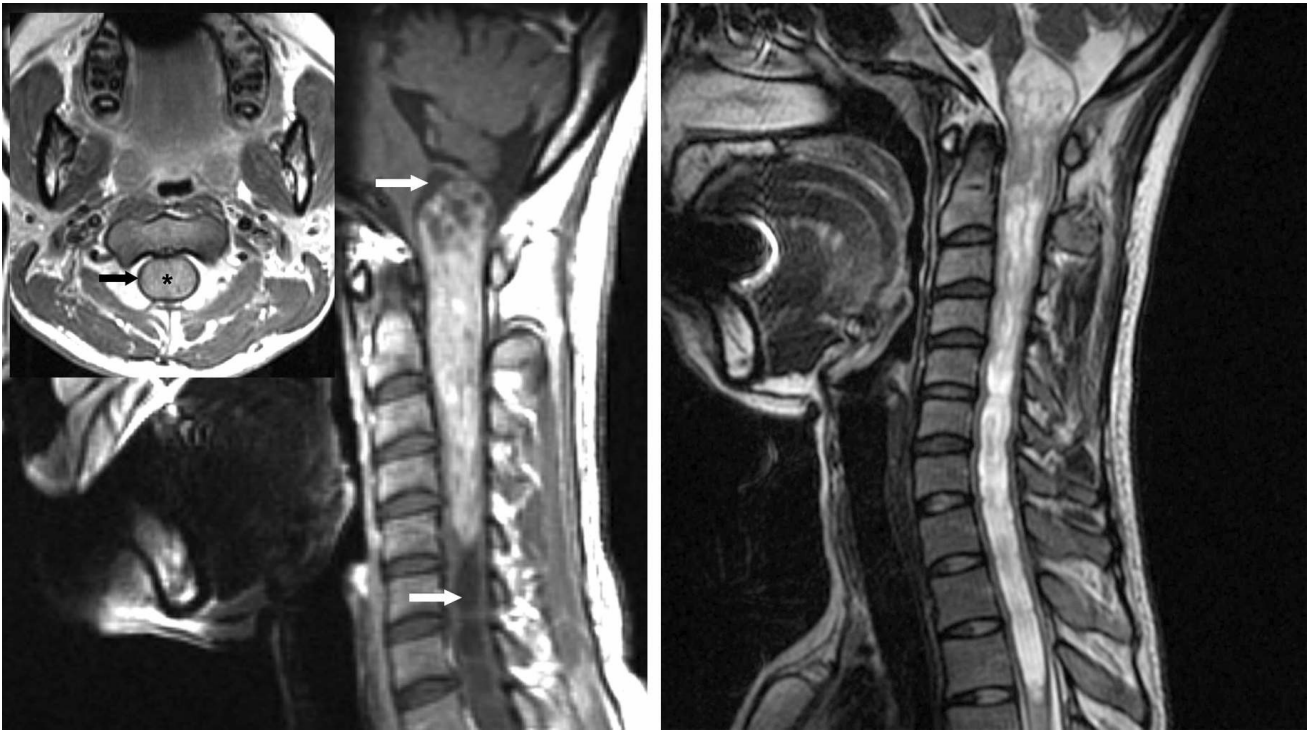
Figure 1: Typical imaging presentation of a cervical ependymoma: heterogeneous enhancement, syringomyelia above and below tumor mass (white arrows), and symmetrical cord expansion, which is flattened to a small rim (black arrow; left image). Note the extensive T2 signal change within the cord beyond the syrinx (right image).
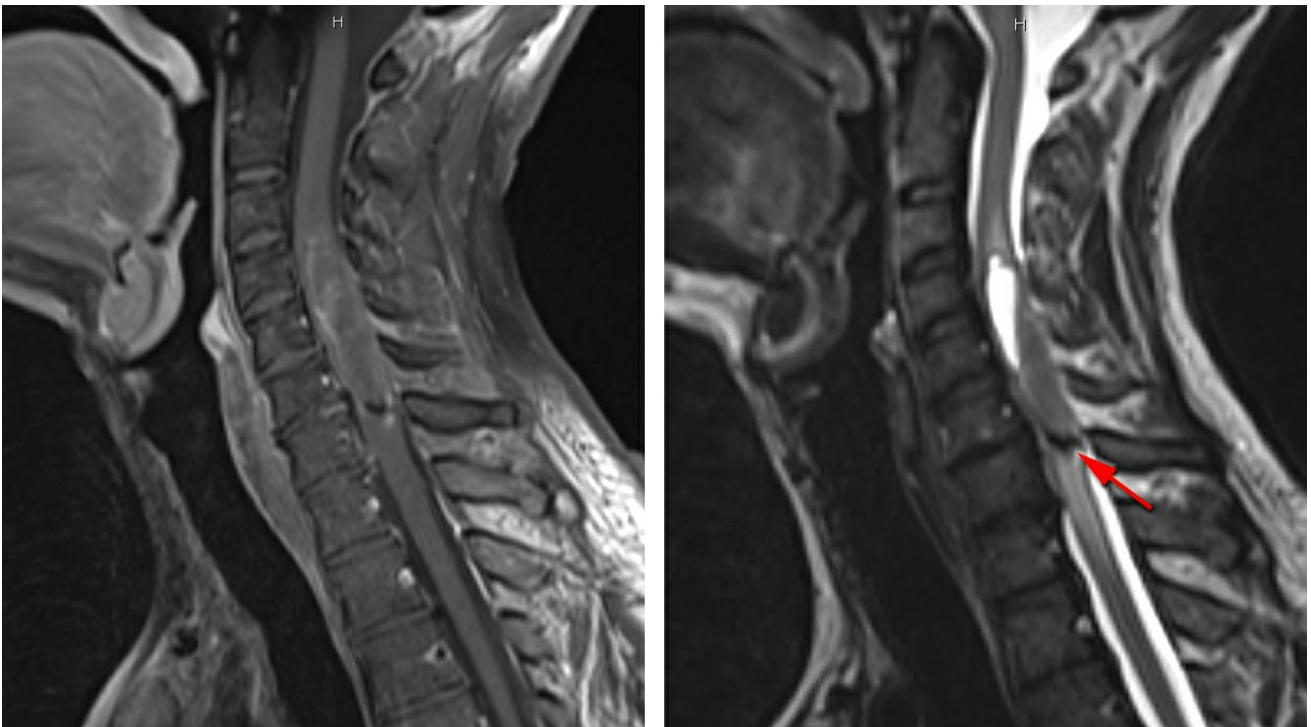

Figure 2: A cervical ependymoma with minimal enhancement (left image) associated with a small nidus of hemorrhage along its lower pole (red arrow, right image).

Figure 3: Imaging of a thoracic hemangioblastoma is shown in a patient with VHL and multiple lesions. Note that the large tumor has an eccentric position in the spinal cord associated with a large-size cyst.

Figure 4: A thoracic pilocytic astrocytoma with an associated cyst is shown. This tumor type typically has an eccentric dorsal location within the cord. Unlike higher-grade astrocytomas, pilocytic astrocytomas almost always are well defined and tend to displace rather than infiltrate the cord.

Spinal X-rays have a limited role, but are still recommended in select cases to investigate the bony anatomy, the occurrence of scoliosis, and spinal instability in the spectrum of recurrent tumors. Pedicle thinning is rarely seen.

Finally, cranial CT or MR imaging should be undertaken if hydrocephalus is suspected. There is a rare, but reported, association of this condition with intramedullary tumors.

