Surgery of Intramedullary Spinal Cord Tumors, without using CUSA and IOM? Our Experience

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Abstract

Background: Intramedullary spinal cord tumors are rare lesions. In the past the treatment emphasized biopsy and radiation/chemotherapy. Nowadays, due to the improvement of the tools of microsurgery and intraoperative neurophysiological monitoring (IOM), radical surgery excision became the primary modality of treatment.

Objective: Report our experience of microsurgical resection of intra medullary spinal cord tumors without using CUSA and IOM.

Method: Forty one patients were diagnosed with primary intramedullary tumors. Complete microsurgical removal was attempted whenever possible. Both the IOM and the CUSA are not available in our department further both were never used.

Results: Neurological outcome remains the same after aggressive microsurgical resection in the patients with moderate preoperative deficits.

Conclusion: Despite the rarity of intramedullary spinal cord tumors, they remain important clinical entities which should be diagnosed early and treated conveniently and efficiently thanks to imaging tools and surgical advances.

Keywords: Intramedullary spinal cord tumors; Microsurgery; Neurological status

Introduction

Intra medullary spinal cord tumors (IMSCTs) are rare and account for only 2-4% of all central nervous system tumors. The standard treatment for most cases of IMSCT remains essentially surgical resection, which has been improved with the adequate neurosurgical instrumentation, the use of a performed operating microscope, as well as the intraoperative monitoring of motor and somatosensory evoked potentials [1-11]. However, the amount of resection almost depends on the presence or absence of a clear plane of dissection between tumor and tracts. Often ependymomas like Hemangiolastoma have a clear plane, at the opposite astrocytomas except pilocytic entity are more infiltrative, this makes any attempt at gross-total resection at high risk and mays leading to postoperative neurological aggravation. Most patients with IMSCTs present with some neurological deficit at the time of the diagnosis may get these complications aggravated by surgery and/or radiotherapy [12-20].

Gliar tumors represent more than 70% of IMSCTs, including ependymoma and astrocytoma. These entities are generally slow-growing. The correlation between clinic history and MRI informations allows predicting a histological approach of the tumor before surgery which might dictate the surgical management of the tumor and also predict properly the functional outcome after surgery [12,19-30].

In this paper we report our experience about a prospective study, realized in the optic of thesis work untitled “intramedullary spinal cord tumors diagnosis, treatment and fellow up” in the department of neurosurgery UDL SIDI BEL ABBES.

Materials and Methods

This study was conducted prospectively, between 2008 and 2017. It concerns all patients diagnosed with IMSCTs by MRI examination and confirmed by histological study. Medical history was focused on the type of the symptoms, onset's modality, and the delay of diagnosis, the completed neurological examination and the functional assessment by using the McCormick scale. Basically, the imaging assessment by MRI was used to determine the presence of IMSCTs as well as its characteristics. Histological analysis was based on the WHO classification. Statistical analysis was focused on anamnestic and clinical data, quality of resection, histological type, and functional status outcome.

Results

Forty one patients with IMSCTs were managed during our neurosurgical practice since 2008 until 2017. The age range was 2 to 71 years (mean 32 years). The female to male ratio was 1.73 (26 females to 15 males) and more than 1/3 was children. Only 3 cases were found before 2 years as well as 3 cases after 60 years old. The incidence of IMSCTs is almost equal in pediatric, young and adult population. The tumor location was thoracic in 21 patients (51.22%), cervical or cervicothoracic in 14 patients (34.15%), and conus medullaris in 6 cases.
The mean duration of symptoms before diagnosis was 22.5 months, ranging between few days and 124 months. In the pediatric population this delay was not clear and not identified in the majority of cases. A rapid onset was noted in one case due to acute decline of a pediatric cervicothoracic lipoma. In contrast, the case with the longest interval between symptoms onset and diagnosis (124 months) was observed in a C3-C6 ependymoma occurred in 67 years old male. The most common symptom was of course pain, but the weakness/gait disturbances was the most complaint which brought the patients to physician in (87%) of cases, followed by bladder and/or bowel troubles in (70%) and sensitive deficits in 18 patients (43.9 %). At the opposite in the pediatric population complaints were not easy to identify and two children manifested before diagnosis a spinal deformity and the third an acute hydrocephalus treated beforehand by ventriculo-peritoneal shunt.

