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CASE REPORT

Five-level cervical corpectomy for neurofibromatosis-associated spinal deformity: case report

Scott L. Parker · Jean Paul Wolinsky · Anthony P. Tufaro · Ziya L. Gokaslan · Timothy F. Witham

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Abstract
Purpose To describe a successful five-level cervical corpectomy and circumferential reconstruction in a patient with a plexiform neurofibroma causing a severe kyphotic deformity.
Methods Case report.
Results 43-year-old man with history of Neurofibromatosis presented with signs and symptoms of myelopathy with spastic lower extremities and gait difficulties. Imaging studies demonstrated a severe kyphotic deformity of the cervical spine with associated cord compression secondary to an anteriorly positioned plexiform neurofibroma. Two-stage surgical procedure was designed to treat this lesion. Stage I consisted of tracheostomy placement, transmandibular, circumglossal approach to the anterior cervical spine, C2–C6 corpectomies, and C1–C7 reconstruction with a custom titanium cage/plate. Stage II consisted of suboccipital craniectomy, C1–C2 laminectomies, and occipital-cervical thoracic instrumented fusion (O-T8). There were no operative complications, but the patient did develop a small pulmonary embolism post-operatively treated with anticoagulation. Patient required two-weeks of inpatient rehabilitation following surgery. Gastrostomy tube and tracheostomy were successfully discontinued with preserved swallowing and respiratory function. Patient-reported outcome measurements revealed significant and sustained improvement post-operatively.
Conclusions Five-level cervical corpectomy including C2 can be safely and successfully performed via a transmandibular, circumglossal approach. Circumferential reconstruction utilizing a custom anterior titanium cage and plate system manufactured from a pre-operative CT scan was utilized in this case. Long segment occipital-cervical-thoracic reconstruction is recommended in such a case. Using such a technique, improvement in myelopathy, correction of deformity, and improved quality of life can be achieved.

Keywords Cervical deformity · Corpectomy · Transmandibular, circumglossal approach · Neurofibromatosis

S. L. Parker (✉)
Department of Neurosurgery, Vanderbilt University Medical Center, Nashville, TN, USA
e-mail: slparker7@gmail.com

J. P. Wolinsky · Z. L. Gokaslan · T. F. Witham
Department of Neurosurgery, The Johns Hopkins Hospital, 600 N. Wolfe Street, Meyer 7-109, 21287 Baltimore, MD, USA
e-mail: twitham2@jhmi.edu

A. P. Tufaro
Department of Surgery, The Johns Hopkins Hospital, Baltimore, MD, USA

Introduction

Severe deformities of the cervical spine remain a challenging pathology to manage, regardless of etiology. Successful surgical outcomes are the result of extensive pre-operative planning, adequate decompression, spinal reconstruction using instrumented constructs tailored to the specific pathology, and ultimately bony fusion. Cervical corpectomy in the setting of multilevel myelopathy allows for extensive neurologic decompression and has been shown to be associated with excellent clinical results [1]. Reports of cervical corpectomies extending over as many as four levels have been reported in the literature [2–4]. There have also been reports of six-level corpectomy that extend across the cervico-thoracic junction [5, 6].

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However, five-level corpectomy in the cervical spine has not been reported in the literature to date. Such extensive surgical reconstruction can be associated with a high risk of early and late complications; in particular, hardware failure is an important possible early complication and pseudoarthrosis a potential late risk following multi-level cervical corpectomy. We describe a successful five-level cervical corpectomy and circumferential reconstruction in a patient with a plexiform neurofibroma causing a severe kyphotic deformity.

Case report

History

A 43-year-old male with a known history of Neurofibromatosis type 1 presented with a three month history of progressive myelopathy. Symptoms included bilateral upper and lower extremity stiffness and balance difficulties, including a fall secondary to transient paralysis and numbness. The patient also reported dizziness, constipation, back pain, and neck pain, but denied radicular complaints. Past medical history was significant for asthma and parathyroid disease, both of which were well controlled.

Physical exam

Physical exam demonstrated mild weakness in bilateral upper extremities: deltoids 4+, right hand intrinsics 4+, and left hand intrinsics 4+ out of five motor strength. His left lower extremity demonstrated weakness with dorsiflexion 4+ and extensor hallucis longus three out of five motor strength. Spasticity was present in all four extremities and the patient walked with a spastic gait.