Indications for Surgery

Symptomatic tumors constitute the major surgical indication for their resection. Similarly to their extramedullary counterparts, there is still controversy regarding the appropriate treatment plan for asymptomatic patients. I usually recommend a conservative strategy for asymptomatic tumors that have at least a rim of normal spinal cord overlying them.

Radiation therapy is an unproven option for patients who have undergone subtotal resection of their intramedullary ependymoma. As expected, there is an important adjuvant role for this modality in cases of high-grade astrocytomas.

Preoperative Considerations

MR imaging should include T1-, T2-weighted, contrast-enhanced, and 3D-CiSS (Constructive Interference in Steady State) or FIESTA (Fast Imaging Employing Steady-state Acquisition) sequences in at least two planes.

Digital subtraction angiography (DSA) is an option for studying feeding and draining vessels of hemangioblastomas. I do not believe embolization has a role because of its risk to the collateral *en passage* vessels.

After diagnostic imaging, patients are classified according to their clinical symptoms and functional capabilities. The clinical parameters prognosticate their postoperative outcome. The main scoring systems for intradural extramedullary tumors are the Nurick scale and the Klekamp and Samii neurological score system.

For a detailed discussion about the above clinical classification, please refer to the [Extramedullary Spinal Cord Tumor](#) chapter.

Patient counseling before surgery is very important. Patients should be aware of the high risk of temporary postoperative neurologic deterioration, which affects up to two-thirds of the patients at discharge after surgery. Even though such occurrence is mostly transient, approximately 30% of patients develop permanent morbidity, albeit mostly minor.

RESECTION OF INTRAMEDULLARY SPINAL CORD TUMORS

Exposure

For a detailed discussion of perioperative patient care, surgical positioning, and surgical approach, please refer to the [Extramedullary Spinal Cord Tumor](#) chapter. Somatosensory and motor evoked potentials are monitored. Nerve root stimulation is recommended for patients with tumors arising from cervical (C5-T1) or lumbosacral (L2-S1) levels.

For intramedullary tumors, a posterior midline approach is all that is needed for safe exposure. Ligamentum flavum is removed all the way to the next bony segment cranially and caudally. Laminotomy is a reasonable alternative, especially for extensive tumors that require multilevel bone work.

Immaculate epidural hemostasis using gelfoam powder soaked in thrombin is paramount to achieve uninterrupted pristine microsurgery after dural opening.

Analogous to extramedullary tumors, intraoperative ultrasound is also a helpful adjunct in select cases for localizing the exact position of the tumor and the adequacy of the dural exposure before the spinal cord is manipulated.

Operative Approaches for Central Tumors

Centrally located tumors are not readily identified after the dural opening. Occasionally, superficial discoloration may be present as a result of tumor hemorrhage or cystic degeneration. Converging small vessels and avascular areas suggest the position of the posterior median sulcus.

I then examine the dorsal surface of the cord at the area of greatest enlargement for the site of the myelotomy. When the spinal cord is rotated and/or the midline is difficult to identify, the bilateral dorsal root entry zones may be used as landmarks. Intraoperative dorsal column monitoring can be useful in determining the location for the midline myelotomy.

Next, I cut the pia mater with microscissors or an arachnoid knife to develop a midline myelotomy. I avoid excessive coagulation to preserve most of the posterior spinal vessels. The posterior white mater fibers overlying the tumor are subpially mobilized/dissected along their axis using the spreading action of fine forceps.