The neurologic status at the time of diagnosis as assessed using the McCormick scale was as follows: grades III and IV (severe deficit) in 25 patients (60.1%) and grades I and II (normal or mild deficit in 16 patients (39.9%).

In MRI, all IMSCTs presented a spinal cord expansion at different degrees. The "cap sign" was present in ependymomas more than in hemangioblastomas. However, it was absent in the rest of IMSCTs in particular in astrocytomas. Ten cases had satellite and/or syrinx cysts, including 6 bipolar cysts. Eight others had intratumoral cysts whose intraoperative macroscopic appearance was xanthochromic, unlike the polar cysts which were predominantly fluid. The hematoma was observed in 8 cases. Peritumoral or polar edema was present in 7 cases, well demonstrated in T2 and flair as hypersignal.

In most tumors, we found varied degrees of contrast enhancement. However in 8 cases: 1 astrocytoma II, 2 lipomas, 2 cavernomas, 2 dermoid cysts, and 1 neuroglial cyst, did not manifest any contrast enhancement.

Forty six surgical procedures were undergone; four patients underwent more than surgery stage. The surgery procedure was based only on the microscope and micro instrumentation, neither CUSA nor IOM were available in our department.

The extent of resection was determined on my own appreciation and on the postoperative MRI findings. Great total resections (GTR) like partial resection have been realized each one in 45% of cases (Figures 1 and 2 and Table 1).

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<th>Resection</th>
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<td>% Histological Distribution</td>
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<td>37.5</td>
<td>9.76</td>
<td>4.88</td>
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Table 1: Resection quality and histological type (GTR: Great Total Resection; STR: Subtotal Resection; P: Partial Resection; EP: Ependymoma; AST: Astrocytoma; HEM: Hemangioblastoma; DC: Dermoid Cyst; LIP: Liopma; CAV: Cavernoma; SHW: Shwannoma; ATRT: Atypic Teratoid rhabdoid Tumor).

Glial tumors were present in 68.29% of our patients (astrocytoms and ependymomas) with astrocytoma predominance at 39.02%. The malignant gliomas represent 1/3 of astrocytoma. The hemangioblastomas present in 9.76%. The rest of 21.95% patients presented with lipomas, dermoid cysts, cavernomas, teratoid rhabdoid atypic tumor, and neuroglial tumor (Table 1).

The radiation and/or chemotherapy were prescribed for only 4 patients harboring malignant tumors and for one disseminated glioma II occurred in 2 years old girl.

Postoperatively, no surgery related mortality was encountered. Nine complications without impact on mortality with 7 CSF leakages, these patients were treated with lumbar drainage, and/ or wound revision, 1 thrombophlebitis and 1 pulmonary atelectasis. Three complications by spreading have occurred in astrocytoma II (Figure 3), glioblastoma, and benign ependymoma and were responsible of death in the 2 last cases after acute intracranial hypertension.

Figure 1: Sagittal MRI before and after surgery showing great total resection of dorsal Ependymoma.
The final functional evaluation has concluded that intermediate grades II and III were most susceptible to change, either to aggravation especially in high-grade malignant gliomas, or to improvement, especially in low-grade of malignancy IMSCTs; however Grades I and IV, changed little. None of our patients with severe preoperative bladders disorders have improved after surgery.

Discussion

IMSCTs are rare lesions and considered for a long time as inoperable due to the high surgical risk. So the surgical procedure being often limited to a biopsy.

In most series there is no predominance of sex [7,9,20], whereas in our own one, there is a clear female predominance with a sex ratio of 1.7, close to that in the series of Krifa and Woodworth [21-37], while Raco and Nakamura [28-30] found a male predominance.

The average age in our series is 32 years, lower than that reported in the literature ranging from 36 to 43 years [10,17,28,30-38].

If we consider the upper limit of the pediatric age 20 years as reported in Epstein et al. series [11], our pediatric population concerns 15 cases (36.58%), which represents more than a third of our series; with an average of 15 years, Canstantini in his pediatric and young adults series, found an average of 10 years.

