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Original Investigation

Surgical Strategies and Functional Outcome of Intramedullary Cervicomedullary Ependymoma

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ABSTRACT

AIM: To study the surgical strategies and functional outcomes of intramedullary cervicomedullary ependymoma (ICE).

MATERIAL and METHODS: The authors retrospectively reviewed the clinical and imaging data of 28 ICE patients who underwent surgical resection, including 11 males and 17 females and the average age was 37.9 years (range:11-69 years). The modified McCormick grading system (mMG) was used to evaluate the neurological functions before surgery, at discharge and at long-term follow-up.

RESULTS: Gross total resections (GTRs) were performed in 21 patients (75.0%), and subtotal resections (STRs) were achieved in the other 7 patients (25.0%). An ill-defined tumor border was a significant factor leading to STR (p=0.026). At discharge, the mMG improved in 11 patients (39.3%), stabilized in 14 (50.0%), and worsened in 3 (10.7%). The 3 patients with deterioration were all in the STR Group and the neurological deterioration rate was significantly higher in the STR Group than that in the GTR Group (p=0.011). The follow-up period ranged from 9 to 77 months (mean 49.6 ± 26.9 months). Two patients (7.1%) had tumor recurrence during this period. The long-term follow-up mMG was improved in 25 patients (89.3%), stabilized in 3 (10.7%), and no one worsened, compared with preoperative mMG.

CONCLUSION: GTR of ICE could be achieved in majority of patients, especially for those with well-defined tumor boundaries. GTR lead to a better outcome in the short term after surgery. A favorable functional outcome could be achieved for most ICE patients in the long term.

KEYWORDS: Ependymoma, Cervical spinal cord, Medulla oblongata, Intramedullary, Prognosis

INTRODUCTION

Ependymomas are uncommon central nervous system (CNS) neuroectodermal tumors and constitute approximately 2-8% of all primary CNS tumors and 30%–88% of primary spinal intramedullary tumors (6, 9, 21, 26). The majority of previous studies regarding ependymomas are either cerebral or intraspinal ependymomas, and intramedullary cervicomedullary ependymoma (ICE) is a really rare entity (15). Due to the location of the tumor, ICE may result in neurological dysfunction, such as motor weakness, respiration disorders, dysphagia, and so on.

Most ICEs are benign tumors that grow slowly, and usually lead to compression of the adjacent brain stem rather than infiltration. The plane of these tumors was relatively clear and gross-total resection (GTR) could be achieved in most cases. However, because of the tumor location, the surgical resection of the tumor could still result in disability even for experienced surgeons. Although radical resection is the goal of surgery, every effort should be made to preserve the functional status (22, 27).

Due to the rarity of this tumor, the treatment strategies, functional outcome and the prognostic factors are still unclear.

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Therefore, the authors conducted this retrospective study to analyze the surgical strategies and functional outcomes of the ICE patients.

■ MATERIAL and METHODS

Patient Population

This study was approved by the Institutional Review Board of Beijing Tiantan hospital, Capital Medical University. The authors performed a retrospective study on 28 consecutive ICE patients, who were admitted in our institute between January 2009 and December 2014. All patients enrolled in this study underwent surgical resection and histological examination, and all patients were pathologically diagnosed with ependymoma (WHO Grade II) or anaplastic ependymoma (WHO Grade III). The definition of ICE was intramedullary ependymoma, which both rostrally involved the medulla oblongata and caudally involved the cervical cord.

According to the criteria, as shown in Table I, there were 11 males and 17 females enrolled in this series (ratio 0.65:1.00). Ages ranged from 11 to 69 years with an average of 37.9±13.4 years. The clinical features, imaging findings, surgical treatment, complications and outcomes at short-term and long-term follow-up were analyzed.

Clinical Evaluation

Functional status was assessed based on the modified McCormick grading (mMG) system (Table II) preoperatively, at discharge (1-4 weeks after surgery), and at long-term follow-up (9-77 months after surgery) (5, 12, 18). The patients were followed up by telephone, outpatient and inpatient interviews.

Imaging Study

All patients underwent contrast-enhanced T1-weighted magnetic resonance imaging (MRI) of the cervical spinal cord preoperatively, 1-4 weeks after surgery, 3 to 6 months after surgery, and annually thereafter. The imaging findings included tumor location, tumor volume, tumor boundary, and presence and location of syringomyelia. Tumor volume was defined as: greatest anteroposterior dimension×greatest mediolateral dimension× greatest craniocaudal dimension×0.5 (19). The tumor boundary was assessed according to preoperative T2-weighted and contrast-enhanced T1-weighted MR images. On T2-weighted MRI, we identified if there was a clear subarachnoid space between the tumor and cord; on contrast-enhanced T1-weighted MRI, we determined if there was a smooth contour line between the tumor and the cord. If one or two of the above two conditions were met, it was defined as a well-defined tumor boundary (Figures 1A-D, 2A-D, 3A-D). If neither condition was met, it was considered as an ill-defined tumor boundary (Figure 4A-D) (7, 15). The length of syrinx was measured on sagittal T2-weighted MRI.