The initial myelotomy should be only 1 to 2 cm over the most enlarged segment of the cord. I then examine the interface between the tumor and the cord to determine the feasibility of resection. If reasonable planes are present, I further expand the myelotomy along the entire length of the tumor.
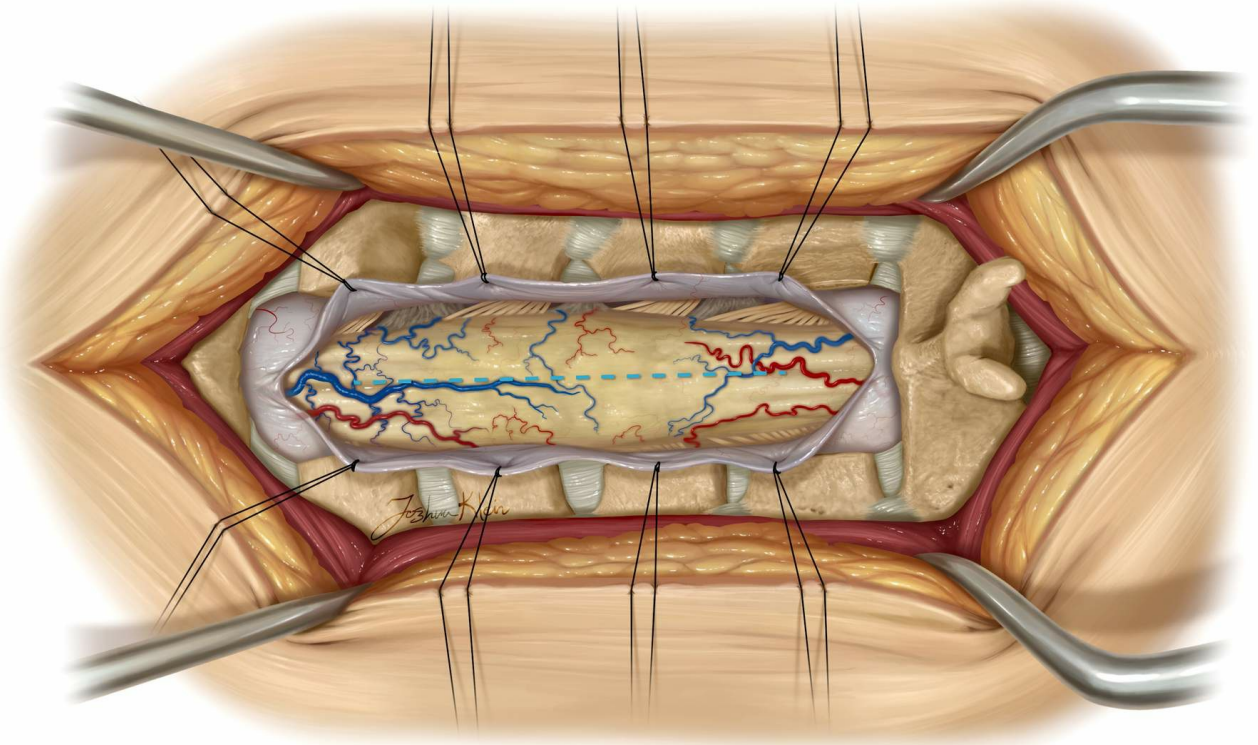
