

## Spinal cord ependymomas

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Ependymomas are the most frequent spinal cord tumors in adult patients. Although magnetic resonance imaging can be a highly accurate diagnostic tool, it does not always provide accurate differentiation between ependymomas and astrocytomas. This is why the authors recommend surgical resection and histological evaluation in all intraspinal cord tumors. It cannot be said that a tumor is unresectable without first attempting to remove it. Complete removal should be accomplished whenever possible, and patients should undergo operation before they become neurologically impaired. Quality of life depends on preoperative neurological status. Postoperative radiotherapy should be avoided in all low-grade ependymomas even after partial removal. Radiotherapy may be used to treat anaplastic ependymomas, which are quite rare in the spinal cord. Patients with low-grade ependymomas must be followed for years and undergo reoperation if necessary. In our experience treating 93 spinal cord ependymomas, complete removal was achieved in 86 patients and only one patient underwent reoperation 18 years later for tumor recurrence. The gold standard in treatment protocol is gross-total resection without adjunctive radiation therapy. Good long-term outcomes have been achieved by using this strategy.

**Key Words \* intramedullary spinal cord tumor \* ependymoma**

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Ependymomas are the most frequent spinal cord tumors in adult patients.[14] These tumors have no typical clinical presentation; however, when the major symptomatology consists of subjective peripheral sensory disorders of the limb or trunk, a diagnosis of ependymoma can be suspected.[5]

Preoperative diagnosis of ependymoma is not easy. Magnetic resonance (MR) imaging, which has a high degree of accuracy for visualizing nonglial tumors (such as hemangioblastomas, dermoid and epidermoid tumors, and lipomas), cannot determine with certainty the differential diagnosis between ependymomas and astrocytomas, except when typical features (described below) are present.[1]

Most spinal cord ependymomas are classified as World Health Organization (WHO)[11] Grade II tumors. In view of their good prognosis they could be classified as Grade I. Grade III malignant ependymomas located in the spinal cord are very rare. Subependymomas are classified as WHO Grade I tumors.[11]

The spinal cord ependymoma is, most often, a benign, slowly developing tumor that may grow to a considerable size, sometimes affecting the entire spinal cord (holocord ependymoma) before becoming clinically detectable. In rare cases, the tumor may be exophytic.[2]

Complete surgical removal of ependymomas whenever possible is the best treatment, and this can be achieved with success in more than 90% of cases. Although some authors advocate adjunctive radiotherapy when only incomplete tumor removal is achieved,[12,22] we disagree with them.

## CLINICAL MATERIAL AND METHODS

### *Patients Selection*

Our experience is based on the surgical removal of 93 spinal cord ependymomas in 43 women and 50 men. Patient age ranged from 18 to 73 years (mean  $41 \pm 12$  years). We did not treat any children.

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Three cases are illustrated on the video clip.

The first patient is a 67-year-old man with a cervical ependymoma extending from C3-6. Three months before operation, the tumor bled, which explains the dark color seen on the left side at gross macroscopic inspection (patient in prone position, head down, on video). The dorsal median sulcus is opened with microscissors. Both posterior columns are gently separated over the entire length of the tumor, which has a brown color. Pial traction suturing eliminates dangerous manipulation of the cord. A biopsy with forceps and scissors is performed, without coagulation. Then, using an ultrasonic aspirator, the tumor is debulked, and careful dissection of the tumor from the spinal cord is performed, without bleeding, and with the help of cottonoids to allow visualization of the correct cleavage plane. Several feeding vessels arising from the anterior spinal artery are cautiously coagulated. Complete removal of the tumor is achieved. The dorsal columns are then released from pial traction, brought together again with caution, and approximated with No. 6-0 pial sutures. The arachnoid is reconstituted.

The second patient is a 57-year-old woman with a C5-7 solid tumor and satellite cysts from C1-T1. After separation of posterior columns, the tumor capsule is grasped with forceps to facilitate dissection, which was quite easy. Complete removal of the ependymoma is achieved.

The third patient is a 40-year-old woman with a C5-T1 friable ependymoma without any capsule. The brown color is very helpful in identifying the tumor and distinguishing it from normal white spinal cord tissue. After tumor debulking, the cleavage plane is followed and complete removal is achieved without any neurological worsening.

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### *Histological Evaluation*

In our series, all tumors were classified as WHO Grade II except two that were classified as Grade III ependymomas. Among the low-grade tumors, two were subependymomas, the others being cellular, tanycytic, or cystic. There was no myxopapillary form typical of the ependymoma of the filum terminale and cauda equina, thus of extramedullary tumors.

