doi: 10.2176/nmccrj.cr.2021-0086

NMC Case Report Journal 8, 705–711, 2021

Postoperative Cervicothoracic Kyphosis Following Infantile Intramedullary Tumor Resection Accelerates Neurological Deterioration

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Abstract

Intramedullary spinal cord tumors are rare in children. Regardless of the type of tumor, surgical removal is thought to improve progression-free survival. However, postoperative kyphosis is a serious problem in children, who can expect long-term survival. We present a pediatric case of neurofibromatosis type 2-related spinal ependymoma at the cervicothoracic regions where acute neurological deterioration was developed due to a combination of tumor recurrence and postoperative kyphotic deformity. In the first surgery, subtotal tumor resection was performed via osteoplastic laminotomy. Postoperative radiological evaluation at several months showed cervicothoracic junctional kyphosis, which subsequently made a significant improvement by lifestyle instructions. However, 22 months after the surgery, he exhibited rapid neurological deterioration caused by the regrowth of the recurrent tumor and re-emergence of kyphotic deformity, which led to the fixed laminar flap sank into the spinal canal. Therefore, a second surgery was performed 23 months after the first surgery, and gross total removal was achieved. Osteoplastic laminotomy is presumed to reduce the occurrence of postoperative kyphosis compared with laminectomy, but there have been no reports on the spinal cord compression by plunging of the re-fixed laminar flap into the spinal canal. The kyphosis deformity increases the chance of re-fixed laminar flap coming off, thereby accelerating neurological injury on top of the neural damage by tumor recurrence itself. Therefore, pediatric patients with spinal cord tumors should be carefully managed in terms of recurrent tumors and postoperative kyphosis, and timely surgical intervention is necessary before kyphotic deformity becomes evident.

Keywords: osteoplastic laminectomy, postoperative deformity, postoperative kyphosis, intramedullary spinal cord tumors, ependymoma

Introduction

Primary spinal cord tumors are very rare for the pediatric age group with an overall incidence rate of 0.26 per 100000 person-year.[1)] However, spinal cord tumors are commonly prevalent and present in approximately half of all patients with neurofibromatosis type 2 (NF2).[2–5)]

The development of postoperative spinal deformity, primarily post laminectomy kyphosis, particularly in the pediatric population is a relatively common problem, with reported rates between 24% and 100%.[6,7)] Progression of spinal deformity leads to adverse symptoms, such as pain, numbness, weakness, pulmonary complications, and negative physiological responses.[8)] Osteoplastic laminotomy has been developed to prevent postoperative spinal deformity.[6,9,10)] This procedure involves en bloc removal of spinous processes and laminae with the ligaments (interspinous ligaments and ligamentum flavum) and refixing them by titanium plates after the tumor removal.