Figure 5: Before intraoperative images are presented, I will review the salient operative maneuvers via illustrations. The laminectomy/-otomy should be inclusive of the entire length of the tumor. The midline myelotomy (dashed blue line) should preserve as many of the dorsal vessels as possible.
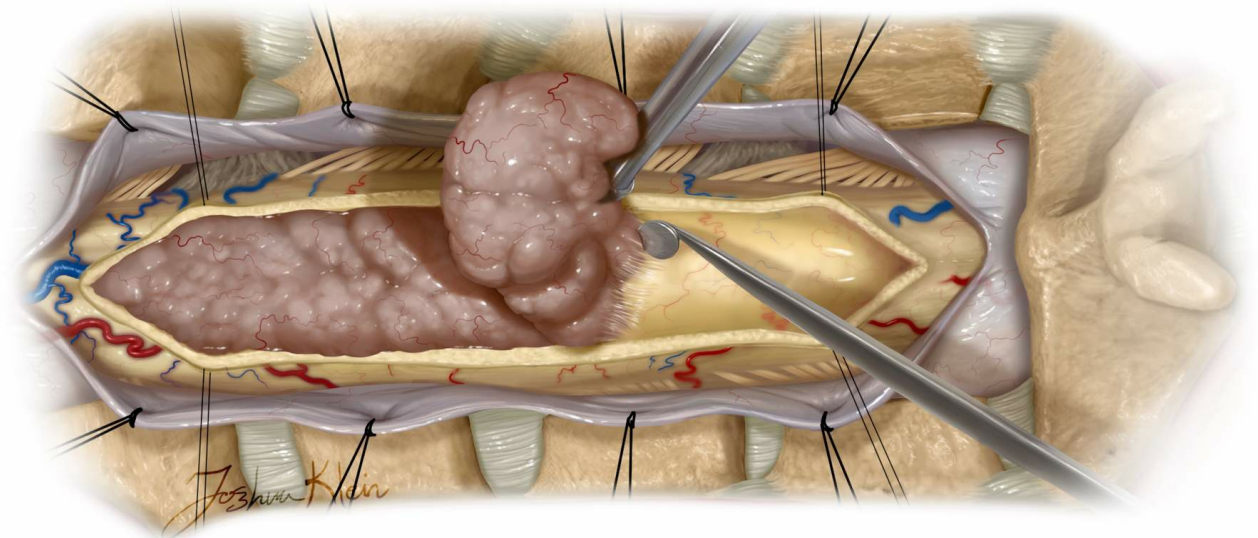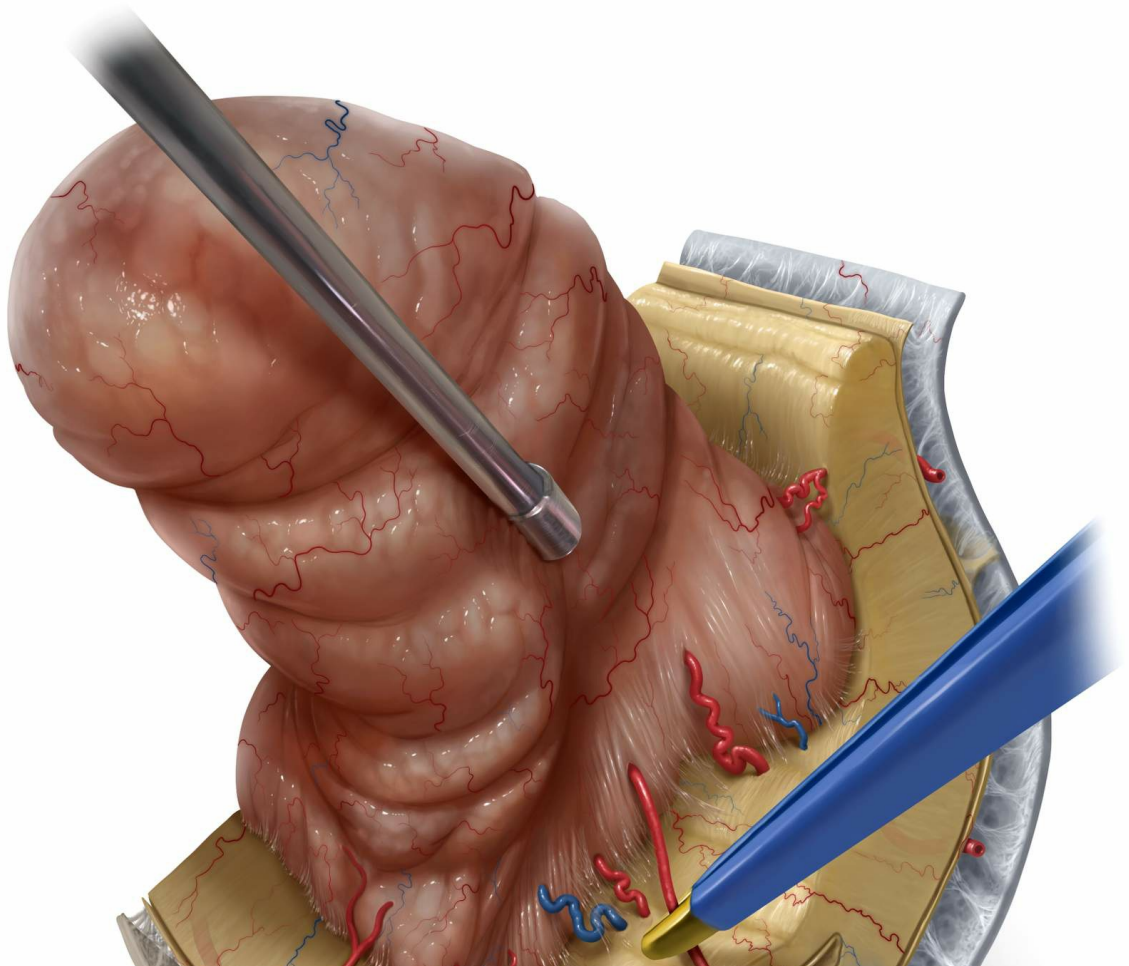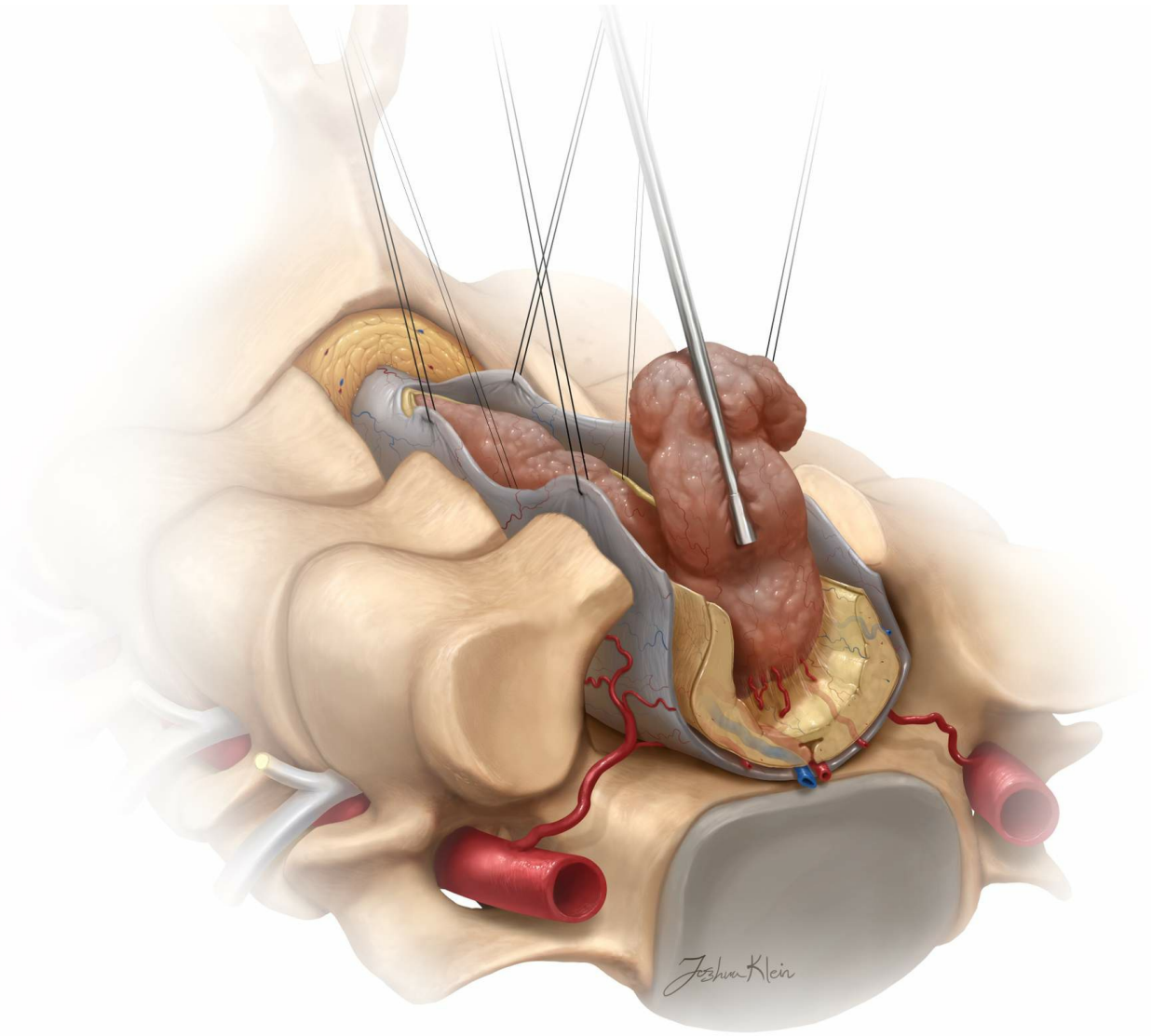

