

Surgery of Intraspinal Cord Tumors

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Since the pioneer work of Elsberg,¹⁰ several authors have greatly contributed to spinal cord surgery, including Cooper,⁹ Epstein,¹¹ Guidetti,¹⁴ McCormick,²⁰ Malis,²¹ Stein,^{26–28} and others.^{2,6,8,12,13}

Strategy has been deeply modified, mainly with magnetic resonance imaging (MRI), ultrasonic aspiration, and, recently, with preoperative neurophysiology. But, one should not forget the value of clinical history because MRI alone does not guarantee an accurate diagnosis in every case.⁵ That is the reason why we recommend taking care of anamnesis and neurological examination to avoid performing unnecessary surgery on multiple sclerosis lesions or vascular myelitis. If there is any doubt, a complementary brain MRI scan should be performed to search for demyelinated plaques. Cerebrospinal fluid (CSF) analysis and, if there are any doubts, clinical and MRI follow-up, may also be of great importance and wiser than performing unnecessary surgery.

Spinal Cord Tumors Have No Typical Clinical Presentation

The initial presentation may consist of sensory disorders, torticollis, motor disorders, urinary dysfunction, scoliosis, myoclonus, rarely papilledema, subarachnoid hemorrhage or hydrocephalus, especially in children. Most commonly, adult patients complain of back or radicular pain, or paresthesias. Children present with scoliosis or neurological complaints. The clinical course may be insidious (lasting several years), abrupt in onset, or may progress episodically.

Our strategy differs from gliomas to vascular tumors.^{3,12,13} In gliomas (ependymomas, astrocytomas), we favor a midline approach with gentle separation of posterior columns. In vascular tumors (hemangioblastomas, cavernomas), which are mostly subpial lesions, we approach them directly from where they are seen under the microscope.³

GENERAL CONSIDERATIONS

The patient is positioned prone on bolsters, freeing the abdomen and the thorax from any pressure. We do not use the sitting position for intramedullary tumors, even in the cervi-

comedullary junction. General anesthesia is performed using intravenous opioids and a continuous administration of propofol. Dosage adjustments are necessary to keep the patient's invasively monitored blood pressure close to the one measured supine and awake. Halogenated volatile anesthetics should be avoided because they modify sensory evoked potentials (SEP). Short-acting muscle relaxants are used only to facilitate the intubations, but are not readministered, allowing the use of motor evoked potentials (MEP) during the entire procedure. To prevent inadvertent cervical flexion and pressure on the face, we use a three-point head fixation. Before starting surgery, SEP and MEP are monitored. When the location of the tumor is cervical, we take care to avoid any cord compression due to head position. At the beginning of the procedure, we prefer giving the patient a large dose of methylprednisolone (30 mg/kg), following the spinal cord trauma protocol (30 mg/kg bolus followed by 5.4 mg/kg/h for 23 h).

A midline skin incision is made, centered at the level of the lesion extending above and below it, cutting across the "raphe," and allowing symmetrical retraction of muscular masses. Laminectomy or laminotomy are performed gently and patiently, avoiding any damage to the adjacent spinal cord and preserving the lateral masses, to diminish the risk of postoperative kyphosis. Bone opening is limited one level above and one level below the solid part of the tumor.

In pediatric patients, we always do a laminotomy. In infants, one can perform a unilateral incision of the soft tissues, and unilateral laminotomy is possible. However, when an extensive laminectomy or laminotomy has to be performed in children or in adults, we recommend the preservation of one posterior arch every 5 to 6 vertebrae. Hemostasis in the epidural space requires as much care as that in the soft tissues. A very clean field without any bleeding must be obtained before opening the dura.

We like opening the dura under the microscope, keeping the arachnoid intact when possible. Dural suspension by simple traction sutures may often be sufficient, but sutures to adjacent muscles allow better surgical room. The arachnoid is then opened separately with microscissors, and delicately freed from the posterior or lateral spinal cord, keeping it intact for closure at the end of surgery. Careful inspection of the spinal cord under magnification may show a subpial color