Almost two third of our patients were in worse neurological status which has condemned their postoperative outcome, it is the same finding in the series of developing countries of KRIFA and Taricco [21-35]. At opposite Harrop [14] found 88% of patients without neurological deficit on the day of surgery, which explain the favorable outcome.

Only MRI is able to give a direct image of the intramedullary lesion, also provide a histological approach which will be an important element in the surgical decision and particularly in the quality of the resection [8].

The IMSCTs in our series were thoracic in 51% of cases, with an average range of 3-4 levels, which has been reported by several authors [9,21,35,36]. This is due to the low rate of the ependymomas in our series, which frequently occurred in the cervical region [1,12,20], so when the seat is thoracic like own series, it is a priori pediatric series dominated by the astrocytomas, that preferentially locate there [9].

During the surgical procedure, the CUSA, IOM, intraoperative ultrasound and far away the laser are mandatory and also used by several authors; unfortunately these tools in particular CUSA and IOM did never belong to our technical platform. Indeed this equipment offers surgeons more security as explained by Nadkarni et al. [27], but they remain complementary to microsurgery procedure.

In the literature, postoperative MRI is performed at 72 hours and/or 3 months after surgery for a better assessment of the quality of the resection. Total excision in our series has primarily and essentially concerned low-grade of malignancy IMSCTs.

For more comprehension we may discuss separately and succinctly the predominant histological entities found in our series: astrocytomas, ependymomas and hemangioblastomas.

The astrocytomas are considered to be the most frequent IMSCTs in children and young adults; they were dominant in our series with 16 cases, 39% of IMSCTs with an average age of 24 years and ranges from 2 to 55 years. These figures are close to those of the literature since the

In the follow-up, we noted five deaths in medium term and six patients were lost of view. Patients in our series had postoperative follow-up ranging from 3 months to 9 years with an average of 30 months; during that period, the functional status according to the McCormick scale was evaluated periodically, and the data were compared to the preoperative status. Then we have noted 8 worsening which 2 irreversibles, it concerns one thoracic glioblastoma and 1 thoracic ependymoma II totally resected, in which we have suspected a persistent compressive residual syrinx which has been evacuated without improvement at the last control at 12 months. Only 6 cases of benign IMSCT had improved.

So as final results of McCormick Scale, we have 28 cases unchanged, 11 cases improved and 2 worsened.
average age varies between 21 years for Sandler et al. [32] and 28 years for Hurth [16]. In our series 1/3 of the cases are of pediatric age, very close to that reported by Sandler. [32] This predominance of astrocytomas is also reported in adult series [30].

The potential for neurological aggravation following total excision of low-grade astrocytomas is considerable and their optimal treatment remains controversial, in particular the role of radical surgery as well as radiotherapy in terms of survival [4].

Of the 16 astrocytomas in our series, 10 cases were of low grade malignancy and the surgery was macroscopically total only in 2 pilocytic astrocytomas, representing a macroscopically total excision rate of 12.5 %. The partial resection interested the rest. Total resection could not be performed because of the lack of distinguishable boundary between the tumor and the spinal cord. This attitude is in agreement with the recommendations of the majority of authors who favor radical surgery as long as the cleavage plan is detectable, a common occurrence in pilocytic astrocytomas [18,19], which avoid radiotherapy, especially in children, given the frequency of astrocytomas in the pediatric population, the effects of which may be dismal [17-34].

McGirt et al. [24] insisted on the possibility of complete excision in anaplastic astrocytomas. They were able to perform total resection in 44% and subtotal in 56% of cases.

The rate of malignant astrocytomas in our series is relatively high; Raco found a ratio close to 1/4. [30]

The poor functional status in astrocytomas is directly related to their natural evolution, especially for those of high grade of malignancy which represent 6 cases in our series; as well as the diagnosis delay since the 3 glioblastomas arrived with a functional grade IV, also for 2 anaplastic astrocytomas and one astrocytoma II.

Unfortunately, and because of the altered functional and neurological status, aggressive surgery even decompressive surgery did not improve these patients and the adjuvant treatment could not be systematically instituted [3,26,27,39]. Our results are close to those of the literature where all glioblastomas died seven months on average after surgery alone. Radical surgery of glioblastoma, followed by radiotherapy, is associated with a low survival rate, which is similar to those with biopsy and radiotherapy. All glioblastoma died within a mean survival of 9 months [24]. It must be concluded that the prognosis of high grade astrocytomas is still very poor. Raco et al. [31] finds that surgical treatment does not improve postoperative neurological status, at the contrary in some cases aggravates it, and radiotherapy and chemotherapy do not have an obvious impact on survival rates.

