Neurofibromatosis is often complicated by multiple spinal tumors such as schwannomas and neurofibromas. The C-2 nerve root frequently arises in such tumors. The C-2 nerve root tumors occupy the spinal canal compressing the spinal cord and nerve roots, causing various neurological symptoms. When patients with spinal cord tumors present with paralysis, tumors directly compressing the spinal cord are clearly observed on MRI. Goel et al. presented a series of 60 patients with C-2 nerve root tumors. Fifty-six of the 60 patients had unilateral C-2 nerve root tumors, whereas 4 patients had bilateral tumors. In 9 patients, including all 4 with bilateral tumors, there was evidence of NF. Bilateral C-2 tumors were often observed in patients with NF. In the present report, we describe 2 cases of NF presenting with cervicomedullary myelopathy. In both patients, conventional MRI of the cervical spine revealed tumors located at the bilateral lateral portion of the C1–2 interlaminar space without direct compression of the spinal cord. The spinal cord exhibited an I-shaped deformity at the same level as the tumors in one case and a trapezoidal deformity at the same level as the tumors in the other case. Computed tomography myelography and MRI on rotation of the cervical spine revealed bilateral intracanal protrusion of the tumors compressing the spinal cord from the lateral side. The tumors were successfully excised and occipitocervical fusion was performed.

The authors describe 2 patients with C-2 nerve root tumors in whom the lesions were located bilaterally in the lateral portions of the C1–2 interlaminar space and compressed the spinal cord when the atlantoaxial joint was rotated. The patients were adult men with neurofibromatosis. Each presented with clumsiness of both hands and motor weakness of the extremities accompanied by spastic gait. Magnetic resonance imaging of the cervical spine performed with the neck in the neutral position showed tumors at the bilateral lateral portion of the C1–2 interlaminar space without direct compression of the spinal cord. The spinal cord exhibited an I-shaped deformity at the same level as the tumors in one case and a trapezoidal deformity at the same level as the tumors in the other case. Computed tomography myelography and MRI on rotation of the cervical spine revealed bilateral intracanal protrusion of the tumors. The tumors were successfully excised and occipitocervical fusion was performed.

The tumors were pushed out into the spinal canal from the bilateral lateral portion of the interlaminar spaces due to rotation of the atlantoaxial joint. This was caused by a combination of postero-medial displacement of the lateral mass on the rotational side of the atlas and narrowing of the lateral portion of the interlaminar space on the contralateral side due to the coupling motion of the lateral bending and extension of the atlas. The spinal cord underwent compression from both lateral sides in a one-way rotation. Without sustained spinal cord compression, intermittent long-term dynamic spinal cord compression from both lateral sides should induce a pathognomonic spinal cord deformity and the onset of paralysis. To the authors’ knowledge, there have been no reports of the present conditions—that is, the bilateral protrusion of tumors from the bilateral lateral portion of the C1–2 interlaminar spaces into the spinal canal due to atlantoaxial rotation.

**Key Words**
- spinal cord tumor
- dumbbell tumor
- neurofibromatosis
- atlantoaxial joint
- oncology

**Abbreviations used in this paper:** CTM = CT myelography; JOA = Japanese Orthopaedic Association; NF = neurofibromatosis.

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Case Reports

Case 1

History and Examination. In 2002 this 58-year-old man with NF Type 1 began to experience an unstable gait. Since early 2008, he had felt numbness and clumsiness of both hands and gait disturbance that rapidly progressed. He therefore visited our clinic. Neurological examination revealed lower-extremity muscle weakness. Tendon reflexes of the upper and lower extremities were accelerated bilaterally. We established a diagnosis of myelopathy of the upper cervical spine. The patient’s JOA score for cervical myelopathy\(^4\) was 8 (combined score of 2, 1, 1, 1, 1, and 2) of a possible 17 points. The symptoms did not change with flexion, extension, or rotation of the neck. Plain lateral radiography of the cervical spine showed no spinal canal stenosis or instability. Midsagittal T2-weighted MRI performed with the patient in the neutral position showed spinal cord swelling with a higher signal intensity region at C1–2. Axial MRI revealed tumors at the bilateral lateral portion of the C1–2 interlaminar space and slight indentation of the dural tube. Although there was adequate subarachnoid space around the spinal cord, the cord exhibited an “I”-shaped deformity at C1–2 (Fig. 1). The spinal cord was not compressed directly and seemed to show atrophy. We performed MRI and CTM under flexion, extension, and rotation of the cervical spine. The flexion and extension MRI and CTM studies did not show any spinal cord compression. However, the rotation MRI and CTM studies revealed bilateral intracanal protrusion of the tumors compressing the lateral side of the spinal cord (Fig. 2). We considered that the paralysis and spinal cord deformity were caused by protrusion of the tumors from the bilateral lateral portion of the C1–2 interlaminar space into the spinal canal following rotation of the cervical spine.