modification by the tumor. If there is any doubt, we use intraoperative ultrasonography, which is helpful in locating solid and cystic parts of the tumor according to MRI data. But, most of the time, we don't need it. Now, it has to be decided if the approach will be posterior through the midline or lateral. In our view, the midline surgical approach is an absolute rule, with one exception: when the lesion is located in one dorsal column and is apparent on the surface without any cortical "mantle." Ependymomas are centrally located tumors (Figs. 22.1 and 22.2). They will benefit of surgery through the posterior midline.^{3,6,12} Astrocytomas are eccentric tumors. Sometimes, they are exophytic and may be followed from outside to inside the spinal cord. But, in most cases, the approach will be similar to the one advocated for ependymomas, all the more so because it is difficult to know the nature of the tumoral process before getting a biopsy.^{12,13} Some rare deep located hemangioblastomas may also be approached through the midline, but most of them are superficial and subpial, and so are cavernomas. There is no need to enter the spinal cord though the midline in such a situation. It is safer to go where the tumor has already prepared the way.³

EPENDYMOMAS AND ASTROCYTOMAS

The first difficulty the surgeon has to face is to secure with the midline. It is not always easy, especially with asymmetrically developed astrocytomas. Identification of the

midline may sometimes be difficult, with a need to search for it above or below the tumor where the spinal cord is normal. High-powered microscopic magnification will allow localization of the dorsal median sulcus, which appears as a distinct median "raphe," over which the very tortuous posterior spinal vein runs. Sometimes, this sulcus is identified only by the convergence of vessels toward the midline. Those vessels of varying size running vertically over the dorsal columns are dissected and mobilized laterally to expose the posterior sulcus, trying to spare all the thinnest arterial or venous vessels in the sulcocommissural region. The dorsal columns are carefully retracted and opened progressively with microforceps and scissors, not with a laser, over the entire length of the solid portion of the tumor, as if they were pages of a book. This maneuver is continued to expose the rostral and caudal cysts, if present.

Pial traction suturing improves the surgical exposure and reduces the severity of repeated trauma due to dissection (Fig. 22.3). This can be accomplished using a 6-0 suture without tension to hold the median pia mater and the dura mater close together, instead of using suspension with sutures and weights at the ends of the sutures. We are aware of SEP recording during that handling to be sure keeping good function of posterior columns. Sometimes, the pressure of the tumor itself helps to keep the posterior columns separate and no pial traction is needed.

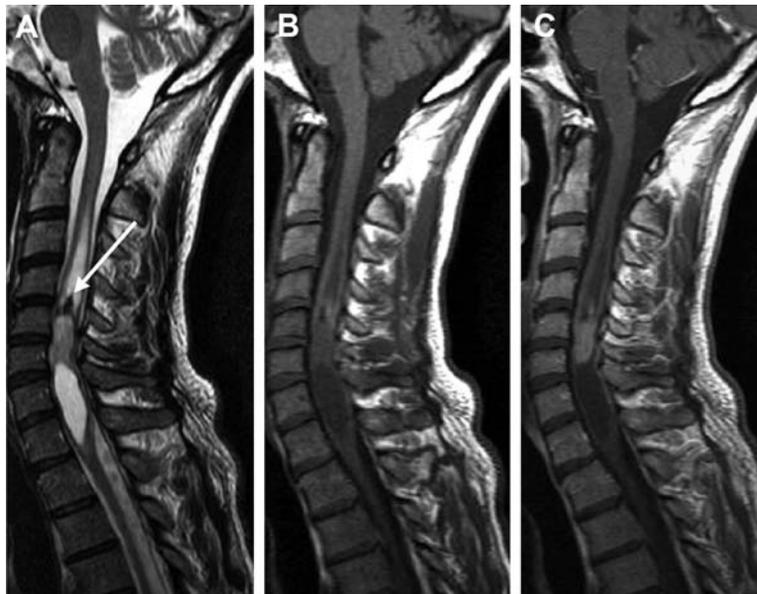


FIGURE 22.1 Cervical ependymoma. A, sagittal T2-weighted image. B, sagittal T1-weighted image. C, sagittal gadolinium T1-weighted image. A solid nodule is seen at the level of C5-C6 which is hyper-signal on T2, iso-signal on T1 and enhancing intensely and homogeneously after gadolinium administration. Above the tumor, there is a small hypointense area (arrow) visible on all pulse sequences corresponding to hemosiderin deposits due to chronic bleeding, the so-called "cap sign" typically seen in ependymomas. Moreover, associated edema is seen above the tumor and as a moderately hyperintense lesion on T2-weighted images and hypointense on T1-weighted images, extending as high as the C2 level. An associated cyst is seen at the level of C7-T1 exhibiting the same signal behavior as CSF. The borders of those "reactive cysts" do not enhance.