Received March 20, 2021; Accepted July 26, 2021

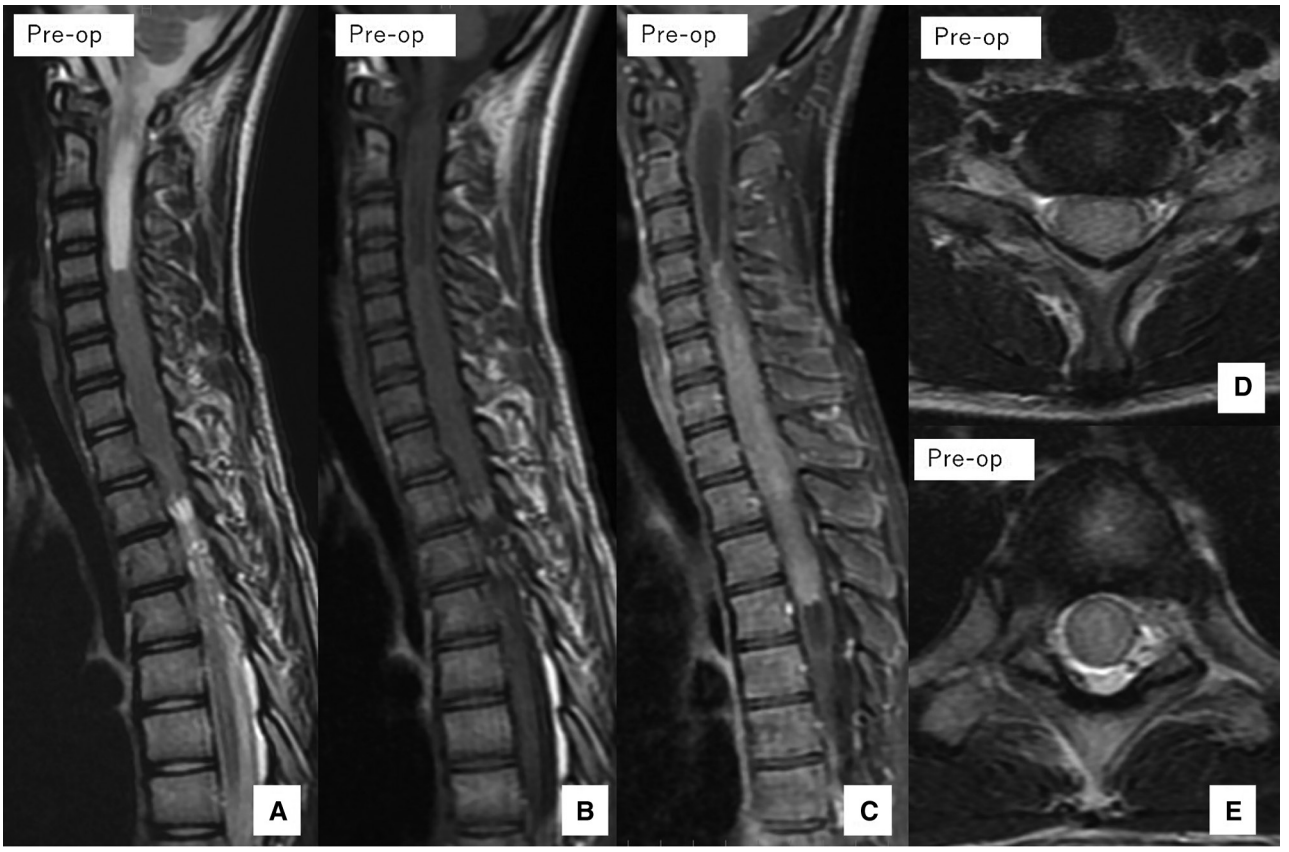

Fig. 1 Sagittal plane of MRI showing an intramedullary mass lesion associated with the syrinx extending above and below the tumor from C4 to Th4. (A) A T2-weighted MRI showing that the tumor is iso-intense. (B) A T1-weighted MRI showing that the tumor is slightly hypo-intense. (C) A gadolinium-enhanced T1-weighted MRI showing homogenous tumor staining. Axial image T2-weighted MRI at C6–C7 (D) and Th3–4 (E) showing well-demarcated central intramedullary tumor.

We report a pediatric case of an NF2-related spinal ependymoma at cervicothoracic regions where acute neurological deterioration was developed due to a combination of postoperative spinal deformity and recurrence of the tumor following tumor removal via osteoplastic laminotomy. The fixed laminar flap sank into the spinal canal because of postoperative kyphotic deformity at the cervicothoracic junction, by which neurological symptoms caused by tumor recurrence was additionally aggravated. The patient was successfully treated with re-do total removal of the recurrent tumor via laminectomy. We discuss the careful management of postoperative spinal deformity particularly observed in pediatric patients following the removal of spinal tumor.