### *Clinical Signs and Symptoms*

The spinal cord ependymomas in our patients had no typical clinical presentation, except the prevalence of sensory disorders, which were observed in 62% of patients. Motor disorders occurred in less than 40% of cases. Urinary disturbance was infrequent. The mean preoperative course was 3.9 years.

### ***Tumor Location***

Ependymomas most commonly affect the cervical cord (75% in our series) either exclusively or with extension to the thoracic area or medulla. The thoracic site alone was affected in 16% of our cases, and 9% of tumors were located solely within the conus medullaris. (Table 1)

Tumor Location	Total No. of Tumors
cervicomedullary	9
cervical	33
cervicothoracic	22
thoracic	15
conus	8
holocord	6

### ***Findings on Magnetic Resonance Imaging***

Although MR imaging reveals some intratumoral cysts, more often it shows unipolar or bipolar cysts sometimes with associated hydrosyringomyelia or with lower brainstem cyst. The solid portion of the tumor may be isointense on T<sub>1</sub>-weighted images in 50% of cases. A slight T<sub>1</sub> hyperintensity occurs rarely and suggests the presence of intratumoral hemorrhage. On T<sub>2</sub>-weighted images, ependymomas may exhibit hyper- or isointensity. A hyperintense signal area on T<sub>2</sub>-weighted images, consistent with tumor-associated edema, was observed in 60% of our cases. Gadolinium enhancement of the solid part of the tumor was seen in 80% of cases, with a homogeneous signal in 70%. When this occurred, contrast uptake in the solid portion was complete in 87% of cases and partial in 13%. On axial views, two out of three ependymomas were located in the center of the spinal cord. A "cap-like" appearance (that is, an absence of signal at both poles of the solid part of the tumor) was observed in 30% of ependymomas in our series, and this feature can only be considered typical of ependymoma because we have never observed it in astrocytomas.

In summary, a diagnosis of ependymoma may be claimed when the tumor is located in the center of the spinal cord, when contrast enhancement is homogeneous, when the margins of the tumor are clearly defined, and when a "cap" sign is observed at both poles of the solid portion. Otherwise, MR image diagnosis may be hazardous.

Finally, on MR imaging one can never say if a good plane will be found between the tumor and the spinal cord but when all the signs are present together, it may be suspected.

### ***Surgical Technique***

The entire surgical procedure is performed while somatosensory potentials are recorded. A midline incision is made, centered at the level of the lesion. Bone removal, either through a laminectomy or

laminotomy, should provide sufficient access to the solid part of the tumor. As a rule, we recommend exposing one additional vertebra above and below the tumor. It is not necessary to extend the opening far over the cysts. At this stage, intraoperative ultrasonography may be helpful in locating solid and cystic areas.

We try to open the dura without opening the arachnoid; the latter is opened separately, when possible, with microscissors, which helps in closing it after tumor removal. High-powered microscopic magnification will allow localization of the dorsal median sulcus over which the very tortuous posterior spinal vein runs. In ependymomas, the midline surgical approach is the absolute rule, except in the rare cases of exophytic tumors. We do not use lasers to cut the tissue because we prefer to spread the posterior columns apart with microscissors and microdissectors and to carefully retract them. The surgical field is gently opened progressively over the entire length of the solid portion of the tumor, as if it were pages of a book.[10] This maneuver is continued, exposing the rostral and caudal cysts, if present. The opening of the spinal cord must allow exposure of the poles of the lesion and evaluation of the cyst walls but should not extend further than this. Pial traction suturing improves the surgical exposure and reduces the severity of repeated trauma due to dissection. This can be accomplished using a No. 6-0 suture without tension to hold the median pia mater and dura mater close together.

The first surgical maneuver consists of exposing a sufficient portion of the tumor to obtain a biopsy sample with forceps and scissors, without coagulation. This is immediately followed by histological examination. Any information suggesting an infiltrating or malignant tumor, or both, can be crucial in deciding whether tumor removal should be continued. Removal of tumor begins by reducing the volume of the tumor using an ultrasonic aspirator. The tumor must be debulked before looking for a cleavage plane. Intratumoral resection is performed from inside to outside. After strict control of hemostasis, dissection can be started laterally, on the side on which resection proves easiest. If the tumor is not too friable, or if there is a capsule, it can be grasped, allowing visualization of the correct cleavage plane. However, common sense and patience are necessary. If there is any difficulty, we prefer to move the microscope to another area and to come back later; for example, to leave one pole and go to the other one. The same policy is adopted when somatosensory evoked potentials start to alter. In our experience, we have always found a clear cleavage plane. The purple-blue or brown color is very helpful in recognizing the ependymoma and distinguishing it from the normal white spinal cord tissue. Most ependymomas have their vascular pedicles arising from the anterior spinal artery. Some large tumors may separate both sides of the spinal cord, resulting in a true diastematomyelia with a high risk of injuring the anterior spinal artery, which can lead to catastrophic operative results if caution is not exercised. Whereas several authors[3,8,19] routinely use ultrasonography for the detection of residual tumor, we do not because we have observed that bleeding spontaneously stops when tumor removal is macroscopically complete. However, in case of doubt, ultrasonography may be useful. On the other hand, in the absence of a polar cyst, it is not always easy to distinguish between the filiform end of the tumor and the increasingly dense fibrous bend into which it merges. The fibrous gliotic bend should be cut where it enters the center of the cord, but we recommend that the last portion of resected tissue undergo histological examination.