Imaging

Subsequent imaging studies revealed a severe kyphotic deformity of the cervical spine associated with C2–C7 cord compression secondary to an anteriorly positioned plexiform neurofibroma, Fig. 1. The neurofibroma measured $10.2 \times 5.3 \times 9.3$ cm (transverse $\times$ AP $\times$ craniocaudal), was centered within the left neck from the left mastoid process to the level of the thyroid cartilage, and completely encased the left internal carotid and vertebral arteries. The mass entered the cervical canal at multiple levels via left-sided neural foramina and caused remodeling of C2–C4 vertebral bodies. The kyphotic deformity was centered at C3–C4 with approximately 90° of angulation, Fig. 2.

Operation

A two-staged procedure was planned with an anterior approach first to resect the plexiform tumor and reduce the cervical kyphosis followed by a posterior approach to stabilize the occipital-cervical-thoracic spine.

Stage 1

The patient was intubated with awake, fiberoptic technique. Continuous motor- and sensory-evoked potential monitoring was employed during the case. He was positioned supine on the operating room table with his head placed in a halo ring with traction in extension to correct the deformity. A tracheostomy and exposure to the spine via a transmandibular, circumglossal approach was first performed [7]. A midline incision from the patient's lower lip through the submental area to nearly the right mastoid process was made. From the right neck, subcutaneous tissue and muscle flaps were elevated and the dissection continued anterior down to the patient's mandible. An osteotomy was performed just anterior to the mental nerve. The lesser segment of the mandible on the right side was retracted posteriorly while the tongue base was retracted to the left and inferiorly. Next a curvilinear incision was performed along the length of sternocleidomastoid muscle to gain exposure to the patient's cervical spine from C1 to C7. The esophagus was mobilized and retracted to the left and a 5 cm incision in the midline of the patient's posterior pharyngeal wall was made to clearly visualize the large plexiform neurofibroma. A large portion of the neurofibroma was resected to expose the anterior spine from C1 to T1. A gross total resection was not planned.

Next a high-speed air drill was used to perform corpectomies at C2–C6 down to the posterior longitudinal ligament. The posterior longitudinal ligament was dissected off of the anteriorly located dura to decompress the spinal cord, which was under significant stenosis from C2 to C6. Once the spinal cord was well decompressed, further traction was placed on the patient's extended head to correct the kyphosis. Next a cage filled with allograft bone and demineralized bone matrix was placed between C1 and C7 and the plate component was fastened with two C1 lateral mass screws and four additional screws placed in C7. This custom cage plate was manufactured based on pre-operative 3D CT imaging. Intraoperative AP and lateral radiographs showed excellent position of the cage-plate construct. After closure, the patient remained in a halo post-operatively for 3 days until stage 2 was to be performed. Time from skin incision to completion of wound closure for stage 1 lasted 830 min, and estimated blood loss totaled 600 mL.

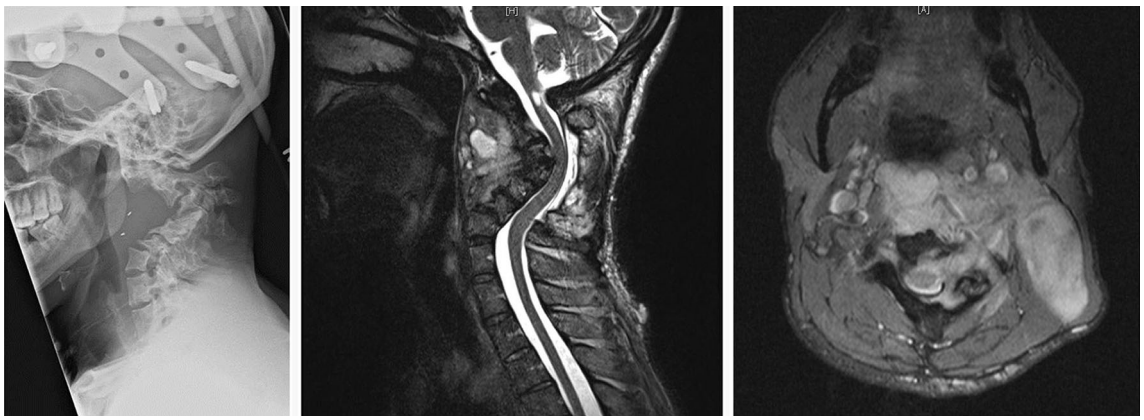

Fig. 1 Pre-operative radiograph and MRI depicting cervical kyphotic deformity and remodeling of the C2–C4 vertebral bodies