Case Presentation

An 11-year-old boy was referred to our hospital with a 4-month history of progressive neck pain, bilateral arm weakness, hand clumsiness, and gait disturbance. He had a known history of NF2 with bilateral acoustic schwannomas. Physical examination revealed weakness of the upper and lower limbs, numbness or insensitivity below the Th4 level, and bladder and bowel movement dysfunction. The muscle strength of both the lower arms and lower limbs was of grade 4/5. Hyperreflexia and pathological reflexes were positive in bilateral arms and legs. The modified McCormick Scale[11,12] was grade III. Cervical and thoracic spine magnetic resonance imaging (MRI) showed an intramedullary mass lesion ranging from C4 to Th4 (Fig. 1A–1E). The tumor was located centrally within the spinal cord and was associated with the syrinx that extended above and below the tumor. The tumor was isointense on T2-weighted MRI (Fig. 1A, 1D, and 1E) and slightly hypointense on T1-weighted MRI (Fig. 1B). Gadolinium-enhanced T1-weighted MRI showed homogeneous tumor staining (Fig. 1C). His cervical spine was lordotic curvature of 6 degrees. The tumor was removed via the midline posterior approach. Laminotomy from C4 to Th4 was performed by separating muscle and ligamentous attachments from the spinal arches leaving the periosteum and

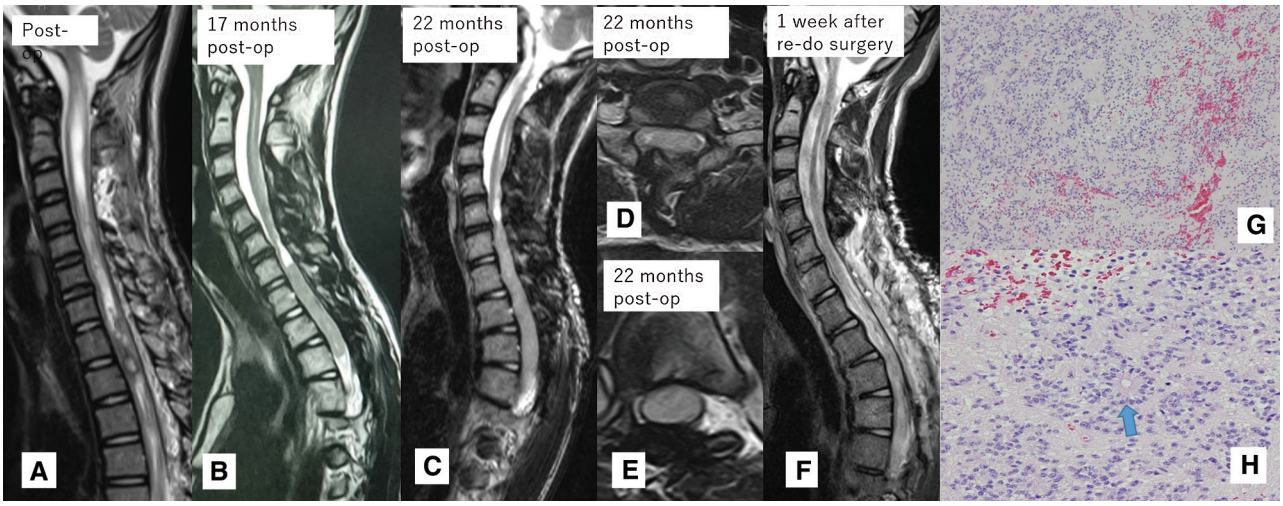

Fig. 2 (A) Postoperative T2-weighted MRI showing a small amount of residual tumor. (B) A T2-weighted MRI taken 17 months after the first surgery showing that the tumor has grown back from C7 to Th3. (C) A T2-weighted MRI taken 22 months after the first surgery showing enlargement of the recurrent tumor. (D and E) Operated laminar and spinous processes sinking into the spinal canal and compressing the spinal cord at C6–C7 (D) and Th3–4 (E). (F) A T2-weighted MRI taken after the second surgery showing no residual tumors. (G and H) Histopathological examination showing ependymal rosettes and perivascular pseudorosettes, without mitoses, vascular proliferation, or necrosis (hematoxylin and eosin, G: original magnification ×100, H: original magnification ×200). The tumor had a low Ki-67 index (up to a maximum of 8% in focal areas) and was classified as an ependymoma, WHO grade II. WHO: World Health Organization.

