Motion-related vascular abnormalities at the craniocervical junction: illustrative case series and literature review

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The craniocervical junction (CCJ) functions within a complicated regional anatomy necessary to protect and support vital neurovascular structures. In select instances, vascular pathology can be attributed to this complicated interplay of motion and structure found within this narrow space. The authors report 3 cases of complex vascular pathology related to motion at the CCJ and detail the management of these cases. Two cases involved posterior circulation vascular compression syndromes, and one case involved a vascular anomaly and its relation to aneurysm formation and rupture. The patient in Case 1 was a 66-year-old man with a history of syncopal episodes resulting from the bilateral vertebral artery becoming occluded when he rotated his head. Successful microsurgical decompression at the skull base resulted in patent bilateral vertebral artery V3 segments upon head movement in all directions. The patient in Case 2 was a 53-year-old woman who underwent elective resection of a right temporal meningioma and who experienced postoperative drowsiness, dysphagia, and mild right-arm ataxia. Subsequent MRI demonstrated bilateral posterior inferior cerebellar artery (PICA) strokes. Cerebral angiography showed a single PICA, of extradural origin, supplying both cerebellar hemispheres. The PICA exhibited dynamic extradural compression when the patient rotated her head; the bilateral PICA strokes were due to head rotation during surgical positioning. In Case 3, a 37-year-old woman found unconscious in her home had diffuse subarachnoid hemorrhage and evidence of a right PICA aneurysm. A right far-lateral craniectomy was performed for aneurysm clipping, and she was found to have a dissecting aneurysm with an associated PICA originating extradurally. There was a shearing phenomenon of the extradural PICA along the dura of the foramen magnum, and this microtraumatic stress imposed on the vessel resulted in a dissecting aneurysm. This series of complex and unusual cases highlights the authors’ understanding of vascular pathology of the CCJ and its management.

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Manipulation of the neck was first implicated as a potential source of vertebral artery compression in 1947. Manipulation-induced compression can occur through various mechanisms; rotation, extension, or flexion of the neck may also cause compression. Extrinsic compression of the vertebral artery by osteophytes during head rotation was demonstrated initially in cadaveric specimens and subsequently in living patients. The atlas loop segment of the extracranial vertebral artery is susceptible to mechanical compression when the neck is rotated toward the contralateral side. This can result in hindbrain ischemia and/or vessel injury. The resulting vertebrobasilar insufficiency can cause syncope or near syncope, blurred vision, dizziness, vertigo, drop attacks, tinnitus, hypacusis, and sensory or motor deficits. The symptoms are typically alleviated once the head is returned to a neutral position. In 1978, initially described the phenomenon of a reversible neurological deficit after head turning (bow hunter’s syndrome) in a patient who suffered a posterior fossa stroke after archery practice.

We present various symptomatic cases of vascular pa-
thology related to motion at the craniocervical junction (CCJ). We include two illustrative cases of extradural arterial compression—one of a patient with bilateral V₃ segment occlusion occurring upon head rotation and one of a patient with bilateral posterior inferior cerebellar artery (PICA) strokes caused when the artery as a solitary feeder with an extradural origin became compressed during patient positioning for an unrelated surgical procedure—and one case highlighting an extradural PICA origin in which stress at the edge of the dura of the foramen magnum resulted in a ruptured dissecting PICA aneurysm.

**Case Reports**

**Case 1**

**Clinical Presentation and Imaging**

A 66-year-old man presented with repeated episodes of syncope and loss of consciousness that occurred with head rotation. A CT scan demonstrated extradural vertebral artery compression by the occipital bone and posterior arch of C-1. The compression was exacerbated when the patient turned his head, causing the symptoms. Cerebral angiography confirmed these findings, demonstrating bilateral occlusion of the vertebral arteries at the V₃ segment upon head rotation (Fig. 1).

**Surgical Intervention**

The patient was positioned prone on the operating table with his head in a three-point clamp. Intraoperative angiography was performed to rule out iatrogenic vertebral artery occlusion. Intraoperative somatosensory evoked potentials and motor evoked potentials were monitored throughout the entire procedure, and no significant change was observed.

