69 Indications and Technique for Intradural Intramedullary Lesions

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69.1 Introduction

Intradural intramedullary neoplasms are extremely rare. Only 5–10% of all spinal and 2–4% of all CNS tumors are located intramedullary.

The most common entities are spinal cord gliomas-intramedullary ependymomas (WHO grade I-III) and astrocytomas (WHO grade I-IV) – with approximately 80–90% of all intramedullary tumors [1–3]. Their incidence is higher in childhood [4]. Men are affected more often. The majority are benign or low grade lesions, whereas the incidence of higher graded tumors is higher in children. Due to the benign natural history and slow growing patterns, especially of ependymomas, the clinical signs are usually mild and non-specific, which delay the correct diagnosis. The average duration of symptoms up to the first diagnosis is more than 2 years [5], but less than 1 year for astrocytomas [6].

Other less common entities are hemangioblastomas (5–10%), metastatic lesions (<5%), and cavernomas (5–10%) [1, 7, 8].

Prospective data and thus clear evidence for optimal treatment are missing.

69.2 Case Description

69.2.1 Case 1

A 28 year-old female patient presented with neck and diffuse arm pain, bilateral distal arm paresis and mild gait ataxia. The symptoms were quickly progressing over the last 2 weeks. The next day after the hospitalization the patient showed an acute worsening of her symptoms developing tetraparesis, pronounced gait ataxia and bladder dysfunction (ASIA C).

MRI showed an intramedullary contrast enhanced tumor at C5/6 with an extensive edema of the cervical spinal cord (Fig. 69.1).

Resection of the contrast enhanced tumor resection was urgently performed under neuromonitoring (motor and sensor evoked potentials) via a right-sided hemilaminectomy C5 and partly C4 and C6, durotomy and myelotomy at the dorsal root entry zone C6, as the tumor reached the cord surface at this point.

After the surgery, the patient showed a partial improvement of the motor and vegetative dysfunction. The postoperative MRI confirmed the gross total resection (Fig. 69.2). Histological examination revealed an astrocytoma WHO

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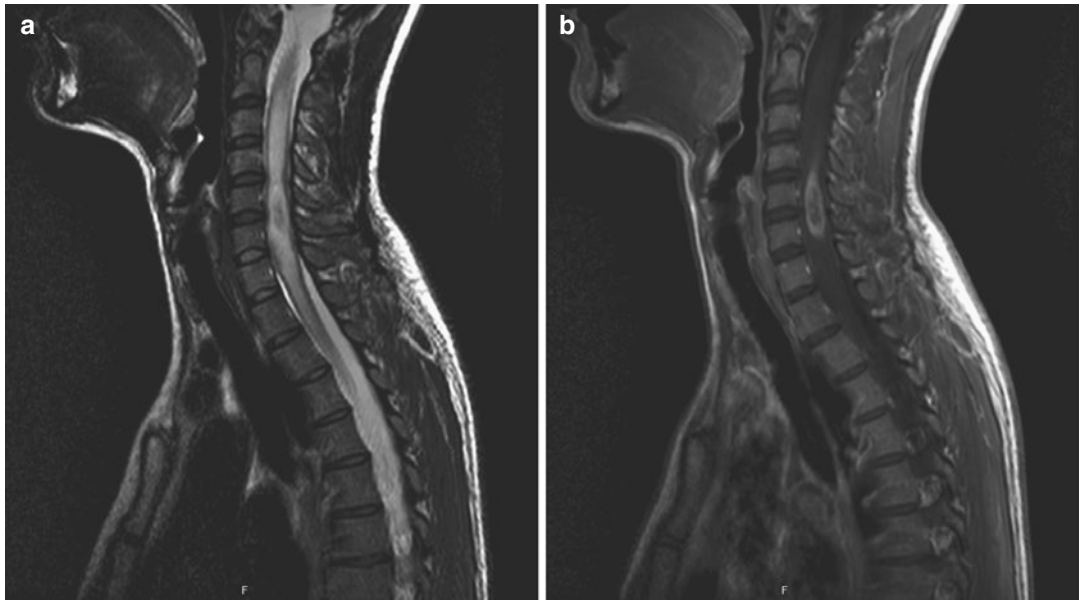

