



Management and outcome of primary spinal ependymomas: A single center experience from Taiwan



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ABSTRACT

Background: Surgical treatment of spinal ependymomas requires careful consideration of the relative risks of neurological worsening from surgery. Our aim was to determine the risk factors of neurological deterioration after surgery for spinal ependymomas.

Material and methods: This 20-year study included 17 patients (seven men and 10 women; 44.65 ± 13.62 years) with histologically confirmed spinal ependymomas. The basic features were reviewed and the functional status was assessed by using the modified McCormick classification. We subdivided the patient population into two groups according to whether neurological deterioration occurred after primary tumor resection ($N=5$) or not ($N=12$), and compared their clinical characteristics.

Results: The average duration of presenting symptoms in the 17 patients was 23.53 ± 21.45 months. Three (17.6%) patients underwent subtotal or partial resection and 14 (82.4%) patients underwent gross total resection. The incidence of neurological deterioration after primary resection of spinal ependymomas was 29.4%. There were five (100%) and two (16.7%) male patients in the neurological-deterioration and no-deterioration groups, respectively ($p=0.003$). The duration of presenting symptoms was 24 months or over in all the patients with neurological deterioration and five of the 12 patients with improved or stabilized function ($p=0.044$).

Conclusion: The risk associated with surgical resection of spinal ependymomas should not be overlooked because of the significant incidence of neurological deterioration. The male gender and long-standing symptom (≥ 24 months) are risk factors of postoperative neurological worsening. Early diagnosis and surgery are therefore critical for successful treatment of spinal ependymomas.

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1. Introduction

Ependymomas are neuroectodermal tumors and are thought to arise from the ependymal lining of the ventricles and central canal. These tumors constitute approximately 30–88% of the spinal gliomas [1]. They can be intramedullary or extramedullary; the cervical region is the most common level of intramedullary occurrence [2]. Most ependymomas are benign and slow growing, and tend to compress the adjacent cord parenchyma rather than infiltrate it [3,4]. Thus, such tumors present a relatively clear surgical plane and are curable with gross total resection. This surgical preference is mainly based on the progressive nature of neurological deficits in patients with these tumors and the fact that

complete resection can be achieved in around 90% of the cases [5].

However, even after technically successful surgical excision, significant improvement in severe or long-standing preoperative neurological deficit rarely occurs [6]. Preservation rather than restoration of neurological function is the reasonable aim of surgery. More importantly, surgery unavoidably carries the risk of spinal cord injury, and some patients experience persistent worsening of the neurological grade following surgery. The incidence of postoperative neurological deterioration in patients with prior neurological deficits of variable severity is reportedly 13.9%; in patients with no objective neurological deficit preoperatively, there is still a 10% chance of postoperative worsening [5].

Although most authors conclude that gross total resection of ependymomas can be safely performed [7,8], surgeons should not overlook the morbidity resulting from surgery. We retrospectively analyzed neurological function, complications, and recurrence in patients who underwent surgery for spinal ependymomas to determine the risk factors of postoperative neurological deterioration.

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Table 1
Modified McCormick classification.

Grade	Definition
I	Neurologically normal Gait normal Normal professional activity
Ib	Tired after walking several kilometers Running is impossible, or moderate sensorimotor deficit does not significantly affect the involved limb Moderate discomfort in professional activity
II	Presence of sensorimotor deficit affecting function of involved limb Mild to moderate gait difficulty Severe pain or dysesthetic syndrome impairs quality of life Independent function and ambulation maintained
III	More severe neurological deficit Requires cane and/or brace for ambulation or maintains significant bilateral upper-extremity impairment May or may not function independently
IV	Severe neurological deficit Requires wheelchair or cane and/or brace with bilateral upper-extremity impairment Usually not independent

2. Materials and methods

From 1990 to 2010, 21 patients with spinal ependymomas underwent primary surgery at Kaohsiung Chang Gung Memorial Hospital in Taiwan. The inclusion criterion was histologically confirmed ependymoma (World Health Organization Grade II) with primary spinal localization. We excluded one patient with myxopapillary ependymoma (World Health Organization Grade I), and three patients with incomplete medical records. Finally, 17 patients were enrolled. Their medical records were reviewed after approval by the Institutional Review Board of our Hospital.

We applied a modified McCormick classification [9] (Table 1) to evaluate the neurological function. Grades were assigned on the basis of documented neurological examinations. The functional assessments were conducted at the preoperative stage, immediate postoperative stage, and regular follow-up period.