Figure 6: The myelotomy is complete and tumor resection is conducted. Fine pial sutures may be used under spinal cord monitoring to hold the cord open and minimize repetitive cord manipulation via dynamic retraction.
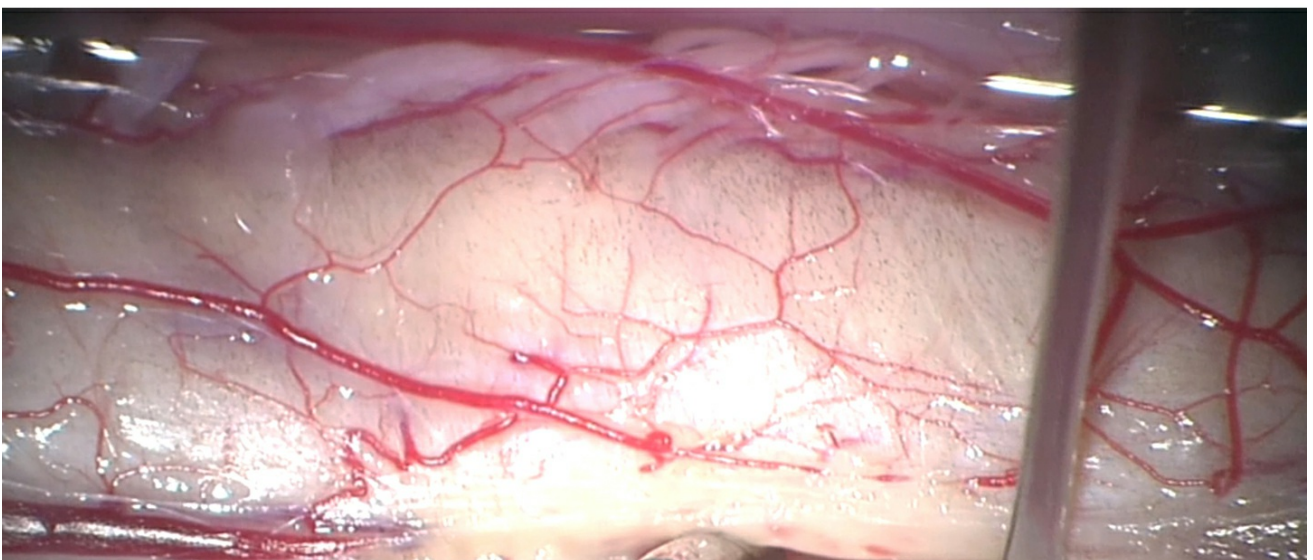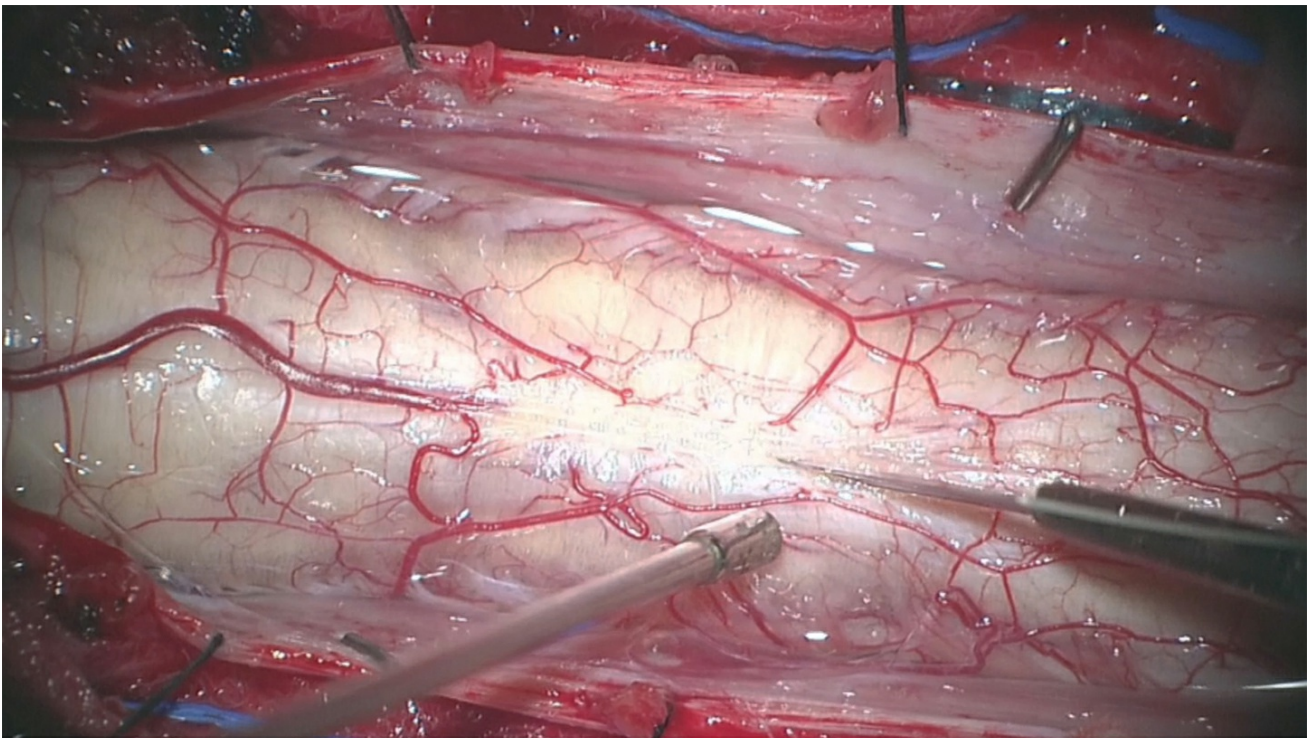
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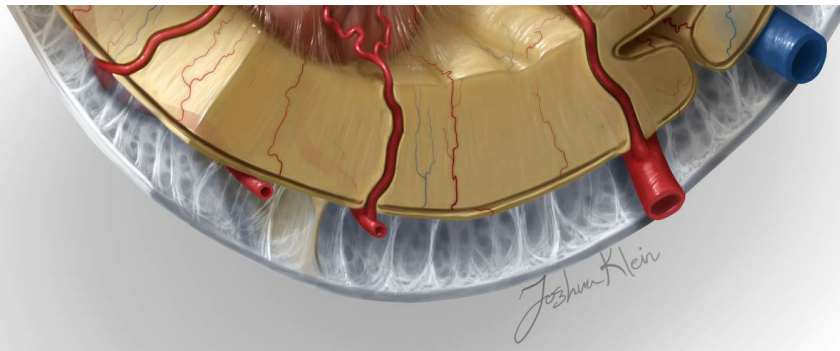
Figure 7: For large tumors, the feeding vessels from the anterior spinal artery should be carefully handled and cauterized/severed. Their avulsion can lead to ventral cord and anterior spinal artery injury.
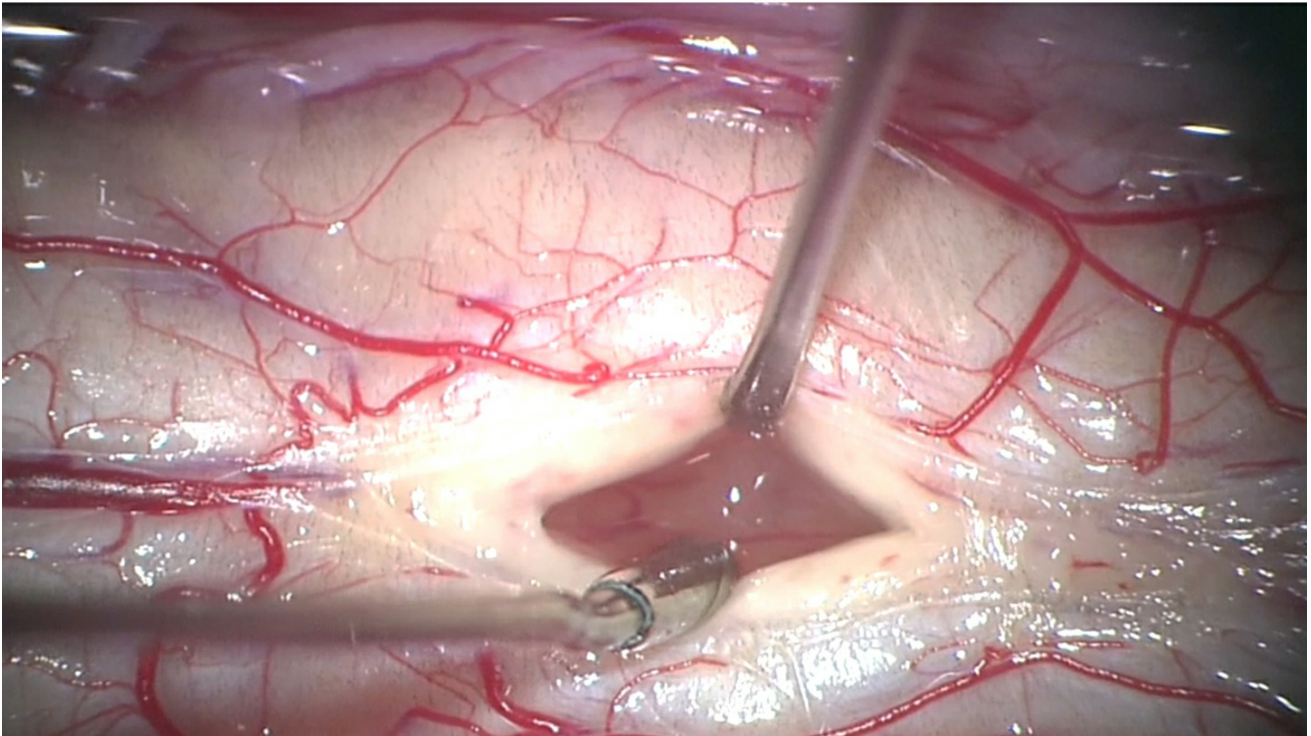
Figure 8: A midline myelotomy is completed for resection of a giant cervical ependymoma (see Figure 1 for preoperative images). After careful coagulation of the midline pia and its incision via an arachnoid knife, a slightly curved fine dissector or a pair of fine forceps are used to gently split and displace, rather than transect, dorsal column fibers (top two images). The countertraction vector of the suction is utilized to uncover the tumor (bottom image).