Fig. 69.1 Initial MRI scan. The MRI scan shows an intramedullary astrocytoma at C5/6. Sagittal T2 (**a**) and contrast T1 (**b**)

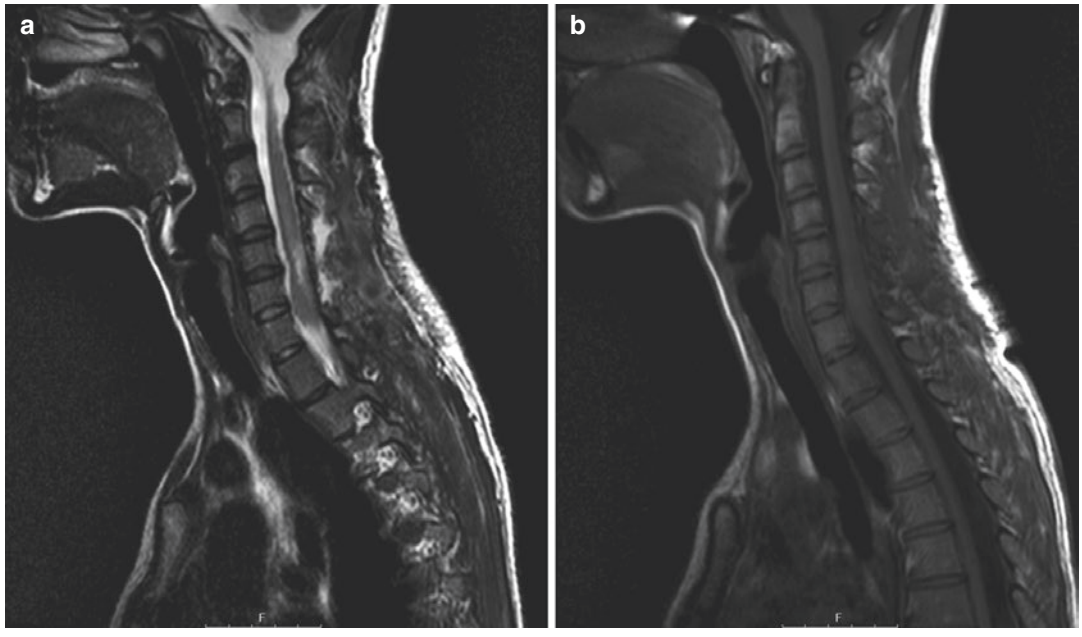

Fig. 69.2 Postoperative MRI scan I. The MRI scan shows postoperative sagittal T2 (**a**) and contrast T1 (**b**) rendering gros total tumor resection