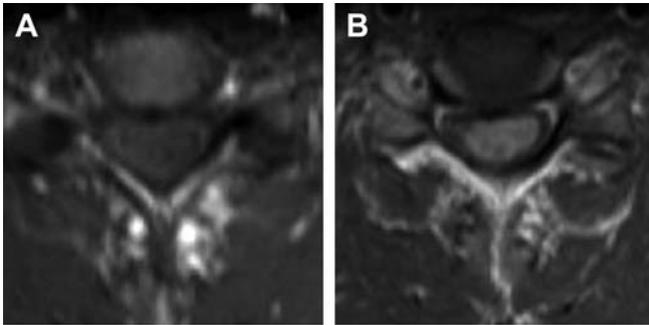


FIGURE 22.2 Cervical ependymoma (same patient as in Fig. 28.1). Axial T1-weighted images at the level of C5–C6 without (A) and after (B) gadolinium injection. The enhancing tumor is centrally located within the cervical spinal cord, a typical finding in the case of ependymoma.

Intraoperative monitoring of the functional integrity of the spinal cord during intramedullary procedures has been recognized as a promising adjunct that may help in intraoperative decision-making and prediction of neurological outcome.^{17,22} Historically, SEPs were the first to be used, but MEPs have gained the foreground. The preservation of the SEPs must encourage us to proceed an aggressive intervention, but loss of SEPs is of little value for motor deficit. On the other hand, MEPs correlate more closely with the postoperative motor function. A decline of 50% or more in amplitude of MEPs is a warning sign to the surgeon,²² almost when the tumor is infiltrative. Nevertheless, when there is a good plane of separation, after waiting a few minutes, we go ahead, even if MEPs are bad.

The first surgical maneuver consists of exposing a sufficient portion of the tumor to perform a biopsy with forceps and scissors, without coagulation. This is followed by immediate histological examination, whereas careful hemostasis is carried out before proceeding with surgery. Any information suggesting an infiltrating or malignant tumor, or a non-tumoral process such as sarcoidosis, can be crucial in deciding whether tumor removal should be continued or not, especially when the infiltrative character of the lesion is obvious. Otherwise, we adopt the same policy with ependymomas and astrocytomas: we perform a debulking of the tumor, which is the second surgical manoeuvre, before searching for a cleavage plane. That is essential to avoid any traction or pressure on the spinal cord (Fig. 22.3). Reducing the volume of the tumor is done by using ultrasonic aspiration after setting the suction at the lowest possible level and using vibratory force to a suitable low degree, because of the fragility of the location. Intratumoral resection is performed from inside to outside, and this is sometimes facilitated by the presence of a cyst or an intratumoral hematoma. It is also important to remember that no one ultrasonic aspirator makes the

difference between tumor and spinal cord. Only the surgeon does. We are not in favor of laser surgery, which chars the tissues in a procedure in which the best landmark between tumor and normal cord is the color. We prefer to keep that landmark clean under our eyes. Thereafter, it is essential to look for a cleavage plane, which exists in most ependymomas^{3,6,7,20,25} and in 30 to 40% of astrocytomas.^{3,4,7,15,24} That is the third surgical maneuver. Finding the cleavage plane makes the tumor removal possible, with the objective being total removal. The best dissection is made with two micro-forceps, like we do when separating an acoustic schwannoma from the brainstem. It is safer than using microdissectors that may hurt the spinal cord. We also prefer using cottonoid moistened with saline at 37°C or a protection at the tip of the sucker. It is essential to keep the field clean from blood and not to lose the plane. If there is a capsule, or if the tumor is not too friable, it can be grasped, allowing visualization of the correct cleavage plane, which must be respected. We have observed that the difficulties are often different on each side or on each pole of the tumor. That is the reason why, instead of persisting in a particularly difficult area, or when SEP or MEP give us warning signs, we recommend moving the microscope either from one side to the other, or from one pole of the tumor to the other, and coming back later after further reduction of the tumor bulk has been carried out with the ultrasonic aspiration.