Smaller tumors may be resected *en bloc*, but generally internal decompression by debulking tumor mass or draining cystic components is needed to minimize the risk of traction injury on the cord during resective maneuvers.
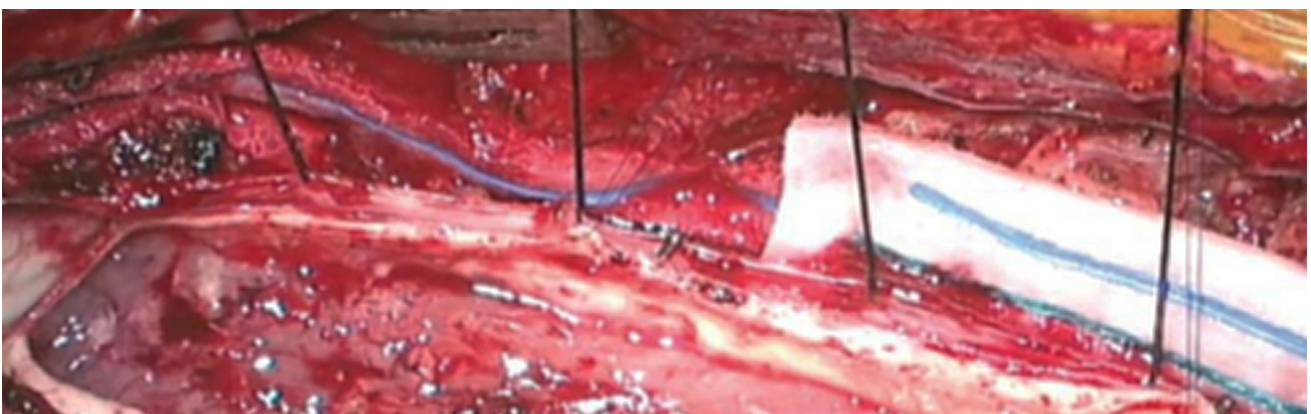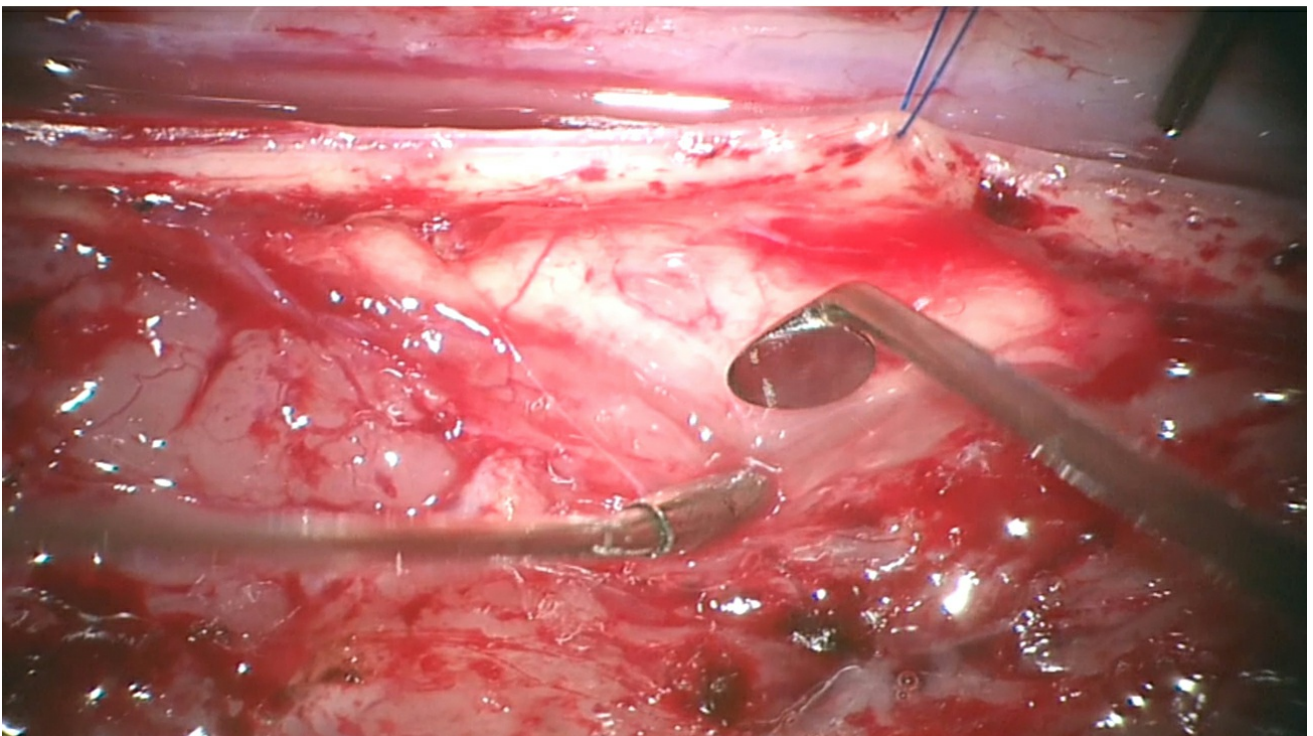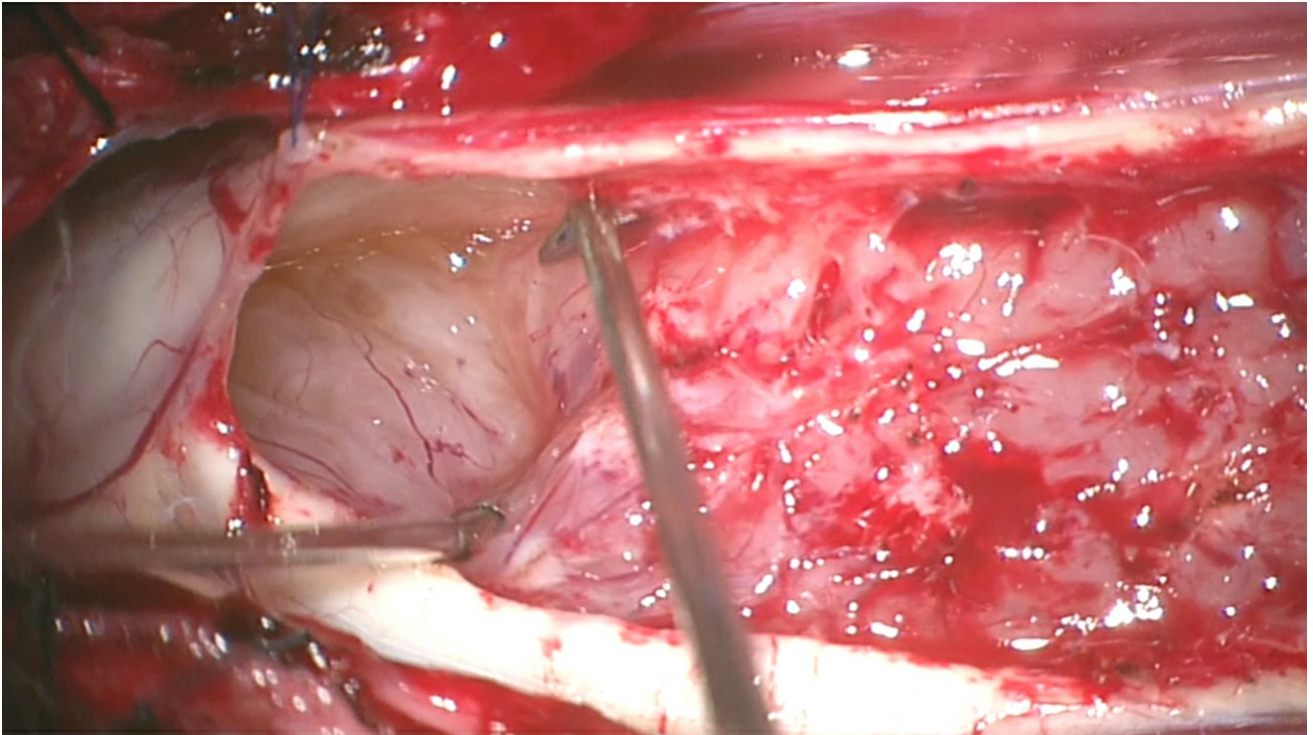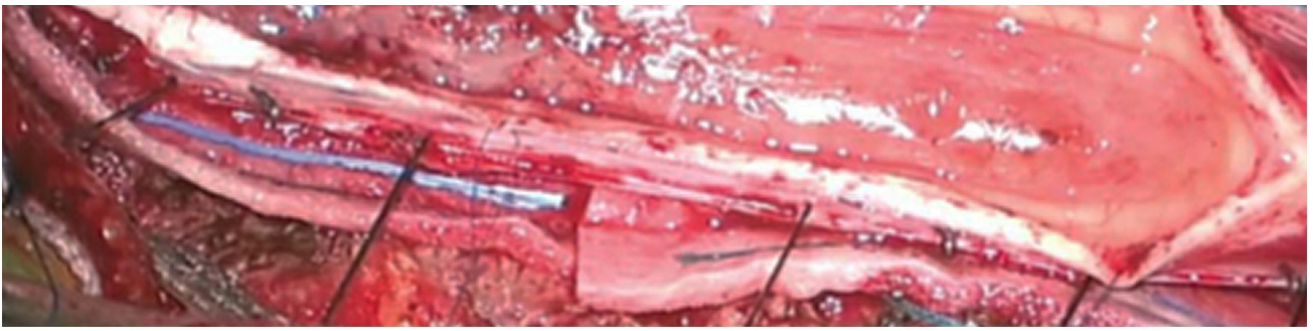
Figure 9: Pial retraction using 6.0 sutures mobilizes the pia mater, increases surgical exposure, and minimizes repetitive cord manipulation (top image). The cyst at the superior pole of the tumor facilitates the initial stages of dissection (middle image). An angled dissector mobilizes