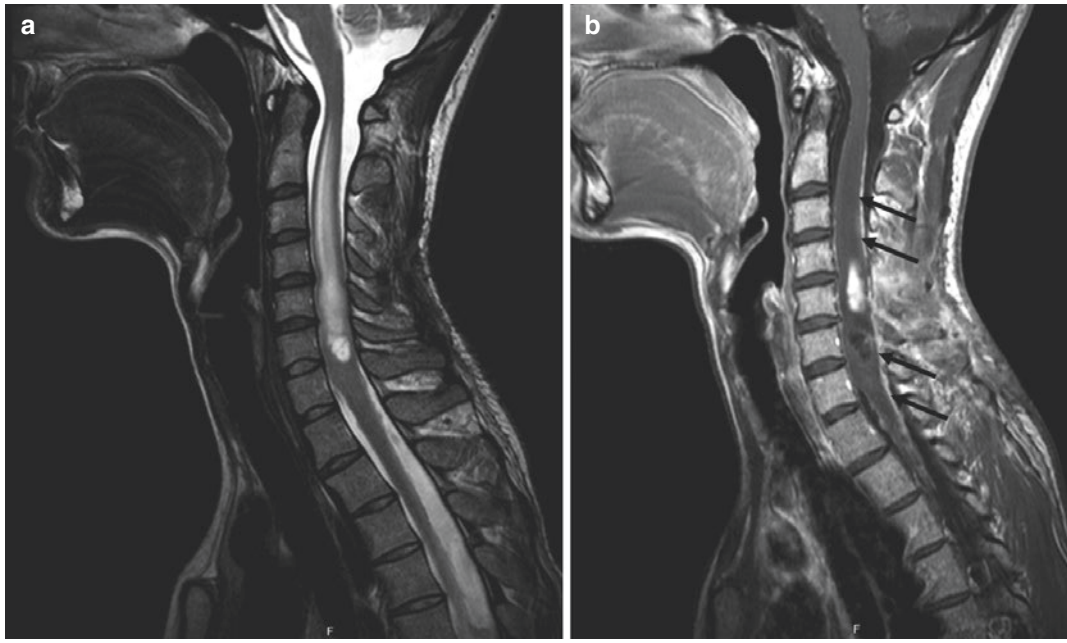

Fig. 69.3 Follow up MRI scan 6 weeks after surgery. The MRI scan shows postoperative sagittal T2 showing progressive edema of the cervical spinal cord (**a**) and contrast T1 (**b**) showing local tumor recurrence and diffuse pial enhancement along the cervical spine (arrows)

grade II. Holospinal MRI and CSF cytology were negative for tumor dissemination. The patient was assigned to receive adjuvant radiation therapy after the neurorehabilitation.

Six weeks later the patient was transferred back emergently to our department from the rehabilitation clinic because of secondary worsening of the right sided hemiparesis. The new MRI revealed a recurrent contrast enhanced lesion at the initial tumor site, additionally diffuse pial enhancement along the cervical spine (Fig. 69.3). The tumor was subtotally re-resected (Fig. 69.4), the histology was anaplastic astrocytoma WHO grade 3. Additional holospinal and cerebral MRI confirmed the suspected leptomeningeal tumor spread (Fig. 69.5).

The patient was referred to radiation oncology center for palliative radiation therapy. The patient died 5 months after the last surgery.

69.2.2 Case 2

A 44 year-old man presented with neck pain and a mild myelopathy involving slightly impaired fine motor skills, hypesthesia of the right hand and foot, and gait ataxia. Initially misdiagnosed as suffering from polyneuropathy, the patient was treated by his neurologist with Vitamin B12 without any success. The symptoms were slowly progressive over the last 3 years. A finally performed MRI revealed a large intramedullary tumor of the craniocervical junction (Fig. 69.6).

The tumor was gross totally resected via laminectomy C1–3, durotomy and median myelotomy under neuromonitoring with motor and sensory evoked potentials.

Immediately after the surgery the patient was transferred for 1 week to the intensive care unit due to the transiently impaired tetraparesis, difficulties with swallowing and ventilation. Over the

