Intramedullary hemangioblastomas: timing of surgery, microsurgical technique and follow-up in 23 patients

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Abstract Hemangioblastomas are highly vascularised tumors of the central nervous system and account for 1.5–2.5% of all spinal cord tumors. Because of the rarity of these tumors, surgical experience is often limited and, therefore, treatment and indications for timing of surgery are discussed controversial. The authors reviewed their data of 23 consecutive patients with respect to timing of surgery, microsurgical technique, and follow-up. Clinical records of 23 consecutive patients with intramedullary hemangioblastomas who underwent first surgery in our department between 1990 and 2005 were reviewed. In three cases the tumors were localised at the craniocervical junction; four patients had a single tumor in the cervical spine, six patients multiple tumors in the cervical and thoracic spine, eight patients in the thoracic spine only, one patient in the conus region, and one patient had multiple tumors located in the thoracic and lumbar spine. In eight patients, a von-Hippel-Lindau disease (VHL) was associated. The neurological follow-up was evaluated according to the classification of McCormick. Operation was recommended to every symptomatic patient as early as possible. In asymptomatic

patients with a sporadic tumor surgery was discussed for diagnostic purposes at any time. In VHL patients, surgery was recommended if tumor growth was observed on MRI in the next practicable time. All tumors were diagnosed by magnetic resonance imaging and in all cases but one a DSA was performed. All patients were treated microsurgically through a posterior approach. The tumors in the spinal cord were removed microsurgically through a partial hemilaminectomy ($n = 1$), a hemilaminectomy ($n = 15$), or laminectomy ($n = 4$) and at the craniocervical junction ($n = 3$) through a suboccipital craniotomy. During follow-up after 6 months, 18 patients remained neurologically stable (17 in McCormick grade I and 1 in McCormick grade II) and 5 patients recovered to a better status (3 from grade III to II, 2 from grade II to I). There was one complication with a CSF fistula and one recurrence/incomplete removal. Following the above-mentioned principles of microsurgical removal of intramedullary hemangioblastomas, operation is possible with a low procedure-related morbidity and can be recommended especially in VHL patients with progressive symptoms or tumor growth during follow-up. Patients without VHL most frequently require hemangioblastoma resection for diagnostic purposes and/or because symptoms prompted an imaging work-up that lead to the discovery of the tumor.

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Introduction

Hemangioblastomas are highly vascularised tumors of the central nervous system and account for 1.5–2.5% of all spinal cord tumors [2]. They occur in 20–30% of

VHL disease and are often multiple [5]. More than 50% of spinal hemangioblastomas are accompanied by a syrinx [6].

Magnetic resonance imaging (MRI) is the diagnostic tool of choice and is helpful in preoperative planning and the differential diagnosis of spinal cord neoplasms and vascular lesions [11]. Hemangioblastomas are most common isointense on unenhanced T1-weighted images, hyperintense on T2-weighted images, and show a homogeneous intense enhancement. Larger hemangioblastomas tend to be hypointense or mixed hypo-isointense on T1, heterogeneous on T2, and show an intense heterogeneous enhancement [1]. In addition, cystic components of the tumor can be associated with a nonenhancing rim of the cyst. In large hemangioblastomas, flow voids may be present along the perimedullary circumference.

Because of the rarity of these tumors, surgical experience is often limited and, therefore, indications for treatment and timing of surgery are still unclear. Most of the recently published series recommend microsurgical removal in symptomatic patients or in cases with tumor growth during follow-up [3, 7, 10]. The authors reviewed their data of 23 consecutive patients with respect to timing of surgery, microsurgical technique, and follow-up.

Patients and methods

Between 1990 and 2005, 23 patients underwent first microsurgical treatment for an intramedullary hemangioblastoma in our department. The clinical records and operative reports of these patients were analyzed retrospectively. In eight patients (35%) a von-Hippel-Lindau disease and in seven patients (30%) a syrinx was associated. In eight (35%) patients the tumors were multiple ($n > 2$)—all of them were associated with VHL. Interestingly, two of these patients were identical twins and both were symptomatic because of a hemangioblastoma at the same level (TH 11).