Surgical Procedures

All patients were microsurgically treated through a standard suboccipital midline approach with the lateral decubitus position. After laminectomy, the dura mater and the arachnoid membrane were opened with the assistance of microscope. Then, a myelotomy was performed along the midline of the spinal cord. The plane between the tumor and the cord was carefully dissected (15). To prevent neurological deficits, intraoperative monitoring of evoked potentials was applied, including spinal cord somatosensory evoked potentials (SSEPs), motor-evoked potentials (MEPs), and brainstem auditory evoked potentials (BAEPs) (16). After tumor resection, the dura mater was closed and then laminae replacement was performed to avoid future spinal instability. The syrinx was not manipulated directly during the operation.

Extent of Tumor Resection

The extent of surgery was classified as GTR, subtotal resection (STR), or partial resection. GTR was defined as complete removal of the visible tumor under microscope and no evidence of remnant on postoperative contrast-enhanced T1-weighted MRI (Figures 1A-D, 2A-D, 3A-D). It was taken as STR when at least 80% of the tumor was removed (Figure 4A-D). If less than 80% of the tumor was excised, it was considered as partial resection (14, 15).

Statistical Analysis

All statistical analyses were performed with SPSS (Windows version 18.0, IBM). Wilcoxon signed rank test was performed to compare the mMG before surgery, at discharge and at the last follow-up. Logistic regression analysis was done to study factors associated with STR, including gender, age, duration of symptoms, syrinx, tumor volume, and tumor border. Chi-square test for the R×C contingency tables was used to analyze the difference in neurological outcome between the GTR Group and the STR Group. Odds ratio with 95% confidence interval (CI) was presented. A probability value <0.05 was considered statistically significant.

■ RESULTS

Clinical Characteristics

On admission, all patients presented with neurological deficits of variable severity. As shown in Table I, the main clinical symptoms included motor deficits in 14 patients (50.0%), neck pain in 13 (46.4%), sensory changes in 12 (42.9%), headache in 6 (21.4%), lower cranial nerve (CN) deficits (dysphagia, hoarseness, and bucking) in 4 (14.3%) and dizziness in 2 patients (7.1%). Clinical progression was relatively slow, and the mean duration of symptoms was 14.2 ± 19.2 months (range: 1-72 months).

Neurologically, according to the mMG system, 1 patient (3.6%) was categorized in Grade I, 8 (28.6%) in Grade Ib, 12 (42.9%) in Grade II, 5 (17.9%) in Grade III and 2 (7.1%) in Grade IV (Table I).

Radiological Features

The mean of the tumor length was 5.5 ± 4.8 cm (range: 1.0–22.0 cm), and the caudal end of the tumor ranged from cervical 1 (C1) to thoracic 4 (T4) (medium C2). The mean tumor volume was 9.9 ± 11.5 cm^3 (range: 0.3-49.5 cm^3). Syrinx was observed in 15 patients (53.6%) and the length of syrinx ranged from 1.0 cm to 15.0 cm (mean 7.3 ± 4.5 cm). Eighteen patients

(64.3%) presented with a well-defined tumor boundary and an ill-defined tumor boundary was detected in the other 10 patients (36.2%) (Table I).

Surgery and Morbidity

Overall, GTR was achieved in 21 patients (75.0%) (GTR Group), STR was conducted in 7 patients (25.0%) (STR Group), and no patient underwent partial resection. Out of the 7 patients who underwent STR, in 6 patients had an ill-defined tumor boundary and 1 patient lost SSEPs intraoperatively, which prevented GTR. Furthermore, the factors leading to STR were studied using logistic regression analysis, and results showed that an ill-defined tumor border was a significant factor (p=0.026, OR=202.937, 95% CI: 1.870, 22022.498). The other factors were not significantly associated, including gender (p=0.574), age (p=0.115), duration of symptoms (p=0.315), syrinx (p=0.987), and tumor volume (p=0.378).

Twenty-six patients had a benign ependymoma (WHO Grade II) and 2 patients had an anaplastic ependymoma (WHO Grade III) (7.1%), demonstrated by pathological examinations. Neurological function impairment was observed in 16 patients (42.1%), including respiratory difficulty (n=3, 10.7%), lower cranial nerve function impairment (n=3, 10.7%), intracranial infection (n=2, 7.1%), pneumonia (n=2, 7.1%), and pneumothorax (n=1, 3.6%).