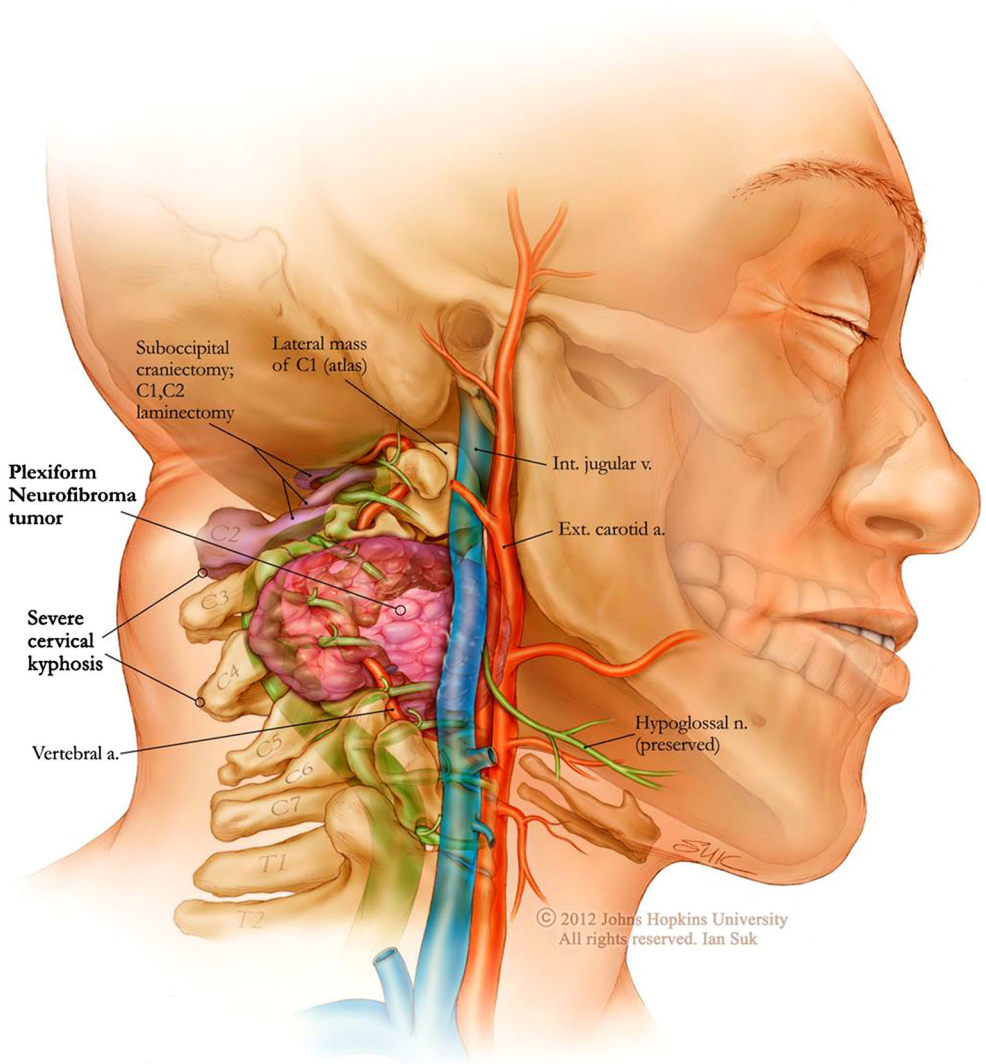

Fig. 2 Artist's illustration depicting anatomical location and disruption caused by the lesion