In three (13%) cases the tumors were localised at the craniocervical junction; three (13%) patients had a single tumor in the cervical spine, seven (30%) patients multiple tumors in the cervical and thoracic spine, eight (35%) patients in the thoracic spine only, one (4%) patient in the conus region, and one (4%) patient had multiple tumors located in the thoracic and lumbar spine. There were 11 men and 12 women with a mean age of 44 years (range 22–71 years).

Only patients with new onset of neurological deficits or with tumor progress in the follow-up of conservative treatment were treated by surgery.

All patients were routinely examined 6 months after the operation both clinically and by MRI to detect tumor

recurrency, or in case of multiple localization, additionally tumor progress. The neurological follow-up was evaluated according to the classification of McCormick [4] (Table 1). Mean follow-up was 27 months (range 6–120 months).

Patients with VHL disease were routinely screened for other manifestations of the disease by a set of additional investigations like CT of thorax and abdomen and ophthalmologic consultation and blood tests.

Treatment strategy, microsurgical technique

In the cases with a single tumor operation was indicated if a new onset of neurological deficit or tumor progress was encountered. In the patients with multiple tumors surgery was performed either in patients with neurological deficits related to one tumor/level or with tumor progression during follow-up. Duration of symptoms ranged between several weeks and years. Operation was recommended to every symptomatic patient as soon as possible. In asymptomatic patients with a sporadic tumor surgery was discussed for diagnostic purposes at any time. In VHL patients, surgery was recommended if tumor growth was observed on MRI in the next practicable time.

All patients were treated microsurgically through a posterior approach. Sitting position was favored if the tumor was localized at the craniocervical junction or cervical, prone position if the tumor was localized at the thoracic or lumbar spine. The spinal cord tumors were removed through a partial hemilaminectomy ($n = 1$), a hemilaminectomy ($n = 13$) or laminectomy ($n = 4$), and at the craniocervical junction through a suboccipital craniotomy with widening of the foramen magnum. Midline dural incision was made over the dorsal surface of the spinal cord followed by lateral dural sutures. The strategy of microsurgical removal of the tumor was to coagulate the dominant arterial feeders at the entrance of the tumor—identified by DSA—by bipolar coagulation with low power first, then coagulate and shrink the tumor and to coagulate the venous drainage last. Finally the tumor was excised carefully en bloc (Fig. 1).

Table 1 Clinical presentation prior surgery in correlation to patients' outcome according to the McCormick classification

	Grade	Follow-up		
		I ($n = 19$)	II ($n = 4$)	III ($n = 0$)
Prior surgery	I ($n = 17$)	17	0	0
	II ($n = 3$)	2	1	0
	III ($n = 3$)	0	3	0

Fig. 1 Intraoperative view of an intramedullary hemangioblastoma in the cervical spinal cord before (a) and after (b) complete resection

Results

Clinical data

Pre-operatively 17 patients (74%) showed only mild focal deficits not significantly affecting the function of the limbs and were classified in McCormick grade I. Three (13%) patients were classified in grade II, and three patients (13%) in grade III.

All tumors were primarily diagnosed by MRI. Imaging protocol consisted of T1-weighted sagittal and axial views of the spinal cord with and without gadolinium and a T2-weighted scan. In all but one patient a digital subtraction angiography (DSA) was also performed (Fig. 2). In the case without DSA the lesion on the pre-operative MRI was misinterpreted as a meningioma and, therefore, a DSA was not performed (Fig. 3).