After surgery, tracheal intubation was routinely kept to prevent suffocation or respiratory failure in all patients. Intubation was removed in 25 patients (89.3%) in 1.5 ± 1.2 days (range: 1–6 days). The other 3 patients (10.7%) who had respiratory difficulty underwent tracheostomy and required ventilator assistance for 3, 30, and 41 days, respectively. All these patients improved and could breathe independently before discharge. The tracheostomies were closed at 17, 40, and 60 days after surgery, respectively. Besides, nasogastric feedings were performed in 6 patients (21.4%) for 15.4 ± 20.5 days (range: 1–50 days) due to lack of cough reflex.

We recommended radiotherapy to patients with STR and anaplastic ependymomas. A total of 7 patients underwent postoperative adjuvant conventional radiotherapy, including 3 patients with STR, 2 patients with anaplastic ependymomas, and 2 patients with benign tumors and GTR.

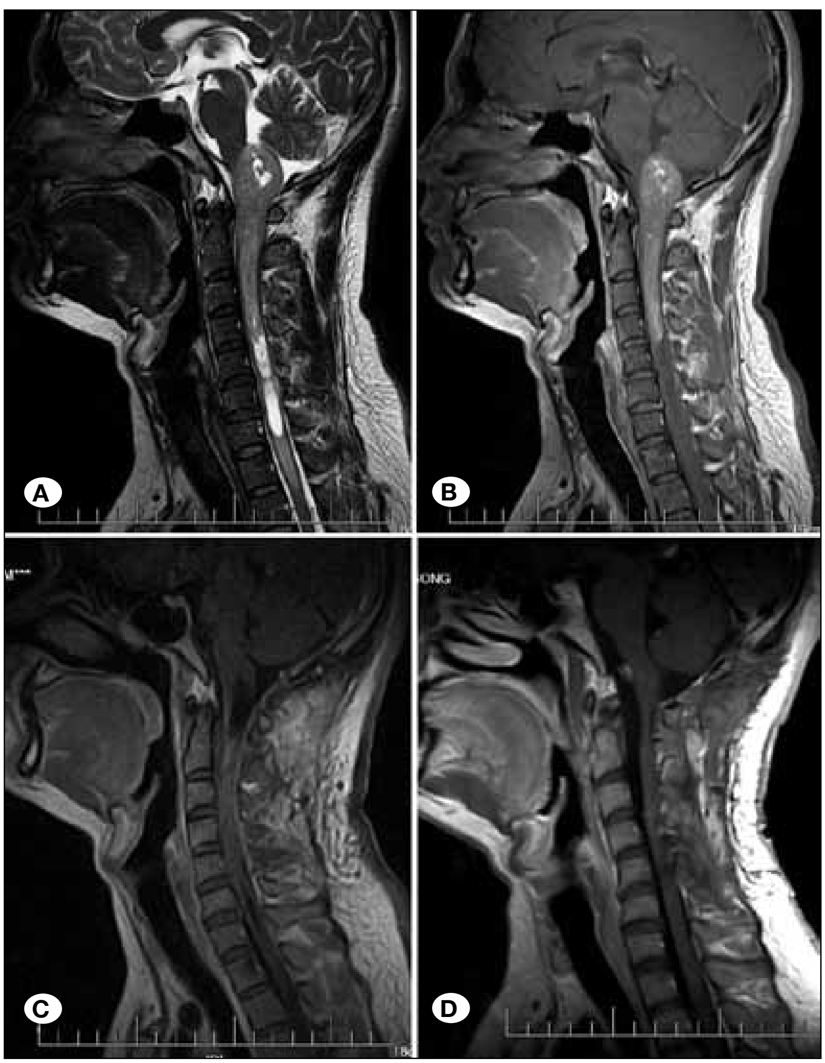

Figure 1: Pre- and postoperative MRI scans of patient No. 10.
Preoperative sagittal T2-weighted MRI **(A)** and T1-weighted imaging with contrast enhancement **(B)**, showing the ependymoma from medulla oblongata to fourth cervical vertebra with a well-defined tumor boundary and a cervical syrinx. Postoperative MRI scans with contrast enhancement at 1 week **(C)** and 3 years after operation **(D),** demonstrating complete resection of the lesion (C) and no evidence of recurrence (D). And reduction of the syrinx could also be observed (D).