Operation. We excised the tumors of the C1–2 interlaminar space following resection of the C-1 posterior arch and posterior fusion (occiput–C3) with segmental instrumentation. The tumors seemed to originate from the C-2 nerve root ganglion, and they did not adhere to the dura mater. The tumors were excised in a piecemeal manner. Histologically the tumors were determined to be neurofibromas.

Postoperative Course. At 4 years after surgery, the patient had improved neurologically, and his JOA score had increased to 13. Bone fusion was complete 1 year postoperatively. Magnetic resonance images showed restoration of the spinal cord shape with enlargement of the higher signal intensity region on T2-weighted sequences. Tumors at the bilateral lateral portion of the C1–2 interlaminar space had not recurred (Fig. 3).

Case 2

History and Examination. In early 2008 this 32-year-old man with NF Type 1 experienced numbness and clumsiness in both hands and gait disturbance. He visited our clinic. Neurological examination revealed upper-extremity muscle weakness. The triceps tendon reflex and knee and ankle jerks were accelerated bilaterally. We made a diagnosis of cervical myelopathy in the upper cervical region. The patient’s JOA score was 13 (combined score of 3, 3, 1, 2, 1, and 3). The symptoms did not change when the patient moved his neck. Plain lateral radiography showed no unusual findings in the cervical spine. Sagittal T2-weighted MRI and CTM studies at C1–2 showing an “I”-shaped deformity of the spinal cord despite adequate subarachnoid space around the spinal cord.

Fig. 1. Case 1. A: Sagittal T2-weighted MR image taken in the neutral position showing spinal cord swelling with a higher signal intensity region at C1–2. B: Axial T2-weighted MR image at C1–2 showing tumors at the bilateral lateral portion of the C1–2 interlaminar space (asterisks) and slight indentations of the dural tube. B and C: Axial T2-weighted MRI and CTM studies at C1–2 showing an “I”-shaped deformity of the spinal cord despite adequate subarachnoid space around the spinal cord.
MRI with the patient’s neck in the neutral position showed spinal cord swelling and a higher signal intensity region at C1–2. Axial MRI demonstrated tumors in the bilateral lateral portion of the C1–2 interlaminar space and adequate subarachnoid space around the spinal cord. Although the spinal cord was not compressed directly, it showed a trapezoidal deformity at C1–2 (Fig. 4). Both MRI and CTM performed on rotation of the cervical spine revealed bilateral intracanal protrusion of the tumors with direct lateral compression to the spinal cord (Fig. 5). We determined that the patient’s paralysis and spinal cord deformity were induced by the protrusion of tumors into the spinal canal.

**Operation.** We performed tumor excision and occiput–C3 posterior fusion. The tumors seemed to originate from the C-2 nerve root ganglion. Most of the tumors were excised in a piecemeal manner. The histological diagnosis of the tumors was neurofibroma.

**Postoperative Course.** At 4 years after surgery, the patient showed an excellent neurological improvement, and his JOA score increased to 16. Magnetic resonance imaging demonstrated restoration of the spinal cord shape. The tumors did not recur during follow-up (Fig. 6).

**Discussion**

We have reported that spinal dumbbell tumors accounted for 18% of all spinal cord tumors. The most frequent originating nerve root is C-2, which passes through the lateral portion of the interlaminar space between the C-1 posterior arch and the C-2 lamina. It is necessary to recognize that tumors located at the bilateral lateral portion of the C1–2 interlaminar space may protrude into the spinal canal on rotation of the cervical spine, even though the tumors are not observed in the spinal canal in the neutral position. On the occasion, the spinal cord is compressed from the lateral side by a protruding tumor, so that the spinal cord shows the characteristic form of an I-shaped elongation in the anteroposterior direction. This is a clue to the diagnosis of this unusual pathological entity.