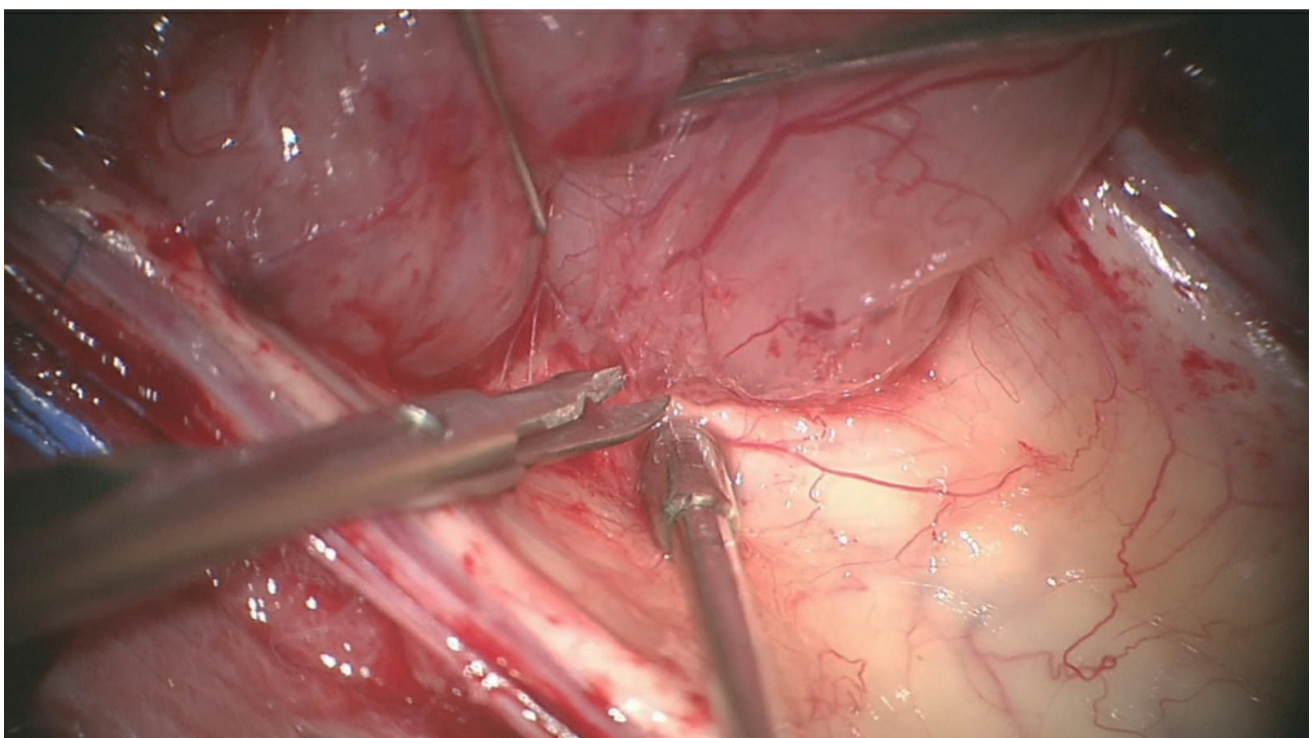the centrally debulked tumor away from the cord (bottom image).
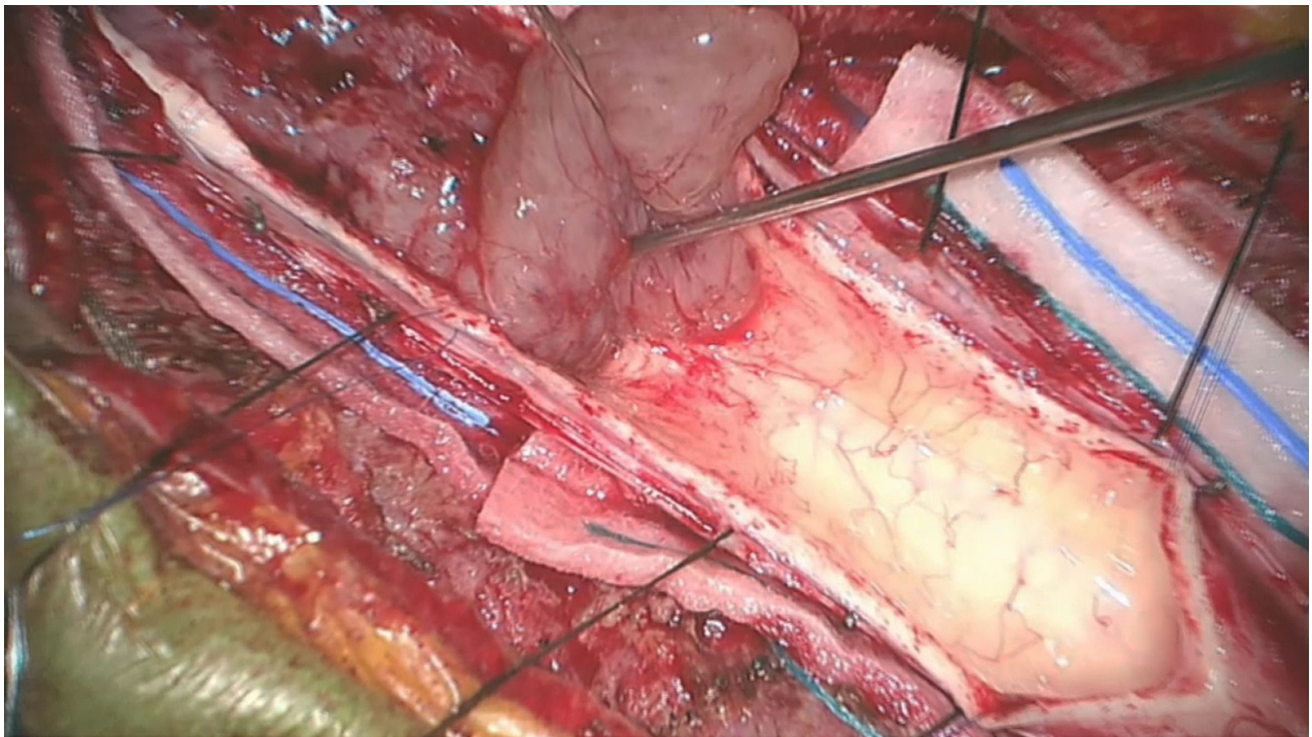
Figure 10: Standard microsurgical methods are used to dissect the tumor capsule away from the spinal cord parenchyma. A gliotic wall as a result of tumor hemorrhage or a syrinx facilitates atraumatic development of cleavage planes. Most ependymomas provide a reasonably identifiable tumor-cord interface. Small feeding vessels deriving from the anterior spinal artery are generally found in the anterior-most aspect of an ependymoma mass. These perforating tumor arteries should be carefully isolated, coagulated, and cut. Indiscriminate traction leads to their