Table I: Clinical, Imaging and Follow-up Data of the 28 ICE Patients

No.	Age (yrs)	Sex	Main Symptoms	DOS (months)	Tumor Location	Tumor Volume (cm³)	Tumor Border	Syrinx Length (cm)	Surgical Removal	Trache-otomy	mMG			Radio-therapy	FUP (months)	P/R
											Pre-	Post-	Recent			
1	11	M	Motor weakness	26	Medulla-T4	49.5	WD	7.0	GTR	-	IV	IV	III	-	77	+
2	46	M	Headache	7	Medulla-C2	1.9	WD	-	GTR	-	Ib	Ib	I	-	77	-
3	53	M	Headache	2	Medulla-C2	31.5	WD	-	GTR	-	Ib	Ib	I	+	77	-
4	42	F	Dysphagia	18	Medulla-C2	12.5	WD	-	GTR	-	II	Ib	I	-	76	-
5	24	M	Motor weakness	12	Medulla-C6	30.0	ID	14.0	GTR	+	II	II	I	-	74	+
6	55	M	Headache	6	Medulla-C1	2.8	ID	-	GTR	-	II	Ib	Ib	+	74	-
7	39	F	Motor weakness	4	Medulla-C5	6.5	ID	-	GTR	-	III	III	Ib	-	73	-
8	39	M	Neck pain	2	Medulla-C2	1.1	WD	-	GTR	-	Ib	Ib	Ib	-	73	-
9	23	F	Neck pain	4	Medulla-C2	10.9	WD	-	GTR	-	Ib	Ib	I	+	73	-
10	31	M	Neck pain	24	Medulla-C4	20.0	WD	7.0	GTR	-	Ib	Ib	I	-	71	-
11	32	M	Neck pain	12	Medulla-C7	10.9	WD	8.0	GTR	-	IV	IV	Ib	-	69	-
12	54	F	Numbness of limbs	72	Medulla-C2	3.4	WD	7.0	GTR	-	II	II	Ib	-	57	-
13	54	F	Dizziness	6	Medulla-C1	5.0	WD	-	GTR	-	Ib	I	I	+	46	-
14	22	F	Motor weakness	6	Medulla-C4	6.8	WD	15.0	GTR	-	III	Ib	I	-	28	-
15	44	M	Motor weakness	9	Medulla-C1	1.1	WD	-	GTR	-	II	I	I	-	27	-
16	49	F	Motor weakness	10	Medulla-C2	2.3	WD	2.0	GTR	-	II	I	I	-	27	-
17	31	M	Dysphagia	2	Medulla-C1	6.0	WD	-	GTR	+	III	II	II	-	26	-
18	30	F	Motor weakness	12	Medulla-C6	9.9	WD	13.0	GTR	-	III	III	I	-	23	-
19	69	F	Motor weakness	50	Medulla-C2	1.5	WD	-	GTR	-	II	II	I	-	9	-
20	37	F	Headache	1	Medulla-C2	2.6	WD	4.0	GTR	-	II	Ib	I	-	9	-
21	39	F	Numbness of limbs	1	Medulla-C1	0.3	ID	7.0	GTR	-	I	I	I	-	9	-
22	12	F	Headache	8	Medulla-C1	6.0	WD	-	STR	-	Ib	I	I	-	77	-
23	37	F	Numbness of limbs	72	Medulla-C5	22.5	ID	10.0	STR	+	III	IV	II	-	74	-
24	45	F	Headache	3	Medulla-C1	9.4	ID	3.0	STR	-	II	II	Ib	-	72	-
25	22	F	Motor weakness	14	Medulla-C3	2.0	ID	2.0	STR	-	II	III	II	+	36	-
26	42	F	Numbness of limbs	6	Medulla-C1	1.5	ID	1.0	STR	-	II	Ib	Ib	+	24	-
27	44	M	Numbness of limbs	6	Medulla-C2	2.6	ID	10.0	STR	-	Ib	II	I	-	18	-
28	36	F	Motor weakness	2	Medulla-C1	16.2	ID	-	STR	-	II	I	I	+	12	-

***DOS:** Duration of symptoms; **FUP:** Follow-up period; **GTR:** Gross total resection; **ID:** ill defined; **mMG:** Modified McCormick grade; **Pre-:** Preoperative; **Post-:** Postoperative; **STR:** subtotal resection; **WD:** well defined; **"-":** negative; **"+":** positive.*

Short-Term Outcomes

As shown in Table I, compared to the preoperative mMG, the mMG at discharge improved in 11 patients (39.3%), stabilized in 14 (50.0%), and deteriorated in 3 (10.7%). Wilcoxon signed rank test showed the mMG at discharge was significantly better than the preoperative mMG (p=0.018). All the 3 patients with deterioration worsened 1 grade after surgery and they were all in the STR Group. And the neurological deterioration rate was significantly higher in the STR Group than that in the GTR Group (p=0.011, by Fisher's exact test). Chi-square test showed the other factors were not significantly associated with neurological deterioration, including gender (p=1.000), age (p=1.000), syrinx (p=0.226), and tumor length (p=1.000).