A standard posterior midline approach to the CCJ was performed. The arch of C-1 was obscured by the inferior portion of the occiput narrowing the interlaminar space between the occiput and C-1, which resulted in compression of the vertebral arteries at the foramen magnum. A suboccipital decompression with C-1 laminectomy was performed. The laminectomy was carried out laterally on C-1 until the vertebral artery was visualized within the J-groove. The suboccipital decompression was performed to ensure full decompression of the course and intradural entry of the vertebral artery.

**Outcome**

The patient tolerated the procedure well. A dynamic cerebral angiogram after the surgery confirmed adequate decompression without further motion-related occlusion of the vertebral arteries (Fig. 2).

**Case 2**

**Clinical Presentation and Imaging**

A 53-year-old woman presented with worsening headaches, forgetfulness, and a recent decline in her work performance. MRI demonstrated a 4-cm, enhancing, dural-based mass over the right temporal lobe with internal gradient susceptibility, consistent with a meningioma with areas of calcification (Fig. 3). Because of these findings, the patient agreed to the recommendation that she undergo a right temporal craniotomy for resection of the mass.

**Surgical Intervention**

The patient was placed supine on the operating table with her head rotated 45° to the contralateral side and slightly extended in a three-point head clamp. A right-side temporal craniotomy and resection of the meningioma were performed in the standard fashion, and there was no significant blood loss, hypotension, or hemodynamic lability. The patient was extubated in the operating room.
and taken to the intensive care unit for close observation and monitoring.

Postoperative Imaging and Follow-Up

Although the patient could follow simple commands postoperatively, she remained drowsy and had difficulty swallowing and mild right-arm ataxia. Postoperative MRI demonstrated matched regions of T2-weighted/FLAIR signal hyperintensity and diffusion restriction in the bilateral medial cerebellar hemispheres and the cerebellar vermis, with sparing of the deeper cerebellar parenchyma and the brainstem (Fig. 4). These findings were concern-

FIG. 2. Postoperative cerebral angiograms. After suboccipital decompression with head rotated and neck extended, the left and right vertebral arteries are patent.

FIG. 3. Contrast-enhanced T1-weighted image demonstrating a large right temporal meningioma.

FIG. 4. Routine MR image, obtained on postoperative Day 1, demonstrating diffusion restriction in the bilateral medial cerebellar hemispheres and vermis. These regions had matched T2/FLAIR signal abnormality consistent with an acute stroke.

FIG. 5. Right vertebral artery injection angiograms, anteroposterior (left) and lateral oblique (right) views, depicting an extradural origin of the right PICA, which supplies both cerebellar hemispheres.

ing for an acute infarction involving the distribution of both PICAs.

Cerebral angiography demonstrated a large right PICA supplying both cerebellar hemispheres, with an extradural origin below the skull base (Fig. 5). Angiography performed with the patient’s head extended and turned toward the left and right demonstrated occlusion of the extradural portion of the right PICA; there was contrast stasis in the intradural portion of the vessel and delayed filling of both cerebellar hemispheres (Fig. 6).

Further workup for detection of a stroke, including an echocardiogram, demonstrated no other significant findings. The patient was started on aspirin (81 mg daily) and subsequently discharged to an inpatient rehabilitation facility.
Clinical Presentation and Imaging

A 37-year-old woman presented with a Hunt and Hess Grade IV/Fisher Grade 4 subarachnoid hemorrhage, with blood in the posterior fossa and basilar cisterns. She had complained of neck pain 2 days prior to her hemorrhage. Angiography demonstrated an irregular 10-mm aneurysm along the proximal right PICA (Video 1).

Case 3

Clinical Presentation and Imaging

A 37-year-old woman presented with a Hunt and Hess Grade IV/Fisher Grade 4 subarachnoid hemorrhage, with blood in the posterior fossa and basilar cisterns. She had complained of neck pain 2 days prior to her hemorrhage. Angiography demonstrated an irregular 10-mm aneurysm along the proximal right PICA (Video 1).

VIDEO 1. Operative video demonstrating a far-lateral approach, dissection, and clipping of dural-associated PICA aneurysm. Copyright Department of Neurosurgery, University of Utah. Published with permission. Click here to view with Media Player. Click here to view with Quicktime.

