CLINICAL EVOLUTION OF PRIMARY INTRAMEDULLARY TUMORS IN ADULTS

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ABSTRACT

Objectives. The objectives of our clinical study are the evaluation of preoperatory myellic involvement and postoperative follow up, establishing in the mean time the interval and capacity of recovery for adults with primary intramedullary tumors.

Material and method. Between January 2001 and December 2008, 14 adult patients diagnosed with intramedullary tumors representing 20,6% of all primary spinal tumors treated in Targu Mures Neurosurgery Clinic, were studied. The majority of intramedullary tumors were represented by ependimomas (64.3%), followed in equal numbers by astrocytomas and hemangioblastomas (14.3% each) and a rare case of intramedullary epidermoid cyst. Each sign and symptom was monitorized with the help of a grading scale going from 5 (asymptomatic patient) to 0 (paraplegia, total absence of functional capacity). Aditionally, for monitoring the functional capacity outcome we used the Karnofsky score. The resulted quantitative data of our study was analysed using the T-student test.

Results. The major signs of debut were spinal back pain (50%) and motor deficit (35%). The interval between disease debut and diagnostic was variable scaling from one month to 15 years with an average of 28.8 months ± 45.4 months. The clinical status was towards progressive neurological deterioration. At the diagnostic moment all patients presented motor deficit, 12 of them (86%) presented walking disorders, 64% presented sensory deficit and only 12% presented sphincter disturbances. Postoperative evolution was caracterized by a transitory neurological deterioration in 57% of cases, the deterioration being recovered during the first 3 months in 75% of cases, in 25% of cases the neurological recovery to preoperatory status extended during a period of 6 months postoperatively. Beyond the interval of 3 months postoperatively, the neurological status was stationary. The main result of surgical intervention was maintenance of preoperative neurological status.

Conclusions. The clinical evolution of primary intramedullary tumors in adults was towards progressive neurological deterioration and severe myelic involvement. The surgical intervention maintains rather than improves the preoperative neurological status thus early diagnosis and treatment of this pathology represent the major favorable prognostic factors.

Key words: spinal cord, motor deficit, back pain, Karnofski score, myelic involvement

INTRODUCTION

Spinal cord tumors represent about 15% of tumors of the central nervous system (CNS). Most have as a starting point cells forming the spinal cord, philum terminale nerve roots or meninges. Metastatic damage to the intradural compartment is rare (1).

Intradural tumors of the spinal cord are classified by their relationship with the latter. A small number of tumors can have a mixed intra-and extramedullary component, which usually communicates through the dorsal root entry zone in the spinal cord or through the conum medullare – philum terminale transition zone. Some intradural tumors can spread along nerve sheath in the extradural space (2, 3).

In adults about 2/3 of spinal cord tumors are extramedullary. Nerve sheath tumors, meningiomas and ependymomas, the philum terminale tu-

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mors represent up 95% of extramedullary tumors in adults. Metastases, tumors inclusion cysts, paragangliomas and melanotic tumors are extremely rare (4).

With few exceptions, extramedullary tumors are histologically benign and complete surgical resection is suitable.

One third of spinal cord tumors in adults are intramedullary tumors. Over 80% are benign glial primary tumors (Table 1) (1).

TABLE 1. *Intramedullary tumors in adults (1)*

Intramedullary tumor in adults	
Ependymomas	45%
Astrocytomas	40%
Hemangioblastomas	5%
Other tumors: lipomas, cavernomas, sarcoidosis	10%

MATERIAL AND METHODS

This prospective study comprised 68 patients operated in the Neurosurgery Clinic University Emergency Hospital of Targu-Mures from January 2001 until December 2007. Intramedullary tumors represented 20.6% of all primary spinal tumors operated. (Table 2).