interspinous ligaments intact. Laminar osteotomy was done with a high-speed drill just medial to an imaginary line separating the lamina from the lateral mass, and the laminar flap was temporarily removed. After opening the dura, the gray-colored tumor was identified under a surgical microscope. Subsequently, the tumor was almost resected, but we found that a small part of the tumor was left behind in the floor of the tumor cavity because it appeared to adhere to the spinal cord parenchyma as well as to the small vessels branching from the anterior spinal artery. After the spinal dura was sutured watertight, the laminar flap was brought into its anatomical position and fixed with titanium plates and screws at bilateral C5, Th4, left C7, Th2, and right Th1 levels. Completing the reconstruction of the laminar flap, interspinous ligaments were adapted and muscle insertions reconstructed segmentwise. Postoperative T2-weighted imaging revealed a small amount of residual tumor (Fig. 2A). Histological analysis revealed ependymal rosettes and perivascular pseudorosettes, without mitoses, vascular proliferation, or necrosis. The tumor had a low Ki-67 index (up to a maximum of 8% in focal areas) and was classified as ependymoma (World Health Organization [WHO] grade II) (Fig. 2G and 2H). After surgery, the ambulatory disorder recovered to modified McCormick Scale Grade II due to the rehabilitation. He was discharged with his neck immobilized in a cervical soft collar for 4 weeks to minimize pseudarthrosis and postoperative kyphosis. However, he started to develop kyphotic deformity at the cervicothoracic junction and compensated cervical lordosis several months after surgery because he spent a lot of time in doing smartphone game with his neck strongly flexed (Fig. 3B). He was strictly instructed to wear the cervical collar again and refrain from using a smartphone. His cervical spine alignment dramatically improved following the instruction (Fig. 3C and 3D) and neurological symptoms recovered in parallel. He remained stable neurologically without kyphosis progression until 20 months postoperatively even as the tumor grew back. However, his ambulation, bilateral leg weakness, and bladder and bowel movements rapidly became exacerbated to the level of modified McCormick Scale Grade IV at 22 months postoperatively as the tumor and associated cyst quickly grew to a considerable size and kyphosis at the cervicothoracic junction obviously progressed because of back muscle weakness (Fig. 2A–2E). He was completely confined to a wheelchair with progression of urinary retention. His condition was much worse than 2 years prior despite the smaller size of the tumor. A CT scan showed that screws and plates came off the fixed operated laminar flap, which sank into the spinal canal from C6 to Th4 because of kyphotic deformity at the cervicothoracic junction (Fig. 4). Finally, a repeat surgery was