Stage 2

3 days later the patient was positioned prone on the operating room table with the halo in place and secured to the Mayfield frame. The patient's head was positioned in a neutral position. The posterior spine was exposed from occiput to T8. A small suboccipital craniectomy and C1 and C2 laminectomies were performed for additional decompression of the spinal cord. The T4 pedicles could not be cannulated, so DePuy Expedium pedicle screws were placed bilaterally in T1–T3 and T5–T8. Then lateral mass screws were placed bilaterally C3–C6 with the exception of C3 on the left. Next an occipital plate was placed in the midline. Tapered rods contoured from the occiput to T8 bilaterally were placed and secured to the screws. A third rod from the occiput to T8 was placed in the midline after removal of the T1–T7 spinous processes. This third rod was crosslinked to the lateral tapered rods at several positions for additional construct stability. Decortication of the occiput, lateral masses of C2–C7, bilateral lamina of C3–C7, and the bilateral lamina and transverse processes of T1–T8 were performed. Bone morphogenic protein, local autograft, allograft, and demineralized bone matrix were placed over the decorticated surfaces once the instrumentation was secured. Prior to closure, an intraoperative radiograph confirmed excellent position of both the anterior and posterior instrumentation. Time from skin incision to completion of wound closure for stage 2 lasted 500 min, and estimated blood loss totaled 800 mL. Postoperative cervico-thoracic radiograph, Fig. 3, and artist's depiction, Fig. 4, of construct.

Post-operative course

The patient's length of stay was 20 days, which included 10 days in the neuro-critical care unit. The patient's postoperative course was significant for decreased oxygen saturation secondary to pulmonary emboli on post-operative day two, which was managed with heparin. On postoperative day seven, the patient became febrile secondary to ventilator-associated pneumonia, which was managed with vancomycin, piperacillin, and tazobactam. The patient spent 2 weeks in an inpatient rehabilitation facility after discharge.

Follow-up/imaging

Patient-reported outcome questionnaires assessing pain (visual analog scale), disability (neck disability index), and quality of life (EQ-5D) demonstrated significant improvement from patient's baseline status pre-operatively, Fig. 5. The short-term improvement observed at the 4-month follow-up was maintained at 24-month follow-up. The patient has resumed normal activities and returned to work. He has full motor strength in all extremities and normal gait. The patient reports only minimal pain that is relieved with ibuprofen. Radiographs show stable position of the hardware from the occiput to T8 at long-term follow-up, Fig. 6.

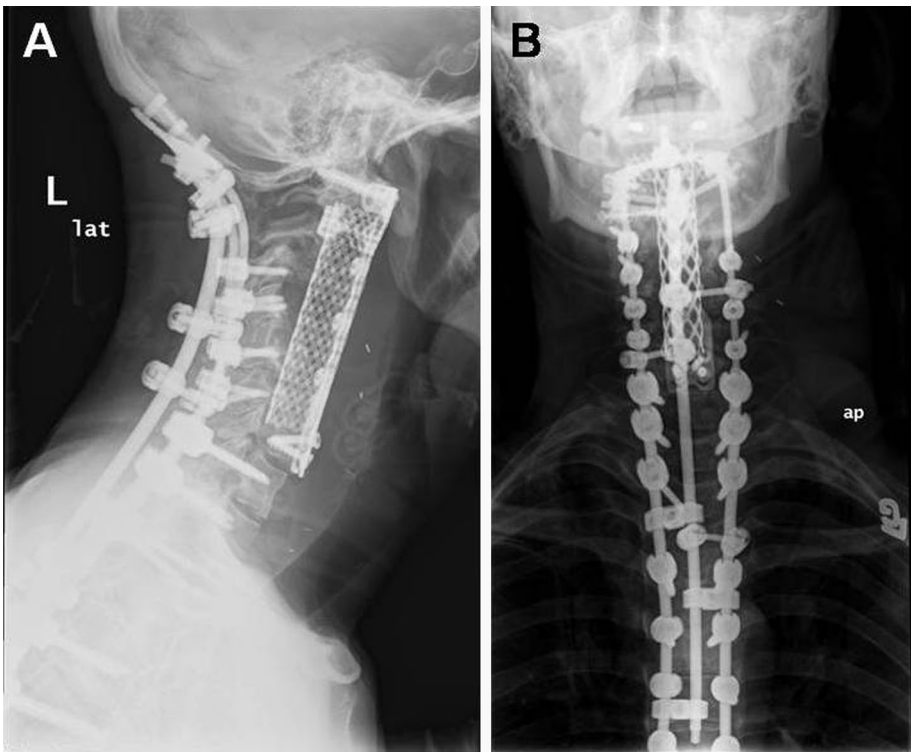

Fig. 3 Immediate post-operative sagittal (**a**) and frontal (**b**) radiographs demonstrating correction of kyphotic deformity and intact hardware in place