Moreover, the transition of different preoperative symptoms was also studied, and the results were shown in Table III. At discharge, neck pain was relieved in all the 13 patients (100.0%), headache was relieved in 5 of the 6 patients (83.3%), lower cranial nerve deficits was improved in 3 of the 4 patients (75.0%), numbness of limbs and dizziness was relieved in 50.0% of the patients, and the improvement rate of motor deficits was 35.7%. Therefore, the improvement rate of neck pain was significantly higher than that of motor deficits (p=0.001, by Fisher's exact test), and numbness of limbs (p=0.005, by Fisher's exact test). No significant difference was found between other symptoms. Only 3 patients with motor deficits deteriorated after surgery and the other symptoms were improved or stable. New onset of paresthesia, motor weakness, and ataxia occurred in 3 (10.7%), 2 (7.1%), and 1 (3.4%) patient, respectively.

Long-Term Outcomes

The mean duration of follow-up was 49.6 ± 26.9 months (range: 9–77 months), and no patient died at the most recent

Figure 2: Pre- and postoperative MRI scans of patient No. 18.
Preoperative sagittal T2-weighted MRI **(A)** and T1-weighted imaging with contrast enhancement **(B)**, showed the ependymoma from medulla oblongata to sixth cervical vertebra with a well-defined tumor boundary, and a cervicothoracic syrinx could also be observed. Postoperative MRI scans with contrast enhancement at 1 week **(C)** and 2 years **(D)**, demonstrating complete resection of the lesion (C) and mild myelatrophy without recurrence, and reduction of the syrinx (D).

follow-up. Two (7.1%) patients (No.1 and 5) had tumor recurrence after a mean period of 30 months, which were both local recurrence. These 2 patients were closely followed up at present and a second operation was not performed because they stayed clinically stationary. As shown in Table I, at the most recent follow-up, there were 17 Grade I patients (60.7%), 7 Grade Ib patients (25.0%), 3 Grade II patients (10.7%), 1 Grade III patient (3.6%), and no Grade IV patients. Compared to the preoperative mMG, the mMG at long-term follow-up improved in 25 patients (89.3%), stabilized in 3 (10.7%), and no one worsened. The long-term neurological function was significantly better than that of the preoperative mMGs ($p<0.001$, by Wilcoxon signed rank test), and also significantly better than that of the mMGs at discharge ($p<0.001$, by Wilcoxon signed rank test), indicating that the operation could lead to better neurological functions in the long run.

The improvements in the symptoms detected at the most recent follow-up were detailed in Table III. In patients with preoperative motor dysfunction, neck pain, numbness of limbs, headache, lower cranial nerve deficits, and dizziness, the corresponding symptom improved in 78.6%, 100.0%, 75.0%, 83.3%, 100.0%, and 100.0% of the patients, respectively.

Moreover, although the syrinx was not directly manipulated during the operation, syrinx reduction could be observed in the follow-up MR images on all the 15 patients with syrinx (Figures 1A-D, 2A-D, 4A-D).

■ DISCUSSION

Epidemiology and Clinical Features

Ependymomas are an uncommon and relatively rare entity, which accounts for 2-8% of all primary CNS tumors and 30%–88% of primary spinal intramedullary tumors (6, 9, 21, 26). Ependymomas involving the medulla oblongata is rather rare. One report showed the average age at diagnosis is in the fourth decade of life (15), which is also confirmed by our results. Our study seemed to support that there might be a female predominance in ICE patients, with a male-to-female ratio of 0.65:1.00.

It is reported that the most common clinical manifestations was pain, followed by paresthesia, motor weakness, lower cranial nerve deficits, bladder dysfunction and headache (15). Our study revealed similar results, showing that motor weakness (n=14, 50.0%), neck pain (n=14, 46.4%) and numbness of limbs (n=12, 42.9%) are the most common symptoms. Although neck pain is not a specific manifestation for ICE patients and may occur in other diseases (cervical disc herniation, for example), we found it is important for ICE

Table II: Modified McCormick Grading Scale for Neurological Function*

Grade	Grade Definition
I	Neurologically normal; gait normal; normal professional activity
I b	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 impairing patient's quality of life; still functions and ambulates independently
III	More severe neurological deficit; requires cane/brace for ambulation or significant bilateral upper extremity impairment; may or may not function independently
IV	Severe deficit; requires wheelchair or cane/brace with bilateral upper extremity impairment; usually not independent

******From McCormick et al., 1990.*

Table III: Transition of Different Symptoms After Surgery

Preoperative Symptoms	At discharge			Recent		
	Improved (%)	Stable (%)	Deteriorated (%)	Improved (%)	Stable (%)	Deteriorated (%)
Motor deficits (n=14)	5 (35.7)	6 (42.9)	3 (21.4)	11 (78.6)	3 (21.4)	
Neck pain (n=13)	13 (100.0)			13 (100.0)		
Numbness (n=12)	6 (50.0)	6 (50.0)		9 (75.0)	3 (25.0)	
Headache (n=6)	5 (83.3)	1 (16.7)		5 (83.3)	1 (16.7)	
Lower cranial nerve deficits (n=4)	3 (75.0)	1 (25.0)		4 (100.0)		
Dizziness (n=2)	1 (50.0)	1 (50.0)		2 (100.0)		

patients because almost half of ICE patients had neck pain and it could be improved in all patients in a short term after surgery.