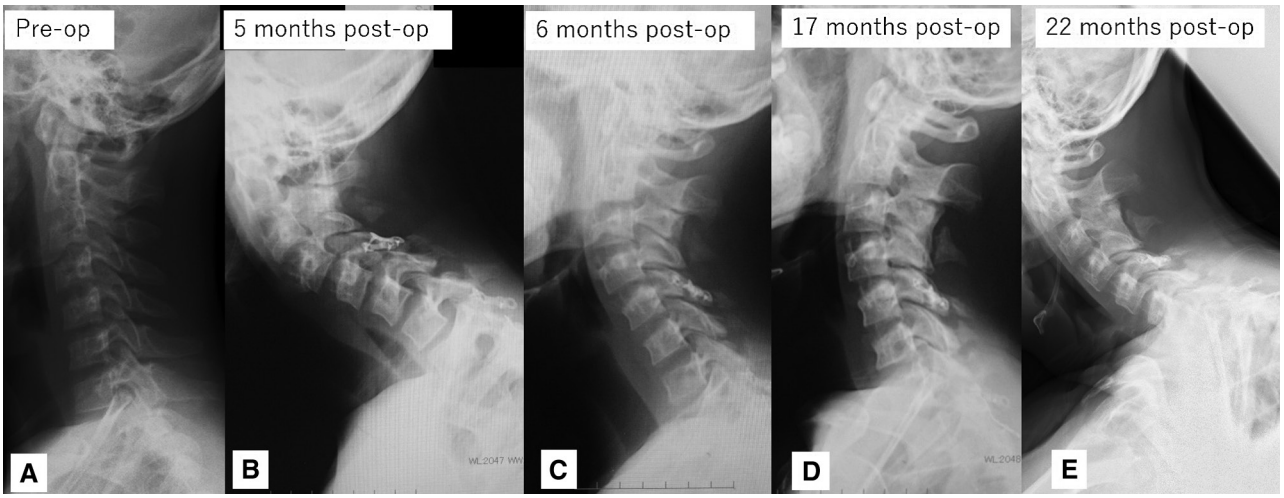

Fig. 3 Cervical spine radiograph. (A) A radiograph taken preoperatively did not show any signs of kyphosis. (B) He developed severe cervical spine kyphosis 5 months after surgery. (C) A radiograph taken 6 months after the first surgery showing improvement of kyphosis at the cervicothoracic junction brought by placement of cervical collar and lifestyle guidance. (D) A radiograph taken 17 months after the first surgery showing compensatory lordotic curvature of cervical spine with mild cervicothoracic junctional kyphosis. (E) A radiograph taken 17 months after the first surgery showing re-emergence of severe kyphotic deformity secondary to back muscle weakness caused by tumor recurrence.

performed 23 months after the first surgery. The operated laminar flap was attached to the scar tissues over the spinal canal. The screws and plates were removed, and the sinking laminar flap was drilled out under the microscope with great care to avoid applying a compression force. The scar tissue was carefully removed, and the dura was incised. A gross total resection of the tumor was performed including the portion that was left in the first surgery via re-do midline myelotomy because the tumor was successfully separated from the spinal cord parenchyma with meticulous dissection. Postoperative T2-weighted imaging showed no residual tumor (Fig. 2F). The pathological result was ependymoma (WHO grade II), with no malignant transformation. The patient is in an excellent physical condition and recovering to modified McCormick Scale Grade III with no progression of kyphotic deformity with cervical immobilization by the cervical soft collar at four-month postoperatively. He is strictly following our lifestyle guidance to prevent spinal deformity, such as doing back muscle training and prohibition of smartphone games with his neck flexed in a sitting position.

Discussion

There are few reports on the long-term outcomes of intramedullary tumors in pediatric population. Tumor malignancy and tumor excision rate are significantly associated with long-term survival[13,14]; therefore, early therapeutic interventions is recommended for extension of overall survival.[13–17] However, postoperative spinal deformity following pediatric intramedullary tumor resection is another serious problem, particularly if long-term survival can be expected by appropriate management of the tumor. Ahmed et al. reported that 20 of 55 cases with intramedullary tumors showed postoperative progression of spinal deformity, of which 55% (11/20) required instrumented spinal fusion.[18] The risk factors for postoperative spinal kyphosis were cervical spine tumors, pre-existing kyphoscoliosis, and laminectomy/laminoplasty at more than four levels as well as pediatric population. Nori et al. described the horizontal cervical spine facets, the lax ligamentous structures, and the growing vertebral column as anatomical characteristics of pediatric populations leading to postoperative spinal deformity.[19] Furthermore, pediatric patients with large tumors in the cervical spine like the present case tend to have neck muscle weakness even before surgery, which could be another risk of postoperative deformity.