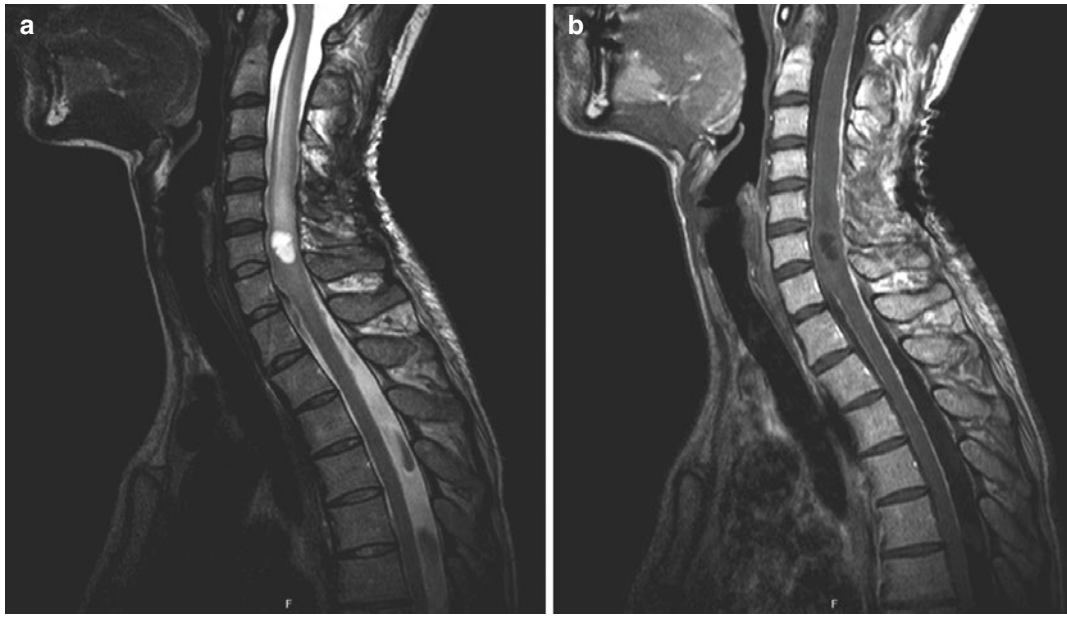

Fig. 69.4 Postoperative MRI scan II. The MRI scan shows sagittal T2 (**a**) and contrast T1 (**b**) after the subtotal resection of the recurrent astrocytoma

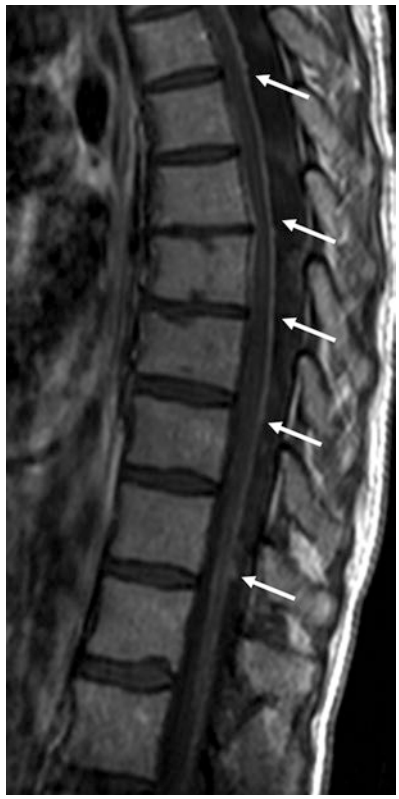

Fig. 69.5 Holospinal MRI. The holospinal contrast T1 MRI scan (here shown: thoracic spine) demonstrates diffuse contrast enhancement along the whole spinal axis corresponding to leptomeningeal tumor spread (arrows)

next 2 weeks the new deficits were fully recurrent and at discharge his clinical status was unchanged to that before surgery. Over the next 3 months the symptoms improved. The patient walks without assistance. He is back to his full-time job as sales manager with a slight residual gait ataxia and hypesthesia of his right hand.

The histological examination revealed a WHO grade II ependymoma. The postoperative holospinal MRI showed no residual tumor (Fig. 69.7). The CSF cytology and cranial MRI were negative for tumor dissemination. According to the tumor board decision the patient received no adjuvant radiotherapy. The follow up examinations proceeded every year. The patient is progression-free for almost 10 years now after the surgery (Fig. 69.8).