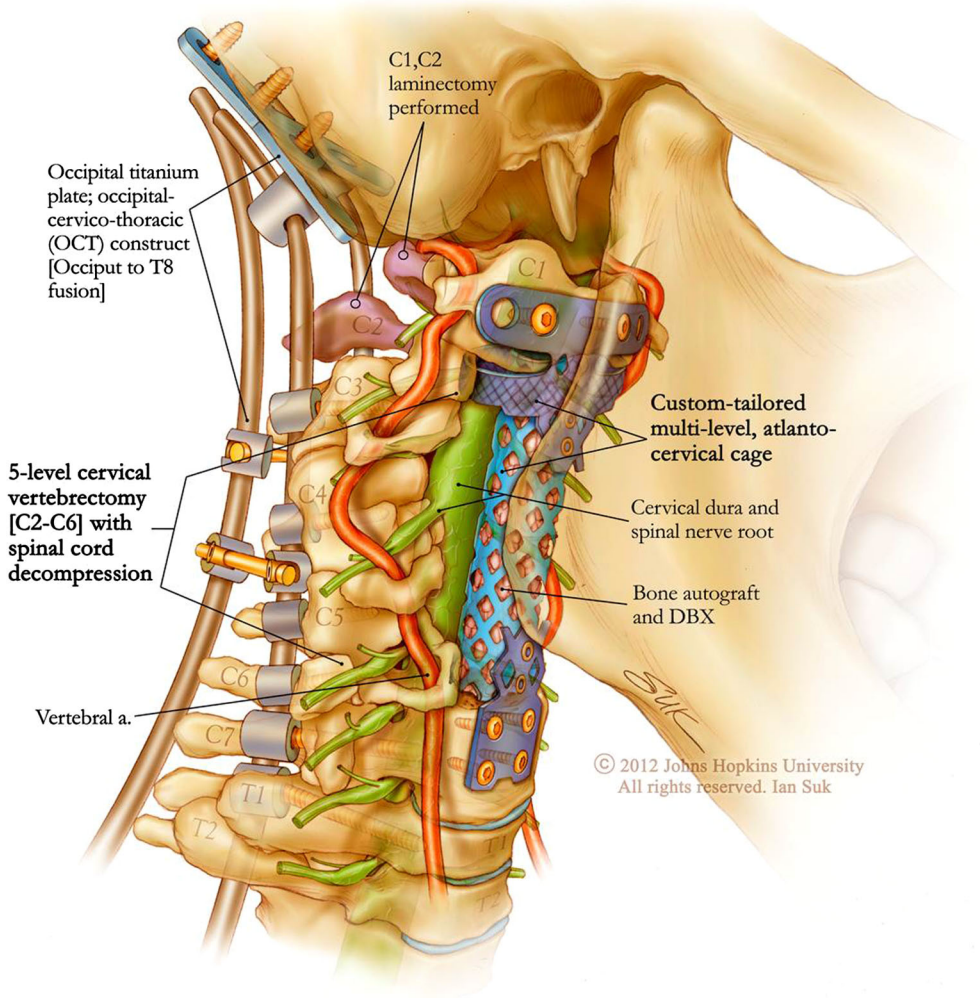

Fig. 4 Artist depiction of spinal construct

Discussion

There have been multiple reports of circumferential instrumented fusion for correction of cervical kyphotic deformity in the literature [8–10]. However, this is the first five-level cervical corpectomy with circumferential reconstruction to be described. Extreme cervical deformities represent a unique challenge to successful deformity reduction. In this case of severe kyphotic deformity of the cervical spine associated with C2–C7 cord compression and C1–C2 instability secondary to an anteriorly positioned plexiform neurofibroma, a two-staged procedure was planned. First, an anterior approach designed to resect the plexiform tumor and reduce the cervical kyphosis was performed. This was followed 3 days later by a posterior approach to stabilize the occipital-cervical-thoracic spine. Also, utilization of Halo vest-ring immobilization was mandatory to maintain the progressively improved alignment achieved between each of the staged surgeries.

