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Surgical management of a huge intramedullary ependymoma from oblongata to T4: A case report

Dongao Zhang^{a,1}, Wayne Fan^b, Xingang Zhao^a, Cong Liang^a, Tao Fan^{a,*,1}^a Spine Center, Sanbo Brain Hospital, Capital Medical University, Beijing, People's Republic of China^b Faculty of Science, University of British Columbia, Canada

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ABSTRACT

INTRODUCTION AND IMPORTANCE: Huge intramedullary tumor is a rare condition. Surgical management of such huge ependymoma is technically challenging. We reported one case of 300 mm long intramedullary spinal cord ependymoma from oblongata to T4, which was satisfactorily en bloc gross total resected and the cervical spine alignment was successfully maintained by laminoplasty.

CASE PRESENTATION: A 30-year-old man presented to our service with two-year progressive neck pain combined with bilateral sense disorders of both the upper and lower extremities. The neurological grade was ASIA D. Repeat MRI imaging demonstrated a “medulla oblongata to T4” intramedullary spinal cord tumor complicated with cervical physiological curvature loss.

CLINICAL DISCUSSION: Laminectomies were made at the C1–T4 level by a milling cutter. The tumor was gently separated bilaterally. The ventral part of the tumor was separated from the cranial and the caudal ends until reaching the middle site of the tumor. The whole tumor was isolated en bloc. Titanium miniplates were used bilaterally to fix both sides of the laminoplasty section on each level. At the 2-year postoperative follow-up, the muscle strength of both the upper and lower limbs were grade IV. No tumor recurrence was detected.

CONCLUSION: The total removal of an intramedullary ependymoma should be recommended as soon as the diagnosis is made, regardless of its extension. Long-level laminoplasty could be an effective alternative option to maintain the proper spine alignment.

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1. Introduction

Intramedullary spinal cord tumors account for approximately 16% of the spinal tumors and approximately 2–4% of all central nervous system tumors [1]. With the administration of intraoperative neurophysiological monitoring and microsurgical techniques, surgeons can achieve more aggressive resection and have the potential for total excision of intramedullary spinal cord tumors. It is indicated that the prognosis of intramedullary ependymomas is closely related with the total resection rate [2,3]. Tumors that involve more than 5 vertebral segments can be defined as long-level intramedullary tumors [4]. In this condition, surgical treatment of such long-level intramedullary tumors becomes more complicated and challenging [5]. Herein, one case of 300 mm long intramedullary spinal cord ependymoma from oblongata to T4 is reported. The operation was performed by the corresponding

author, who had over 20 years of specialized surgical experiences. This report was guided by the Surgical Case Report (SCARE) guidelines [6].

2. Case presentation

A 30-year-old man presented to our service with two-year progressive neck pain combined with bilateral sense disorders of both the upper and lower extremities. The patient started experiencing difficulty of urination in the last 2 months. The neurological grade was ASIA D. Hyperreflexia, Romberg sign and Babinski signs were positive. Repeat MRI imaging demonstrated a “medulla oblongata to T4” intramedullary spinal cord tumor complicated with cervical physiological curvature loss (Fig. 1a–d). No relevant drug history, family history or psychosocial history was found.

The patient was willing to accept the operation for tumor resection. Through a midline incision, laminectomies were made at the C1–T4 level by a milling cutter. The interspinous ligaments were separated at the end of the laminoplasty section, leaving the interior supraspinal ligament and spinous processes intact, and the whole laminoplasty section was separated en bloc. After recognizing and incising the dorsal middle line of the spinal cord, the reddish-gray colored tumor was encountered. The tumor was gen-

* Corresponding author at: Spine Center, Sanbo Brain Hospital, Capital Medical University, NO.50 XiangshanYikesong Road, Haidian District, Beijing, People's Republic of China.

E-mail address: fant@ccmu.edu.cn (T. Fan).

¹ Dongao Zhang and Tao Fan contributed equally to the article and should be considered as the co-first authors.