69.3 Discussion of the Cases

69.3.1 Indication

Due to the rare occurrence of intramedullary gliomas and a predominantly benign behavioral pattern of the majority of them, there are no randomized data available regarding the optimal therapy. The largest retrospective

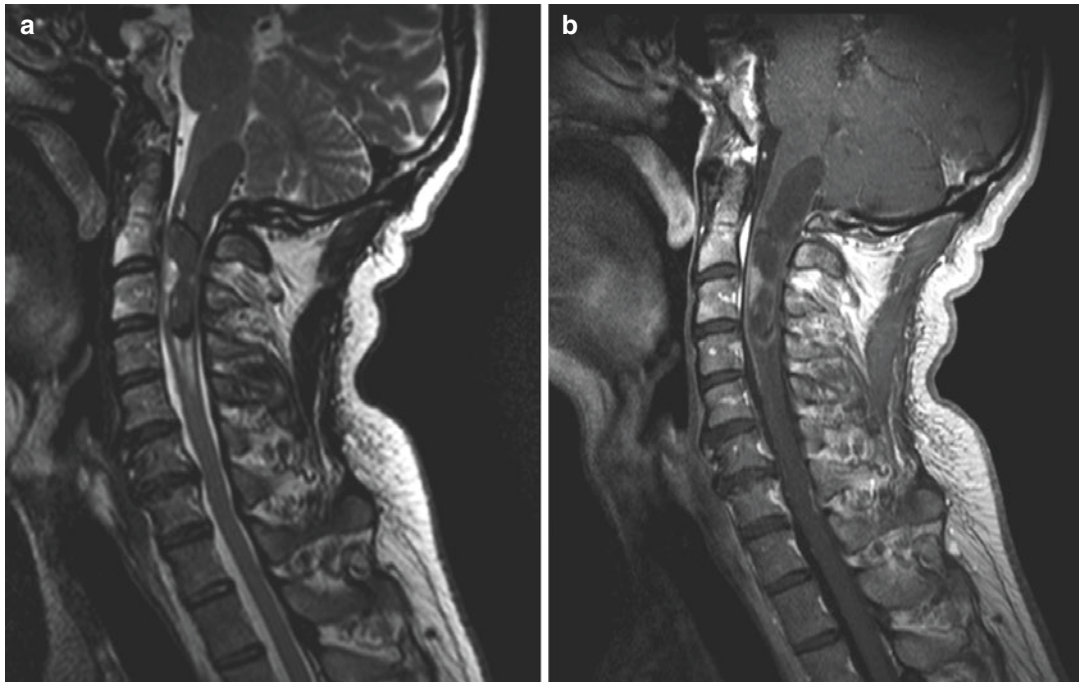

Fig. 69.6 Initial MRI scan: Preoperative MRI showing a contrast enhanced spinal cord tumor between the medulla oblongata and the C3 level with an associated syrinx formation (Sagittal T2 in **a**, sagittal contrast T1 in **b**)

series included maximum 100–150 cases [6, 9–12]. Therefore, there are no clear guidelines for the indication of the specific therapeutic modality.

According to the results of larger clinical series and expert opinions, a gross tumor resection represents the gold standard in the treatment of spinal ependymomas and [9, 10] pilocytic astrocytomas [13].

Higher-grade infiltrative astrocytomas cannot be treated by surgical treatment alone. As demonstrated by our first case, even the complete resection of the contrast enhanced tumor mass appears to be not sufficiently effective due to the tumor infiltration of the surrounding spinal cord tissue and a high tendency of these tumors to recur. Some authors find that surgical resection is associated with a poorer neurological outcome and poorer overall survival [6, 11, 14]. Only few series show an advantage of radical tumor removal in terms of oncological prognosis, even in malignant astrocytomas [15]. In any case, operative debulking to reduce the space-occupying effect and to obtain histological samples plays an important role in the treatment of infiltrative spinal astrocytomas.

Regarding the optimal timing of surgery, the majority is convinced that early resection should be attempted when symptoms are mild, because of a then clearly better prognosis for a neurological recovery and a lower risk for new postoperative deterioration [1, 16, 17]. Opinions vary as to the timing of the operation of inicidental findings, but the majority tends to follow up these patients first at close intervals and to proceed with surgery in cases of tumor progression and/or a development of a neurological deficit.

Spinal gliomas occasionally show drop metatases or disseminated manfestation at the first diagnosis. In such cases, the operation should focus primarily on the resection of the main tumor, since no additional benefits appear from the resection of the metastatic lesions [18]. Due to the ability of spinal cord glioma cells to spread along the neural axis, the perioperative diagnostics should include holospinal and cerebral MRI as well as CSF cytology.