Because there was significant cord signal change at C2 and the superior aspect of the tumor extended up to the level of C1, a transmandibular, circumglossal approach was required in this case. Further important anatomic factors of this case include presence of a severe kyphotic deformity at the level of C2, the apex of the cervical deformity being at C3, and instability at C1-2 (as demonstrated on pre-operative flexion–extension radiographs). Therefore, to achieve appropriate decompression, stabilization, tumor resection, and alignment; it was felt that C2 needed to be resected.

Multi-level cervical corpectomy comprising greater than two levels involves a complex surgical exposure and has been demonstrated to be associated with complications such as graft migration and pseudoarthrosis [3, 11–13]. In this case, a custom titanium cage and plate system manufactured from a pre-operative CT scan was utilized to optimize the fit of the cage and reduce the likelihood of cage displacement. As has been previously demonstrated,

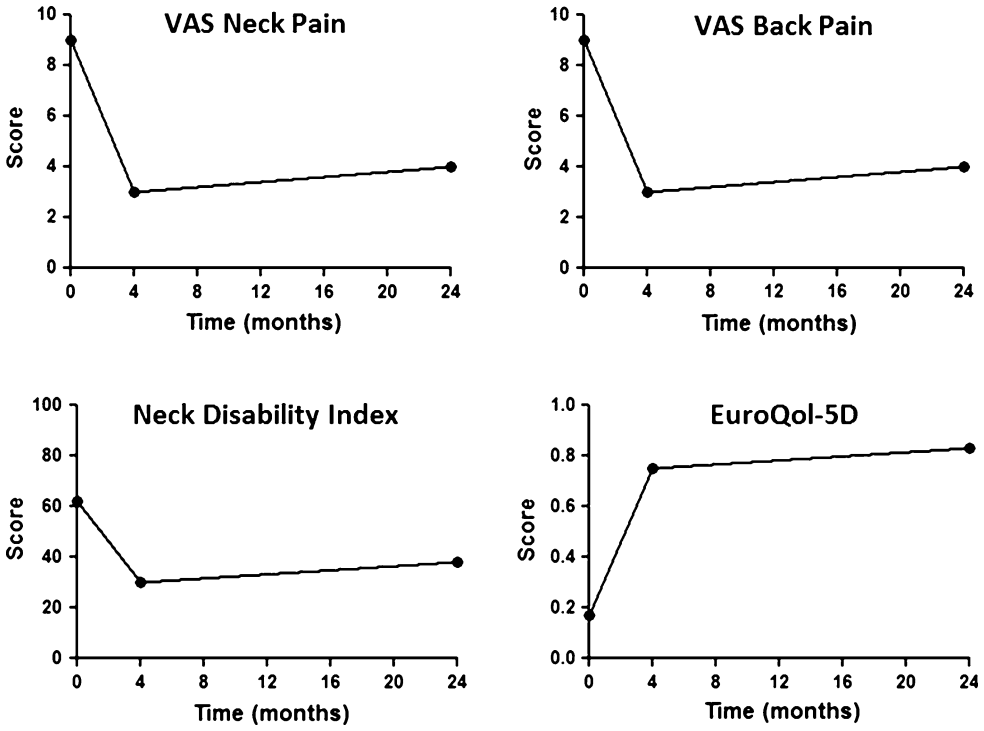

Fig. 5 Patient-assessed pain [visual analog scale (VAS)], disability [neck disability index (NDI)], and quality of life [EuroQol-5D (EQ-5D)] pre-operatively as well as 4 and 24 months post-operatively. The patient experienced improvement in each outcome measure