At the end of the procedure, it is necessary to inspect the wall of the cyst or cysts adjacent to the tumor bed. When normal spinal cord tissue can be seen through a transparent cyst wall, surgery can be terminated, because the cyst wall adjacent to ependymoma does not contain tumor. After tumor removal, the dorsal columns are released from pial traction and carefully brought together again. We like to approximate the cord with No. 6-0 pial sutures whenever possible. The arachnoid may also be partially

reconstituted if it was preserved on opening. Finally, the dura is always closed in a watertight fashion.

No patient who underwent surgery for low-grade ependymoma in whom a complete or subtotal removal was achieved received postoperative radiation therapy.

## RESULTS

The operative mortality rate was 3.2% (three patients). The first patient died from pulmonary embolism 10 days after operation for a malignant cervicomedullary extension of a thoracic ependymoma that had initially been treated surgically 3 months earlier; the second patient died from respiratory distress 22 days after surgery for a cervicomedullary Grade I ependymoma; and the third patient, also Grade I, died on the 20th day postsurgery from severe septicemia.

Four additional deaths occurred later: one patient died from a malignant recurrence at 12 months postsurgery, one patient died from complications related to his neurological condition 4 years after surgery; another from intercurrent disease 5 years after surgery; and a fourth patient died from ependymoma recurrence 20 years after incomplete removal at surgery.

All the other patients are alive, but one male patient will probably die during the next several months, having multiple subpial metastasis of a Grade III ependymoma. This man underwent subtotal tumor removal (confirmed by MR imaging) of an exophytic thoracic Grade II ependymoma with unexpected recurrence 6 months later, a second subtotal resection was performed, confirming the same histological type, and a new recurrence 13 months later was determined to be Grade III at the time of the third surgery. Fifty-one patients were followed for more than 5 years (Table 2).

Follow-Up Duration	No. of Patients
5-10 years	10
11-15 years	21
16-20 years	9
21-25 years	3
>26 years	8

Except the two patients related above who died 5 and 20 years after surgery, all the others remain alive. Only three tumor recurrences were observed among these 51 patients followed for 5 years or more: one died 20 years after surgery (incomplete removal) in spite of adjunctive radiotherapy (60 Gy) received at another institution; another who underwent operation for a recurrence 18 years postsurgery with a good result; and a third patient who underwent reoperation 7 years postsurgery. Among the patients who underwent complete surgical removal of their tumors, only the last two described above required a second operation for in situ recurrence, 7 and 18 years later, respectively. In treating ependymomas, we favor complete removal, which was possible in 86 (92.5 %) of our 93 cases. We do not treat ependymomas, except malignant ones, with adjunct radiotherapy and we have observed only one recurrence 18 years after complete removal. These results have led us to banish postoperative radiotherapy in low-grade ependymomas and to try to remove the tumor totally, which has always been our aim in one-stage or in two-stage operations in particularly difficult or holocord tumors. Therefore, for us, radiotherapy is not indicated in low-grade spinal cord ependymomas.

We observed one postoperative complication only: a cerebrospinal fluid fistula with no effect on the mortality and morbidity rates.

### ***Surgical Results***

We achieved complete tumor removal in 86 of 93 cases; in the seven patients (7.5%) whose tumors were subtotally resected, complete tumor removal was impossible because we could not find a clear cleavage plane or because the presence of a previous hemorrhage several weeks or months before made dissection more difficult. In nine patients, complete tumor removal was achieved after a second operation. Special attention should be drawn to holocord ependymomas. We encountered seven such cases, two of which were exophytic. All of these tumors were totally removed, either in one (four cases) or in two operative procedures (three cases).