avulsion, placing the adjacent vital vessels at risk during the process of securing hemostasis.

I employ certain techniques during tumor resection. These include preferential retraction on the decompressed capsule rather than the cord to develop the dissection planes. The operative field is kept pristine and meticulously dry so that the plane between the tumor and the spinal cord can be readily identified. The ultrasonic aspirator provides the most atraumatic method of tumor decompression so that the tumor capsule can be mobilized without placing the cord under tension.

I do not attempt to remove the thin walls of the <u>cysts</u> associated with ependymomas because these walls are not neoplastic and their dissection places the integrity of the cord at risk.

The surgeon's intraoperative judgment regarding the decision to attempt gross total resection should be based on the appearance of the tumor-cord interface rather than the histologic diagnosis of the tumor. Some low-grade astrocytomas are well- circumscribed and resectable. However, an intraoperative histological diagnosis of an ependymoma should encourage the surgeon to find a reasonable plane of dissection.

An inadequate myelotomy may cause the surgeon to underestimate the resectability of the tumor. Moreover, a suboptimal biopsy specimen may confuse the pathologist.

Ependymomas are the archetype of centrally located tumors. Centrally-situated astrocytomas undergo resection using the same surgical strategies. Infiltrative tumors (mostly astrocytomas) are debulked until the transitional peritumoral zone is encountered. Resection should stop if no clear dissection plane is detected.

Any change in the evoked potentials should alert the surgeon to stop, relieve the retraction vectors, and apply papaverine-soaked gelfoam pledgets to relieve vasospasm. Gentle systemic hypertension can also lead to the return of potentials. Irreversible but minor changes in the potentials do not usually prevent me from continuing the operation, but a significant

loss of evoked potentials will lead me to abandon the procedure and awaken the patient for definitive evaluation.

Appropriate interpretation of changes in evoked potentials and subsequent surgeon's judicial response rather than "knee-jerk" overcautious reaction are important considerations that demand surgical experience.

Operative Approaches for Eccentric Tumors

Eccentrically located tumors are generally accessed through the dorsal root entry zone or the area where exophytic tumor reaches cord surface. The pia mater is cut and a combination of gentle blunt and sharp dissection is used to circumferentially dissect the tumor capsule. Unnecessary coagulation is avoided.

Astrocytomas are generally infiltrative tumors, which can possess relatively safe cleavage dissection planes. Internal debulking with <u>an ultrasonic aspirator or fine tumor forceps</u> is the first surgical step. Extensive tumors over several cord levels (holocord astrocytomas) may occasionally need a two-staged resection strategy. In the case of malignant astrocytomas, conservative debulking is prudent because a cleavage plane cannot be reliably developed.
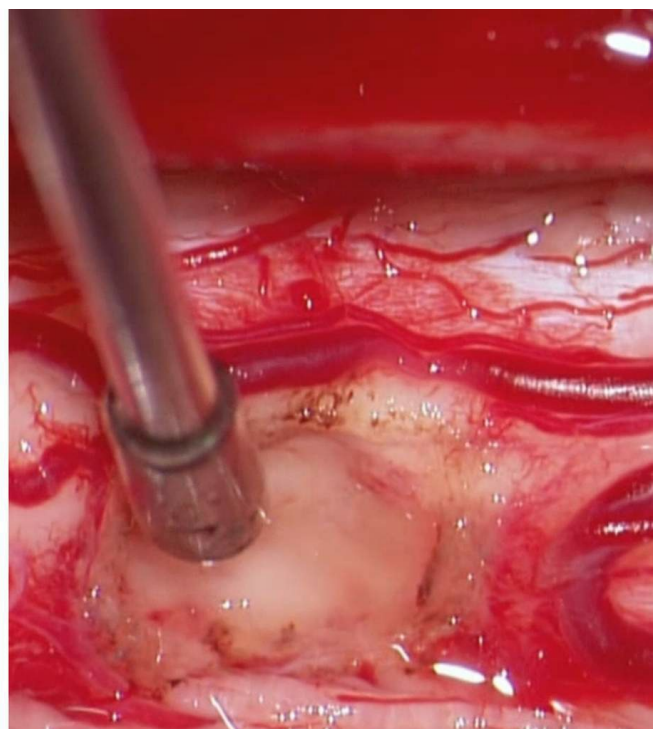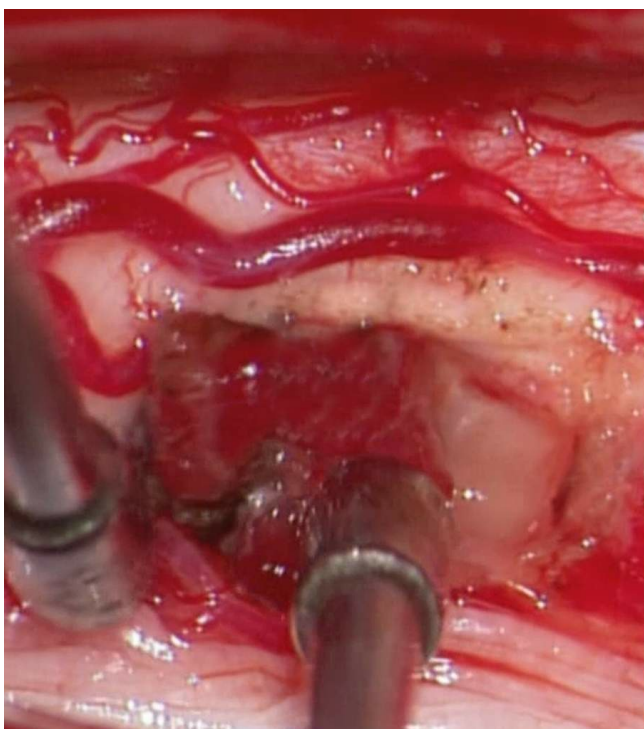
Figure 11: Hemangioblastomas are highly vascularized tumors that are primarily epipial and not intraparenchymal masses. Therefore, their dissection should also be confined within epipial planes when possible to avoid cord injury. They are best removed in an *en bloc* manner, given that any attempt at their debulking is associated with profuse bleeding. The tumor is usually positioned at the posterolateral sulcus (dorsal root entry zone). These "arteriovenous malformation-like" masses should be carefully devascularized around their capsule before their dissection. After tumor definition, bipolar coagulation at a lower power is useful to shrink the tumor mass so that circumferential dissection is made progressively more possible. The occurrence of cystic components or a syrinx facilitates the creation of the deep dissection plane. There is also a gliotic interface between the cord and the deep aspect of the tumor.

The steps in resection of spinal cord hemangioblastomas involve: 1) arachnoid and nerve root dissection, as well as cauterization of the surface or tumor-feeding vessels and identification of the tumor/pial margins; 2) circumscribing pial and polar myelotomy incisions for exposure of the tumor poles, especially for large elongated lesions; 3) the use of 6-0 prolene pial traction sutures for gentle mobilization of the adjacent cord in large lesions, followed by coagulation of the deeper feeding arteries while preserving the draining veins until the completion of devascularization. Finally, the lesion is circumferentially disconnected.

If the patient is affected by VHL, only the symptomatic tumor is removed.

Closure

The dura is closed with running sutures in a watertight manner. Duraplasty is necessary to repair the resultant dural defect if the dural edges are not readily approximated.

Postoperative Considerations

The patient is observed in the intensive care unit overnight for frequent neurologic evaluations as well as pain and blood pressure control. An immediate postoperative MR scan is obtained. Steroids are weaned slowly

postoperatively.

Patients are kept lying flat for 24 hours after the operation and then gradually mobilized.

Pearls and Pitfalls

- The most common histologic types of intramedullary spinal cord tumors are ependymomas, astrocytomas, and hemangioblastomas.

- The goal of surgery is preservation of function followed by tumor debulking and gross total removal if reasonably safe tumor-cord interface dissection planes can be developed.

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