Case Report

Correction of cervical kyphoscoliosis, bisected spinal cord, and vertebral artery to epidural vein fistula in neurofibromatosis type 1

ABSTRACT

Neurofibromatosis-1 (NF1) presents complex challenges due to its multisystemic effects, including kyphoscoliosis, dural ectasia, and arteriovenous fistulas (AVF). We present a case of a 31-year-old male with NF1 exhibiting severe cervical kyphoscoliosis, dural ectasia, a bisected cervical cord, and an arteriovenous fistula, highlighting the intricacies of managing such intricate cases. Rapid weakening in the patient's right arm and leg prompted imaging revealing severe cervical kyphotic deformity and a dural fold dividing the spinal cord. Surgical intervention addressed a high-flow arteriovenous fistula involving the right vertebral artery and an epidural vein, necessitating sacrifice of the artery. Posterior fusion and laminectomy were performed, resulting in stable neurological status postoperatively and significant improvement in sensory loss and weakness at three months. This case underscores the importance of a tailored posterior-only approach, involving dural fold release, to allow the spinal cord to relocate to a less tense position, thus demonstrating effective decompression in complex NF1 cases with concurrent kyphotic deformity and vertebral artery AVF.

Keywords: Arteriovenous fistula, diastematomyelia, endovascular, kyphosis, neurofibromatosis type 1, posterior fixation, split cord

BACKGROUND AND IMPORTANCE

Neurofibromatosis type 1 (NF1) is an inherited condition characterized by café-au-lait spots and nerve tumors called neurofibromas. Some patients may also experience severe spinal deformities such as kyphoscoliosis,[1,2] and a rare complication known as dural ectasia, which usually occurs in the lower spine.[3] While AVFs, an even rarer complication, have been reported in the cervical spine,[4] they are usually discovered incidentally during imaging for spinal deformities, and their symptoms are primarily related to the pressure they exert rather than bleeding.[5]

CLINICAL PRESENTATION

This case report was deemed exempt from our institution's internal review board (IRB) approval. The patient consented to the procedure and was granted permission to describe their case. We have removed all critical patient identifiers.

Presentation

A 31-year-old male with NF1, initially showing few external signs, presented with months of numbness and tingling in his

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extremities, along with radicular pain during cervical flexion or extension movement. While being assessed for elective surgery, he suddenly experienced a rapid deterioration in his spinal cord function over 48 h. This resulted in significant muscle weakness, particularly in the right arm (2/5), and notable neurological signs including bilateral ankle clonus and a positive right-sided Hoffman's sign.

Imaging

Magnetic resonance imaging (MRI) imaging revealed a severe kyphotic deformity from the upper cervical to the midthoracic spine. There was severe cord compression at the apex of the kyphosis corresponding to the C4-5 region with significant malformation of the vertebral bodies. The spinal cord was found to be bisected at the C3-4 level from a posterior dural fold originating at C1 and terminating at C7 [Figure 1].

Surgical management and postoperative course

The surgical intervention was necessitated by a thick dorsal dural fold causing compression of the spinal cord. The chosen approach involved an extensive posterior fusion spanning from C2 to T4, coupled with a laminectomy from C2 to T3. The goal was to release the dura, allowing the spinal cord to shift backward into the region affected by dural ectasia. To determine the stopping point for the laminectomy, a vertical plumb line was drawn on a sagittal MRI image, guiding the removal of the lamina obstructing the cord's transition from cervical to thoracic regions [Figure 2].

Throughout the procedure, pre- and postpositioning neuromonitoring readings remained consistent. Due to anatomical constraints, pedicle screw fixation was viable only at C2 and T2-4. A trough laminectomy was executed from C2 through T3, revealing a decompressed thecal sac and a notable structure resembling a dilated epidural vein with brisk arterial-like flow along the right lateral recess. Ultrasound confirmed sufficient bony decompression, while Doppler ultrasound indicated arterial flow within the vessel, hinting at the presence of an arteriovenous fistula (AVF).

The vertical dural crease dividing the cord was carefully separated and sutured, progressively unfolding the dura and securing it laterally to the rod spanning from C2 to T4. This maneuver successfully decompressed the thecal sac and spinal cord, resolving the tethering issue. Subsequently, the remainder of the surgery was halted for thorough evaluation of the vascular anomaly, retaining temporary rods with provisional fixation.