Post-operatively, all patients in McCormick grade I ($n = 17$) remained stable. One patient with grade II

Fig. 2 On T2-weighted (a) and enhanced T1-weighted images pathological vessels could be identified (b). Only by DSA the exact feeding vessels could be identified and the diagnosis of a hemangioblastoma underlined (c)

Fig. 3 In this patient, MRI was more typical for a meningioma; therefore, a DSA was not performed. As intraoperatively the tumor was presenting like a hemangioblastoma, the surgeon tried to follow the principles of microsurgical technique for removal of hemangioblastomas and the tumor could be removed without massive bleeding

recovered to a better status and one showed a temporary worsening of his symptoms but recovered to a better grade (I) during the 6 months' follow-up. One grade II patient remained also stable, but this patient had multiple spinal hemangioblastomas. He showed a worsening of his symptoms due to progress of the remaining tumors 2 years later and was in grade IV before the second operation. All patients with grade III ($n = 3$) recovered to grade II.

Complications

In one patient a CSF fistula occurred. This was treated successfully by a lumbar drainage. In one patient follow-up

MRI demonstrated incomplete removal. As the patient was free of any symptoms he was followed-up during the next years. Six years after the first operation, the tumor showed a progress on MRI and a second operation was performed.

In one patient the tumor could not be identified during the first operation. Postoperatively, a MRI was performed showing that the tumor was localized more inferior with respect to the approach. The patient underwent a second operation and the tumor could be removed finally. No tumor recurrence was encountered so far.

Discussion

Intramedullary hemangioblastomas are rare lesions and there are not many reports about the microsurgical removal in larger series [3, 10, 12]. They have been considered to pose severe problems during surgery because of the risk of massive bleeding [3]. In our series we did not come across this problem. We think this might be due to the surgical principle to coagulate the dominant and then the other feeding arteries first followed by shrinking the tumor and finally coagulating the venous drainage. Therefore, this surgical strategy is in our opinion the key to successful and safe removal of these highly vascularised intramedullary tumors.

Most of the authors recommend a DSA prior to the operation and emphasised the need for a precise angiographic study to delineate the feeding arteries and draining veins associated with the hemangioblastomas [3]. We agree with these authors, although nowadays MRI has become the diagnostic method of choice and is recommended by several authors in the diagnosis of hemangioblastomas [1, 8–10]. However in the diagnosis of the feeding and draining vessels of intramedullary tumors, MRI technique is still insufficient and, therefore, in our opinion DSA is mandatory for a successful and safe microsurgical treatment.

In our series MRI with T1- and T2-weighted images was performed in all patients for the preoperative planning, screening and follow-up of the patients. DSA was performed in all but one patient. In this case the MRI was misinterpreted as a meningioma and DSA was thought to be unnecessary.

The entity of a tumor-associated cyst or syrinx has been reported to be present between 50 and 80% [6, 7, 10]. Only one author reported in his series a smaller proportion with 16% [3]. In our series a syrinx in 7 of the 23 patients (30%) was present. No tumor cyst was associated like known in cerebellar hemangioblastomas.

To the best of our knowledge this is the first report about identical twins with VHL who had been symptomatic for a hemangioblastoma at the same level (TH

11). After the first one became symptomatic for this hemangioblastoma, MRI revealed multiple spinal and intracranial lesions. Then the second twin was screened for lesions and MRI revealed a single spinal lesion at the same level and multiple intracranial lesions. Finally, VHL was diagnosed in both. Several months later the second twin became symptomatic for the spinal hemangioblastoma and underwent surgery.

Conclusions

Using the aforementioned principles of microsurgical removal of spinal hemangioblastomas, microsurgical removal is feasible with a low procedure-related morbidity and can be recommended, especially in VHL patients who were symptomatic or with tumor growth during follow-up. Patients without VHL most frequently require hemangioblastoma resection for diagnostic purposes and/or because symptoms prompted an imaging work-up that lead to the discovery of the tumor. Thus, in patients with sporadic intramedullary hemangioblastomas, radiographic progression is rarely used as an indication for surgery.

Occlusion of the feeding arteries, shrinking of the tumor by bipolar coagulation with low power and its en bloc removal are the key points for a safe microsurgical treatment. MRI is the method of choice in the diagnosis of these tumors, but our experiences underline that DSA is mandatory for the preoperative planning.

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