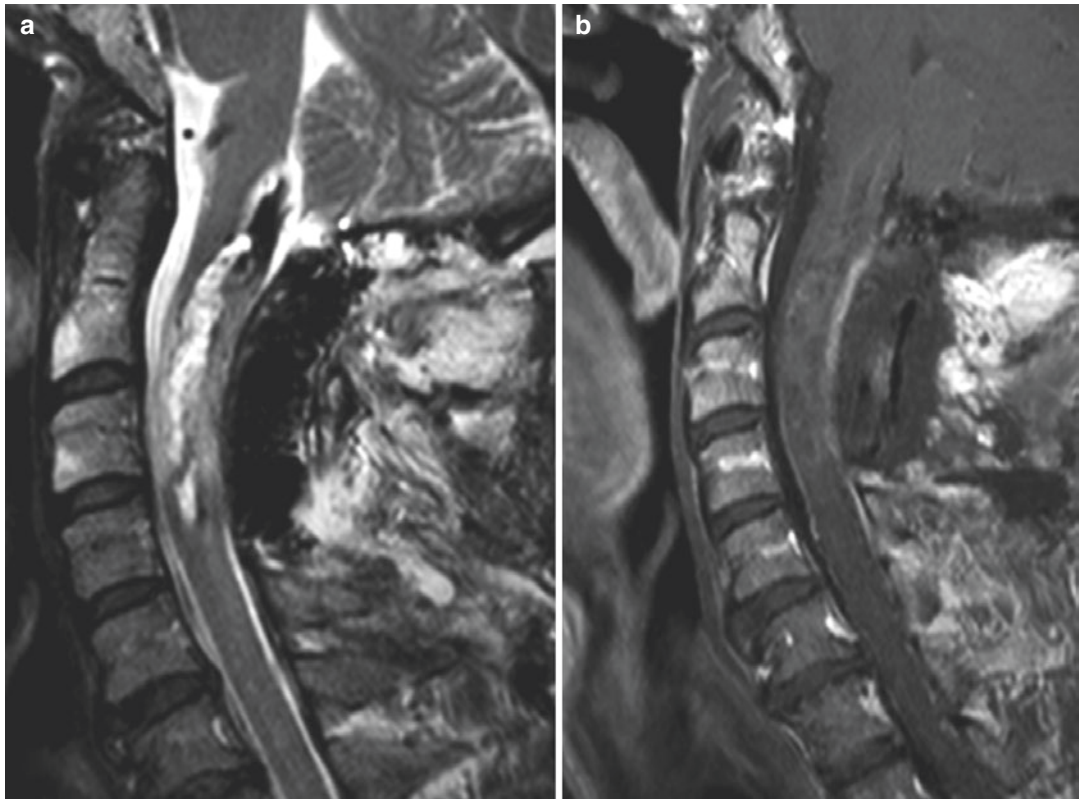

Fig. 69.7 Postoperative MRI scan. The MRI scan shows postoperative sagittal T2 (**a**) and contrast T1 (**b**) rendering gros total resection of the ependymoma

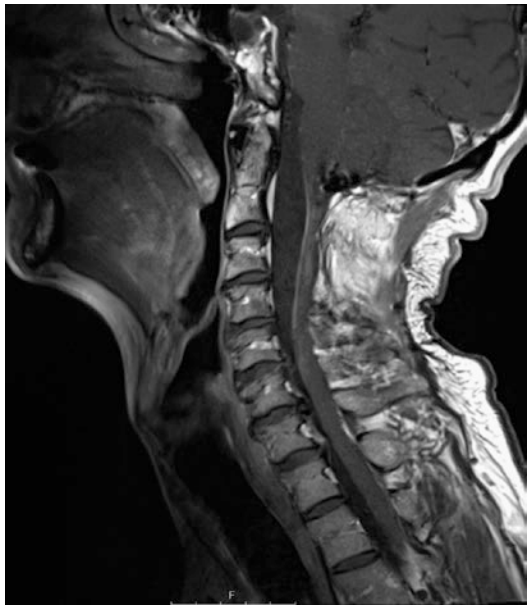

Fig. 69.8 Follow up MRI 9 years after surgery. The contrast T1 MRI 9 years after surgery shows no signs of a local tumor recurrence