A postoperative MRI revealed persistent lateral to medial compression from the arterialized epidural vein in the upper

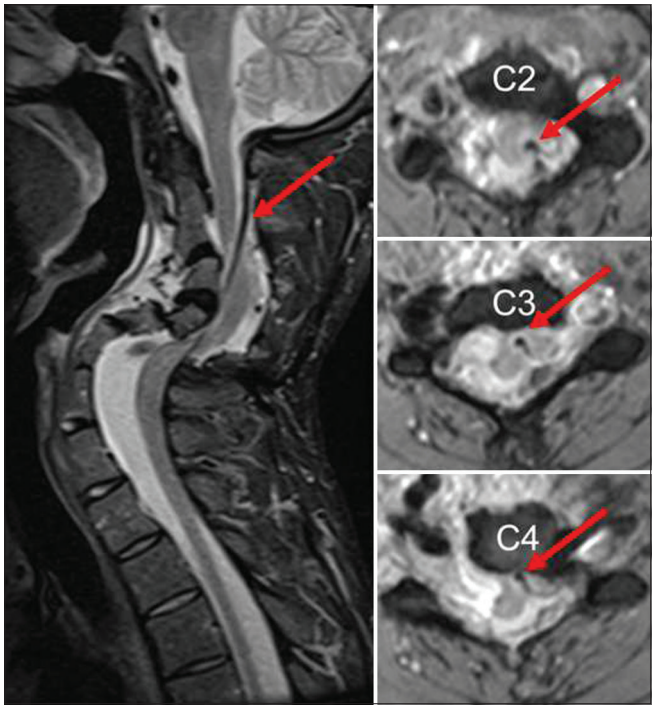

Figure 1: Left. Sagittal T2-weighted magnetic resonance imaging of the cervical and upper thoracic spine. Right. Axial T2-weighted magnetic resonance imaging centered behind the C2, C3, and C4 vertebral bodies. Red arrows. Dural fold originating at C1 and terminating at C7 bisecting the spinal cord at the C3-4 level

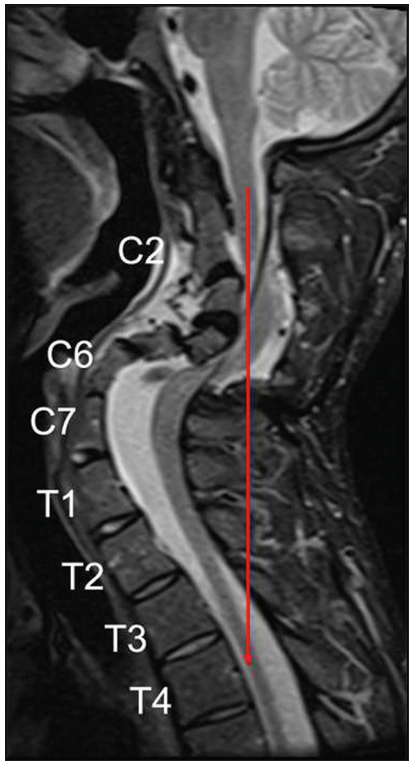

Figure 2: Sagittal T2-weighted magnetic resonance imaging of the cervical and upper thoracic spine. Red arrow. Plum line from the center of the spinal cord demonstrating the extent of laminectomy which must be performed for adequate spinal cord decompression

cervical spine. However, the lower spinal cord was completely decompressed, having migrated posteriorly beyond the spinal column [Figure 3]. Digital subtraction angiography exposed a high-flow direct right vertebral artery to the epidural vein fistula spanning multiple cervical segments. This necessitated the complete sacrifice of the V2 segment of the vertebral artery through catheter-based embolization with coils [Figure 4].

Within 1 day postsurgery, the patient exhibited significant strength improvement in the right upper extremity (4-/5), right lower extremity (5/5), and with stable left-hand grip (4/5). A subsequent operation was conducted to finalize the arthrodesis, implement bone grafting, and install a robust four-rod construct. In addition, a dorsal metal mesh was positioned to serve as a protective barrier over the spinal cord during potential future posterior cervical procedures in this area [Figure 5].

Three months postsurgery, the patient has shown significant improvement in most preoperative symptoms. While his left hand, previously affected by severe numbness and some loss of function, remained stable and did not worsen, his right hand regained full strength.