If the plane is not easy to find, particularly if the intraoperative appearance is suggestive of an infiltrating tumor, we exercise caution and we don't continue tumor removal at any cost because this may be both dangerous and unnecessary for the patient. At that stage, MEPs are of great

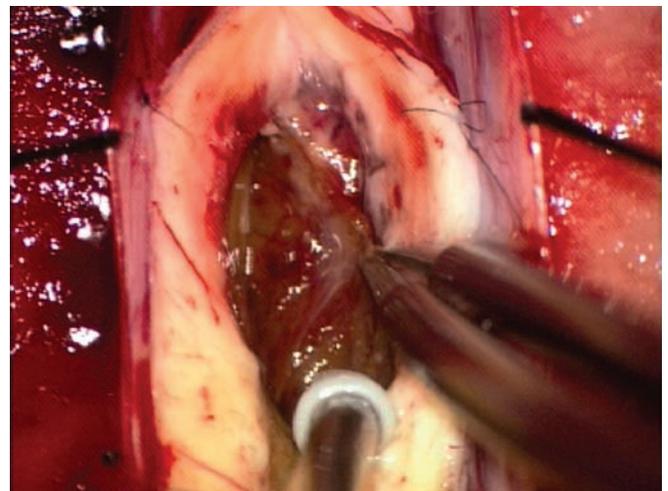


FIGURE 22.3 Intraoperative image showing a cervical ependymoma. The spinal cord opened with pial sutures to lateral dura. Separation of the tumor from spinal cord.

help to safely go as far as possible, always keeping quality of life in mind. If no plane of separation is found, as is often the case with many astrocytomas (Fig 22.4), no aggressive surgery should be done.

The last difficulty lies in the vascular pedicle or pedicles that supply the tumor and are connected to the anterior spinal artery, which may not be injured. Careful coagulation and division of the small arterial feeders makes hemostasis easy. In fact, when removal is macroscopically complete, there is no more bleeding. In view of the small size of the cord vessels, which stop bleeding spontaneously, it is unusual to have to coagulate an area outside the tumor. When normal spinal cord tissue can be seen through a transparent cyst wall, surgery can be terminated, as the cyst wall is similar to that seen in syringomyelic cavities.

After tumor removal, the dorsal columns are released from pial traction and brought together again with caution. Whenever possible, we like to approximate the cord with non-absorbable silk 6-0 pial sutures, putting gently together both posterior columns at the condition the tumor has been totally removed. The arachnoid may also be partially reconstituted with the same suture, if it has been preserved on opening. Indeed, after tumor removal, there is often enough arachnoid because the cord volume is smaller than when

surgery started. In partial or subtotal removal, we only close the arachnoid.

HEMANGIOBLASTOMAS AND CAVERNOMAS

Those lesions are mostly superficial, lying over and into the spinal cord (Fig. 22.5). When they are purely intra, surgery is done through the midline, as described above, but no debulking is made, otherwise massive bleeding may compromise the surgery. When those tumors are seen after opening of the dura on the posterior or posterolateral surface of the spinal cord (Fig. 22.6), they are directly approached.²³ In some small lateral hemangioblastomas associated with a huge and tense syringomyelic cyst, it is useful to gently aspirate the fluid through a 22-Gauge needle to flatten the spinal cord and get an easy access to the solid tumoral nodule.¹⁸ In huge hemangioblastomas, one should pay attention not to coagulate the draining vein at the beginning of the procedure. Under magnification, we start by searching for the limit of the tumor at an area with few vessels. This is the starting point of our dissection. Cautious coagulation on the tumor surface enables to obtain slight retraction that helps detach the lesion from the cord and its vascular connections that are step by step coagulated and divided, ending by the draining vein. The latter, as seen in arteriovenous malformations, becomes blue



FIGURE 22.4 Low-grade glioma extending from C3 to T4. A, sagittal T2-weighted image. B, sagittal T1-weighted image. C, gadolinium T1-weighted image. The tumor infiltration is clearly hypersignal on T2-weighted images, shows almost isosignal on T1-weighted images, and does not enhance after contrast administration. D and E, axial T2-weighted images best show the tumor infiltration predominating to the right part of the medulla.