Osteoplastic laminotomy, which was first described by Raimondi et al. in 1976,[9] is the procedure of refixing the laminar flap with supra- and interspinous process ligaments to its anatomical position. This procedure is presumed to preserve the posterior elements effectively and to maintain spinal stability. While Papagelopoulos et al. reported that 33% of pediatric patients undergoing

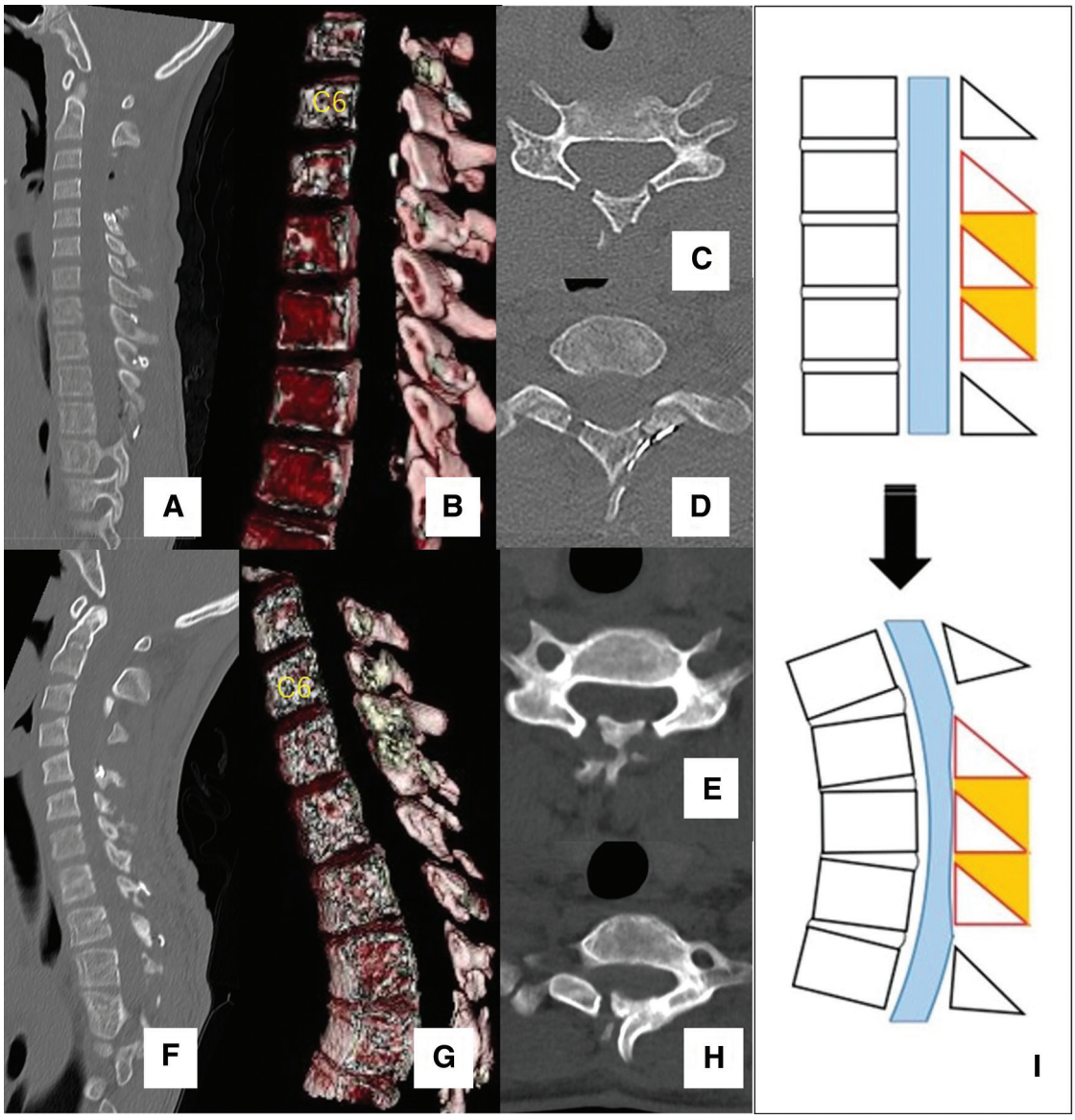

Fig. 4 CT scans of the spine on the day after the first surgery showing the osteoplastic laminotomy, where the laminae and spinous processes with attaching ligaments were excised en bloc and placed back in with titanium plates, following tumor removal. (A–D) CT image before the second surgery showing kyphotic deformity at the cervicothoracic spine and operated laminae and spinous processes sinking into the spinal canal and compressing the spinal cord. (E–I) This is a schema of the sagittal image of the spine. The kyphotic deformity caused the re-fixed laminar flap (red: spinal process and laminar, orange: posterior ligaments) to come off the residual laminar around 5 months postoperatively as a result of several months of kyphosis. While kyphosis made a significant improvement by placement of cervical collar and lifestyle guidance, plunge of laminar flap into the spinal canal caused by re-emergence of kyphotic deformity accelerated neurological deterioration combined with tumor recurrence.

laminectomy for intramedullary spinal tumors developed spinal deformity,[20] McGirt et al. described that osteoplastic laminotomy significantly decreased the incidence of spinal deformity progression from 30% to 5%.[6] Several other studies have also recommended osteoplastic laminotomy for pediatric cases.[18,21]

However, osteoplastic laminar flaps plunged into the spinal canal and compressed the spinal cord in the present case, which has never been reported to date. Raimondi et al. and other authors used high-speed drill to remove the laminar flap,[9,10] but there can be a small gap between the laminar flap and the residual laminar. Although T-saw or osteotome

is an ideal tool to reduce the gap,[6,22] there are some risks of spinal cord injury when the tool is inserted under the laminar under high spinal cord pressure like the present case. Considering the bone gap could delay bony union, attaching titanium plates and screws to all laminar or bone grafting to fill the gap may be necessary when the high-speed drill is adopted. Alternatively, by using myoarchitectonic spinolaminoplasty reported by Kim et al., spinal canal enlargement can be achieved while preserving muscle tissue.[23] This method might decrease the chance of postoperative spinal deformity.