TABLE 2. *Pacients with intra/extra medullary tumors operated from january 2001 until december 2007*

Tumor type	**Number of patients**	**Number of tumors.**
Intramedullary	14	14
Extramedullary – Intradural – Intra-extradural – Extradural	 45 4 5	 51 4 5
Total	68	74

All patients were diagnosed with MRI and were operated on using microsurgical techniques. Treatment recommendations and description of surgical techniques used were based on the experience of the Neurosurgical Clinic in Targu Mures and reflect the current views of this group. The objectives of this study were:

1. Evaluation of preoperative myelic damage and postoperative follow-up of different signs and symptoms.

2. Setting range and capacity of recovery of patients with intramedullary tumors.

3. Follow-up of imaging features and of different histopathological entities in an attempt to anticipate the nature of the tumor and preoperative surgical strategy to follow.

4. Description of surgical techniques used to determine the consequences they have on spinal stability and clinical course of patients.

Patients were examined neurologically before and after surgery, and outpatient follow-up was based on imaging and clinical follow-up records at intervals of 3, 6, 12 months.

Each sign and pre-and postoperative neurological symptoms were documented and analyzed individually using a scoring system (Table 3) (5).

Additionally, the clinical evolution and functional independence level were followed by Karnofski score (6).

RESULTS

Clinical aspects

In our series of patients with intramedullary tumors, 7 patients (50%) had back pain as the first symptom, in the other 5 (35.7%) the disease started with motor deficits and one patient accused difficulty in walking and impaired sensitivity (Fig.1).

The time from onset to diagnosis was highly variable and ranged from 1 month to 15 years with an average of 28.8 ± 45.4 months.

Clinical course over this period was characterized by slowly progressive deterioration of the neurological function.

TABLE 3. *Patient's neurological status evaluation*

Score	**Sensory disturbances and pain**	**Motor deficit**	**Walking disorders**	**Sphincter disturbances**
5	No symptoms	Absence of motor deficit	Normal walking	Normal sphincter function
4	Present but mild	Movement against ressistance	Unstable walking without help	Minimal sphincter disfunction without vesical catheter
3	Singnificant without functional restrictions	Movement against gravity	Mobile, but with help	Residual bladder without catheter
2	Some functional restrictions	Movement with gravity eliminated	A few steps with help	Mild incontinence
1	Severe functional restrictions	Flicker or trace contraction	Mentains ortostation with help	Often vesical catheter
0	Functional incapacity	Plegia	Plegia	Permanent vesical catheter

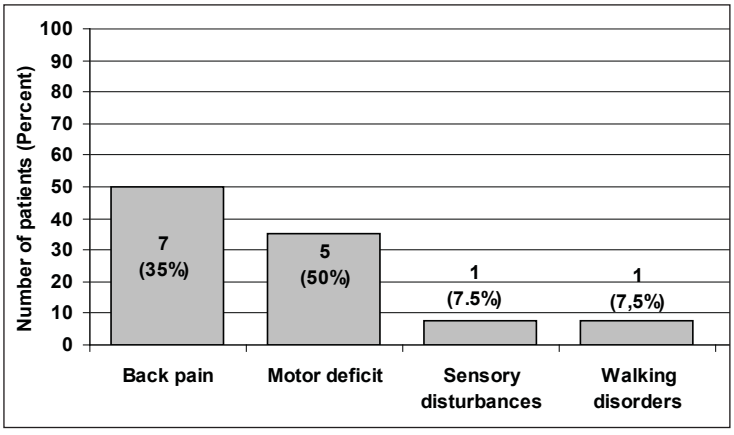

FIGURE 1. *Patients' symptoms at the moment of disease debut*

The mean age at diagnosis was 49.1 ± 12.3 years with an equal distribution between the sexes.

Of the tumors, 50% were located in the cervical segment (7 cases), 29% (4 patients) dorsal, and 21% (3 cases) in the medullary cone.

Upon admission, the motor function was affected in varying proportions in all patients. Thus, all patients had motor deficits, and 12 (86%) had difficulty walking. Karnofski average preoperative score was 52.1 ± 9.8. Pain persisted at diagnosis in 3 patients (21%). A total of 9 cases (64%) had abnormal sensitivity and only 2 patients (14%) complained of abnormal sphincter (Fig. 2).