The lateral atlantoaxial joint consists of the lower articular surface of the lateral mass of the atlas and the upper articular surface of the axis. The rotational range of motion at the atlantoaxial joint is the largest in the spine. In a 3D kinetic analysis of the atlantoaxial joint, the atlas shows the coupling motion of right bending and extension to the axis during left rotation and the coupling motion of left bending and extension to the axis during right rotation. In the present cases, the tumors were in the bilateral lateral portion of the interlaminar space at C1–2. They were pushed out into the spinal canal from the bilateral lateral portion of the interlaminar spaces by the rotation of the cervical spine. This was caused by a combination

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**Fig. 3.** Case 1. Sagittal (A) and axial (B) T2-weighted MR images demonstrating restoration of the spinal cord shape and enlargement of the higher signal intensity region 4 years after surgery.

**Fig. 4.** Case 2. A: Sagittal T2-weighted MR image taken in the neutral position showing spinal cord swelling with a higher signal intensity region at C1–2. B: Axial T2-weighted MR image at C1–2 demonstrating tumors at the bilateral lateral portion of the C1–2 interlaminar space (asterisks). B and C: Axial T2-weighted MRI and CTM scans at C1–2 revealing a trapezoidal deformity of the spinal cord despite adequate subarachnoid space around the spinal cord.

**Fig. 5.** Case 2. Axial T2-weighted MRI and CTM images at C1–2 on rotation of the cervical spine showing bilateral intracanal protrusion of the tumors compressing the lateral side of the spinal cord (rotation to the right [A and C] and rotation to the left [B and D]). The arrowheads show the indentation of the dural tube.
of posteromedial displacement of the lateral mass on the rotational side of the atlas and narrowing of the lateral portion of the interlaminar space on the contralateral side due to the coupling motion. In other words, the spinal cord underwent compression from both lateral sides in a one-way rotation. The head and neck are considered to be oriented either to the left or right most of time. Without sustained spinal cord compression, intermittent long-term dynamic spinal cord compression can induce a pathognomonic deformity of the spinal cord and the onset of paralysis. To our knowledge, there have been no reports of the present conditions—that is, bilateral protrusion of tumors into the spinal canal from the bilateral lateral portion of the C1–2 interlaminar spaces due to atlantoaxial rotation.

Kokubun\textsuperscript{6} has reported that 30% of patients with dumbbell tumors originating from C-2 nerve roots present with electric-like shocks extending down the trunk that are triggered by rotation of the neck, and 10% of these patients present with transient muscle weakness of the arm. Therefore, we chose occipitocervical fusion to excise the tumors. We thought that the Magerl screw technique and the posterior C-1 lateral mass–C2 pedicle screw fixation technique were also not feasible because of C-1 posterior arch resection necessary for one-way rotation. The head and neck are considered to be oriented either to the left or right most of time. Without sustained spinal cord compression, intermittent long-term dynamic spinal cord compression can induce a pathognomonic deformity of the spinal cord and the onset of paralysis. To our knowledge, there have been no reports of the present conditions—that is, bilateral protrusion of tumors into the spinal canal from the bilateral lateral portion of the C1–2 interlaminar spaces due to atlantoaxial rotation.

The excision of the tumors alone or in combination with atlantoaxial fusion should be performed. Excision of the tumors may be incomplete because of excess bleeding, or the tumors in NF may regenerate over many years. Some improvement of the paralysis over the long term would be expected by adding atlantoaxial fusion. There are several methods available for atlantoaxial fusion. Posterior fusion techniques including the use of Gallie wires, Brooks wires, and Halifax interlaminar clamps were unequable because of the bone erosion of the atlas and the upper part of the axis. Therefore, we chose occipitocervical fusion to maintain the frail spinal cord in complete repose for an extended period. Both patients showed an excellent improvement after surgery. On the other hand, we must take the difficulty of reoperation into consideration when the residual tumors grow and paralysis recurs.

Conclusions

We described 2 cases of C-2 nerve root tumors located in the bilateral lateral portion of the C1–2 interlaminar space and compressing the spinal cord on rotation of the cervical spine. It is important to recognize that tumors in the bilateral lateral portion of the C1–2 interlaminar space can protrude into the spinal canal by rotation of the atlantoaxial joint, even though the spinal cord is not compressed in the neutral position.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Ozawa, Kusakabe, Aizawa, Nakamura. Acquisition of data: Ozawa, Kusakabe. Analysis and interpretation of data: Ozawa, Kusakabe. Drafting the article: Ozawa, Kusakabe. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Ozawa. Study supervision: Ozawa, Kusakabe, Ishii, Itoi.

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