The ependymomas represented 29.27 % of all IMSTs and 42.85 % of gliomas with average age of 39 years and range from 2 to 67 years. Hausmann [15] finds the same ratio astrocytoma /ependymoma, but in most series the ependymoma is considered as the most frequent IMSTs. This is related to the age of our patients who are in the majority children and young adult; the age of choice for astrocytomas.

Our series confirms the primary therapeutic objective of ependymomas, which is total excision with preservation of neurological function. Indeed these lesions have a good cleavage plan to achieve this goal.

Unfortunately, the improvement or preservation of the functional status depends essentially on the preoperative conditions. Indeed, we achieved a total resection in 75% of cases; this rate is much lower than reported in the literature. This is due to the presence in our series of an ependymoma grade III, and in 2 other cases the surgery was interrupted because of appearance of hemodynamic disorder in thoracic ependymomas and the presence of adherence to anterior tracts in C2C3 ependymoma. Radical surgery remains the only effective weapon for better survival (Figure 2). While the effect of radiotherapy remains uncertain. Oh et al. [29] found, that the 5-year progression-free survival (PFS) is 97.9% in the total excision, 63.3% in the partial excision followed by radiotherapy and 45.1% in the subtotal excision without radiotherapy; which means a 20% improvement in PFS with adjuvant radiotherapy. On the other hand, only the total excision improves the survival rate. Needless to say, his paper did not take into account unlike our series the favorable preoperative neurological status which is an important prognosis factor?

The functional results of our patients were not satisfactory because of the bad neurological status before surgery. One case that holds a low thoracic (Th8 to Th10) ependymoma worsened. Nevertheless, functional prognosis is still poor in patients with severe preoperative paralysis and also in those with a tumor arising in the thoracic cord. This could be attributable to irreversible changes in the spinal cord caused by prolonged tumor compression and to poor microcirculation within the thoracic spinal cord both increasing the vulnerability of the spinal cord against intraoperative maneuvers. It is, therefore, concluded that early diagnosis and early surgery, before the paralysis becomes severe, are important to obtain good functional outcomes particularly in cases with the tumors arising in the thoracic cord, even if the neurological deficit is mild. These results are very consistent with the literature [1,6,13,22].

The hemangioblastomas were the third highest in frequency, with a rate of 9.76%, and an average age of 46.5 years in agreement with the literature [5,23,30]. We performed total resection in three-quarters of cases and one case taken for ependymoma, whose surgery was partial because of the abundant bleeding after attempting resection by fragmentation. In the literature total resection has reached up to 100% of cases, sometimes even after several attempts [23,25,33]. As advocated by the literature to eliminate VHL disease, all our patients underwent brain MRI in addition to spinal MRI, which did not object other locations.

Our study has achieved results that relatively corroborate those reported in the literature. The early surgical aggressive resection of primary intramedullary tumors would be suggested for patients with mild neurological complaints harbored well delimited tumors with great caution for those located into the low thoracic spinal cord. Several points still need to be reassessed; the effective useful of intraoperative electrophysiological monitoring especially in the tumors with well plane of dissection and in the patient with severe neurological deficit; on the other hand, the advantage in using CUSA for small tumors, knowing that it could induce micro trauma to the spinal cord and increased the eventuality of motor deterioration. In fact the good analysis of the natural history of the IMST, their imaging features, adequate knowledge of surgical anatomy and the correct use
of microsurgical techniques allowed total resection of the majority of well demarcated tumors with minimal morbidity.

Conclusion

IMSCIs are best treated before major neurological deficits appear, with GTR being the primary aim of treatment. Before surgery a histological approach could be predicted by combining MRI features with medical history and clinical examination. This allows an approximate surgical management of these lesions and providing pertinent predictors of the functional outcome. Preoperative neurological status remains the best predictor of postoperative function. The use of adjuvant radiotherapy and/or chemotherapy is an option when GTR cannot be achieved especially in malignant tumors.

References