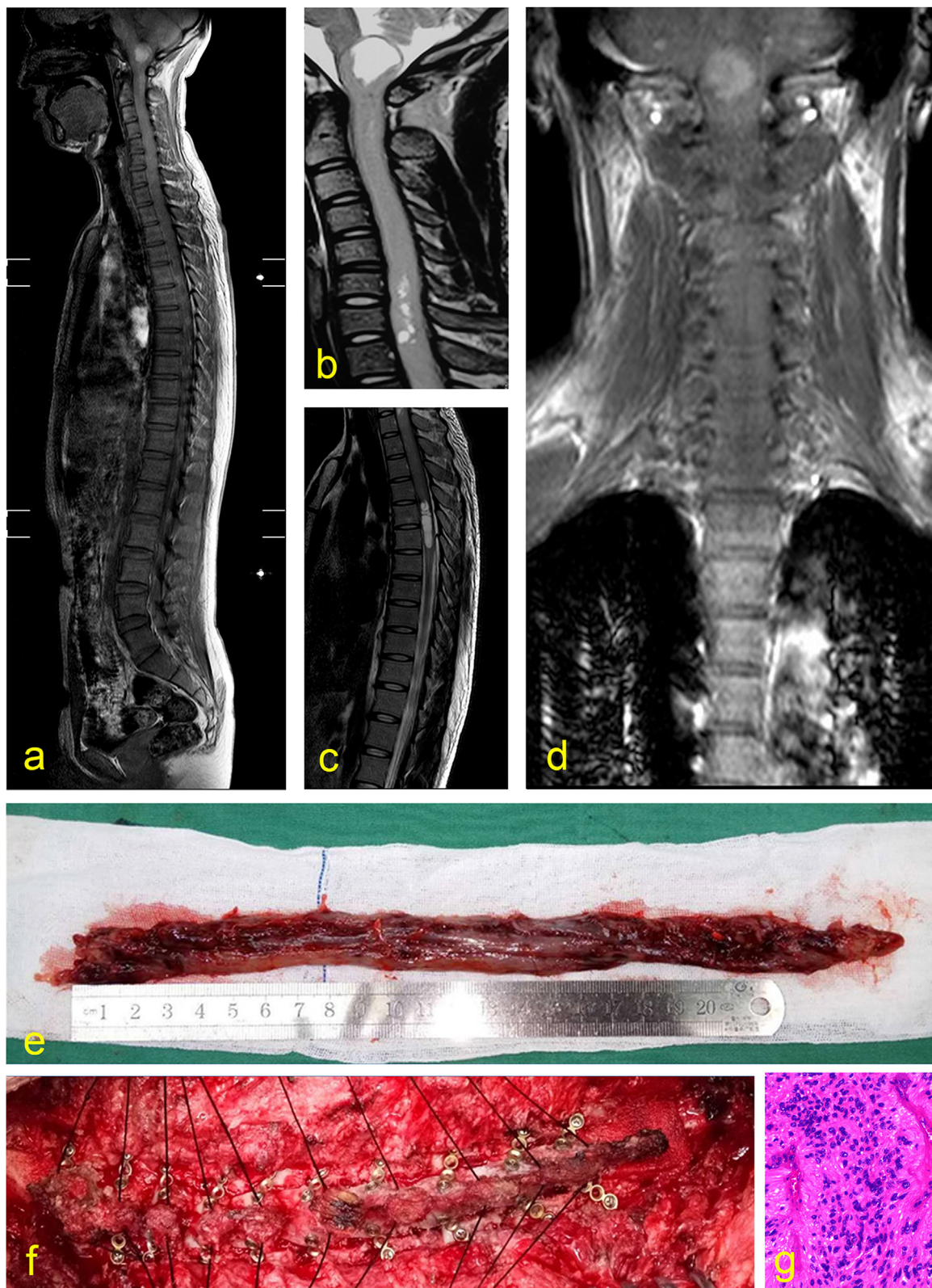


Fig. 1. A 30-year man diagnosed with intramedullary spinal cord tumors. MRI imaging demonstrated a “medulla oblongata to T4” intramedullary spinal cord tumor complicated with cervical physiological curvature loss (a–d). Intraoperative image of the excision of the tumor. Size 300 mm long (e). Laminoplasty with miniplates (f). Pathology images indicated an ependymoma (WHO II) (g).