Magnetic resonance imaging (MRI) data of the spine were obtained in all cases at admission, and interpreted according to the findings including the vertebral level spanned by the tumor, size of the tumor measured at the solid part with enhancement, presence of syringomyelia, or intratumoral hemorrhage (Fig. 1). Follow-up MRI studies were performed in the event of new onset of neurological deficits or for routine postoperative evaluation.

Surgery was performed under microscopic assistance. The midline of the cord was grossly estimated by inspecting the bilateral dorsal root entry zones and myelotomy was performed at the area of the maximal cord enlargement. The tumor in entirety was exposed rostrally and caudally. In most cases, a well-defined tumor was easily identified, and excision was continued. If an infiltrating tumor was encountered, further tumor removal was not warranted. All specimens were obtained to establish a histological diagnosis (Fig. 2). The extent of surgery was classified as follows. Gross total resection (GTR) was defined as gross macroscopic removal of the visible tumor, or no evidence of a residual tumor on postoperative MRI. Tumor removal was considered subtotal resection (STR) when at least 80% of the tumor was excised. Partial resection (PR) was consistent with a less than 80% resection. There is no standard for postoperative adjuvant radiotherapy at our institute.

The clinical and radiological findings were integrated to evaluate tumor recurrence or progression. Reappearance or worsening of neurological symptoms in the patients who underwent GTR implied recurrence, and the diagnosis was established by an MRI scan showing a new-growing tumor arising from the vertebral levels spanned by the primary tumor. Progressive disease after STR

or PR was indicated by deterioration of the previously stable neurological signs and symptoms, and volumetric progression was confirmed radiologically.

The general characteristics of the 17 patients were documented. The postoperative complications consisted of cerebrospinal fluid (CSF) leakage, infection of the wound or central nervous system, urinary tract infection, pneumonia, and respiratory failure, defined as the need for tracheostomy and mechanical ventilation. The follow-up period was terminated either by the end of the study (July 2010) or owing to the patient's death. All the patients were followed up at the outpatient department to determine the neurological outcome.

All statistical analyses were conducted by using SPSS version 12.0 (SPSS, Inc., Chicago, IL). The quality of the means for continuous variables was assessed by using the Mann–Whitney *U*-test. Categorical variables were analyzed by using the chi-square test or Fisher's exact test. A *P*-value of 0.05 or less was considered statistically significant.

3. Results

There were seven men and 10 women. Their mean age was 44.65 ± 13.62 years (range, 16–73 years). The follow-up period ranged from 4 months to 191 months (54.00 ± 52.50 months). The patient characteristics are summarized in Table 2.

3.1. Clinical manifestations

The duration of symptoms before diagnosis ranged from 1 month to 6 years, and in one patient, the tumor was incidentally diagnosed during the health examination. The average duration of presenting symptoms was 23.53 ± 21.45 months. The most frequent symptom, weakness of the extremities, was reported by 13 (76.5%) patients. Eleven (64.7%) patients had sensory disturbance and the location of deficits varied by the tumor site. Pain localized to the neck or back was noted in four (23.5%) patients and gait disturbances were documented in 10 (58.8%) patients. Other neurological deficits included bowel or bladder dysfunction in five (29.4%) and hyperreflexia in 10 (58.8%) patients.

3.2. MRI findings

The MRI scans showed the tumor in the cervical region in six (35.3%) patients, cervicothoracic region in four (23.5%) patients, thoracic region in four (23.5%) patients, thoracolumbar region in one (5.9%) patient, and lumbar region or conus medullaris in two (11.8%) patients. The tumors spanned one vertebral level in one patient, two levels in four patients, three levels in seven patients, four levels in three patients, and five or more levels in two patients. On average, the solid portion of the tumor extended over 3.24 ± 1.56 vertebral bodies and the size was 46.63 ± 30.63 mm in length. The presence of syringomyelia was identified in 10 (58.8%) patients. No intratumoral hemorrhage was found by the MRI examination.

3.3. Surgery and adjuvant therapy

GTR were performed in 14 (82.4%) patients. Among them, there were three patients with complete resection confirmed by neurosurgeons. The other 11 patients underwent postoperative MRI and showed no evidence of residual tumor. STR was achieved in one (5.9%) patient, and PR was performed in two (11.8%) patients. Post-surgically, two (11.8%) patients underwent adjuvant radiotherapy: one patient who underwent PR received a total dose of 1800 cGy in 18 fractions, and the other one had GTR and subsequent radiotherapy. However, as part of the latter patient's medical records

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