Comparing the patients' onset complaints with complaints at the time of diagnosis there was a significant increase of sensory and motor function impairment, and a decrease in pain-related complaints. This shows the slowly progressive neurological deterioration for patients (Fig 3).

IMAGISTIC ASPECTS

The gold standard for intramedullary tumor diagnostic is represented by spine MRI (4). There are several criteria for positive diagnosis of intramedullary tumors:

- increasing the volume of MS (100% of cases);
- almost obligatory presence of contrast enhancement, with the exception of intramedullary epidermoid cyst;
- association between intramedullary tumors and syringomyelic cavities is common (42.8% of cases) (7).

In adults, pathognomonic for preoperative diagnosis of intramedullary ependymoma was the association between an intramedullary tumor and polar hemosiderin deposits ("cap sign").

Diagnosis of intramedullary hemangioblastoma was suggested by the association between an intensely T2 enhanced tumor signal and satellite cysts sometimes disproportionate to the size of the tumor (8, 9).

SURGICAL TREATMENT

In recent years there have been notable advances in intramedullary tumor surgery. The aim of surgery is spinal decompression and obtaining biopsy material for histological diagnosis (9,10,11).

In our series, 71% of intramedullary tumors were completely resected, and in the remaining 29% subtotal resection was achieved . Regarding the quality of resection, there were significant differences related to tumor histology. Thus, complete resection was mostly realised in ependymomas (78%), in hemangio-