As shown in Fig. 4, the kyphotic deformity causes fixed plates and screws to come off and to sink into the spinal canal, which aggravates neurological symptoms combined with tumor recurrence. The kyphotic deformity is considered to be triggered by back muscle weakness as a result of surgical muscle damage and tumor regrowth. Given the present case exhibited rapid neurological deterioration corresponding to kyphosis progression, tumor should have been timely removed before back muscle weakness became clearly visible. Postoperative kyphosis in the present case was nicely corrected at 6 months postoperatively by a cervical collar and lifestyle instructions because of flexibility and reversibility in pediatric patients; however, spinal cord compression caused by re-emergence of kyphosis should have been expected as the plates and screws had probably come off around 5 months postoperatively. It is considered necessary to limit the range of motion with the cervical collar at least until the bony fusion of the laminar flap is observed. Progressive cervicothoracic junctional kyphosis also increases the chance of rapid neurological deterioration by the posterior shift of severely damaged spinal cord and could result in demyelination and neuronal loss in the anterior horn associated with both continuous mechanical compression and vascular changes.[24] Therefore, careful management and early therapeutic intervention may be necessary once neurological symptoms become evident, even if the recurrent tumor is relatively small.

There is another option to perform concurrent spinal instrumented fusion; however, there are associated risks of postoperative complications such as prolonged surgery, surgical site infections, postoperative limited range of motion, and adjacent segment disease. Furthermore, metal implants produce artifacts and hinder the visibility of MRI, which makes early detection of recurrent tumors extremely difficult. Therefore, we recommend surgical intervention without instrumented fusion followed by cervical immobilization with a cervical collar and close follow-up with general lifestyle guidance.

Conclusions

Postoperative kyphotic deformity following intramedullary tumor removal in pediatric patients is a serious concern. Osteoplastic laminotomy is generally thought to reduce the risk of kyphosis; however, multi-level laminotomy for large tumors could be a risk factor in pediatric patients. Kyphotic deformity might lead to sinking of the operated laminae and accelerate neurological deterioration caused by recurrent tumors. When the tumor recurs and neurological symptoms appear in pediatric patients, early assessment of spinal alignment is necessary and careful follow-up should be conducted to prevent rapid deterioration.

Conflicts of Interest Disclosure

None.

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