69.3.2 Surgical Aspects

For the resection of most intramedullary tumors, a mono- or multi-segment laminotomy/laminectomy is suitable. For circumscribed pathologies with a side-emphasis, hemilaminectomy is often sufficient. The risk of secondary postoperative instability of the spine after removal of intradural tumors varies between 10% in adults and up to more than 50% in pediatric series [19–21]. Therefore, laminoplasty is often thought to prevent secondary deformity, especially in childhood. However, no statistically clear evidence for benefits of this approach to secondary stability exists [22].

In cases with multi-segmental laminectomies for large intramedullary tumors additional stabilization with internal fixation may be considered. However, difficulties would occur in assessment of the follow up MRIs.

For the resection of spinal gliomas, the median myelotomy is usually chosen. After opening the dura in the midline, the edges are held apart with sutures (Fig. 69.9a). After the opening of the pia, the myelon is opened between the posterior branches. The degree of resection is determined by the demarcation of the tumor against the surrounding spinal cord tissue, the histological findings and any changes to the IONM. In infiltrative higher graded astrocytomas a complete resection can rarely be achieved. Rather, it is a debulking operation for decompression, relief of syrinx and recovery of histological samples. In contrast, in benign processes a gros total resection should be attemptes, provided there is no permanent neurological damage. Since almost 40% of postoperative deficits are due to surgical manipulation before or after tumor resection [23], ultrasonic aspirator debulking of the central parts of the tumor is performed as the first step to prevent further spinal cord injury due to the traction of larger tumor masses (Fig. 69.9b). Subsequently, tumor-supplying vessels are coagulated and severed; the capsule and remaining tumor tissue are removed in toto if possible. After the adaptation of the Pia a watertight dural closure takes place.

Surgery of intramedullary pathology without the use of IONM is obsolete. With the introduction of the IONM, the extent of resection of the tumors was significantly increased, while the rates of postoperative new deficits were reduced by continuous monitoring of the motor and sensory pathways [24]. The standard IONM includes cortical derivation of SEP after peripheral stimulation, and MEP monitoring after transcranial electrical stimulation. In recent years, more and more attention has been drawn to the benefits of direct epidural MEP derivation in terms of the D wave as the strongest predictor of the occurrence of postoperative neurological deterioration [25]. When resecting the ependymomas in the area of the conus medullaris or cauda equina, an intraoperative electromyography for monitoring the sphincter function and individual nerve roots may be used.

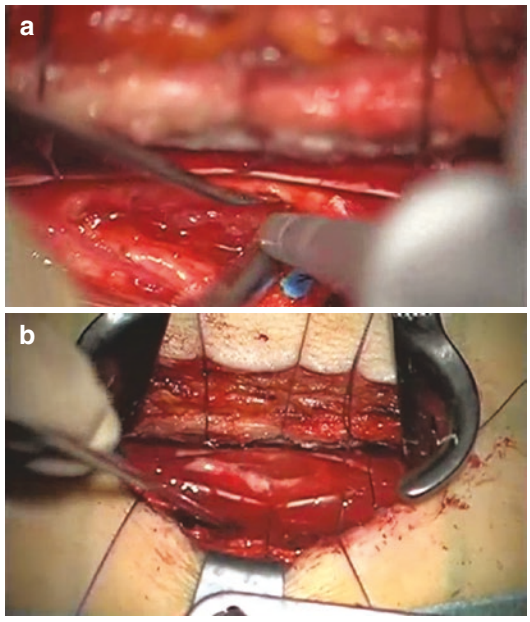

Fig. 69.9 Intraoperative photographs of ependymoma resection. Midline approach with the dura being held apart by tenting sutures (**a**); tumor mass reduction by an ultrasonic aspirator to prevent additional traction during the further resection (**b**)