tly separated bilaterally until reaching the cranial and caudal ends of the tumor. Then, the ventral part of the tumor was separated from the cranial and the caudal ends until reaching the middle site of the tumor. The whole tumor was isolated en bloc (Fig. 1e). The

spinal pia mater was discontinuously sutured and the dura was watertight sutured. Titanium miniplates were used bilaterally to fix both sides of the laminoplasty section on each level (Fig. 1f). The surgical wound closure was then completed. The whole operation

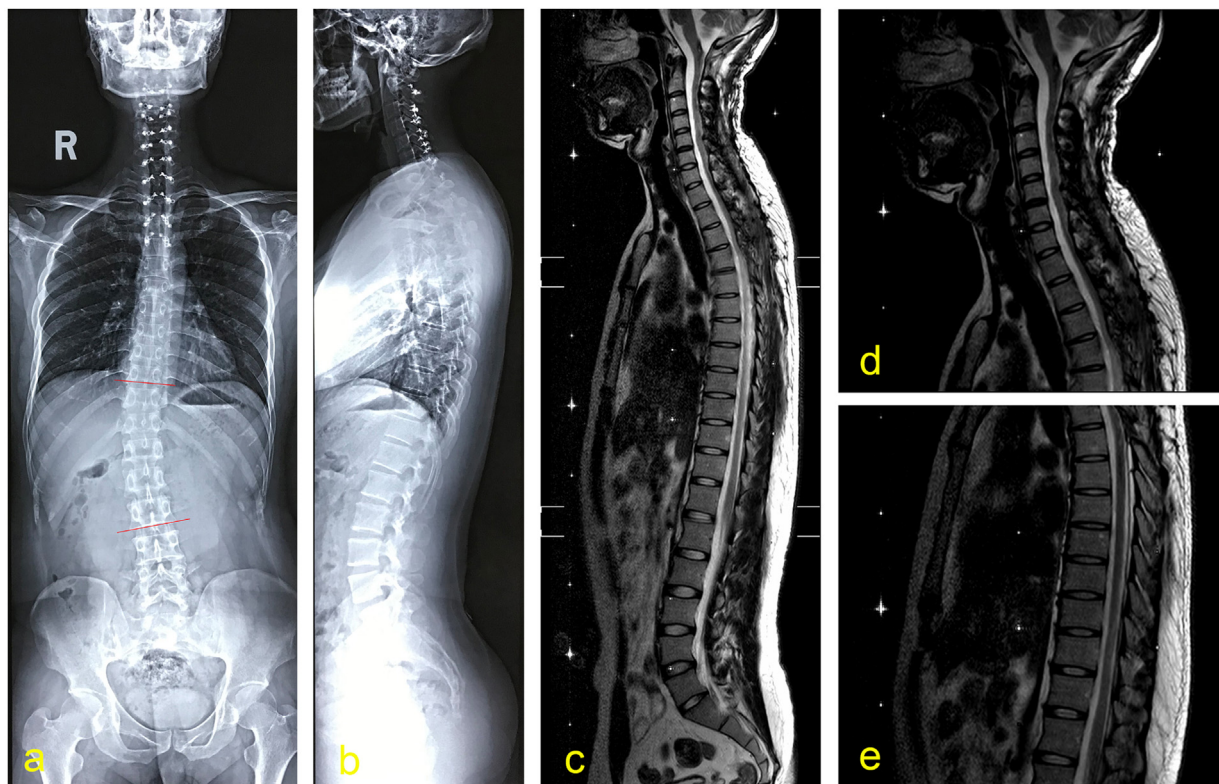


Fig. 2. Postoperative images at 2-year follow-up. X-ray images indicate a new emerging slight scoliosis, and the coronal Cobb angle was 14° (a–b). Sagittal MR images of the total spine showed satisfying tumor excision with no recurrence (c). Upper level of the T2 MRI (d). Lower level of the T2 MRI (e).

lasted 9 h, with approximately 2300 mL blood loss. Autotransfusion was 1000 mL, allogeneic erythrocyte transfusion was 400 mL, and allogeneic plasma transfusion was 200 mL. Somatosensory evoked potentials and motor evoked potentials were both used to monitor neurological function during the operation. The pathology results indicated ependymoma WHO II (Fig. 1g).