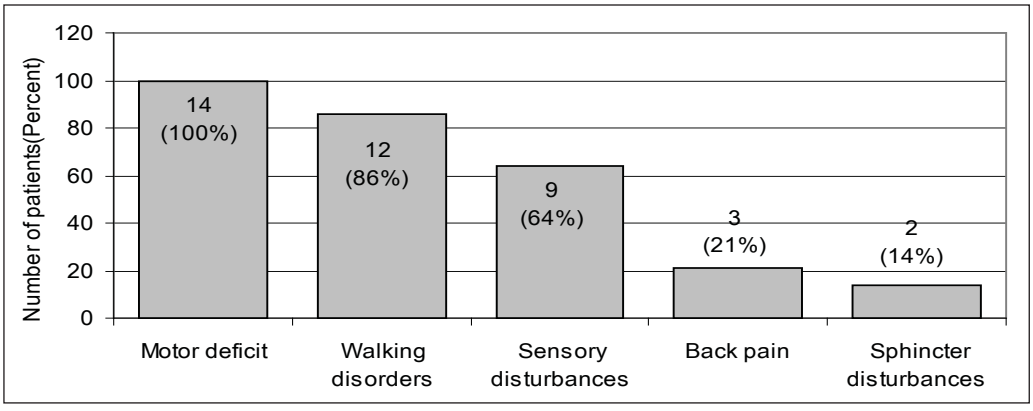

FIGURE 2. *Patients' symptoms at the moment of their diagnosis*

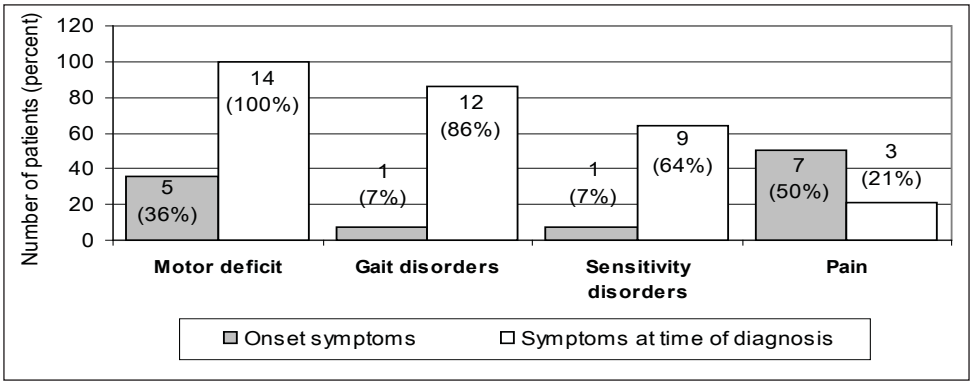

FIGURE 3. *Comparison between onset symptoms and symptoms at time of diagnosis*

blastomas (100%) and epidermoid cyst. In contrast, none of the two cases of intramedullary astrocytoma was completely resected. Depending on the location, 86% of cervical tumors were completely resected compared to 60% of thoracic and 50% of those located in the medullary cone (Fig. 4).

Evaluation of quality resection using MRI is sometimes difficult because of postoperative changes of intramedullary signal. However, persistent postoperative volume changes argue for the existence of a spinal tumor rest. Sometimes, just following the dynamic aspects on postoperative MRI allows accurate assessment of the extent of resection.

POSTOPERATIVE CLINICAL EVOLUTION

The postoperative evolution was characterized by transient neurological deterioration in 57% of patients (8 patients). Of these, 75% were recovered to preoperative neurological deficits within 3 months after surgery. The remaining 25%, needed 6 months to regain the initial neurological status. We attribute this transient damage to the myelotomy and handling of medullary tissue and also to medullary edema which occurs postoperatively. Neurological deterioration was comprehensive and included motor function, gait, sensitivity and sphincter function.

At 3 months postoperatively, pain was about the same average score as in preoperative period. Sensitivity was regained after postoperative deterioration at the preoperative level. Walking and motor function were significantly improved compared to the preoperative period. Sphincter function had minor damage at 3 months postoperatively. Comparing average symptoms at 3 months and 1 year postoperatively, there is minimal difference between the two intervals. This development demonstrates the potential recovery of patients in the first 3 months postoperatively. Patients' progress beyond the first 3 months is insignificant or slightly deteriorated. In conclusion, the 3 months after surgery may be considered borderline for clinical evaluation of patients operated on for intramedullary tumors (Fig 5).

Finally, we analyzed the clinical course compairing the Karnofski score preoperatively and postoperatively. For patients with preoperative Karnofski score between 60-100 and those with scores below 60, the clinical course was similar, and consisted in stabilization or improvement of preoperative neurological status. In other words, even for patients with severe preoperative neurological status, surgery preserves or improves slightly the preoperative status.

SHORT-TERM COMPLICATIONS

Postoperative complications occurred in 21.4% of cases. (Table 4)

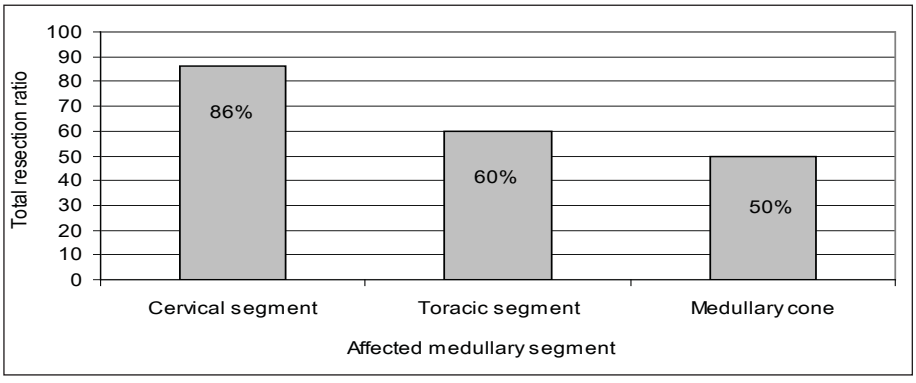

FIGURE 4. *Total resection ratio of intramedullary tumors depending on the affected medullary segment*