The right PICA originated extradurally from the vertebral artery and exhibited contoured grooving at the point where it made its intradural entry.

Surgical Intervention

The patient underwent a right far-lateral craniotomy for aneurysm clipping and decompression of the dura over the PICA. The vertebral artery was mobilized from its attachment in the J-groove of C-1. The course of the vertebral artery was followed, and the extradural PICA takeoff was identified. The PICA aneurysm was visualized at the dural edge. It appeared that shearing forces between the dural band and the PICA had contributed to the formation of a dissecting aneurysm. The aneurysm was primarily clipped using a single curved clip. Distal PICA flow was determined with direct visualization, Doppler ultrasound, and indocyanine green angiography (Video 1).

Discussion

We have presented three unusual and complex cases of symptomatic vascular lesions of the CCJ that further our understanding of these infrequently encountered pathologies.

Bilateral Dynamic Arterial Compression

Diagnosing vertebrobasilar insufficiency and ischemia due to motion requires a thorough evaluation and careful exclusion of cardiac and other potential systemic causes that may mimic the symptomatology. Dynamic digital subtraction angiography of the cerebral vasculature should be performed as part of the initial workup. Treatment may entail an aggressive anticoagulation regimen or surgical decompression based on the extent of compression, severity of symptoms, and/or etiology of occlusion.

Approximately 75 cases of bow hunter’s syndrome have been described in the literature, with 4 large series having been reported (Table 1). The pathophysiology associated with this entity is thought to be transient compression of the dominant vertebral artery against a fibrous band or bony prominence that occurs on rotation of head. The dynamic occlusion of the dominant vertebral artery in the setting of a hypoplastic or occluded contralateral vertebral artery can lead to hemodynamic compromise in the vertebrobasilar territory. These symptoms may be attributable to asymmetrical excitation of the bilateral labyrinth induced by transient ischemia or by disinhibition from inferior cerebellar hypoperfusion. Affected patients classically exhibit stenosis or anomaly of the vertebral artery on the affected side, with hypoplasia or termination of the contralateral vertebral artery in the PICA. The dominant vertebral artery, thus, becomes compressed at the C1–2 level during contralateral head rotation resulting in compromised blood flow and subsequent ischemia.

We described one patient who experienced bilateral compression of the vertebral arteries at the level of the foramen magnum with head motion. There have been three previously described cases of bilateral vertebral artery compression in this setting. Healy et al. described the successful treatment of a 58-year-old man with 90% stenosis of the right vertebral artery at C4–5 and complete occlusion of the left vertebral artery at C1–2 with head rotation to the right. The authors successfully treated the patient by performing a C1–2 left transverse foramen decompression and C2–6 fusion. Tsutsumi et al. reported on a case of bilateral vertebral artery compression at C5–6 and C6–7 in a patient who was successfully treated with anterior decompression and fusion at both levels. Vilela et al. reported a case of bilateral vertebral artery compression at C1–2 that was treated initially with decompression and then with C1–2 fixation for persistent symptoms and the patient subsequently improved after fusion. To our knowledge, ours is the first report of bilateral bow hunter’s syndrome resulting from bony compression at the CCJ compressing the vertebral arteries at the level of the foramen magnum.

Treatment for bow hunter’s syndrome largely depends on the level and severity of compression. The most commonly affected levels are C1–2 (50%) and C5–7 (50%).
A posterior approach is generally used for craniocervical vertebral artery compression (as in Case 1), while an anterior approach is often used for subaxial compression. Typically, decompression without fusion is sufficient to relieve the symptoms of motion-induced ischemia. While the syndrome describes the classic etiology, cases of rotational vertebral artery occlusion can have atypical patterns. Patients may have compression of the vertebral artery at other cervical levels simultaneously compression of bilateral vertebral arteries, compression of the dominant vertebral artery during ipsilateral head rotation or tilt, or compression of the nondominant vertebral artery terminating in the PICA. In our two cases of extradural dynamic compression, neutral imaging demonstrated normal vertebral arteries with bilateral compromise on head rotation, a phenomenon that has been previously described. Given these findings, the diagnosis of dynamic vertebral artery compression should be considered and investigated with provocative imaging.