The majority of ICE are benign tumors and have a relatively long duration of illness (1, 2). In this series, the duration of illness ranged from 1-72 months with an average of 14.2 ± 19.2 months, which reflected the slow growth pattern of the tumor. No patient presented with acute symptoms suggestive of tumor hemorrhage.

Surgical Management

There is an agreement that surgical resection is the main treatment for spinal cord ependymomas, and the primary goal is GTR. Previous studies have shown that GTR is the most significant factor influencing the prognosis for spinal cord ependymomas and GTR was found to be superior to partial resection with radiation (10, 11, 17, 24, 25). Meanwhile, it is also a general consensus to perform conservative STR rather than aggressive GTR, to prevent neurological function deficits (3, 13, 14, 20).

We agree with the above viewpoints. The primary goal of surgical resection of ICE is GTR without severe function deterioration. When GTR is safe to conduct, every effort should be made to achieve GTR. Intraoperatively, if infiltrating growth of the tumor and severe adhesion to the surrounding cord tissues is found, or the monitor shows abnormalities, STR should be considered rather than aggressive GTR. In our experience, with the improvement of microsurgical skills and the introduction of monitoring techniques such as SSEPs, MEPs and BAEPs, the removal of ICE has become associated with even lower morbidity and mortality rates, making GTR the primary goal in the treatment of ICE. The rate of GTR in treating spinal cord ependymomas in other series varies considerably from 50% to 92% (2, 8, 15, 17, 24). In our series, 75.0% of ICE underwent GTR.

Moreover, we found the significant factor leading to STR was an ill-defined tumor border on preoperative MRI (p=0.026, OR=202.937, 95% CI: 1.870, 22022.498). Of the 18 patients with a well-defined tumor border, 17 patients underwent GTR and only 1 patient had STR. Meanwhile, of the 10 patients with an ill-defined tumor border, 4 patients underwent GTR