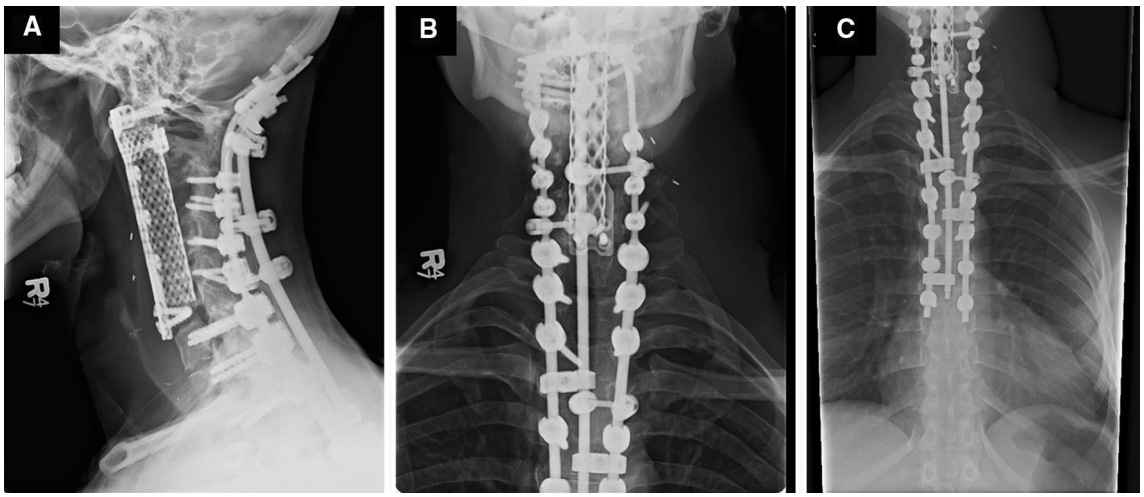

Fig. 6 Sagittal (**a**) and frontal (**b**, **c**) radiographs demonstrating stable correction at 2-year post-operatively

titanium mesh cages allow for optimal correction of sagittal alignment, better endplate purchase, variability in sizes, and the ability to be packed with auto- or allograft [5, 14, 15]. The use of anterior plate fixation has been shown to augment the stability and rigidity of the construct [1, 2, 16]. In this case the anterior plate was fastened with two C1 lateral mass screws and four additional screws placed in C7. Additionally, to help promote solid anterior fusion across five levels, the construct was supplemented with posterior fixation with pedicle screws which has been demonstrated to further reduce the rates of graft-related complications and pseudoarthrosis [1, 12, 17–19]. In our experience, tumor resections of this magnitude are particularly susceptible to construct failure. Although fixation three levels below the anterior construct is a valid option, we felt that extension of the construct posteriorly to the level just below the thoracic kyphosis in this particular patient would provide the most stable construct that would be least likely for hardware failure. As a result, the hardware was extended to T8 which, on pre-operative standing radiographs, represented the first level below the thoracic kyphosis.

The patient in this case series experienced a post-operative pulmonary embolus, a well-known complication following extensive surgical procedures. The immediate risk of adverse pulmonary complications related to the embolus required the use of anticoagulation, regardless of the potential for increased bleeding. It is our practice to initiate DVT prophylaxis (subcutaneous heparin) on all spine patients by post-operative day 3. Patients who develop DVT or PE within 72 h of surgery have to be treated on an individualized basis taking into account the type of procedure performed (for example, levels of cord exposed in the epidural space), time after the surgery, and whether there has been a DVT or PE. Certainly early post-operative DVTs may be treated with IVC filter placement, but those with a PE will often require heparinization despite recent spinal surgery and some inherent risk of epidural hematoma formation.

While longer-term follow-up will ultimately judge the overall success of this procedure, initial clinical results have been favorable. Patient-reported outcome measures reveal significant improvement in pain, disability, and quality of life. The patient's improvements have been durable through last follow-up at 24 months. The patient has been able to return to work and has limited pain treated with over the counter medications. Radiographically, the hardware construct remains stable with no signs of pseudoarthrosis or graft subsidence. Subsequent long-term follow-up CT has not been performed in this patient, as there has been no clinical indication. As a result, we cannot confirm radiographic solid fusion; however, if pseudoarthrosis is present, it has remained a symptomatic to date.

Five-level cervical corpectomy including C2 can be safely and successfully performed via a transmandibular, circumglossal approach. Circumferential reconstruction utilizing a custom anterior titanium cage and plate system manufactured from a pre-operative CT scan was utilized in this case. Long segment occipital-cervical-thoracic reconstruction is recommended in such a case. Using such a technique, improvement in myelopathy, correction of deformity, and improved quality of life can be achieved.

Conflict of interest None of the authors has any potential conflict of interest.

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