Lu et al. have reported on the largest series in which 9 patients suffering from rotational vertebral artery occlusion underwent surgery. Seven patients had left-sided occlusion and two patients had right-sided occlusion. In their series, 4 of the 9 patients had C-1 compression. In contrast with our two cases, none of their patients had bilateral compression. The authors used an anterior cervical approach in patients with lesions at or caudal to C-4 (n = 4), a far-lateral approach in patients with lesions rostral to C-4 (n = 4), and a minimally invasive approach in 1 patient with a suboccipital/C-1 lesion. None of their patients required a suboccipital decompression for vertebral artery decompression.

In 2013, Choi et al. reported on 21 patients with rotational vertebral artery occlusion. One case involved a 37-year-old man with fainting spells and blurred vision, who had bilateral compression at C1–2. Cerebral angiography showed normal findings, the patient was treated conservatively, and there was no evidence of stroke in the 13-month follow-up period. In their series, 19 of the 21 patients were treated conservatively and had favorable outcomes, as was the case in our patient (Case 2).

Vilela et al. reported on 10 patients treated surgically for rotational verteobasilar ischemia. The procedures were tailored for each patient depending on the location, pathogenesis, and mechanism of compression. Five patients underwent removal of osteophytes at the level of the subaxial cervical spine; 1 patient, a discectomy; 2 patients, C1–2 decompression; and 2 patients, decompression and fusion at C1–2. Our patient (Case 1) was symptomatic for some time with dynamic occlusion documented on angiography. Thus, surgical intervention was deemed necessary to prevent further, irreversible neurological deficit.

Conservative, surgical, and endovascular therapies have been described as a treatment for bow hunter’s syndrome. In the series by Darkhabani et al., 3 patients underwent stenting of the dominant vertebral artery and 1 underwent stenting of the nondominant vertebral artery.

**Extradural PICA Take-Off With An Associated Dissecting Aneurysm**

The origin of the PICA from the vertebral artery is generally intradural and anterolateral to the medulla at the anterior medullary cistern. In Case 3, the patient had a PICA that originated extradurally on the right side, with an associated dissecting aneurysm. An extracranial, extradural origin of the PICA from the vertebral artery is seen in approximately 5% of cases. Aneurysms arising from an extracranial, extradural PICA are exceedingly rare. In 2009, Bhat et al. reported a case of a 35-year-old woman who presented with giddiness and headaches and was found to have fourth ventricular hemorrhage. Subsequent vessel imaging demonstrated a bilateral extracranial, extradural origin of the PICAs between C-1 and C-2 and associated saccular aneurysms. This patient underwent a suboccipital craniectomy, C-1 arch excision, and clipping of both aneurysms.

In our case, we identified a fibrous ring at the entry of PICA into the foramen magnum and dura. We propose that vessel contact with the fibrous ring may have caused a shearing mechanism, with subsequent vascular trauma resulting in aneurysm formation. Whether the patient had a preceding traumatic event is unknown. There are no similar cases published in the literature. There have previously been 13 cases of extracranial distal aneurysms of the PICA. The location of the aneurysm and its intimate relationship with the fibrous band at the craniovertebral junction is a rare example of the variable vascular anatomy and biomechanical interplay at this location. The imposed microtraumatic stress on the vessel resulted in a
dissecting aneurysm. We theorize that similar cases exist and a low threshold must be held for such pathology in this setting. Knowledge of the origin of the PICA and associated pathology is crucial, as key structures, including the medulla oblongata, lateral brainstem, and cerebellar hemispheres, are fed by perforating branches.

Conclusions

We report three unusual cases of vascular pathology at the CCJ: two cases of dynamic arterial compression and one case of an extradural PICA origin with an associated dissecting aneurysm. Vascular pathology of the CCJ is complex and may be difficult to diagnose. Provocative imaging should be considered as part of the workup when dynamic vascular pathology is suspected. Depending on the etiology and symptoms at presentation, surgical intervention may be required.

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Supplemental Information
Videos


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