The first day after the operation, body examination revealed mild numbness of the trunk and extremities. The muscle strength of lower limbs was grade II, while the upper limbs measured grade IV. The neurological grade was ASIA C. After postoperative rehabilitation exercise, the muscle strength of the lower limbs had improved to grade IV at the latest 2-year follow-up, with no muscle strength loss of the upper extremities. However, the feeling of numbness had no obvious improvement. The sphincter function was normal. The neurological grade was ASIA D at the 2-year follow-up. X-ray revealed a newly emerging mild thoracic scoliosis. The coronal Cobb angle was approximately 14° in X-Ray (Fig. 2a–b). No tumor recurrence was detected (Fig. 2c–e). The patient was satisfied with the results.

3. Discussion

With complete removal of an intramedullary ependymoma, tumor recurrence is rare, and the patient can expect a good prognosis [7–9]. The preoperative status of the patient, the diameter and size of the tumor, and the skill and maturity of the surgeon could be the main factors affecting the postoperative outcomes of intramedullary ependymoma patients. Before the 1980s, giant intramedullary spinal cord tumors had been reported respectively by Cushing H in 1927, Horrax G in 1938, Thurel R in 1946, David H in 1964, Laine E in 1965, and Fischer G in 1971 and 1973 [10]. However, those diagnoses were based on lumbar myelography. In 2015, Unverdi Z. et al. reported a huge intramedullary ependymoma at the T4–L2 levels [11]. The patient was diagnosed by biopsy

but refused further surgery. Our case in this report was a 300 mm long intramedullary ependymoma that was diagnosed by MRI and was satisfactorily en bloc gross total resected. We recommended that the spinal cord should be opened over the entire tumor as the first step of the dissection. This management can largely minimize the traction of the spinal cord during tumor resection and facilitate the separation between tumor and spinal cord tissue. This report demonstrated that the total removal of an intramedullary ependymoma should be recommended as soon as the diagnosis is made, regardless of its extension.

Long-level laminoplasty could be an effective alternative option to maintain the proper spine alignment. It should be mentioned that this ependymoma patient showed a slight lower thoracic to lumbar scoliosis with a Cobb angle of 14° , which emerged far away from the operative section. This may be secondary to paravertebral muscle denervation, and the patient will be further followed up. During the reinsertion of the laminae, we recommended a seamless connection of all the laminae at one side, which will promote bone fusion at one side of the laminae and may also facilitate the stability of the spine. Even after reinsertion of the lamina, atrophy and abnormal innervation of neck and back muscles may cause muscular imbalance, which may be sufficient to induce spinal instability despite reinserted laminae. However, reinsertion of the laminae could avoid stiffness of the neck and back muscles caused by an epidural scar and could also make a reoperation easier in case of tumor recurrence. Reinsertion of the laminae in all patients with miniplates after removal of the tumor to restore the anatomy as far as possible is quite reasonable to keep the spinal alignment [12].

4. Conclusion

The total removal of an intramedullary ependymoma should be recommended as soon as the diagnosis is made, regardless of its

extension. Long-level laminoplasty could be an effective alternative option to maintain the proper spine alignment.

Declaration of Competing Interest

The authors report no conflict of interest.

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Ethical approval

The paper is a case report, and therefore does not require ethics approval.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Dongao Zhang: Data curation and Writing - original draft.
Wayne Fan: Investigation.
Xingang Zhao: Methodology.
Cong Liang: Writing - review.
Tao Fan: Conceptualization, Funding acquisition, Supervision.

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Not applicable.

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