69.3.3 Outcome and the Role of the Adjuvant Treatment

Diffuse and malignant astrocytomas of the spinal cord grow infiltratively, which limits the resectability and surgical safety of postoperative deficits: while gros total resection can be achieved in maximum 15% of cases, new permanent deficits are expected in up to 50% [6]. In contrast, pilocytic astrocytomas and ependymomas are well circumscribed, and thus, are well operable tumors with a rate of complete removal of 70–90% and a likelihood of severe residual deficiency of <10% [10]. The oncological prognosis of ependymomas is generally favorable: the median progression-free survival after a gros total resection is about 7 years on average (6 years for grade I, 15 years for grade II and 4 years for grade III ependymomas) [12, 26, 27]. In terms of overall survival, ependymomas and pilocytic astrocytomas also have a good prognosis with a 10-year survival of about 80% [11, 28]. Higher-grade astrocytomas have a significantly worse prognosis with a median survival of 17 months in

anaplastic astrocytomas and 9–10 months in patients with spinal glioblastomas [6, 15].

Adjuvant radiotherapy is not recommended following gross total resection of grade I-II ependymomas and spinal pilocytic astrocytomas [10, 11, 17]. In cases of partial tumor resection, recurrence, disseminated and anaplastic ependymomas, as well as in cases of astrocytomas grade II-IV, a fractionated radiotherapy is recommended [11, 29], although there is no clear evidence for that either, and decisions for the radiotherapy are usually made on a case-by-case basis.

69.3.4 Accordance with the Literature Guidelines

As discussed above, guidelines cannot be derived from the literature. However, the indication for treatment as well as the surgical approach were most probably not in accordance with the current common consensus of the majority of peers. Yet the same accounts for the authors' preferred method.

Level of Evidence: C
The level of evidence available to date is low. Only several large restrospective series of more than 50 cases (cited above) are available on surgical and adjuvant treatment of intramedullary tumors.

69.4 Conclusions and Take Home Message

Primary spinal cord tumors are extremely rare with about 3% of all primary CNS tumors. Prospective data and thus clear evidence for optimal treatment are missing. The most common entites are intramedullary ependymomas (> 60% in adults) and astrocytomas (15–20% in adults, >50% in children). The majority of intramedullary tumors are benign or low grade (WHO grades I-II). Diffuse and malignant astrocytomas of the spinal cord grow infiltratively, which limits the resectability and surgical safety of postoperative deficits: at a complete resection rate of maximum 15%, new permanent deficits are expected in up to 50%. In contrast, pilocytic astrocytomas and ependymomas are well circumscribed, and thus, are well operable tumors at a rate of complete removal of 70–90% and a likelihood of a severe persistent deficiency of <10%. Therefore, a complete resection represents the gold standard in the therapy of the latter entities. In terms of overall survival, ependymomas and pilocytic astrocytomas have a good prognosis with a 10-year survival rate of more than 80% after a gros total resection. Higher-graded astrocytomas have a significantly worse median survival with 17 months in anaplastic astrocytomas and 9–10 months in patients with spinal glioblastomas. Adjuvant radiotherapy is not recommended following gross total resection resection of grade I-II ependymomas and spinal pilocytic astrocytomas. In cases of recurrence, disseminated and anaplastic ependymomas, as well as after any surgical treatment of spinal astrocytomas grade II-IV, a fractionated radiotherapy should be performed.

Pearls
- Ependymomas and pilocytic astrocytoma: go for a gross total resection
- Infiltrative or malignant astrocytoma: go for a biopsy
- Intraoperative IONM is mandatory resp. highly recommended
- Radiation therapy for grade II-IV astrocytomas and grade III ependymomas

Editorial Comment
Intramedullary tumors are an orphan disease and should therefore be treated in specialized centers only according to us. It is a benign disease in unsually younger patients with a high probability of "cure" and a potential for devastating operative complications.

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