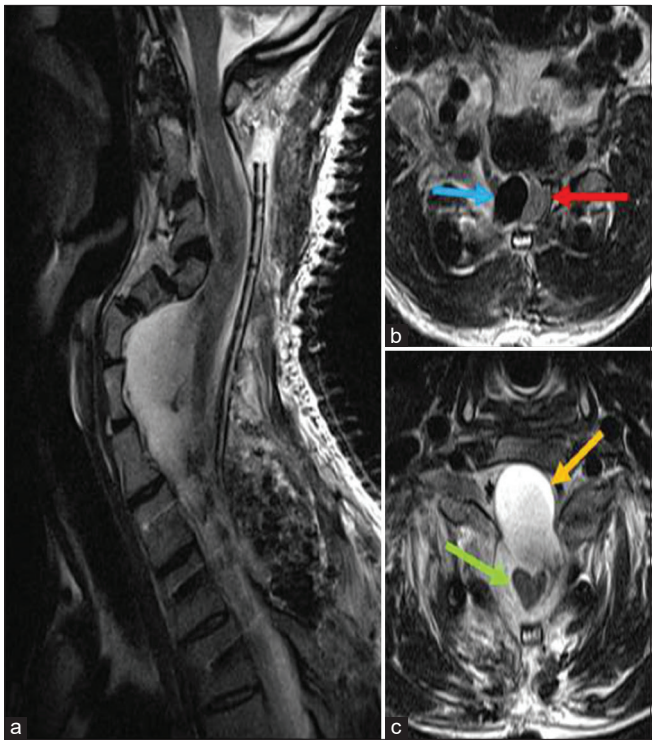

Figure 3: (a) Postoperative sagittal T2-weighted magnetic resonance imaging of the cervical and upper thoracic spine showing decompression and migration of the spinal cord posteriorly. (b) Axial T2-weighted magnetic resonance imaging of the cervical at the C2 level. Blue arrow. Large arterialized epidural vein not noted on prior imaging. Red arrow. Spinal cord now significantly compressed by arterialized epidural vein which was allowed to expand secondarily to cervical decompression. (c) Axial T2-weighted magnetic resonance imaging of the cervical at the C7 level. Yellow arrow. Empty spinal canal. Green arrow. Posteriorly migrated spinal cord post laminectomy

DISCUSSION

NF1 can lead to skeletal deformities, such as the dystrophic kyphoscoliosis[2] observed in this patient. While this manifestation typically occurs in the thoracolumbar spine, it can uncommonly present as a cervical deformity.[6] Early surgical intervention is advised to prevent progressive instability and neurological complications.[7] While a combined anterior–posterior approach is an option, posterior-only instrumentation, as employed in this case, has shown efficacy for initial stabilization in NF1 patients with dystrophic kyphosis.[7]

NF1 can also manifest with vascular abnormalities, including venous AVFs. Endovascular techniques are the primary treatment, resulting in symptom improvement with low morbidity rates.[8] Dural ectasia,[9] occurring in only 10% of NF1 cases, is often accompanied by spinal deformities. This case is unique as it combines a cervical kyphotic deformity, vertebral AV fistula, and severe dural ectasia causing spinal cord bisection, a rare occurrence not previously reported.

This case is the first documented instance of simultaneous cervical kyphotic deformity, vertebral AV fistula, and severe dural ectasia. Only one other case involving severe kyphoscoliosis and a vertebral AV fistula exists in the literature, treated solely with endovascular coiling.[5] In addition, this is only the third reported case of cervical dural ectasia coexisting with dystrophic kyphosis.[10]

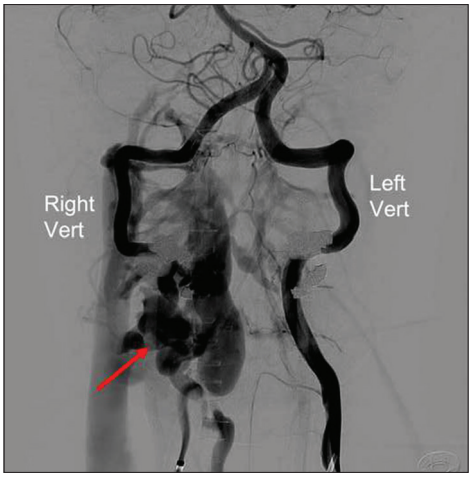

Figure 4: Mid-phase left vertebral artery digital subtraction angiography demonstrating retrograde flow into the right vertebral artery to the arteriovenous fistula. Red arrow. Multilevel vertebral artery to epidural vein fistula with large draining arterialized epidural vein centrally

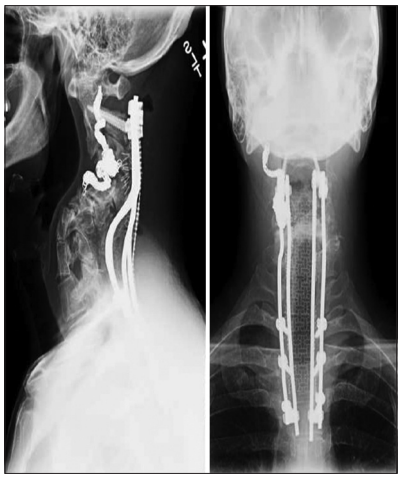

Figure 5: Sagittal and Anteroposterior (AP) postoperative X-rays showing posterior cervical instrumentation from C2 to T4 with 4 rod rigid fixation and coil sacrifice of the V2 segment of the right vertebral artery. Metal mesh can be seen on the AP view protecting the posteriorly migrated spinal cord

CONCLUSION

In the rare case of a patient with NF1 exhibiting severe kyphotic deformity, dural ectasia leading to spinal cord dissection, and a vertebral AV fistula, the primary treatment goals involve stabilizing the spine and addressing the vascular anomaly. While endovascular therapy for AV fistulas is standard in NF1, it is rare for a patient to also require surgical correction for severe kyphosis. The unfolding of the bisected dural leaflet is a safe technique to create adequate space for the spinal cord. This case demonstrates that a posterior-only approach for decompression and fixation can effectively stabilize the deformity, especially in the presence of challenging surgical anatomy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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