FIGURE 22.5 Hemangioblastoma of the lower thoracic cord in a young patient with von Hippel Lindau disease. A, sagittal T2-weighted image. B, sagittal gadolinium T1-weighted image. The tumor nodule is posteriorly located and is inhomogeneously hypointense on T2-weighted images, whereas it enhances strongly after gadolinium injection. There is a rich vascular network on the posterior surface of the cord, as this a hypervascularized tumor. There is a typical associated cyst and edema both superior and inferior to the lesion.

when major feeders have been occluded. Then, it may be coagulated and divided without massive bleeding. Both hemangioblastomas and cavernomas should be removed “en bloc.” The fundamental principle is to avoid debulking and to injure the lesion during dissection. This is mandatory for hemangioblastomas for which we advise against the use of ultrasonic aspiration. The cleavage plane is distinct as long as bleeding does not interfere with the dissection. We like closing the arachnoid layer over the surgical field to avoid spinal cord tethering to dura (*Fig. 22.7*). In huge cystic cavities associated with hemangioblastomas, removal of the tumor nodule with opening of the cyst is sufficient to solve the problem.^{3,18} No shunting of the associated syringomyelia is necessary. We never had to do it, even in a second stage. In von Hippel Lindau disease, only symptomatic lesions should be treated.² Multifocal forms, which may or may not be associated with other neuraxial or visceral lesions in von Hippel Lindau disease, raise the question of operative indications in the absence of clinical manifestations. It is now generally agreed that only symptomatic lesions should be treated surgically.

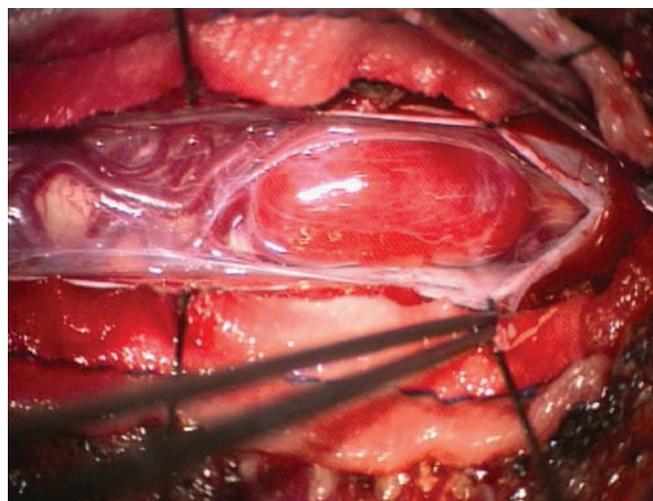


FIGURE 22.6 Intraoperative image showing a thoracic hemangioblastoma, a large tumor with big draining veins.

Some cavernomas should be reduced to be safely removed, sometimes with the help of ultrasonic aspiration, but this maneuver is not generally recommended and a similar approach to hemangioblastomas is advocated.² Many cavernomas are anterolaterally located. They may be safely approached after division of one or two dentate ligament attachments, allowing to gently rotating the spinal cord with the help of 6–0 silk sutures holding the ligament. That provides enough room, giving access to the anterolateral spinal cord from which one may see the cavernoma through a transparent pia-matter.^{3,16} We approach the lesion directly, with a small but sufficient opening of the pia, through which we often put 8–0 silk sutures to keep the path open. Then, with gentle coagulation on the lesion, we shrink it, which gives place to dissect and separate it from the cord. In some huge cavernomas, it may be necessary to design a strategy combining different approaches (posterior and anterolateral) in the same surgical procedure.

Many cavernomas are discovered incidentally. In our opinion, only symptomatic lesions should be operated. We will operate on cavernomas that have bled and those producing symptoms, even mild symptoms such as radicular pain, but we have been quite conservative in recommending surgical removal of incidental cavernomas. In hemangioblastomas and cavernomas, we do not try to close spinal cord opening. It is not possible in most cases. However, we cover the surgical field with the arachnoid that may be approximated and sutured, as done in ependymomas and astrocytomas.

In all the procedures, the dura is closed in a watertight fashion and without tension, with a 5–0 non-absorbable silk. When a biopsy alone has been done, it is better to make a duraplasty. If laminotomy has been performed, the bone is



FIGURE 22.7 Intraoperative image showing complete removal of the hemangioblastoma. All the veins are kept intact. Suture of the arachnoid layer over surgical field.

returned to its place with internal fixation, taking care to avoid any compression of the spinal cord. If needed, a spacer may be placed to gain a larger spinal canal volume. Paraspinal muscles are reapproximated and sutured, as is the fascia

over the raphe, with absorbable Vicryl sutures (Ethicon, Inc., Somerville, NJ). The skin is closed in two layers with a postoperative orthosis in place for 6 months if a laminotomy is performed in children. A Hemovac drain is placed in the muscle layers and removed after 2 days. When the tumor is cervical, the patient is progressively wakened and extubated in the intensive care unit where he stays and is monitored for one night. When the lesion is in the thoracic cord, the patient is wakened and extubated in the operating room, then placed for a few hours in the recovery room before going back to the normal care unit.