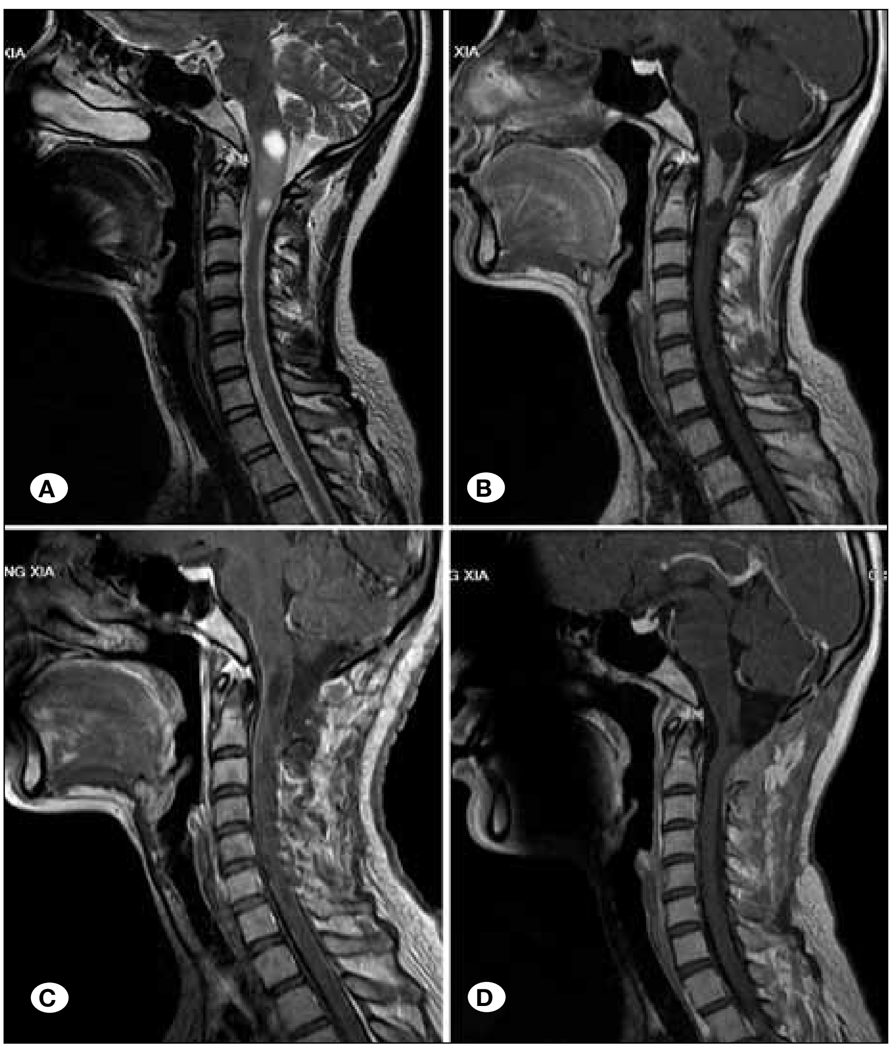

Figure 3: Pre- and postoperative MRI scans of patient No. 20.
Preoperative sagittal T2-weighted MRI **(A)** and T1-weighted imaging with contrast enhancement **(B)** showing the ependymoma from medulla oblongata rostrally to the second cervical vertebra caudally with a well-defined tumor boundary. Postoperative MRI scans with contrast enhancement at 1 week **(C)** and 6 months **(D)** illustrating total removal of the lesion (C) and no recurrence (D).

and 6 patients had STR. Therefore, the tumor border is a very important factor predicting if GTR could be achieved. In determining if the tumor boundary is clear, both T2-weighted MR images and contrast-enhanced T1-weighted MR images are important. A well defined boundary should meet at least one of the following two conditions: on T2-weighted MR images, there is a clear subarachnoid space between the tumor and normal cord; on contrast-enhanced T1-weighted MR images, the boundary line between the tumor and normal cord is smooth. Otherwise the boundary is ill defined. When most of the boundary was ill defined, even if some parts were well defined, the tumor boundary should be regarded as ill defined (14).

Besides, in our experience, it is not necessary to manage the syrinx associated with the ependymoma. We avoided entering or draining the syrinx during the course of resection because the syrinx would collapse after tumor removal (5). In our series, all the 15 patients with syrinx showed syrinx reduction after surgery.

Short-Term Outcomes

Generally speaking, a favorable functional outcome could be achieved in most patients. In the early postoperative course, it is reported that 9–67% of spinal ependymoma patients experienced neurological status aggravation (4, 23, 24, 28). In this series, 25 of the 28 patients (89.3%) had stable or improved mMG grades directly after surgery, and only 3 patients (10.7%) experienced acute neurological decline after surgery, but all these 3 patients returned to baseline within 1 month of surgery. The neurological decline found in this study is similar to that reported in other spinal ependymoma series. The protection of respiratory function is extremely important during operation. Three of our patients (10.7%) had respiratory difficulty after surgery, and underwent tracheostomy and required ventilator assistance. And all these 3 patients recovered their respiratory function before discharge. Every effort should be made to prevent aspiration and pneumonia for patients with lower cranial nerve deficits and respiratory difficulty, which might be fatal for these patients.

Regarding the transition of different preoperative symptoms, an interesting finding of this study is that neck pain was relieved in all the 13 patients (100.0%) in a short term after surgery, followed by headache (5 of 6, 83.3%), lower cranial nerve deficits (3 of 4, 75.0%), numbness of limbs (50.0%), dizziness