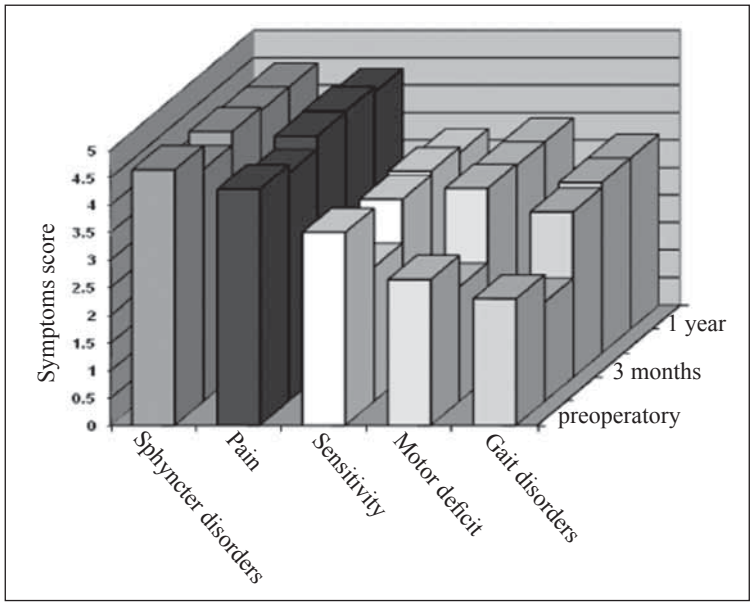

Figure 5. *Clinical evolution of intramedullary tumors in the first year after surgery.*

Prevention of CSF fistula is realised through meticulous closure of the dura, and when necessary with a duroplasty with autologous material.

TABLE 4. *Postoperative complications*

Type of complication	Number of patients
CSF leak	1
Superficial wound infection	1
Subarachnoid haemorrahage in posterior fossa	1
Total	3 (21.4%)

Mortality in the first 30 days after surgery was 7%. It was the case of a 56 years old man operated for intramedullary ependymoma D3-D4. The patient developed a massive CSF fistula through the surgical wound, so that an external lumbar drainage was mounted. At 48 hours postoperatively, after drainage of approximately 700 ml CSF, the patient became drowsy and subsequently fell into a coma. Emergency cranial CT revealed a subarachnoid hemorrhage associated with massive cerebral edema in the posterior fossa indicating a tonsillar herniation through the foramen magnum.

Long-term complications

Surgical approach required the practice of 38 laminectomies to 14 patients with an average laminectomy/patient of 2.7 ± 1.1. Of these, 23 (60.5%) were cervical, 13 (34.2%) thoracic and 2 (5.3%) lumbar. In none of the cases operated instability phenomena or deformation of the spine occur and therefore no spinal stabilization procedure was necessary. Of the 14 patients studied, 2 (14%) developed postoperative paresthesia syndrome described as a burning discomfort or pain. Symptoms occurred at 3-4 months postoperatively, and the control MRI images showed adhesions between the bone and durotomy site. Neither of the two cases required reintervention, and treatment consisted of antiinflamatory pills.

Postoperatively, patients were followed for an average of 29 ± 18.6 months (minimum 6, maximum 57 months). During this period, we observed no tumor recurrence in patients with complete resection, and cases who underwent partial resection remained stable clinically and imagistically. All patients except the patient who died within 48 hours, were alive at the end of follow up.

CONCLUSIONS

The clinical evolution af adult patients with intramedullary tumors is towards progressive neurological deterioration and severe myelic involvement.

The degree of radical resection depends on the hystological type of tumor. The resection is total in ependymomas and hemangioblastomas and subtotal in infiltrative tumors without a cleavage plane. The purpose of surgical intervention is to maintain the preoperative neurological status.

Early diagnosis and treatment of this pathology before severe myelic involvement represent the major favorable prognostic factors for these patients.

Due to the fact that intramedullary tumors are a rare pathology, we consider that their surgical treatment should be realised in regional dedicated centers with trained neurosurgeons.

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