All the patients have an MRI scan performed on the next day (Figs. 22.8 and 22.9). They are aware of temporary aggravation of sensory, and sometimes motor deficit, below the level of the lesion. They also receive analgesics and anti-inflammatory drugs to alleviate often distressing, but temporary, pain in the limbs and the spine. Early postoperative mobilization is advocated to achieve a better functional result.

RESULTS

Mortality and Complications

In our personal series based on 360 patients, the operative mortality is 1.9% with none in the last 149 cases.

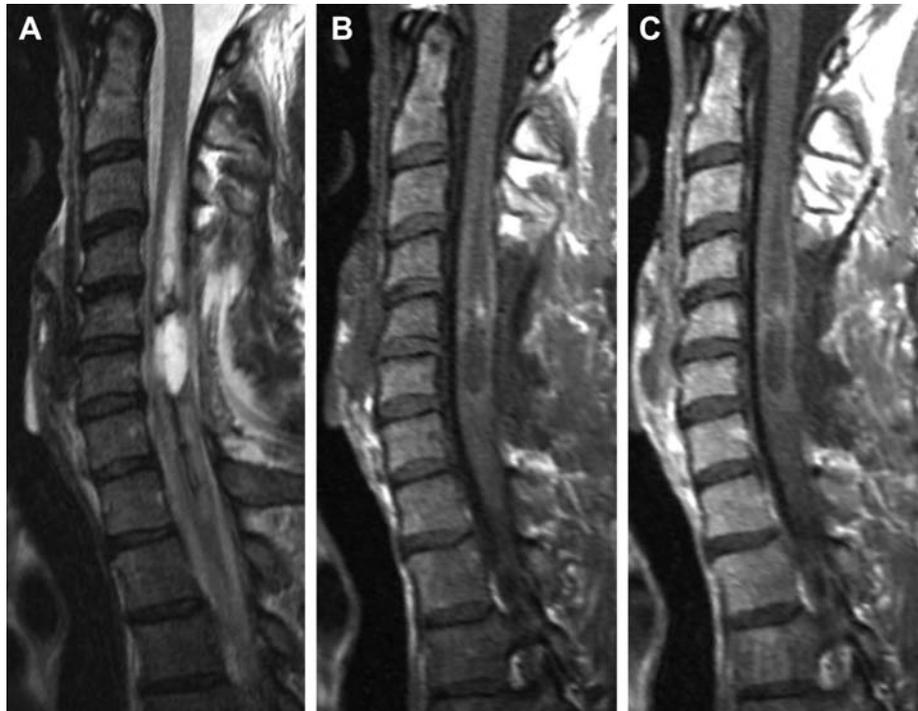


FIGURE 22.8 Ependymoma (same patient as in Figs. 28.1 and 28.2). Postoperative control MRI scan performed 1 day after surgery. A, sagittal T2-weighted image; B, sagittal T1-weighted image; C, sagittal gadolinium T1-weighted image. Complete removal of the tumor is observed and there is no enhancing tumor nodule to be seen. There is a very small hemorrhagic area seen at the level of C5. The underlying associated cyst is already reduced in size.



FIGURE 22.9 Hemangioblastoma (same patient as in Fig. 28.5). Postoperative MRI scan performed 1 day after surgery. A, sagittal T2-weighted image. B, sagittal gadolinium T1-weighted image. The lesion has been completely resected. The enlarged vascular structures are still present, but will disappear on later scans.

Surgical complications (4%) had no effect on the mortality rate, and included the following: hematoma at the operative site or epidural hematoma, arachnoiditis, sepsis, meningocele, cerebrospinal fluid fistula, pulmonary embolism, perforated gastric ulcer, and cervical cyphosis post laminectomy.