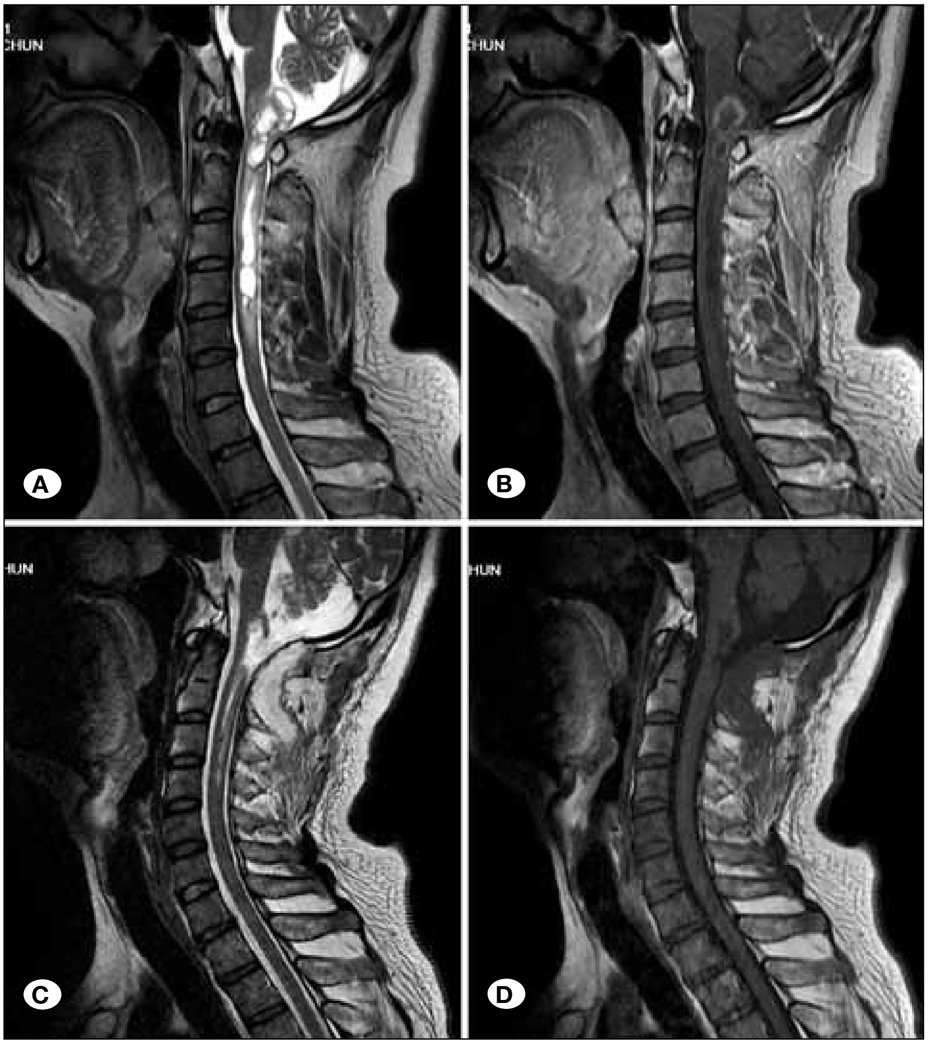

Figure 4: Pre- and postoperative MRI scans of patient No. 27. Preoperative sagittal T2-weighted MRI **(A)** and T1-weighted imaging with contrast enhancement **(B)** showing the ependymoma from medulla oblongata rostrally to the second cervical vertebra caudally with an ill-defined tumor boundary. MRI scans at one week after surgery illustrating subtotal removal of the lesion **(C, D)**.

(50.0%), and motor weakness (35.7%). Therefore, among the three most common preoperative manifestations (motor deficits, neck pain and numbness) for ICE, the improvement rate of neck pain was significantly higher than that of motor deficits (p=0.001), and numbness of limbs (p=0.005). In others words, for patients whose main preoperative symptoms were neck pain, a better clinical outcome could be expected directly after surgery.

Long-Term Outcomes

An excellent outcome was achieved in our series. After a mean follow-up duration of 49.6 months, compared to the preoperative mMG, the mMG at long-term follow-up improved in 89.3% of patients, stabilized in 10.7%, and no one deteriorated. The long-term neurological function was significantly better than that of the preoperative mMGs (p<0.001) and the mMGs at discharge (p<0.001). Therefore, our results showed that operation could lead to better neurological functions in the long run for ICE patients. Compared with previous studies, our series seemed to have a better outcome, as the rate of improvement and stabilization ranged from 81.6% to 95.5% in previous studies (2, 6, 11, 15, 16).

With respect to the transition of different symptoms, among the three most common preoperative manifestations, neck pain is also most likely to relieve (100.0%), followed by motor weakness (78.6%) and numbness of limbs (75.0%). Therefore, compared with neck pain, motor deficits and numbness are relatively difficult to recover.

Prognostic Factors

Most previous studies agree that GTR is a predictor for a better outcome (2, 3, 6, 17, 24). And some authors concluded that poor preoperative status is a significant predictor of a poor outcome (15). Li et al. indicated that the length of spinal ependymoma is a significant factor and patients with longer tumor had worse results (16). Besides, most authors believe adjuvant radiotherapy didn't confer a survival benefit (14, 17).

In our study, GTR is found to be a significant factor leading to a better function outcome in a short-term after surgery, which is similar to previous studies. The other factors are not significantly associated with clinical outcome, such as gender, age, syrinx and tumor length. However, as there is no patient with function deterioration in the long-term follow-up, the analysis of prognostic factor cannot be performed.

Study Limitations

The present study has several limitations. First, it is a retrospective research with limitations inherent to the study design. Second, because the rarity of ICE, a relatively small number of patients was enrolled. This limited the statistical analysis. Because of the small number of patients, we did not analyze the effect of histology (high grade and low grade) and postoperative radiotherapy on the outcomes.

■ CONCLUSION

Gross total resection of ICE could be performed in most patients, especially for those with well-defined tumor boundaries. GTR leaded to a better outcome in the short term after surgery when it is safe to conduct. A favorable functional outcome could be achieved for most ICE patients in the long term. Compared with motor weakness and numbness of limbs, neck pain has a greater possibility to improve after surgery.

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