### ***Functional Results***

Our follow-up time has ranged from 2 months to 33 years. The evaluation of functional results must take into account both motor and sensory function, as proposed by McCormick, et al.[18] When patients wake up in the recovery room, they invariably experience discomfort with diffuse hyperesthesia and paresthesias that can last several days. Due to the separation of posterior columns, they have deep sensory deficits, which can disturb early postoperative reeducation, but in most cases recovery is observed within 6 weeks to 3 months. Nine patients slightly improved (by a single McCormick grade) 1 year after surgery. Fourteen were slightly worse at 1 year, but only one deteriorated from Grade 1 to 3. All other patients were in the same condition at 12 months that they were before surgery.

## **DISCUSSION**

On MR imaging, an ependymoma may look like an astrocytoma and vice versa. Although homogeneous contrast medium uptake indicates an ependymoma,[5,16] it is not absolutely conclusive. After gadolinium injection, the signal of astrocytoma may also be enhanced albeit mostly in a heterogeneous manner. Indeed, by opposition to ependymomas, astrocytomas appear either heterogeneous with a hypo- or isointense signal, or homogeneous with a hypo- or isointense signal. When the tumor is eccentric, this may suggest an astrocytoma. However, in our experience[10] this has been observed in only 57% of astrocytomas but in one-third of ependymomas. The only typical MR imaging sign in favor of an ependymoma is the "cap" sign when it is present at both poles of the tumor. It has been correlated histologically and intraoperatively with areas of chronic hemorrhage.[1] We have never observed a "cap" sign involving a tumor determined to be an astrocytoma. Regarding the limits of the tumor, we may say that after contrast medium injection, tumoral contours appeared poorly defined in 50% of astrocytomas and clearly defined in 72% of ependymomas. In 20% of ependymomas and in 24% of astrocytomas, we observed no contrast enhancement: this is the reason that, except in the four conditions just described, we think that diagnostic MR imaging may be misleading with potentially hazardous ramifications. Histological evaluation remains mandatory, and an intraspinal cord tumor should not be proclaimed inoperable without first attempting operation.[10] Furthermore, some rare lesions with specific treatment may be found, like sarcoidosis[13] as we observed in two patients. One should be reminded that primary spinal cord sarcoidosis may exist and that corticosteroid treatment produced dramatic improvement in both cases. In conus ependymomas, perioperative histological examination may let us know if a myxopapillary tumor is present. One should know that myxopapillary ependymoma is never encountered in the spinal cord. That tumor may be invaginated into the conus and can be incorrectly diagnosed as an

exophytic intramedullary tumor.[9]

The surgical strategy is determined by analyzing the MR image data in three planes, and it is essential to define the limits of the tumor's solid portion, as this will determine the extent of the laminectomy or laminotomy. It is also essential to localize the lesion within the spinal cord with regard to its lateralization and depth. Most ependymomas are centromedullar, but some may be exophytic.[2] Complete removal should be the goal and this can be accomplished in most cases, even in holocord tumors.[10]

With rare exceptions, patients do not recover from severe preoperative neurological deficits, and those patients without disabling deficits after surgery are those who exhibited few or no deficits prior to operation. That is the reason why, like Cooper and Epstein,[4] we favor surgery before the patient experiences neurological deterioration. The postoperative quality of life depends on the preoperative neurological status.[2,4] In our experience, no patient in a paraplegic state at the time of surgery recovered.

We disagree with the use of postoperative radiotherapy except when it is used to treat malignant ependymomas. Of the patients who underwent complete removal (86 cases), only one required a second operation for in situ recurrent tumor 18 years later. Two of our patients who underwent incomplete removal of Grade II ependymoma underwent radiotherapy elsewhere, at 60 and 40 Gy, respectively. One patient developed severe cervical kyphosis. The other continued to deteriorate in spite of radiotherapy; he underwent a second operation 3 months later but died within 10 months.

There is no convincing evidence in the literature that radiotherapy is of any benefit in preventing recurrence of intraspinal cord tumors. The work of Kopelson, et al.,[12] has often been cited in the literature as an additional argument in support of radiotherapy. Their experience is based on a series of 12 ependymomas and 11 astrocytomas of all grades, and they reported eight failures (four ependymomas and four astrocytomas): recurrence with death in five cases. Nevertheless, they recommend radiotherapy at doses ranging from 40 to 50 Gy even after complete tumor removal. On the other hand, several authors[6,7,14,15,17,20,21] do not recommend radiotherapy in patients undergoing surgery for benign intramedullary spinal cord tumors, even after subtotal or incomplete removal. For us, surgery is the gold standard in the treatment of intraspinal cord ependymoma.

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