The risk of postlaminectomy spinal deformity is well known, especially in children. Risk factors often responsible for postoperative deformities are: young age, the presence of preoperative spinal deformity, cervical laminectomy including C2, laminectomy involving at least six vertebrae and malignant neoplasm or adjunctive radiotherapy, or both. Awareness of the risk factors can enable the surgeon to prevent such deformities by avoiding injury to intervertebral joints, by using osteoplastic laminotomy in children followed by at least 6 months of spinal immobilization, by orthopedic and radiological monitoring until the end of the child's growth period, and by not giving radiotherapy to patients with low-grade glial and non-glial tumors.¹⁹

Functional Results

When they waken from surgery in the recovery room, many patients complain of discomfort and are quite anxious, often more so than patients who have undergone surgery for

other conditions. They feel pain everywhere, and have diffuse hyperesthesia and paresthesias. They cannot, or dare not, move. Position sense in the extremities is poorly perceived. Muscle and spinal pain complete this picture of discomfort, which lasts for several days and is quite unresponsive to the usual analgesic drugs. The severity of the postoperative picture depends on the extent of the surgery.

Before the patient's discharge from the hospital, a basic functional evaluation is made using the four-grade system proposed by McCormick²⁰ to document motor and sensory function. Immediately after surgery, an increase in postoperative deficit, of varying severity, is often seen in nearly all patients. Although this early evaluation provides important information, it does not allow any prognosis to be made, even for the medium term, because most of those signs may be transitory.

At 3 months, the neurological situation is more or less definitive. We have observed, in 360 patients, improvement in tumors with satellite cysts only. If we go deeper in the results, according to McCormick's grades, we see that:

- In preoperative Grade I patients, 5% worsened and 95% were unchanged.
- In preoperative Grade II patients, 7% worsened, 86% were unchanged, and 7% improved.
- In preoperative Grade III patients, 18% worsened, 52% were unchanged, and 30% improved.
- In preoperative Grade IV patients, none except one (a child) improved. Most remained unchanged.

This means that intraspinal cord tumors should be operated in Grade I or II for the best results.

A yearly MRI scan is recommended for the follow-up period. When tumor removal has been complete, with a normal initial MRI examination, subsequent examinations are still necessary to make certain that there has been no recurrence. The appearance or reappearance of contrast enhancement suggests tumor recurrence or resumed progression of tumor growth.

Early MRI examination is of particular value when trying to demonstrate the presence of residual tumoral tissue too. Diagnosis of residual tumor is easy when the lesion was found to enhance after contrast injection before surgery. If there was no enhancement preoperatively, the diagnosis is more difficult, and is mainly based on the presence of residual increased signal on T2-weighted images. This examination will evaluate the residual tumor and serves as a baseline for assessing subsequent progression of the lesion.

RELEVANCE OF RADIOTHERAPY

Based on our experience, we are convinced that there is no indication for radiotherapy in benign spinal cord tumors,

even after incomplete removal, recurrence, or progression. Our personal opinion is based on a follow-up period longer than 5 years in 165 patients operated either for a low-grade ependymoma (104 patients) or a low-grade astrocytoma (61 patients), without adjunctive radiotherapy. After complete removal, we observed only four recurrences (one ependymoma after 18 years and three astrocytomas after 5, 6, and 7 years, respectively). Even partially removed low-grade astrocytomas have a very indolent evolution. Seventeen remain stable in spite of partial removal; few show a slow MRI evolution without any clinical impairment. Furthermore, we found very difficult to reoperate several patients who had previously received radiotherapy after biopsy or partial removal. All have been worsened after our surgery as opposed to those who didn't receive it.

In malignant gliomas, the treatment is only palliative as it is in brain. If in low-grade astrocytomas, irrespective of their histological type, we never prescribe adjunctive radiotherapy, we do it for malignant gliomas but whatever treatment was used for malignant astrocytomas, whether repeat surgery, radiotherapy, or chemotherapy, the disease has always been fatal within 9 months to 3 years.

CONCLUSIONS

Surgery is the "gold standard" in the treatment of spinal cord tumors. Complete removal of the lesion is the first goal. Postoperative results are dependant on the surgeon's experience, but also on the preoperative neurological status. Indeed, in our series, no paraplegic patients recovered except one child. In ambulatory patients, the risk of permanent postoperative deficit was 5%. But, in already handicapped patients, surgical risks of permanent worsening jumped to 18%. That is the reason why we say that, for hoping to get a good quality of life after surgery, patients with an intraspinal cord tumor should be operated before harboring heavy neurological deficit.

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